CRITICAL REVIEW

 Interruption of the Aortic Arch: Clinical Features in 20 Patients*

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The clinical and laboratory findings of interruption of the aortic arch are des-
dcribed. These infants present with a clinical picture similar to preductal coac-
tation of the aorta. Only by angiography can the diagnosis be established.
Prompt recognition and diagnosis of interruption of the aortic arch is impor-
tant, since palliative and corrective operative procedures are currently available.
Without operation, death usually occurs in infancy.

Interruption of the aortic arch is an uncommon congenital cardiac malformation which usually leads to death in infancy. In this condition, continuity between the ascending and descending portions of the aorta is not present, and the descending aorta is supplied through a reversing patent ductus arteriosus. In the majority of patients a ventricular septal defect is present and on occasion more complicated cardiac malformations.1,2 With the development of improved cardiac operative tech-
niques for infants, this malformation now has the potential for correction. Aside from case reports and several reviews, most papers concerning interruption of the aortic arch have dealt primarily with pathologic features.

We present the clinical and laboratory features of 20 personally observed patients with interruption of the aortic arch and formulate a composite clinical picture based upon this experience and the available data in the literature.3-40

OBSERVATIONS

Twenty patients with interruption of the aortic arch have been studied at the University of Minnesota Hospitals be-
tween 1952 and 1970. Among the 20 patients, there were 16 boys and four girls. Eighteen of the patients have died; 13 were dead in the first month of life, and the others at two months (two patients), three months, four months and six months respectively. Two patients are alive at ages three months and seven years.

Necropsy studies were performed in the 18 patients who died and in each a ventricular septal defect and patent ductus arteriosus were coexistent with the interruption of the aortic arch. In four of these patients, significant additional cardiac malformations were present: complete transposition of the great vessels (two patients), aortic pulmonic window (one patient), and persistent truncus arteriosus (one patient).

The entire 20 patients will be considered as a group since the presence of the coexistent malformations did not appear to influence the clinical picture.

Clinical Features

Sixteen patients manifested cardiovascular symptoms or signs in the first week of life, and three others by the end of the first month. The remaining patient was first recognized to have cardiac disease when a cardiac murmur was heard at age 2½ months.

The dominant presenting symptoms and signs were cyanosis, cardiac murmur, and respiratory distress. Eighteen patients manifested signs of congestive cardiac failure and were digitalized, ten within the first week of life, seven others before two months, and the remaining patient at eight months. Seventeen gave a history of slow growth and 15 a history of poor feeding.

There was no prenatal history of maternal viral infection and no history of congenital heart disease in the 13 siblings or in the parents. Simultaneous flush blood pressure readings were obtained in 17 patients. In 11, the values in the arm and leg were identical, but in six the pressure in the leg was 40 to 55 mm
Hg lower than that obtained in the arm. Thirteen of the patients were cyanotic, but in none was differential cyanosis evident. The cyanosis was often mild and difficult to distinguish from that present in infants with congestive cardiac failure. In only eight of the patients was a murmur present. These varied from grade 1-3/6 in loudness, were systolic in timing and were located along the left sternal border. The pulmonic component of the second heart sound was accentuated. No patient showed an ejection click or a diastolic murmur. Four patients exhibited peripheral edema and three were jaundiced.

Electrocardiographic Findings

Electrocardiograms were available for review in 18 patients. The QRS axis ranged between +55 and +240 degrees, with seven being to the right of +150 degrees. Right atrial enlargement was present in six; the others showed no atrial enlargement. Seventeen exhibited patterns of right ventricular hypertrophy, eight of whom showed additional left ventricular hypertrophy. ST segment changes of digitalis were commonly observed.

Roentgenographic Features

No specific cardiac contour was evident. Cardiomegaly of moderate to marked degree was present in each patient. The pulmonary trunk and right-sided cardiac chambers were enlarged and the pulmonary arterial markings were accentuated.

**Figure 1.** Selective left atrial injection of opaque material in a patient with interruption of the aortic arch between left carotid and left subclavian arteries. Simultaneous opacification of pulmonary trunk through ventricular septal defect and ascending aorta. Ascending aorta divides into only two branches, the left and right common carotid arteries.

**Figure 2.** Retrograde aortogram in a patient with interruption of the aortic arch beyond the left subclavian artery opacification of ascending aorta and its major branches. No opaque material passes into the descending aorta. Enlarged internal mammary and intercostal arteries.

**Figure 3.** Retrograde aortogram in a patient with interruption of the aortic arch between the left carotid and left subclavian arteries. Right subclavian artery arises from the descending aorta.

Angiographic Appearance

Angiocardiography played an important role in identifying the location of the interruption and the presence of associated anomalies. Injection of contrast into right-sided cardiac chambers showed a reversing ductus arteriosus with opacification of the descending aorta. Opacification of the ascending aorta showed the site of the interruption. This occurred between the left carotid and left subclavian arteries in four of the ten patients studied (Fig 1), and beyond the left subclavian artery in the other six (Fig 2). In two of the first group both subclavian arteries arose from the descending aorta (Fig 3). Each of the patients showed enlarged collateral vessels.
Clinical Course

Operative correction was attempted in seven patients. Five of these died in the operating room either prior to thoracotomy or during the operative procedure. Two patients survived an operation to anastomose the descending and ascending aorta but died in the early postoperative procedure. Ten patients, despite medical management, showed progressive respiratory difficulty and signs of cardiac failure and died. Two patients survived. One is a three-month-old child and the other a seven-year-old boy with coexistent A-P window. Both the survivors are being maintained on digoxin.

Necropsy Findings

Necropsy was performed in 18 patients. The interruption was beyond the left subclavian artery in seven instances and in each of these, all the great vessels arose from the ascending aorta. In the other 11, the interruption was between the left common carotid and left subclavian arteries. Four of these 11 also had anomalous origin of the right subclavian artery from the descending aorta.

A ventricular septal defect and patent ductus arteriosus were present in each patient. The ductus arteriosus was of large caliber in all cases, but was thrombosed in one. Muscular subaortic stenosis above the ventricular septal defect was found in six cases, and bicuspid aortic valves in seven. Complete transposition was present in two, persistent truncus arteriosus in one and aortopulmonary defect in one.

DISCUSSION

Interruption of the aortic arch is an uncommon congenital cardiac malformation which pathologically and clinically resembles preductal coarctation of the aorta. There is a lack of continuity between the ascending and descending portions of the aorta; the latter is continuous with the patent ductus arteriosus and through it receives blood from the right ventricle. Usually no tissue is present between the two aortic segments but in a few patients a fibrous strand connects the proximal and distal portions of the aorta. The latter condition has been termed atresia of the aortic arch.

Interruption of the aortic arch has been classified according to the site of the interruption. Three patterns have been identified: 1) interruption beyond the left subclavian artery (Fig 4), 2) interruption between the left carotid and left subclavian arteries (Fig 5), and 3) interruption between the innominate and left carotid arteries (Fig 6). In both of the first two varieties the right subclavian artery may arise from one of the following sites: a) the innominate artery (Fig 5a), b) the descending aorta (Fig 5b), or c) the right pulmonary artery through a patent ductus arteriosus (Fig 5c).

Usually the interruption of the aortic arch is of one of the first two types. Only four patients have been described with the third type, and it is difficult to explain this type on a developmental basis.

In almost all patients coexistent cardiac malformations are present so that interruption of the aortic arch should be considered as one part of a triad, the other components being ventricular septal defect and patent ductus arteriosus. A unique variation of this triad was described by Becu and associates5 in which subaortic stenosis and biventricular origin of the pulmonary trunk were present. Interruption of the aortic arch has also been described with complete transposition of the great vessels7,9,16,22 persistent truncus arteriosus23,25,29,33,41 and several other conditions.
Although at least 170 cases of interruption of the aortic arch have been reported in the literature, the clinical picture has not been clearly defined. In an attempt to develop a clinical profile of interruption of the aortic arch we have combined the clinical and laboratory data of our 20 patients with that available from 46 published cases.

In almost all patients the clinical, electrocardiographic, and roentgenographic findings are similar regardless of the type of associated malformation. The exception is the type of differential cyanosis in patients with transposition of the great vessels.

The patients present a history of congestive cardiac failure, respiratory distress, mild cyanosis and early death. At the time of reporting only 12 of the 66 patients were living and of the 12 only four were older than five years. Among the 54 deaths, 41 occurred in the first month of life, 11 more during the next 11 months of age and the remaining two died at three and nine years respectively. Cardiac symptoms were initially recognized during the first week of life in 47 of the 66 patients, and cardiac failure developed in most patients usually by one month of age.

This clinical history is not diagnostic for interruption of the aortic arch, but tends to exclude conditions causing major cardiac symptoms in the neonatal period such as aortic atresia and complete transposition of the great vessels. Coarctation of the aorta frequently causes congestive failure, mild cyanosis and respiratory distress in the first month of life, and in this respect is mimicked by interruption of the aortic arch.

Because of the anatomic features of interruption
of the aortic arch two findings would be anticipated on physical examination. One is differential cyanosis. Since the blood flowing into the ascending aorta and thus to the head and arms is derived from the lungs and the blood flowing into the descending aorta is from the right side of the heart through the ductus, the head and arms should appear pink, while the lower part of the body should be cyanotic. But of the 45 patients with evident cyanosis, a difference in the degree of cyanosis was noted in only 14 patients. Among the 14, cyanosis was greatest in the lower part of the body of one patient, and greatest in the upper part of the body in four patients with complete transposition of the great vessels.

The lack of differential cyanosis in so many patients with interruption of the aortic arch may be caused by several factors. The presence of the ventricular septal defect leads to a large left-to-right shunt at the ventricular level. Thus the blood in the right ventricle has a higher saturation than normal, tending to minimize the difference in oxygen saturation between the blood delivered to the ascending and to descending portions of the aorta. Secondly, the presence of collateral vessels between the ascending and the descending portions of the aorta permit flow from the former to the latter raising the oxygen saturation in the descending aorta. Thirdly, the presence of either pulmonary edema from congestive cardiac failure or pneumonia interferes with oxygenation causing desaturation of blood distributed to the ascending aorta.

Equal blood pressures in arms and legs would also be expected because the large ventricular communication and the patent ductus arteriosus should permit equalization of systolic pressures in the ventricles and the aortic compartments. In 17 cases, however, either higher blood pressure or stronger pulses were found in the arms than the legs. The presence of differential blood pressures indicates that the ductus must be narrowed and is acting as a "coarctation". Another possibility is thrombosis of the ductus arteriosus as occurred in one of our patients. Supportive evidence for the difference in blood pressures is found on aortograms which reveal extensive collateral vessels.

Other findings on physical examination included a nonspecific systolic murmur in 43 of the 66 patients. Usually located along the left sternal border, the murmur was soft or only moderately loud. Apical diastolic murmurs were infrequently heard. Because of pulmonary hypertension, the pulmonary component of the second heart sound was regularly accentuated. Hepatomegaly and growth failure, features of congestive cardiac failure, were common.

Electrocardiographic data were available in 47 patients. In 18 right axis deviation for age was observed, while in the remaining patients it was normal. A variety of patterns of atrial enlargement were found: right in seven, biatrial in six and left in two. Since the right ventricle is supplying the descending aorta the right ventricle is hypertrophied. In almost all patients this is manifested electrocardiographically and may be associated with left ventricular hypertrophy, especially in older patients. Left ventricular hypertrophy generally indicates excessive pulmonary blood flow but may also indicate coexistent subaortic stenosis.

Thoracic roentgenograms reveal no distinctive cardiac contour. Generalized enlargement related to right atrial, right ventricular, and left atrial enlargement is observed. The pulmonary trunk is prominent and the pulmonary vasculature markedly increased.

The clinical picture of interruption of the aortic arch thus is characterized by mild generalized cyanosis, respiratory distress, and congestive cardiac failure occurring in the first month of life. The roentgenographic features are cardiomegaly and increase in pulmonary vascular markings and the electrocardiogram reveals right axis deviation and right or combined ventricular hypertrophy. In addition the blood pressure in the legs may be lower than in the arms and differential cyanosis may be present. Except for this last feature, the clinical picture thus tends to resemble that in infants with preductal coarctation of the aorta.

The diagnosis of interruption of the aortic arch can be made only by angiographic studies. These serve to diagnose and localize the site of the interruption and to define coexistent cardiac malformations. Following injection of contrast material into either the right ventricle or pulmonary trunk the descending aorta opacifies indicating a reversing ductus arteriosus. The ascending aorta must also be carefully visualized either on delayed films of a right-sided injection or preferably by ascending aortography, since a severe coarctation of the aorta and aortic atresia may also be associated with a reversing ductus arteriosus. In review of the angiograms, care must be taken to identify the origin of the brachiocephalic vessels, particularly the subclavian arteries. The left subclavian artery may arise either from the proximal or distal aortic segments. The right subclavian artery may arise from one of three sites, the innominate artery, descending aorta with a retroesophageal course or from the right pulmonary artery through a rightsided ductus arteriosus.
As indicated by the clinical and laboratory findings two major physiologic problems exist in these patients: 1) excessive pulmonary blood flow and 2) obstruction to blood flow to the lower extremities. The first is evident by the cardiomegaly present in thoracic roentgenograms and the other by the difference in blood pressure between the arms and legs. Operative procedures should be directed towards relief of both abnormalities but this may be difficult to accomplish in critically ill infants. The operative problems presented by these patients are similar to infants with preductal coarticulation of the aorta. In such patients the operative approach is to remove the aortic block to establish an adequately sized aortic lumen. Similar approaches to interruption are hindered because the proximal and distal aortic segments are separated by a distance, which cannot be bridged by the existing tissue.

Several operative approaches have been suggested, one of which is banding the pulmonary arteries to relieve the excessive pulmonary blood flow. Van Praagh and co-workers47 have recommended a combined approach of banding the individual pulmonary arteries and placement of a Teflon graft between the pulmonary trunk and descending aorta, thus bypassing a narrowed ductus arteriosus. The approach we currently favor is the establishment of continuity between the ascending and descending aorta by a graft, division of the ductus arteriosus and banding of the pulmonary trunk. This requires the use of cardiopulmonary bypass, which is now possible in infants. The operative mortality for each of the procedures mentioned is very high, perhaps because the infants are critically ill. At the time of operation we favor any approach which would establish aortic continuity and protect the lungs.

While the presence of coexistent conditions may complicate operative approach, Gomes and McGoorn48 recently described the successful operative correction of a truncus arteriosus and interruption of the aortic arch through use of an aortic homograft.

References

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The Beginning of Opera

The history of opera as an important art form began in 1607 with the production of L'Orfeo, by Claudio Monteverdi, at the court of Mantua. Monteverdi was already famous as one of the finest creators of madrigals in the late Renaissance manner. He seemed to grasp the new style intuitively, but he also had the superb dramatic instinct to see the limitations of a whole evening of thinly accompanied recitative. The greatness of L'Orfeo lies partly in its canny synthesis of old and new practices. There are madrigalesque choruses interspersed in the action which provide both dramatic and musical contrast. There are moments of passion in which the constantly flexible recitative is subtly altered to a more shapely and regular melody of great poignance. In L'Orfeo there is a duality in the way the story is told: the free, speech-derived recitative sung by the characters as they act out the story itself, and the lyrical aria in which the action stops and the characters sing of their state of mind, and in which purely musical considerations dominate.