Left-sided juxtaposition of the atrial appendages¹ *Report of 4 cases diagnosed by cardiac catheterization and angiocardiography*

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Four cases of juxtaposition of the atrial appendages diagnosed during cardiac catheterization and angiocardiography are described. These cases and 49 previously reported were all associated with severe cyanotic congenital heart disease, especially transposition of the great arteries. Though the presence of left-sided juxtaposition of the atrial appendages may be suspected during cardiac catheterization it can only be shown with certainty by angiocardiography. The possible dangers of performing balloon atrial septostomy in transposition of the great arteries when left-sided juxtaposition is present and the precautions necessary to prevent them are described.

The term juxtaposition of the atrial appendages was coined by Dixon (1954) though the abnormality was first described by Birmingham (1893). In this anomaly the body of the affected atrium, usually the right, is normal, but the atrial appendage is elongated and passes behind the great arteries so that its tip lies adjacent to the opposite normal appendage. When left-sided juxtaposition is present the right atrial appendage is abnormal, and when right-sided, the left is the abnormal one. Melhuish and Van Praagh (1968) reviewed published reports of 21 cases brought to necropsy and a further 21 cases which they collected from 2 centres: The Congenital Heart Disease Research and Training Centre, Chicago, and the Hospital for Sick Children, Toronto. Raghib, Anderson, and Edwards (1966) described 1 case which was mentioned but not reviewed by Melhuish and Van Praagh, and Wagner, Alday, and Vlad (1970) added 6 cases making a total of 49. Forty-four had left-sided juxtaposition and 5 right-sided juxtaposition. All were associated with severe congenital heart disease which in approximately 90 per cent of cases consisted of transposition of the great arteries. The average age at death in Melhuish and Van Praagh's series was 8 months and in the smaller series of Wagner *et al.* it was 4½ years. In only 3 cases was the anomaly demonstrated in life by angiocardiography (Ellis and

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Jameson (1963) 1 case, and Wagner *et al.* (1970) 2 cases).

Eighty-three babies with severe cyanotic congenital heart disease were investigated by us between 1967 and 1970 and 4 were found to have left-sided juxtaposition of the atrial appendages. Three were associated with transposition of the great arteries and 1 with persistent truncus arteriosus. This report presents the clinical and angiocardiographic findings in these 4 infants and the necropsy findings in the 3 babies who subsequently died. The possible difficulties and dangers of balloon atrial septostomy in transposition of the great arteries when left-sided juxtaposition of the atrial appendages is present are pointed out.

Patients and method

The 4 cases in this report were diagnosed during the emergency investigation of 83 infants with severe congenital heart disease between 1967 and 1970. Cardiac catheterization was carried out under general anaesthesia with curarization and intermittent positive pressure respiration throughout. Right heart catheterization was performed via the right femoral vein below the sapheno-femoral junction and intracardiac pressures were recorded using Elema transducers and an Elema Schönderling Mingograph 81 Recorder. Oxygen saturation of the blood samples was measured by a Kipp haemoreflectometer. Single plane cine angiocardiograms were recorded in each case on 35 mm cine film and videotape, using 'Cardioconray' and a Contrac injector, and biplane Elema angiocardiograms were recorded in Cases 2 and 3.

Case reports

Case 1

Male, date of birth 24 July 1967, birthweight 2800 g.

At birth the baby was cyanosed for a short period, but heart disease was not then suspected. At 10 days of age he was admitted to hospital with increasing cyanosis and heart failure. On examination the heart sounds were normal and a systolic murmur grade 3/6 was heard over the praecordium. Chest x-ray (Fig. 1a) showed cardiac enlargement, pulmonary plethora, and a narrow upper mediastinum on the posteroanterior view. The electrocardiogram was within normal limits. A clinical diag-

nosis of transposition of the great arteries was made and the baby was investigated the following day.

A cine angiocardioqram in the anterior ventricle demonstrated transposition of the great arteries and persistent ductus arteriosus (Fig. 1b); other frames from this angiocardioqram showed a ventricular septal defect. Subsequently a cine angiocardioqram was recorded following injection into what was thought, by the catheter position, to be the left atrium. The videotape recording, however, showed that the injection had been made not into the left atrium, but into an elongated right atrial appendage which extended out to the upper left heart border posteriorly (Fig. 1c and 1d). The baby died at the

FIG. 1 a) Chest x-ray, posteroanterior view, showing cardiac enlargement, pulmonary plethora, and a narrow upper mediastinal shadow. b) Cine angiocardioqram, left lateral view, showing the aorta arising from the right ventricle, and persistent ductus arteriosus. c and d) Cine angiocardioqram, posteroanterior and left anterior oblique views, demonstrating the elongation and abnormal position of the right atrial appendage. e) The heart is viewed from its left anterior aspect. The upper pointer indicates the right atrial appendage and its posterior relation to the great arteries. The left atrial appendage, lower pointer, is slightly retracted.

age of 6 weeks and necropsy confirmed the presence of transposition of the great arteries, ventricular septal defect, persistent ductus arteriosus, and left-sided juxtaposition of the atrial appendages. The left atrial appendage was normal, but the right appendage passed behind the great arteries to lie above and adjacent to the left and formed the upper part of the left heart border viewed from the front (Fig. 1e).

Case 2

Female, date of birth 12 December 1969, birthweight 2500 g.

The baby was born by caesarean section after antepartum haemorrhage 4 weeks before term. A systolic murmur was noted at the age of 2 days and the child was admitted to hospital with cyanosis and heart failure at the age of 3 weeks. A systolic thrill was palpable to the right of the sternum in the second intercostal space.

A widely conducted midsystolic murmur grade 4/6 was heard loudest at the aortic area. Chest x-ray (Fig. 2a) showed cardiac enlargement with prominence of the right upper mediastinum. Pulmonary vascularity was increased on the right compared with the left. The electrocardiogram showed left axis deviation and left ventricular hypertrophy. A clinical diagnosis of tricuspid atresia was made and cardiac catheterization was carried out the following day.

A biplane Elema angiocardigram in the left ventricle showed a ventricular septal defect, a hypoplastic right ventricle, persistent truncus arteriosus Type 2, and a right-sided aorta (Fig. 2b and 2c). A right atrial angiocardigram demonstrated tricuspid atresia, an atrial septal defect, and in addition, left-sided juxtaposition of the atrial appendages (Fig. 2d).

The baby died shortly after the investigation was completed. Necropsy confirmed persistent truncus arteriosus with a quadricuspid myxomatous valve, ventricular septal defect, atrial septal defect, rudimentary right ventricle, and left-sided juxtaposition of the atrial appendages (Fig. 2e). In addition, there was a large left-sided pericardial defect which left the main ventricular mass uncovered.

Case 3

Male, date of birth 6 February 1970, birthweight 3500 g.

The baby appeared normal at birth, but became cyanosed at 4 days of age and was admitted to hospital at 15 days of age. The cardiac impulse was forceful and a systolic murmur grade 4/6 was present at the left sternal edge. Chest x-ray (Fig. 3a) showed cardiac enlargement and pulmonary plethora. The left heart border and mediastinum were prominent and the thymic shadow prominent on the right. The electrocardiogram showed left axis deviation and left ventricular dominance. Transposition of the great arteries was diagnosed and the baby was investigated 2 days later. During the investigation the catheter tip was repeatedly passed from the right atrium into the left atrium as confirmed by blood oxygen saturations. At other times the catheter

seemed to advance along an identical path towards the upper left border of the heart, but the oxygen saturation at this site was similar to right atrial blood. The presence of left juxtaposition of the atrial appendages was suspected and confirmed by cine angiocardigrams. The cine and biplane Elema angiocardigrams (Fig. 3b and 3c) also showed the presence of transposition of the great arteries, atrial septal defect, and ventricular septal defect. The baby died suddenly 3 months later and necropsy confirmed the presence of transposition of the great arteries, large atrial and ventricular septal defects, and left juxtaposition of the atrial appendages (Fig. 3d).

Case 4

Female, date of birth 8 May 1970, birthweight 2900 g.

Cyanosis was present at birth and the child was admitted to hospital on the second day of life, in heart failure. A short systolic murmur grade 2/6 was heard at the left sternal border. Portable chest x-ray showed oligæmic lung fields, prominent left heart border, and wide superior mediastinal shadow. The electrocardiogram showed left axis deviation. Pulmonary atresia was thought to be the most probable diagnosis.

Cardiac catheterization was carried out at the age of 4 days. During the investigation the catheter tip, as in Case 3, could be passed from the body of the right atrium either into the left atrium or to the upper left border of the heart from which site desaturated blood was obtained. Juxtaposition of the atrial appendages was again suspected, and selective cine angiocardigrams in the right and left atrial appendage in posteroanterior and lateral projections confirmed this. Cine angiocardiology also demonstrated transposition of the great arteries, ventricular and atrial septal defects, and infundibular and valvar pulmonary stenosis.

Severe cyanosis and dyspnoea persisted and on the 10th day of life a shunt operation was carried out which resulted in considerable clinical improvement.

Discussion

Melhuish and Van Praagh (1968) suggested that, from the morphogenetic standpoint, juxtaposition of the atrial appendages was not a primary cardiac malformation *per se*, but a secondary effect of one or more associated anomalies. All the recorded cases had severe cono-truncal abnormalities which consisted of transposition of the great arteries in more than 90 per cent. In our own small series of 4 cases, 3 had transposition of the great arteries and 1 had truncus arteriosus Type 2: an anomaly not previously described in association with juxtaposition of the atrial appendages.

Diagnosis

There seems to be little possibility of ever diagnosing the anomaly on clinical grounds alone and chest x-rays also seem to be unhelpful. Ellis and

FIG. 2 a) Chest x-ray, posteroanterior view, showing cardiac enlargement, prominence of the right upper mediastinum, and pulmonary vascularity increased on the right side. b and c) Selective biplane Elema angiogram demonstrating ventricular septal defect, hypoplasia of the right ventricle, and persistent truncus arteriosus with right-sided aorta. d) Selective biplane Elema angiogram demonstrating tricuspid atresia, atrial septal defect, and left-sided juxtaposition of the atrial appendages. e) The heart is viewed from its right and anterior aspect. The small right ventricle and the persistent truncus are shown with the juxtaposed atrial appendages at the upper left heart border.

FIG. 3 a) Chest x-ray, posteroanterior view, demonstrating pulmonary plethora, prominent thymic shadow on the right, and prominence of left heart border and left upper mediastinum. b and c) Selective biplane Elema angiogram. The aorta arises from the right ventricle and there is abnormal position of the right atrial appendage. d) A left lateral view of the heart. The posterior relation of the right atrial appendage to the transposed great arteries is shown and the juxtaposition of the atrial appendages.

Jameson (1963) commented on the flat and inconspicuous right heart border in 6 of the 9 published chest x-rays, but the x-rays of our own cases did not show this and actually gave no clue as to the presence of juxtaposition. It is probable that the inconspicuous right heart border described by Ellis and Jameson (1963) was due to the primary cardiac lesion and not to the abnormal atrial appendage. The diagnosis can be made during life with certainty only by angiocardigraphy, though in 2 of our cases it was suspected during cardiac catheterization. In Case 1 a selective 'left atrial' angiogram actually showed that the injection had been

made into the abnormally placed and elongated right atrial appendage. In Case 2 a right atrial angiogram unexpectedly showed the abnormal appendage, while in Cases 3 and 4 it was suspected during catheterization when the catheter tip apparently passed from the right atrium into a left atrial position at the upper left heart border, but the oxygen saturation was similar to that in the right atrium. Angiocardigrams in the right atrial appendage or right atrium confirmed the diagnosis in these 2 cases, and the posterior relation of the atrial appendage to the great arteries was best seen in the lateral projection.

Because of the seemingly identical path the catheter follows on passing on the one hand from the right atrium across the septum into the left atrium and on the other hand from the right atrium along the abnormal appendage to its tip, it is obvious that a potential hazard exists if balloon atrial septostomy is necessary. To inflate the balloon in the right atrial appendage and 'pull back' could well produce either inversion of the appendage into the right atrium or possibly its rupture. It seems essential, therefore, under such circumstances to use a double-lumen Rashkind catheter (Rashkind and Miller, 1966) to determine, by blood sampling, the exact position of the balloon, whether in the left atrium or in the abnormal right atrial appendage.

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