Complete Interruption of the Aortic Arch

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A LOCALIZED NARROWING of the aorta at or distal to the ductus arteriosus (coarctation of the adult type) is one of the most common congenital malformations of the great vessels. A narrowing of the aorta proximal to a patent ductus arteriosus (coarctation of the infantile type) is less common, while complete interruption of the aorta proximal to the ductus is quite rare, and interruption distal to the ductus has not been recorded. Interruption of the aortic arch, also termed absence, interruption, or atresia of the isthmus of the aorta, functionally corresponds to the most severe form of the infantile type of coarctation.

The adult type of coarctation of the aorta is relatively easily recognized by clinical means and the results of surgical treatment are, in general, good. The diagnosis of complete interruption of the aorta, on the other hand, has rarely been made before death or exploratory operation and, although it too is amenable to surgical correction or palliation, few attempts at operative treatment have been made. Three patients with complete interruption of the aortic arch have been studied at the National Heart Institute. In the present report the clinical, hemodynamic, angiographic, and pathologic findings in these patients are described and the information derived from 52 previously reported patients is reviewed.

Embryology

A pathologic classification used by Celoria and Patton¹ of aortic arch interruption, based on the sites of origin of the branches of the aortic arch, has been found applicable in the present study. The embryologic mechanisms by which the various types of aortic arch interruption may occur are illustrated schematically in figure 1. The type A interruption, which occurs immediately distal to the left subclavian artery, appears to result from a regression or atrophy of the segment of left dorsal aorta between the ductus arteriosus and the left subclavian artery (fig. 2A). In the type B deformity, the interruption is just distal to the origin of the left carotid artery and represents a failure of formation of the left fourth arch, since the left subclavian artery arises from the descending aorta (fig. 2B). The pathogenesis of the type C malformation, in which only the right subclavian and right carotid arteries arise from the ascending aorta (fig. 2C), is less clear. Celoria and Patton suggest two possible mechanisms: (1) partial or complete failure of formation of the left third and fourth aortic arches and persistence of the segment of dorsal aorta between these arches as the left common carotid artery, or, (2) a failure of connection of the outgrowth of the aortic sac with the third and fourth arches and fusion of these two arches to form the left common carotid artery.

Four variations of the basic type B interruption have been reported (fig. 3). In two of these (b and d) the ductus persists on the right as well as on the left. Bilateral persistence of the ducti (double ductus arteriosus) occurs when the entire right sixth arch and part of the right dorsal aorta remain. Each ductus connects the subclavian artery with the pulmonary artery on the respective side. According to Wagenvoort et al.,² the embryologic basis for the origin of the right pulmonary artery from the ascending aorta (fig. 3C) is the persistence of part of the right dorsal aorta and of the distal part of the right sixth arch.

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Schematic representation of the great arteries arising from the heart in the fetus. The primitive aortic arches from which the arteries were derived are indicated. Also designated (lined areas) are the sites where defective development produces types A and B complete interruptions of the aortic arch (upper diagram), and type C (lower diagram). (Reproduced with permission of the authors and publisher, C. V. Mosby Co., from Celoria, G. C., and Patton, R. B., Am. Heart J. 58: 407, 1959.)

Clinical Summaries

Case 1
L.N. (no. 01-58-99), a 25-month-old boy, was the product of a full-term normal pregnancy. During the first few months of life he had frequent upper respiratory infections, choking and coughing spells, and occasionally syncope during more severe coughing episodes. At 4 months he was found to have a heart murmur. At 14 months of age cardiac catheterization at another hospital...
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Figure 2
Diagrammatic illustrations of the three principal anatomic types of complete interruption of the aortic arch. P.T., pulmonary trunk; P.D.A., ductus arteriosus; P.A., pulmonary artery; R.S., right subclavian artery; R.C.C., right common carotid artery; L.C.C., left common carotid artery; L.S., left subclavian artery.

Figure 3
Diagrammatic illustrations of four variations of basic type B aortic arch interruption. In b. and d. the ductus arteriosus persists on the right as well as on the left. Abbreviations as in figure 2.

showed equal systolic pressures (76 mm. Hg) in the right ventricle, pulmonary artery, and left ventricle. A ventricular septal defect was crossed by the catheter. Left-to-right shunts entering both the right atrium and the right ventricle were demonstrated by oximetry. In the ensuing 9 months he grew and developed normally and never evidenced cardiac failure. Two months before admission, however, he had several "spells" of choking and cyanosis and his respiration became progressively more difficult.

On admission the child was normally developed.
The radial and femoral pulses were full and equal. The fingertips of the right hand were redder than those of the left, but the nailbeds of the fingers of both hands, as well as the toes, were cyanotic; there was no clubbing. The left precordium was bulging, and both left and right ventricular lifts and a systolic thrill to the left of the sternum were palpable. The second heart sound was louder in the pulmonary than in the aortic area and a grade 4/6 systolic murmur was audible over the entire precordium, loudest in the fourth and fifth intercostal spaces along the left sternal border.

The hematocrit value was 50 per cent. Fluoroscopic and radiographic examinations (fig. 4A) revealed generalized cardiac enlargement with particular prominence of the right ventricle, left atrium, and main pulmonary artery, and increased pulmonary vascularity. The electrocardiogram (fig. 5A) demonstrated right axis deviation, right ventricular hypertrophy, bi-atrial enlargement, and sinus tachycardia. Right heart catheterization disclosed a mean right atrial pressure of 13 mm. Hg, a right ventricular pressure of 88/16, and a femoral arterial pressure of 90/50 mm. Hg. The pulmonary artery could not be entered. Serial blood samples from both venae cavae, the right atrium, and the right ventricle revealed an increase in oxygen content of 2.2 volumes per cent in the atrium and an additional rise of 1.1 volumes per cent in the ventricle. Blood from the femoral artery had an oxygen saturation of 83.3 per cent.

Shortly following these studies the child became severely dyspneic and cyanotic, and atrial flutter developed. With the administration of digitalis the rhythm became regular. One week later bilateral bronchopneumonia developed and he died in heart failure.

Pathologic Findings

At autopsy, the heart (fig. 6) was greatly enlarged, weighing 125 Gm. (anticipated weight, 65 Gm.). The foramen ovale was closed. The right ventricle was dilated and hypertrophied. A defect, measuring 1.1 cm. in greatest diameter and bordered entirely by myocardium, was present in the basal portion of the ventricular septum, directly beneath the pulmonary valve and a portion of the hypertrophied crista supraventricularis and immediately cephalad to the papillary muscle of the conus. The right ventricular outflow tract, pulmonary valve, and pulmonary trunk and all its branches were greatly dilated. The large pulmonary trunk overrode the ventricular septal defect to take origin in part from the left ventricle. A patent ductus arteriosus, measuring 2.0 cm. in circumference and 1.5 cm. in length, arose from the pulmonary trunk and gave origin to the descending thoracic aorta. The intima of the patent ductus was irregular, in contrast to the smooth linings of the adjacent aorta and pulmonary artery. The left subclavian artery arose from the patent ductus. The pulmonary veins, left atrium, and mitral valve were normal. The left ventricle was dilated and hypertrophied. The outflow tract of the left ventricle was narrowed by a muscular ridge immediately cephalad to the ventricular septal defect. The aortic valve, which was bicuspid, measured only 3.2 cm. in circumference while the pulmonary valve was 6.5 cm. in circumference. The origin and distribution of the coronary arteries

*Figure 4*

Posteroanterior roentgenograms of chest in patients L. N. (A) and P. S. (B).
Case 2

R.S. (no. 03-13-50), a male infant, was admitted to the National Heart Institute at the age of 10 days. He was the product of a full-term normal pregnancy and delivery, but at birth was cyanotic, markedly dyspneic, and had a heart murmur. On his eighth day of life he was digitalized because of evidence of congestive heart failure.

On admission he was well-developed, but undernourished (weight 2,550 Gm.), and in moderate distress. The pulse and respirations were rapid. There was mild cyanosis of the nailbeds, similar in the fingers and toes. The brachial pulses were bounding but no femoral pulses could be felt. The systolic blood pressure, recorded by the flush technie, was 100 mm. Hg in each arm and 90
Figure 6

Drawing of heart and great vessels of patient L. N. A. The ascending aorta (Ao.) terminates after giving rise to the left common carotid artery (type B interruption). The descending aorta arises entirely from the pulmonary trunk (P.T.) via the patent ductus arteriosus. The left subclavian artery arises from the "pulmonary-ductus-descending aorta trunk." The pulmonary trunk is markedly dilated and the ascending aorta relatively hypoplastic. The right ventricle is dilated and hypertrophied. B. The right ventricle and pulmonic valve are opened. The ventricular septal defect (V.S.D.) is located below (caudal to) a portion of the crista supraventricularis muscle but above (cephalad to) the papillary muscle of the conus. C. The anterior portion
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mm. Hg in the left leg. The left precordium was prominent, a right ventricular lift and a systolic thrill in the suprasternal notch were palpable. The second sound was single at the base and a third heart sound was audible at the apex. A grade 4/6 harsh systolic murmur, maximal in the fourth left intercostal space, was heard over the entire precordium and also in the neck and back. A grade 2/6 early diastolic murmur was audible in the third left interspace.

The hematocrit value was 45 per cent. The chest roentgenogram showed cardiomegaly and increased pulmonary vascularity. The electrocardiogram (fig. 5B) disclosed sinus tachycardia, deviation of the T waves to the right, and digitalis effect.

At cardiac catheterization the mean right atrial pressure was 8 mm. Hg. An interatrial communication was crossed by the catheter and the mean left atrial pressure was 10 mm. Hg. The right ventricular pressure was 132/18 mm. Hg. A vessel considered to be the main pulmonary artery was entered from the right ventricle and a pressure of 132/48 mm. Hg was recorded within it. The oxygen content (volumes per cent) of blood from various sites was: inferior vena cava, 5.3; right atrium, 5.5; right ventricle, 7.0; right pulmonary artery, 9.3; and left atrium, 14.6. The femoral arterial oxygen saturation was 71 per cent. A selective angiocardiogram was performed with injection into the left atrium. The first film showed opacification of the left atrium, left ventricle, right ventricle, and a common arterial trunk arising from both ventricles. The main pulmonary artery appeared to arise from the common trunk. The aorta, originating also from the truncus, terminated by dividing into two innominate arteries, each of which gave rise to carotid and subclavian arteries bilaterally. From these studies diagnoses of ventricular septal defect, persistent common truncus arteriosus, patent ductus arteriosus, and complete interruption of the aortic arch were made. Shortly following the catheterization procedures the infant became severely cyanotic, left heart failure was evident, and he died 2 days later.

Pathologic Findings

At autopsy, both ventricles were dilated and hypertrophied (fig. 8). A single arterial trunk arose from the base of the heart. The semilunar valve at the base of the trunk was composed of only two cusps, the margins of which were corrugated and thickened. The ostia of both coronary arteries were located behind the right cusp but each artery was normally distributed. The common trunk divided into the aorta and main pulmonary artery. The aorta terminated after giving rise to right and left innominate arteries. The main pulmonary artery divided normally and, through a short, "coarcted" ductus arteriosus, communicated with the descending aorta. A ventricular septal defect, measuring 0.9 cm. in largest diameter, was present just beneath the semilunar valve. It involved the entire membranous septum and a portion of the muscular septum. A large, valvular incompetent, foramen ovale was present. Microscopically, there was thickening of the walls of the pulmonary arterioles and muscular arteries without intimal proliferation (Heath and Edwards, grade 1/6) (fig. 7B).

Case 3

P.S. (no. 03-40-87), a 37-month-old girl, was the product of a full-term normal pregnancy. During the first 3 months of life she ate poorly, gained weight slowly, and had episodes of shortness of breath. At 12 weeks of age she had an episode of extreme dyspnea and cyanosis followed by a convulsion. She was hospitalized, found to be in congestive heart failure, and was digitalized. The femoral pulsations were noted to be diminished. At 15 months of age cardiac catheterization was performed at another hospital and a diagnosis of ventricular septal defect and pulmonary hypertension (pulmonary arterial pressure, 62/30 mm. Hg) was made. At 21 months of age, the nailbeds of the toes were noted to be more cyanotic than those of the fingers. Thereafter, until her admission to the Clinical Center 16 months later, she grew poorly, had dyspnea on minimal exertion, and was cyanotic at rest.

On admission she was undernourished and underdeveloped (weight 10 Kg.). The blood pressure was 115/85 in the arms and 100/80 in the legs. There was generalized cyanosis but the nailbeds of the toes were more cyanotic than those of the fingers. There was early clubbing of the distal digits of the toes, but none of the fingertips. Right and left ventricular impulses were palpable, and the second heart sound in the pulmonic area was accentuated. A third heart sound was heard at the lower left sternal border. A grade 2/6
short ejection-type systolic murmur was heard along the left sternal border. The hematocrit value was 53 per cent. The electrocardiogram (fig. 5C) showed right axis deviation and right ventricular hypertrophy. Fluoroscopic and radiographic examinations disclosed generalized cardiomegaly, dilatation of the pulmonary trunk, and increased pulmonary vascularity (fig. 4B).

Cardiac catheterization at this time showed equal pressures (100/0 mm. Hg) in the right and left ventricles, and, by oximetry, a left-to-right shunt was shown to be entering the right ventricle. The left atrium was entered through an interatrial communication. Blood from both the left ventricle and outflow tract of the right ventricle had the same oxygen saturation (84 per cent). Selective angiograms with both left and right ventricular injections were performed (figs. 9 and 10). The ascending aorta was small and terminated after giving rise to the innominate, left carotid, and left subclavian arteries. A ventricular septal defect was demonstrated immediately below the semilunar valves. The right ventricular injection showed marked dilatation of the right ventricle and pulmonary trunk, which continued as the descending aorta. The ventricular septal defect was again demonstrated.

Surgical correction of the aortic arch anomaly was carried out on March 1, 1961. A left lateral thoracotomy was made, and the ascending aorta and its branches were dissected free. The descending aorta originated from the main pulmonary artery through a large ductus. The intercostal arteries were of normal size and, when the ductus was temporarily occluded, the pressure in the descending aorta fell to 20 to 30 mm. Hg, indicating that no effective collateral circulation to the distal aorta was present. A partially occluding clamp was placed on the ascending aorta at the origin of the left subclavian artery, and the end of a Teflon vascular graft was sutured to the aorta at this point (fig. 11D). The distal end of the prosthesis was anastomosed to the side of the descending aorta immediately distal to the ductus. When the continuity of the aorta had been restored, the ductus was divided and closed. Postoperatively, bounding femoral pulses were palpable and the child's color was good. The day following operation atelectasis of the right upper lobe was evident by x-ray, and a tracheostomy tube was

and of an adjacent arteriole is again evident. There is no intimal proliferation. C. The small muscular pulmonary artery is almost totally occluded by fibrocellular intimal proliferation. In the arteriolar branch (lower left) medial hypertrophy is also present.
inserted. Respiratory distress became more severe, however, and the child died 48 hours after operation.

Pathologic Findings

At autopsy, acute purulent pericarditis and left empyema were found. Pure cultures of a coagulase-positive hemolytic Staphylococcus aureus were grown from these areas and also from the heart blood. The child’s death was clearly attributed to infection.

The heart was greatly enlarged (fig. 11); the walls of each ventricle were markedly hypertrophied. A valvular-incompetent foramen ovale was present. The pulmonary valve and pulmonary trunk were greatly dilated and overrode a ventricular septal defect, which was 1.0 cm. in largest diameter. The defect was in the anterior ventricular septum just beneath the pulmonic valve and a portion of the hypertrophied crista supraventricularis. The membranous ventricular septum was intact. The aortic valve was bicuspid and measured only 2.3 cm. in circumference, while the pulmonic valve was 4.5 cm. in circumference. The ascending aorta was hypoplastic; it terminated after dividing into the innominate, left common carotid, and left subclavian arteries. The prosthetic graft between the ascending and descending aorta was patent and, as observed at operation, there was no evidence of collateral circulation to the descending aorta. The coronary arteries were normal. Microscopically, there were severe medial hypertrophy and fibrocellular intimal proliferation of the smaller muscular pulmonary arteries and arterioles (fig. 7C). These pulmonary vascular changes were of grade 3/6 severity (Heath and Edwards).

Discussion

The first pathologic description of complete interruption of the aortic arch was recorded in the late eighteenth century by Raphael Steidele, Professor of Obstetrics of the University of Vienna and a member of the Royal Council for the Emperor of Austria. Since that time there have been reports of 51 other patients with this malformation. A summary of the clinical and pathologic findings
Figure 9
Selective angiocardiogram with left ventricular injection in patient P. S. In the anteroposterior projection (A) the ascending aorta (A.A.) is seen to terminate with the origin of the left subclavian artery. The pulmonary trunk (P.T.) is opacified also. In the lateral view (B) both ventricles are seen as well as the ventricular septal defect through which a large left-to-right shunt occurs.

In these 52 patients and in the three of the present communication is presented in table 1. The sex was mentioned in 49 of these 55 patients: 23 were male and 26 were female. The age at death was noted in all but one. Forty-one (76 per cent) died in the first month of life or were stillborn, and only six lived for longer than 1 year.

In contrast to coarctation of the aorta, in complete interruption of the aortic arch there is no connection of any kind between the ascending and descending aortas. As an isolated anomaly this malformation is incompatible with life, and the descending aorta always originates from the pulmonary artery through a patent ductus arteriosus. A ventricular septal defect is almost always present as well, having been described in 46 of the 48 patients in which the septum was mentioned. In fact, the association of interruption of the aortic arch, patent ductus arteriosus, and ventricular septal defect is so constant that it has been considered a distinct cardiovascular triology by Everts-Suarez and Carson.33 As pointed out by Becu et al.24 and Neufeld et al.,38 the ventricular septal defect in some, and probably in most of the patients with an interrupted aortic arch has an unusual location. These defects seem not to involve the membranous portion of the ventricular septum but lie anterior to it, above the papillary muscle of the conus, but below the pulmonic valve and a portion of the crista supraventricularis. The pulmonary valve is dilated and overrides the defect. The pulmonary trunk is much larger than the ascending aorta.

Another common associated finding is a bicuspid or deformed aortic valve or a narrowed subaortic outflow tract. Becu et al.24 and more recently Neufeld et al.38 have each described two patients with subaortic stenosis and biventricular origin of the pulmonary trunk.
with complete interruption of the aortic arch. The subaortic stenosis was above the ventricular septal defect in each instance. In each of the two patients with two semilunar valves (patients 1 and 3) in the present report, the left ventricular outflow tract above the ventricular septal defect was narrowed by a muscular ridge. In five of the six patients reported by Kleinerman et al., the aortic valve was bicuspid and in the sixth the valve consisted of one normal and two rudimentary cusps. Two of our patients had bicuspid aortic valves and the third had a bicuspid semilunar valve at the origin of the persistent truncus arteriosus. It seems likely, therefore, that abnormalities of the aortic valve and subaortic stenosis are more frequent than indicated in table 1. Atresia of the mitral valve has also been described in three patients with complete interruption of the aortic arch. The frequent association of obstructive lesions in the left side of the heart with complete interruption of the aortic arch or aortic valvar or subvalvar stenosis or atresia, or mitral stenosis or atresia has prompted Levy and more recently Noonan and Nadas to group these defects under a common term, "the hypoplastic left heart syndrome." There may be justification for this terminology, since in all instances the pulmonary trunk and the right ventricle are disproportionately large. A variety of other intracardiac anomalies has been reported in association with interruption of the aortic arch, and these are listed in table 1.

The site of the interruption of the aortic arch is important from both diagnostic and surgical standpoints. The interruption may occur in one of three sites (fig. 2). In the type A malformation, interruption distal to the left subclavian artery, all branches of the aortic arch are supplied by the left ventricle. The type A anomaly was found in 22 of the 50 patients in whom the site of interruption was precisely stated. In the type B anomaly, interruption immediately distal to the left common carotid artery, the innominate and left carotid arteries are supplied by the left ventricle and the left subclavian artery arises from the "pulmonary-ductus-descend-
Figure 11

Drawing of heart and great vessels from patient P. S.  A. The ascending aorta terminates after giving rise to the left subclavian artery (type A interruption). The descending aorta arises entirely from the pulmonary trunk (P.T.) via the patent ductus arteriosus. The right ventricle and the pulmonary trunk are markedly dilated and the ascending aorta is relatively hypoplastic.  B. The right ventricle and pulmonary valve are opened. The ventricular septal defect (V.S.D.) is apparent.  C. The anterior portion of the heart has been removed. The outflow tract of the left ventricle is compromised by the myocardial ridges immediately below the aortic valve cusps. The root of the aorta overrides the right ventricle. The aortic valve is bicuspid.  D. The surgical repair is illustrated. The patent ductus is closed, an the ascending aorta is anastomosed to the descending aorta by a graft.
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ing aorta trunk," a term coined by Evans\(^{17}\) to include the pulmonary trunk, patent ductus arteriosus, and descending thoracic aorta. The type B anomaly occurred in 26 patients. The type C malformation, in which both the left subclavian and left common carotid arteries arise from the pulmonary-ductus-descending aorta trunk, was specifically described in only two patients. Thus in the majority of patients the interruption was distal to the origin of the left common carotid artery. The phrase "absence of the aortic isthmus" has sometimes been applied to these anomalies. Since the isthmus of the aorta is that portion between the left subclavian artery and the ductus arteriosus, this terminology would correctly apply only to the type A interruption and it would seem best to discard it entirely.

In addition to the variation in the site of aortic interruption, there is frequently abnormal origin of the major branches of the ascending aorta and pulmonary trunk. Thus in six patients (table 1) the right subclavian, as well as the left subclavian artery, arose from the pulmonary-ductus-descending aorta trunk (fig. 3A). Bilateral ducti were present in four patients (table 1). In this situation the right subclavian artery arises from the right pulmonary artery via the right ductus (fig. 3B). Kleinerman et al.\(^{29}\) reported a patient (no. 29, table 1) with complete interruption of the arch and bilateral persistence of the patent ducti who also had a right-sided descending aorta (fig. 3D). The "addendum case" (no. 24, table 1) observed by Becu et al.\(^{24}\) also had a right-sided descending aorta but in this instance the descending aorta took origin from the right-sided ductus, which in turn arose from the right pulmonary artery. Also rarely, a pulmonary artery arises directly from the ascending aorta (fig. 3C) as in the patient reported by Jew and Gross.\(^{21}\) Complete transposition of the great vessels has been recorded twice in association with complete interruption of the aortic arch.\(^{18,85}\)

Because of the variation in the site of interruption and in the origin of the great vessels, it is not surprising that the diagnosis of aortic arch interruption has rarely been made on the basis of clinical study. Indeed, this malformation was recognized before operation or autopsy in only two\(^{30,32}\) of the 52 previously reported patients. In two of the three patients in the present communication, however, the correct diagnosis was established by catheterization and selective angiocardiography.

The symptoms of interruption of the aortic arch are nonspecific and the physical signs are of only limited value. It would seem that differential cyanosis would always be present, but it is apparently the exception rather than the rule. In only 10 of the 38 patients in which the presence or absence of cyanosis was recorded, was differential cyanosis apparent. In seven of these patients cyanosis was evident in the nailbeds of the toes and in the lips. In two patients (nos. 17 and 46, table 1), each of whom also had complete transposition of the great vessels, there was reverse cyanosis, i.e., the nailbeds of the fingers and the lips were cyanotic and the nailbeds of the toes were not. In the tenth patient (no. 26, table 1) there were cyanosis and clubbing of the fingers of the left hand and the toes of both feet, but no cyanosis or clubbing of the fingers of the right hand. This patient, living at the age of 5 years and reported by Dorney et al.,\(^{26}\) had a type B interruption of the aortic arch. In 10 other patients with various forms of interruption of the aortic arch no cyanosis was evident, and in the remaining 18 it was present but presumably generalized.

The frequent absence of differential cyanosis can, of course, be explained by the presence of a ventricular septal defect, which is invariably present and of large size. Arterialized blood from the left ventricle is shunted through the defect and enters the pulmonary trunk and the lower extremities. As a result, the oxygen saturation of blood in the pulmonary artery, ductus, and descending aorta approaches that in the ascending aorta. Greig,\(^{6}\) in 1852, clearly appreciated that the left-to-right intracardiac shunt was responsible for the infrequency of differential cyanosis in these patients. In his paper of over 100 years ago he wrote: "The additional

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Table 1

Clinical and Pathologic Data from 55 Patients with Complete Interruption of the Aortic Arch Proved at Autopsy or Operation

<table>
<thead>
<tr>
<th>Patient number</th>
<th>Author year</th>
<th>Sex</th>
<th>Age at death</th>
<th>Heart murmur</th>
<th>Cyanosis D= differential</th>
<th>Location of interruption</th>
<th>Ventricular septal defect</th>
<th>Patent foramen ovale</th>
<th>Associated cardiovascular defects</th>
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<td>Steidele</td>
<td>M</td>
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<tr>
<td>2</td>
<td>Seidel</td>
<td>M</td>
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<td>B</td>
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<td>B</td>
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<td>A</td>
<td>0</td>
<td>+</td>
<td></td>
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<td>A or B</td>
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<td>+</td>
<td></td>
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<td>M</td>
<td>10 months</td>
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<td>A</td>
<td>+</td>
<td>+</td>
<td>+</td>
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<td>Kohl</td>
<td>F</td>
<td>&quot;hours&quot;</td>
<td>+</td>
<td>A</td>
<td>+</td>
<td>+</td>
<td>(V.I.)</td>
<td>Anerysmal dilatation of asc. aorta</td>
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<td>9</td>
<td>Dick</td>
<td>M</td>
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<td>+</td>
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<td>Anomalous drainage of left jugular, axillary, and azygos vein into R.A.</td>
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<td>30 days</td>
<td>+</td>
<td>+(D)*</td>
<td>A</td>
<td>+</td>
<td>+</td>
<td>Complete transposition of great vessels</td>
</tr>
</tbody>
</table>

* (D) = Ductus arteriosus.
<table>
<thead>
<tr>
<th>Case</th>
<th>Author(s)</th>
<th>Gender</th>
<th>Days</th>
<th>Status</th>
<th>Aorta</th>
<th>Pulmonary Trunk</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>19</td>
<td>Stewart</td>
<td>F</td>
<td>2 days</td>
<td>0</td>
<td>+</td>
<td>A</td>
<td>Origin of right P.A. from asc. aorta</td>
</tr>
<tr>
<td>20</td>
<td>Jew &amp; Gross</td>
<td>M</td>
<td>3 days</td>
<td>+</td>
<td>+</td>
<td>B</td>
<td>Mitral atresia, persistent left S.V.C.</td>
</tr>
<tr>
<td>24</td>
<td>Beene et al.</td>
<td>F</td>
<td>14 days</td>
<td>B</td>
<td>+</td>
<td>+</td>
<td>Aortic stenosis</td>
</tr>
<tr>
<td>25</td>
<td>Jew &amp; Gross</td>
<td>F</td>
<td>3 days</td>
<td>0</td>
<td>+</td>
<td>B</td>
<td>Bicuspid aortic valve</td>
</tr>
<tr>
<td>26</td>
<td>Dorney et al.</td>
<td>F</td>
<td>5 years†</td>
<td>+ +</td>
<td>(D)§</td>
<td>B</td>
<td>Origin of right P.A. via right P.D.A.</td>
</tr>
<tr>
<td>27</td>
<td>Kravtin et al.</td>
<td>M</td>
<td>11 days</td>
<td>+</td>
<td>+ (D)</td>
<td>A</td>
<td>Bicuspid aortic valve</td>
</tr>
<tr>
<td>28</td>
<td>Merrill et al.</td>
<td>F</td>
<td>3½ years†</td>
<td>+ +</td>
<td>0</td>
<td>A</td>
<td>Mitral atresia. Bicuspid aortic valve</td>
</tr>
<tr>
<td>29</td>
<td>Kleinerman et al.</td>
<td>M</td>
<td>8 days</td>
<td>0</td>
<td>0</td>
<td>B</td>
<td>Cor biloculare. Bicuspid aortic valve</td>
</tr>
<tr>
<td>30</td>
<td>Kleinerman et al.</td>
<td>M</td>
<td>3 days</td>
<td>+</td>
<td>+ (D)</td>
<td>B</td>
<td>Origin of both R.S.A. and L.S.A. from ‘ductus-des. aorta trunk.’ Hypoplastic aortic valve. Quadriceps pulmonary valve</td>
</tr>
</tbody>
</table>
Table 1—Continued

Clinical and Pathologic Data from 55 Patients with Complete Interruption of the Aortic Arch Proved at Autopsy or Operation

<table>
<thead>
<tr>
<th>Patient number</th>
<th>Author year</th>
<th>Sex</th>
<th>Age at death</th>
<th>Heart murmur</th>
<th>Cyanosis</th>
<th>Location of interruption</th>
<th>Ventricular septal defect</th>
<th>Patent foramen ovale</th>
<th>Associated cardiovascular defects</th>
</tr>
</thead>
<tbody>
<tr>
<td>34</td>
<td>Kleinerman et al. (1958)</td>
<td>M</td>
<td>21½ hours</td>
<td>0</td>
<td>0</td>
<td>A</td>
<td>+</td>
<td>+ (V.I.)</td>
<td>Bicuspid aortic valve. Single coronary artery</td>
</tr>
<tr>
<td>35</td>
<td>Abrams (1958)</td>
<td>F</td>
<td>10 days</td>
<td>+</td>
<td>+</td>
<td>A</td>
<td>+</td>
<td>+</td>
<td></td>
</tr>
<tr>
<td>36</td>
<td>Noonan &amp; Nadas (1958)</td>
<td>F=5</td>
<td>Five lived for less than 20 days. One lived 3½ months</td>
<td>3</td>
<td>+ (D)=1</td>
<td>A=2</td>
<td>B or C=4</td>
<td>5</td>
<td>Endocardial fibroelastosis=1</td>
</tr>
<tr>
<td>37</td>
<td>Noonan &amp; Nadas (1958)</td>
<td>M=1</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>38</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>39</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>40</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>41</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>42</td>
<td>Foley (1958)</td>
<td>M</td>
<td>6 days</td>
<td>0</td>
<td>0</td>
<td>A</td>
<td>+</td>
<td>+</td>
<td>&quot;Congenital cyst,&quot; tricuspid valve</td>
</tr>
<tr>
<td>43</td>
<td>Evarts-Suárez &amp; Carson (1959)</td>
<td>M</td>
<td>3 days</td>
<td>0</td>
<td>0</td>
<td>B</td>
<td>+</td>
<td>+</td>
<td></td>
</tr>
<tr>
<td>45</td>
<td>Quiie et al. (1959)</td>
<td>M</td>
<td>5½ months!!</td>
<td>+</td>
<td>0</td>
<td>B</td>
<td>+</td>
<td>+</td>
<td>Bicuspid aortic valve</td>
</tr>
<tr>
<td>46</td>
<td>Castellanos et al. (1959)!!</td>
<td>M</td>
<td>9 years</td>
<td>0</td>
<td>+ (D)</td>
<td>A</td>
<td>+†</td>
<td>+ (V.I.)</td>
<td>Complete transposition of the great vessels. Persistent left S.V.C. draining into coronary sinus</td>
</tr>
<tr>
<td>47</td>
<td>Fournier &amp; Zaidi (1960)</td>
<td>F</td>
<td>2 months</td>
<td>+</td>
<td>0</td>
<td>A</td>
<td>+</td>
<td>0</td>
<td>&quot;Coarcted&quot; P.D.A., Bicuspid aortic valve</td>
</tr>
<tr>
<td>48</td>
<td>Tabakin &amp; Hanson (1960)</td>
<td>F</td>
<td>3 months</td>
<td>+</td>
<td>0</td>
<td>B</td>
<td>+</td>
<td>+</td>
<td>Persistent left S.V.C. draining directly into R.A. Origin of both R.S.A. and L.S.A. from &quot;ductus-des. aorta trunk&quot;</td>
</tr>
<tr>
<td>50</td>
<td>Neufeld et al. (1961)** (Case 3)</td>
<td>F</td>
<td>3 days</td>
<td>+†</td>
<td>+ (D)</td>
<td>B</td>
<td>+</td>
<td>+</td>
<td>Subaortic stenosis. Biventricular origin of pulmonary trunk. Origin of both R.S.A. and L.S.A. from &quot;ductus-des. aorta trunk&quot;</td>
</tr>
<tr>
<td>51</td>
<td>Freedman (1961)</td>
<td>F</td>
<td>2 months</td>
<td>+</td>
<td>+</td>
<td>B</td>
<td>+</td>
<td>+</td>
<td>Probable subaortic or aortic valvar stenosis</td>
</tr>
</tbody>
</table>
INTERUPTION OF AORTIC ARCH

54 Present Authors
R.S.††
M 18 days ++ (+D) A ++

55 Present Authors
P.S.††
F 3 years|| ++ (+D) A ++ (V.I.)

*BReverse cyanosis.
†Still living—diagnosis made at thoracotomy.
‡Diastolic murmur in addition to systolic murmur.
§Cyanosis of left hand, both feet; no cyanosis of right hand.
||Died in early postoperative period following ligation of P.D.A. and anastomotic procedure.
**Cases 49 and 50 are siblings.
††Functionally insignificant V.S.D., but anatomically open.
|||Diagnoses made ante mortem (selective angiography).

Abbreviations: V.I.=Valvular incompetent foramen ovale; R.S.A.=Right subclavian artery; L.S.A.=Left subclavian artery; P.A.=Pulmonary artery; P.D.A.=Patent ductus arteriosus; R.A.=Right atrium; L.C.C.A.=Left common carotid artery; S.V.C.=Superior vena cava.

complication, however, of such a large aper-

of the vessels, all parts of the body would be

the two sides of the heart, before it was sent

into the aorta and pulmonary trunk, the out-

blood from the right ventricle to the left, and

right-to-left shunt also was demonstrated in

This child, however, was 3 years old,

in the other two. In summary, an

oxygen content found in the pulmonary trunk has

an infrequent finding in younger children.

Although blood in the pulmonary trunk has

a higher oxygen content than systemic

blood, it is unusual, contrary to the findings

in the ascending aorta. In all four of the

previously reported patients with interrup-

tions of the aortic arch, although the blood in

the ascending aorta was sampled, the oxygen

content was higher than systemic.

Another sign that might be logically ex-

pected in these patients is absence or marked

diminution of the femoral pulses. This

was evident in the other two. In summary,

Differential cyanosis was clinically apparent.

Patient R. S., in the arms, had absent femoral

arterial blood was sampled, distinct from

femoral arterial blood in whom blood from the right ventricular

outflow tract had an oxygen content identical to that sampled from the left ventricle. The

right-to-left shunt also was demonstrated in

right-to-left shunt also was demonstrated in

right-to-left shunt also was demonstrated in

right-to-left shunt also was demonstrated in

right-to-left shunt also was demonstrated in

right-to-left shunt also was demonstrated in
ing aorta. In patient 46, table 1, closure of the patent ductus resulting in disappearance of the femoral pulses, was documented by catheterizations and at autopsy. In patient 52, table 1, there was hypertension (220/120 mm. Hg) in the arms, but the blood pressure in the legs was normal. This patient, reported by Ruiz Villalobos et al., survived operation at the age of 14 years and was doing well 7 months later. Differences in the force of the radial and carotid pulsations, might likewise be expected in type B and C malformations, but this finding has been recorded in only one patient.\textsuperscript{31}

Heart murmurs are probably present in all patients with complete interruption of the aortic arch who live for longer than 1 month. A murmur was present in 22 and absent in 15 of the 37 patients in which this finding was mentioned. All 15 patients who had no murmur died within a few days. The murmur, when present, was systolic in all instances; in addition, however, five patients had diastolic murmurs. The ventricular septal defect is probably the source of the systolic murmur, although the aortic valvular or subvalvular stenosis, if present, would contribute. One would not expect a murmur to result from blood flowing through the ductus, since no pressure differential exists between the pulmonary trunk and the descending aorta. A diastolic murmur can be best attributed to pulmonic regurgitation, secondary to pulmonic hypertension. One patient (no. 28, table 1) had a continuous murmur in the left infraclavicular region.\textsuperscript{28} Collateral vessels were present in this patient between the ascending and descending aortas. Only one other patient (no. 52, table 1), was described with a definite collateral circulation.

The electrocardiogram in these patients usually shows right axis deviation and right ventricular hypertrophy. Biventricular hypertrophy has been noted occasionally. Right ventricular strain and incomplete right bundle-branch block also have been recorded.

Cardiac catheterization was performed in five patients with interruption of the aortic arch previously reported, and in each of the three described herein. All eight had pulmonary arterial and right ventricular hypertension, the peak pulmonary arterial systolic pressure ranging from 50 to 132 mm. Hg and, in general, equal to peripheral arterial systolic pressure. In each, a left-to-right or bidirectional shunt at the ventricular level was found. It is apparent that these catheterization data are not diagnostic, and selective angiocardiography affords the only means by which the definitive diagnosis of complete interruption of the aortic arch can be made. Abrams,\textsuperscript{30} using this method, was the first to diagnose the malformation before operation or autopsy. Selective angiocardiography proved to be the definitive study in Castellanos' patient\textsuperscript{25} and in two of the present ones. In patient P. S. (case 3) both right and left ventricular injections were performed (fig. 9) and the nature of the lesion was apparent in both series of films. By selective angiography, one cannot distinguish complete interruption of the aorta from atresia of the aorta in which there is an anatomic connection by a fibrous band between the ascending and descending aortas. This distinction is of little practical consequence, however, since the method of surgical correction for each is similar.

Complete interruption of the arch of the aorta is potentially amenable to surgical correction. The principal limiting factor, to date, appears to be the early recognition of the lesion, since a majority of the patients die before the age of 1 month. In only two patients has a successful repair been reported. Merrill et al.\textsuperscript{28} corrected the anomaly of the aortic arch in a patient with a type A interruption. He divided the patent ductus and anastomosed the descending aorta directly to the base of the left subclavian artery in an end-to-side fashion. Recently, Ruiz Villalobos et al.\textsuperscript{40} reported the successful closure of the ductus, with anastomosis of the descending aorta to the left subclavian artery by means of a prosthesis, in a 14-year-old boy with a type A interruption. As noted previously, each of these patients was unique in that extensive collateral circulation was present and

\textit{Circulation, Volume XXVI, July 1962}
the descending aorta could be occluded during the repair. Quie et al.\textsuperscript{34} reported correction of a type B interruption, but the patient died in the early postoperative period. The ductus, the left subclavian, and the left common carotid arteries were divided, and the left common carotid artery was anastomosed to the descending aorta. In patient P. S. a procedure similar to that employed by Ruiz Villalobos was performed. The absence of collateral vessels, however, necessitated restoration of the continuity of the aorta, by means of a prosthetic graft, before the ductus could be divided. Blake, Manion, and Spencer\textsuperscript{42} have reported a successful operation of this type in a child with atresia of the aortic isthmus.

The correction of the rare type C interruption, which has apparently not been attempted, probably would involve division of both the left subclavian and left common carotid arteries and an anastomosis of the descending aorta to the base of the right common carotid artery. It would, of course, be necessary to provide distal perfusion to the left carotid until flow to it from the aorta could be restored.

Restoration of continuity of the aorta is obviously only a first step in the treatment of a patient with interruption of the aortic arch, since a ventricular septal defect remains. All these patients have pulmonary hypertension, and the state of the pulmonary vascular bed, as determined at subsequent study, would indicate whether closure of the defect or palliative constriction of the pulmonary artery would be desirable.

**Summary**

The clinical, hemodynamic, angiographic, and pathologic findings in three patients with complete interruption of the aortic arch are presented. In addition, data derived from published reports of 52 previous patients are summarized. This malformation is never an isolated one, since a patent ductus arteriosus is always present and a ventricular septal defect is almost invariably found as well. The diagnosis of absence of the aortic arch is difficult, since differential cyanosis is uncommon, relative systolic hypertension of the arms is rare, and the heart murmur, when present, is of a nonspecific nature. At catheterization the pressure in the pulmonary artery, patent ductus arteriosus, and descending aorta is usually similar to that in the ascending aorta and in its branches. Selective angiocardiography has been shown to be the only means of establishing the presence of the malformation prior to operation or autopsy. Since the majority of patients with absence of the aortic arch die within the month following birth, early recognition of the lesion is imperative if surgical correction is to be of benefit.

**Addendum**

Since this paper was prepared, two additional patients with complete interruption of the aortic arch have been called to our attention. One, a stillborn, reported by Sir William Osler in 1880 (case 4)\textsuperscript{16} had a type B interruption and associated ventricular and atrial septal defects, bicuspid aortic and malformed pulmonic valves. Another patient, a 3-year-old girl, reported by Espino-Vela et al.,\textsuperscript{41} also had a type B interruption and an associated ventricular septal defect. Shortly after this manuscript was submitted for publication, Newcombe et al.\textsuperscript{42} reviewed the surgical and pathologic experiences of the Mayo Clinic in patients with "coarctation" of the aorta, ventricular septal defect, and usually patent ductus arteriosus. The authors used the term "coarctation" in a general sense to include aortic arch interruption as well as aortic narrowing or atresia. The authors mentioned seven examples of aortic arch "coarctation" associated with a distal patent ductus arteriosus, subaortic stenosis, bicuspid aortic valve, and ventricular septal defect. The "coarctation" in at least one of these seven was of the complete interruption form and appears to have already been reported (case 22, tabe 1). In addition, they mentioned a patient with a type B aortic arch interruption who had a persistent truncus arteriosus.

**References**

3. Heath, D., and Edwards, J. E.: The pathology of hypertensive pulmonary vascular disease,


34. Quei, P. G., Novick, R., Adams, P., Jr., Anderson, B. C., and Varco, B. L.: Congenital

Harvey's monumental discovery was given to the world in 1628 but was very slow in winning adherents, particularly in England. Sydenham himself does not once refer to Harvey, although their lives overlapped, and they lived in the same city for a number of years. This attitude of Sydenham is at first sight difficult to understand, but certain facts help us to solve the riddle. Sydenham had no patience with theories and saw little in the speculations founded on the new physiology but carried to an excess not contemplated by Harvey.—DAVID RIESMAN, M.D., Thomas Sydenham, Clinician. New York, Paul B. Hoeber, Inc., 1926, p. 16.
Complete Interruption of the Aortic Arch
WILLIAM C. ROBERTS, ANDREW G. MORROW and EUGENE BRAUNWALD

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