Atresia of the Aortic Arch

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A TRESIA of the aortic arch is a rare anomaly. Evans reported three cases of aortic arch interruption in 19,217 autopsies at the London Hospital. Roberts recently described three patients evaluated at the National Institutes of Health and previously reported patients. Blake reported an additional 18 cases registered in the Armed Forces Institute of Pathology. In all the reported series, aortic arch atresia was never seen as an isolated cardiovascular anomaly, was never distal to a ligamentum arteriosum, and was always associated with a patent ductus arteriosus. Only seven of the 75 cases in Roberts' and Blake's combined series lived longer than 1 year, and the oldest reported case was 14 years old.

This paper presents a 16-year-old girl with interruption of the aortic arch between the innominate and left common carotid arteries, without associated intracardiac anomalies, and without a patent ductus arteriosus. Successful surgical restoration of the continuity between the ascending and descending aorta was accomplished with the use of a prosthetic graft.

Case Report

L. M. (no. 11-60-94) is a 16-year-old white girl, the product of a normal pregnancy and delivery. No heart murmur was heard at birth. The patient's infancy was unremarkable, and she grew and developed normally. At age 6 a vigorous pulsation was noted in the suprasternal notch and a

Figure 1

Chest x-ray showing the normal cardiac silhouette and rib notching on the right side.
heart murmur was heard at that time, but no diagnosis was made. At age 14 a clinical diagnosis of coarctation of the aorta was made.

She has always noted fatigue and shortness of breath with exercise that prevented her from participating in strenuous physical activities. In the past few months she has noted increasing soreness in her calves and tingling in her toes after walking more than one block or after climbing one flight of stairs. The patient has had frequent epistaxes that have been stopped easily with pressure. There was no history of paroxysmal nocturnal dyspnea, hemoptysis, severe headaches, diplopia, edema, syncope, cyanosis, or palpitations.

Physical examination revealed a well-developed, well-nourished girl in no discomfort. The blood pressure in the right arm was 148/100, and no blood pressure could be obtained in the left arm or in either leg. The heart rate was 90 per minute and regular. The skin did not show cyanosis. There was a massively dilated, tortuous, and pulsatile vessel palpable in the suprasternal notch and in the right side of the neck.

A faint pulse was felt in the left common carotid artery. Faint arterial pulses were palpable in both groins and over the right scapular region. No arterial pulses were palpable in the left arm or in the feet. The lungs were clear to percussion and auscultation. The heart was not enlarged. No thrills or heaves were present over the precordium. There was a grade-IV/VI systolic murmur heard loudest over the suprasternal notch and right side of the neck, which diminished in intensity down the left sternal border and could barely be heard at the apex. A systolic bruit was heard over the right scapular region and in the right axilla. The second sound at the pulmonic area was louder than at the aortic area and was normally split. No diastolic murmurs were heard. No bruits were heard over the abdomen. The left hand and arm were slightly smaller than the right. There was no cyanosis, clubbing, or edema. Neurologic examination was normal.

X-ray examination of the chest (fig. 1) showed a normal-sized heart and clear lung fields. The aortic knob could not be seen and there was rib notching only on the right side. The
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Pulmonary vasculature was within normal limits. The electrocardiogram (fig. 2) was interpreted as normal.

A thoracic aortogram was done in August 1963. Renografin (76 per cent) was injected through a catheter placed in the right femoral artery and threaded into the thoracic aorta to the level of the left subclavian artery (fig. 3). The descending aorta was small and did not connect with the ascending aorta. The left subclavian and left common carotid arteries were seen to originate from the descending aorta at its proximal end. A second injection of Renografin was made through a catheter placed in the right axillary artery and threaded into the supravalvular aorta (fig. 4). The aortic valve, the ascending aorta, and the coronary arteries appeared normal. There was no aortic insufficiency. The ascending aorta was seen to be continuous with the innominate artery, which divided into an extremely large internal mammary artery, right subclavian artery, and right common carotid artery. No left common carotid vessel could be seen. A later film (fig. 5) showed massive collateral circulation through the neck, the right shoulder, and the right lateral thoracic vessels. No patent ductus arteriosus was present. No collateral channels were seen in the left chest.

Figure 3
Retrograde aortogram showing the descending thoracic aorta (D.T.A.), and the left subclavian (L.S.C.), and the left common carotid (L.C.C.) arteries.

Figure 4
Supravalvular aortogram showing the aortic valves (A.V.), the internal mammary (I.M.), the right subclavian (R.S.C.), and the right common carotid (R.C.C.) arteries.

Figure 5
Film showing the massive collateral circulation in the right lateral thoracic wall and in the cervical area.
Figure 6

Drawing of the operative findings as seen through a left lateral thoracotomy.

Left lateral thoracotomy was performed with removal of the fourth rib on October 21, 1963 (fig 6). There were no enlarged collateral vessels in the latissimus dorsi and serratus anterior muscles, and the intercostal arteries were small. Dissection of the descending aorta revealed a large left subclavian artery in the normal position and a larger vessel, the left common carotid artery, originating from the aorta just cephalad to the subclavian. None of these three vessels was pulsatile. There was no connection between the descending aorta and the pulmonary artery. Dissection at the base of the heart showed the pulmonary artery and the ascending aorta to be normal in size and position. No thrills were palpable at the base of the heart or over the ventricles. A distance of 3 cm. separated the ascending aorta from the descending aorta, and no fibrous strand connected these vessels. The ascending aorta branched into three trunks: the right common carotid, the right subclavian, and the right internal mammary arteries. The ligamentum arteriosum connected the posteromedial aspect of the ascending aorta to the bifurcation of the pulmonary artery.

Continuity between the ascending and the descending aorta was established with a 10-mm. knitted Dacron graft. A side-biting Potts clamp was used to isolate a portion of the ascending aorta. An end-to-side anastomosis between the ascending aorta and the pre-clotted Dacron graft was made with running 4-0 silk. A similar Potts clamp was applied across the bulbous portion of the proximal descending aorta and an end-to-side anastomosis was completed. The left common carotid vessel was temporarily clamped while the clamps on the aorta were removed to prevent any air trapped in the graft from going to the head.

Postoperatively there was a transient period of hypertension with a pressure rise to 170/110 that subsided in a few hours without treatment. Strong arterial pulsations were palpable in the left radial, left common carotid, both femorals, and the posterior tibial and dorsalis pedis vessels as well as in the right radial and right common carotid vessels. The postoperative convalescence was uncomplicated, and the patient was discharged on the tenth hospital day. On the day of discharge the blood pressure in the left arm...
was 110/80 and in the right arm 120/80.

The patient was seen in the surgical clinic 3 months postoperatively. The claudication and paresthesias in her legs had disappeared. The blood pressure in the right arm was 140/90, in the left arm 120/90, and in both lower extremities 130 mm. Hg systolic.

Discussion

There is a 76-per cent mortality with interruption of the aortic arch within the first month after birth.2,3 This high mortality is thought to be due to associated cardiovascular anomalies. In Roberts2 review of the literature ventricular septal defects were present in 49 of 51 patients in which the integrity of the ventricular septum was cited. Deformity of the aortic valve or of the subaortic outflow tract is seen frequently. A patent foramen ovale or an atrial septal defect is nearly always present. A ductus arteriosus connecting the pulmonary artery to the descending thoracic aorta has been recorded in all the previously reported cases. The large patent ductus plus the associated intracardiac defects lead to early congestive heart failure, pulmonary complications, and death. It has been stated that complete interruption of the aortic arch occurring as an isolated anomaly is incompatible with life.2,3

This patient’s favorable clinical course can be explained by the absence of intracardiac defects and of a patent ductus arteriosus. The blood flow to the descending aorta was by way of collateral vessels in the right side of the chest wall and through retrograde flow in the left common carotid and left subclavian arteries. The retrograde flow of blood in the left common carotid and left subclavian arteries explains the incomplete filling of these vessels on the aortogram. Obliteration of the ductus allowed the massive collateral circulation to develop. This collateral aided in decompressing the left side of the heart and protected the patient against left-sided heart failure and severe systemic hypertension. The lack of any connection between the systemic and pulmonary circulations also prevented the development of pulmonary hypertension and its sequelae.

The possible embryologic explanation of this patient’s defect is pictured in figure 7. Normal regression of the left-sided dorsal aorta between the left third and fourth aortic arches did not occur. This vessel persisted as the large vessel seen connecting the upper part of the descending thoracic aorta with the carotid vessels and is functionally the
left common carotid artery. There was an abnormal regression of the left third, fourth, and sixth aortic arches, which explains the interruption of the aortic arch between the innominate artery and the functional left common carotid artery (persistent left dorsal aorta), and the absence of the left-sided ductus arteriosus. There was a right-sided ductus arteriosus, which became obliterated.

The clinical diagnosis of interruption of the aortic arch is difficult because of the similarities between it and coarctation of the aorta. The associated intracardiac anomalies, usually present, tend to overshadow the interruption of the arch. Cardiac catheterization and angiograms are necessary to demonstrate the entire complex and to make the diagnosis of arch interruption. As demonstrated by our case, the prognosis depends on the severity of the intracardiac anomalies and the presence of a patent ductus arteriosus rather than the arch interruption.

The type of surgical correction depends on the anatomy. An end-to-end anastomosis between the ascending and the descending aorta would be the procedure of choice; however, a prosthetic graft may be needed to bridge the interruption. Merrill reported a successful surgical correction by anastomosing the left subclavian artery to the descending aorta in an end-to-side manner. Quie reported a case in which the left common carotid artery was sutured to the descending aorta, but the patient died 2 days later. Ruiz Villalobos reported the successful correction in a 14-year-old patient by using a Teflon graft between the left subclavian artery and the descending aorta. In Roberts case a Teflon graft was placed between the ascending and the descending aorta, but the patient died postoperatively of pulmonary complications. Blake used a Dacron graft to bridge the interruption between the ascending and descending aorta. In all of these patients the patent ductus arteriosus was divided and sutured.

Summary

A case report of a 16-year-old white girl is presented with the cardiovascular anomaly consisting of interruption of the aortic arch between the innominate artery and the left common carotid artery and distal to a right ligamentum arteriosum. This case is unique because the interruption of the aortic arch was not associated with intracardiac anomalies, a patent ductus arteriosus was not present, and the patient had minimal symptoms. This is the sixth reported attempt at surgical correction of interruption of the aortic arch and represents the fourth successful one.

References

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