Surgical implications of right aortic arch with isolation of left subclavian artery¹

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Few cases have been reported of the combination of right aortic arch with isolation of the left subclavian artery in childhood because of the absence of symptoms from this pairing of defects and the difficulty of its recognition by routine examination. All the reports have been associated with congenital heart disease. We report 4 cases. In one of them, the diagnosis was made during an attempt to create a left Blalock-Taussig anastomosis and the technique was modified. The diagnosis can be made before operation with an aortogram. This procedure should be carried out to show the aortic arch branches in cases where palliative surgery is contemplated.

Among the aortic arch anomalies, the association of a right aortic arch with the left subclavian artery isolated from the aorta is the least common (Stewart, Kincaid, and Edwards, 1964; Stewart, Kincaid, and Titus, 1966; Shuford, Sybers, and Schlant, 1970b; Victorica, Van Mierop, and Elliott, 1970). Few cases have been reported because the diagnosis cannot be made on the chest x-rays or by a barium oesophagogram. Despite the rarity of the anomaly, recognition is important when palliative surgery, especially a Blalock-Taussig anastomosis is contemplated.

Among 231 cases of right aortic arch on record at The Hospital for Sick Children, only 4 cases of this anomaly have been encountered. Three were recognized during life - 2 at surgery and 1 before surgery by angiocardiology - and 1 at necropsy.

Case reports

Case 1

At 6 weeks of age a heart murmur was first detected and breathlessness was noted during feeding. A clinical diagnosis of a ventricular septal defect was made. The peripheral pulses were normal, the systemic blood pressure taken on admission was 21.9 kPa (165 mmHg) in the right arm and 16.6 kPa (125 mmHg) in the left. Chest x-ray showed the heart to be slightly enlarged and the pulmonary arterial vascularity mildly increased, indicating a small left-to-right shunt. An oesophagogram revealed that the aortic arch was right sided. There was no vascular compression of the oesophagus. Cardiac catheterization confirmed a ventricular septal defect and showed infundibular stenosis and a right

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aortic arch. A selective right ventriculogram demonstrated tetralogy of Fallot, with a right aortic arch. The brachiocephalic arteries were not outlined. There was an out-pouching of contrast material from the superior aspect of the pulmonary artery thought to represent the pulmonary end of a ductus arteriosus (Fig. 1A-B). At 21 months of age cyanotic spells developed and a left Blalock-Taussig anastomosis was planned. At operation the left subclavian artery was found to arise from the pulmonary artery, with no obvious connexion with the innominate artery. Because of this, a modified shunt was created, using the left subclavian artery as a free graft, anastomosing to the side of the left carotid artery and the left pulmonary artery near the main pulmonary artery. Postoperatively the shunt murmur was easily audible and the course was uneventful.

Case 2

The child was examined by a cardiologist at the age of 10 days because of a heart murmur. The clinical diagnosis of a left-to-right shunt at ventricular level was made. The peripheral pulses were normal. The chest x-ray showed a slight increase in the pulmonary vascularity and the heart was minimally enlarged. A repeat chest x-ray carried out three days later showed that as well as the shunt there were signs of cardiac failure with pulmonary oedema. Cardiac catheterization demonstrated a large persistent ductus arteriosus, with a moderate size ventricular septal defect, patent foramen ovale, and a right aortic arch. A selective left ventriculogram done in the left anterior oblique position revealed that there was a small membranous ventricular septal defect. With the child in the lateral position the catheter was considered to have passed from the main pulmonary artery through a persistent ductus arteriosus into the aorta and then into the left subclavian artery. Contrast material was injected. There was

FIG. 1 A and B, Case 1. Right ventricular cineangiogram showing infundibular stenosis (→). The lateral angiogram (B) also shows mild valve stenosis (upper arrow). Some contrast material is seen passing into the end of the ductus arteriosus from the main pulmonary artery. PDA, persistent ductus arteriosus.

filling of the left subclavian artery and also some contrast material passed into the left pulmonary artery. An aortogram showed a right aortic arch and there was filling of the main pulmonary artery through what was considered to be a persistent ductus arteriosus. Medical management failed to control the congestive heart failure and surgical treatment was advised at 2 months of

age. During this procedure it was noted that the left subclavian artery was arising from the persistent ductus arteriosus and there was an aortopulmonary window. Both of these were ligated. The postoperative course was uneventful. Review of the angiogram obtained by the injection into the subclavian artery with the child in the lateral position showed filling of the pulmonary

FIG. 2 Case 2. Lateral left subclavian artery injection. The catheter tip was passed through a persistent ductus into the left subclavian artery. There has been some retrograde filling of the left pulmonary artery branch. There is no opacification of the aorta. LPA, left pulmonary artery; LSA, left subclavian artery.

FIG. 3 Case 2. Aortogram in anteroposterior projection demonstrating a right aortic arch. The left and right common carotid arteries are opacified. The catheter is situated within the right subclavian artery. Contrast material is faintly seen, passing superior to the pulmonary artery through a ductus arteriosus and this is connected to the left subclavian artery. RCC, LCC, right and left common carotid arteries; MPA, main pulmonary artery. Other abbreviations as in Figs. 1 and 2.

artery through the ductus, but there was no filling of the aorta (Fig. 2). If the catheter had passed through the ductus into the aorta and then into the subclavian artery there should have been some opacification of the aorta. On the aortic root angiogram a small amount of contrast material was faintly visualized passing from the pulmonary artery into the left subclavian artery. There was opacification of both carotid arteries and the right subclavian artery though there was no visualization of the left subclavian artery from the aorta (Fig. 3).

Case 3

This patient was born at 40 weeks' gestation by caesarian section. From the fifth day he became increasingly cyanosed and was transferred to this hospital on the ninth day. A diagnosis of cyanotic congenital heart disease was made. The peripheral pulses were normal. A chest x-ray showed a slightly prominent right ventricle with decreased pulmonary vascularity and a concave pulmonary artery segment. The aortic arch was right sided. The appearance was consistent with a tetralogy of Fallot. The patient underwent cardiac catheterization to clarify the diagnosis. A selective right ventriculogram demonstrated findings consistent with a tetralogy of Fallot. An aortic root angiogram demonstrated a right aortic arch. The first branch to arise from the aorta was the left common carotid artery and this was followed by the right common carotid and right subclavian arteries (Fig. 4). There is no visualization of the left subclavian artery until a later stage, by which time contrast material had passed retrogradely through the vertebral artery and filled the left subclavian artery. A small amount of contrast material passed from the

proximal portion of the left subclavian artery to the left pulmonary artery through a small persistent ductus arteriosus (Fig. 5). The patient's condition did not warrant surgery at this time and he was discharged to be followed up as an outpatient.

Case 4

At birth this infant had multiple congenital anomalies consisting of a thoracic scoliosis, cleft palate, club feet, clinodactyly, and possible congenital heart disease. Cyanosis was noted at 4 weeks of age. By 3 years the patient was squatting and had fainting spells. At the time of admission the peripheral pulses were symmetrical. The blood pressure in the left arm was 11.3/8.0 kPa (85/60 mmHg) and in the left leg was 12.0/8.6 kPa (90/65 mmHg). A chest x-ray revealed decreased pulmonary vascularity. The heart size was difficult to assess because of a striking scoliosis convex to the left in the lower thoracic region. The aortic arch was right sided. An oesophagogram showed no vascular compression. At cardiac catheterization the diagnosis of tetralogy of Fallot with right aortic arch was made. This was confirmed on a selective right ventriculogram. The arrangement of the brachiocephalic arteries was not clearly defined. Some contrast material was seen projecting superiorly from the main pulmonary artery into what could represent the end of a ductus arteriosus (Fig. 6). At the age of 8 years total correction was carried out with a pulmonary valvotomy and infundibulectomy and patch closure of the ventricular septal defect. During the postoperative course left lung problems arose. At bronchoscopy sanguineous fluid from the left side was noted, but at reoperation only atelectasis of

FIG. 4 Case 3. Anteroposterior aortic root angiogram demonstrating a right aortic arch. The left and right common carotid and right subclavian arteries are outlined. Note the absence of the left subclavian artery.

the left lung was found. Postoperatively, hypoxia continued, with a low cardiac output, anuria, and high blood potassium. The patient was placed on a membrane oxygenator bypass and peritoneal dialysis was started. No improvement took place and she died 2 days later. At necropsy, in addition to the typical postoperative findings of tetralogy of Fallot, which had been satisfactorily corrected, the left subclavian artery was narrowed and arose from a ductus arteriosus which was

connected to the left pulmonary artery near its bifurcation. The ductus was closed over a short segment centrally.

Discussion

Edwards, in his study of aortic arch anomalies, retained those parts of the Rathke diagram depicting normal development of the aortic arch that

FIG. 5 Case 3. Lateral projection following injection into aorta. Contrast material is seen flowing retrogradely down the left vertebral artery into the left subclavian artery and the persistent ductus arteriosus. VA, vertebral artery.

FIG. 6 Case 4. Right ventricular angiogram in the anteroposterior projection demonstrating an enlarged right ventricle with infundibular stenosis, a ventricular septal defect, and right aortic arch. The left common carotid artery is the first branch to arise from the aorta. Contrast material has passed from the pulmonary artery into the lower end of a persistent ductus arteriosus.

remained after earlier changes. Edwards' classical diagram of a hypothetical double arch system as a basic pattern from which all aortic arch anomalies are derived is helpful. Interruption of this arch system at different levels can produce different anomalies (Congdon, 1972; Edwards, 1948; Barry, 1951; Blake and Manion, 1962; Felson and Palayew, 1963; Stewart *et al.*, 1964; Shuford, Sybers, and Edwards, 1970a).

A right aortic arch with isolation of the left subclavian artery can be explained by interruption of the embryonic left arch at two levels; one between the left common carotid and left subclavian arteries and the other distal to the origin of the left ductus. This results in a right aortic arch. The brachiocephalic arteries are so arranged that the first branch is the left common carotid artery in this situation. This is followed by the right common carotid and right subclavian arteries. The left subclavian artery is connected to the pulmonary artery by the left ductus arteriosus.

In previous reports all of the cases were associated with congenital heart disease. Three of the cases presented in this study had tetralogy of Fallot and one had an aortopulmonary window. The frequency of this anomaly in the general population, without associated congenital heart disease, is unknown because of the absence of symptoms and inability to detect the lesion on plain chest x-ray or oesophago-

grams. With this entity, the aortic arch and brachiocephalic arteries lie anterior to the trachea and the aorta descends on the right without producing a posterior defect on the barium-filled oesophagus. There is no way, therefore, of differentiating between the anomaly under discussion from a right aortic arch with mirror image branching by an oesophagogram. In our experience routine examination of the brachial pulses has not been helpful.

Recognition of this anomaly at cardiac catheterization and angiocardiology is possible, though errors of interpretation of the catheter course and of the angiographic findings may give an incorrect diagnosis. It should be stressed that an aortogram be carried out in both the anteroposterior and lateral projections in any child with a tetralogy of Fallot and a right aortic arch so that the anatomy of the brachiocephalic vessels can be clearly defined. This procedure is highly desirable if a palliative procedure, especially a Blalock-Taussig anastomosis, is contemplated.

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