

Creation of an atrial septal defect in transposition of the great vessels

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Four-fifths of the infants with transposition of the great vessels die within the first year of life (Abbott, 1936). Fifty-two per cent. die within the first month and 86% are dead within six months (Keith, Neill, Vlad, Rowe, and Chute, 1953). This lesion in some form occurs in 20% of necropsies on patients with congenital heart disease (Keith, Rowe, and Vlad, 1958). In Birmingham, transposition of the great vessels is the commonest form of cyanotic congenital heart disease in patients admitted to the neonatal cardiac unit. The outcome is frequently fatal. The prognosis is worse if a ventricular septal defect is absent (Noonan, Nadas, Rudolph, and Harris, 1960) and the atrial septal defect is small (Mustard, Keith, Trusler, Fowler, and Kidd, 1964). Any form of therapy is therefore useful if it can prolong life until the patient has grown sufficiently to benefit from corrective surgery. Hanlon and Blalock (1948) and Fontana and Edwards (1962) found that the duration of survival is longest when both atrial and ventricular septal defects are present. The creation of an atrial septal defect therefore appears to be a rational form of palliative therapy (Ochsner, Cooley, Harris, and McNamara, 1961; Mustard, 1964; Mustard *et al.*, 1964; Trusler, Mustard, and Fowler, 1964). Unfortunately, no detailed haemodynamic reports are available to indicate the effectiveness of creating an atrial septal defect, since cardiac catheterization in this group of patients is difficult and dangerous.

We shall describe the findings at cinéangiocardiology before and after the creation of an atrial septal defect in a seriously ill neonate who improved after operation.

CASE REPORT

A.C., a second child, was born at home at full term. Cyanosis was noted at birth. This deepened on the

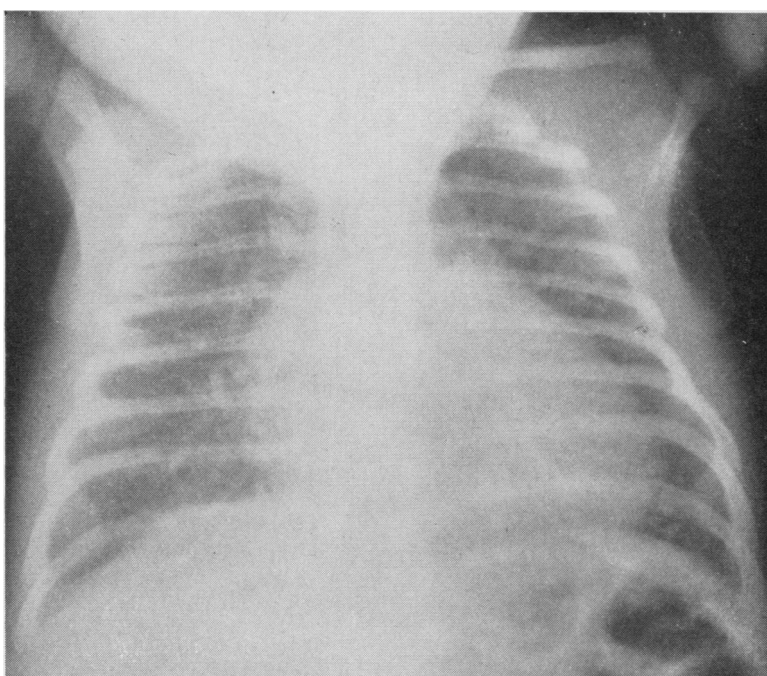
sixth day with a rapid, panting pattern of breathing, and the infant refused to feed.

The dyspnoeic, deeply cyanosed infant was admitted to hospital in gross heart failure on the eighth day of life. The eyelids and feet were oedematous and the liver enlarged 3 cm. below the right costal margin. The child was hypothermic (95° F.). The heart was enlarged with a split second sound and a loud third heart sound. No murmurs were heard. The chest radiograph suggested transposition of the great vessels (Fig. 1), a large heart with pulmonary plethora, and a full left middle segment. An electrocardiogram showed right ventricular hypertrophy. The child had severe metabolic acidosis with alveolar hyperventilation (pH 7.25, Pco₂ 17 mm. Hg, and standard bicarbonate 15.5 mEq/l.).

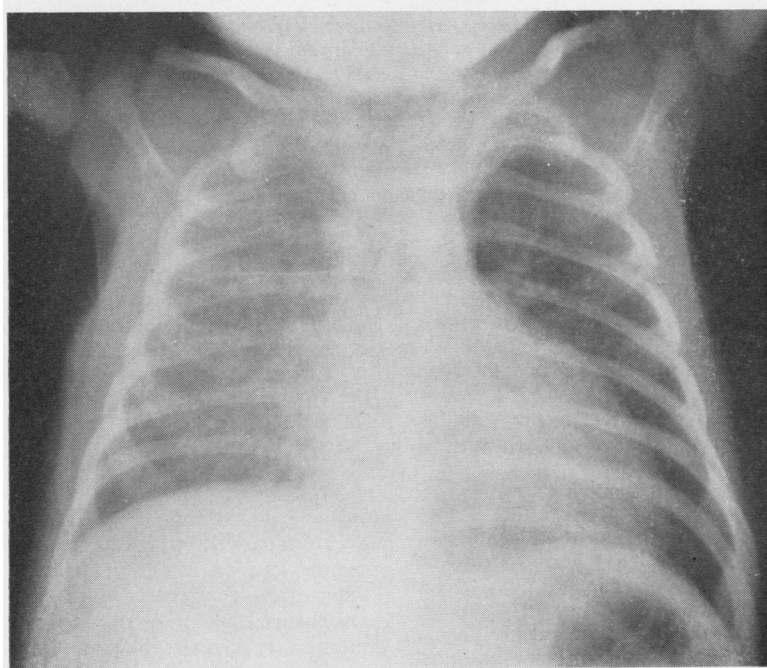
The clinical picture of transposition of the great vessels with an intact ventricular septum, hypoxia, hypothermia, and metabolic acidosis required urgent management. The cardiac failure was treated with digitalis and chlorothiazide, and the metabolic acidosis was corrected with Tris-buffer. The child's condition deteriorated.

A venous cinéangiocardiology was performed. Tracings from the film sequence are shown in Figure 2. Figure 2A shows the patient in the left anterior oblique position. A small quantity of contrast medium passed from the right to the left atrium across a small patent foramen ovale ('pony tail appearance'). Six frames later the foramen ovale was seen again (Fig. 2B). Contrast medium then passed into the right ventricle and an aorta situated anteriorly with the aortic valve at a higher level than normal. Insufficient contrast passed to the left atrium and ventricle to fill these chambers or the pulmonary artery. The ductus arteriosus also filled but formed a stagnant recess without the pulsatile appearance which is seen when the lumen is patent throughout its length. This indicated that the pulmonary end of the ductus had closed. Figure 2C shows the corresponding appearance in the right anterior oblique position. These angiocardiological findings suggested that recent closure of the ductus arteriosus was responsible for the deterioration in clinical status. Urgent surgery was performed on the same day and a Blalock-Hanlon procedure was carried out creating an atrial septal defect 4 mm. by 1 cm. in

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(a)



(b)

FIG. 1. *Chest radiographs (a) before creation of the atrial septal defect. The heart was enlarged with gross pulmonary plethora; (b) two weeks after operation. The heart had become slightly smaller.*

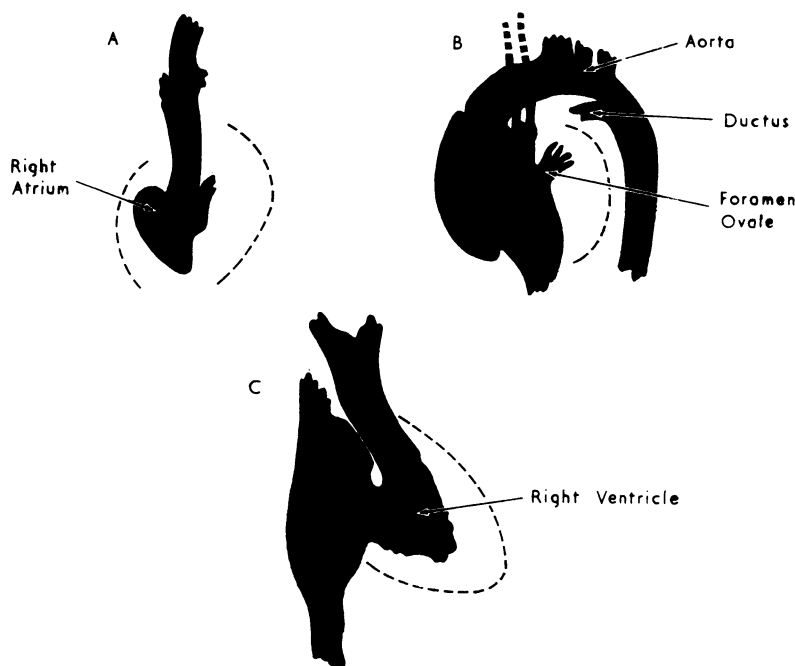


FIG. 2. Venous angiocardigraphy before operation (age 8 days). (A) Left anterior oblique position. Contrast medium has entered a large right atrium and has passed into the left atrium through a small patent foramen ovale ('pony tail' appearance). The transseptal flow occurred only during atrial systole. (B) Left anterior oblique position—six frames later. Contrast passed into the right ventricle and then into an anteriorly situated aorta. The aortic valve lay in an abnormally high position. The foramen ovale was seen again. A stagnant column of contrast medium without 'to and fro' movement filled the aortic end of the ductus arteriosus. (C) Right anterior oblique position. This frame corresponds to Figure 2B. Transposition of the aorta was confirmed. Very little contrast medium entered the left atrium. The left ventricle and pulmonary artery were not seen. There was inadequate mixing of the pulmonary and systemic circuits with a small right to left shunt through the foramen ovale and no indication of the left to right shunt (if any).

size (Blalock and Hanlon, 1950; Ochsner *et al.*, 1961).

The post-operative period was complicated by retention of sputum and severe obstructive airways disease. The PCO_2 increased to 56 mm. Hg. With prolonged intensive care the infant improved and after a fortnight the chest was normal. The cyanosis had also improved, and a week later the child was weaned from the incubator. No murmurs had appeared.

A second venous cinéangiogram was performed to assess the patency of the artificial atrial septal defect. Selected tracings from the film sequence are shown in Figures 3 and 4. Figure 3 shows the patient in the left anterior oblique position. Figure 3A shows the right atrium. The large atrial septal defect with filling of the left atrium can be seen in Figure 3B. Figure 3C shows the large anteriorly situated aorta arising from the right ventricle, and Figure 3D demonstrates good filling of the left atrium and ventricle and pulmonary artery. There was good mixing of the systemic and pulmonary venous return

and no pulmonary stenosis. In addition intermittent opacification of the right atrium suggested that there was a left to right shunt during part of the cardiac cycle with a right to left shunt during the remainder of the cycle. Figure 4 shows the same sequence in the right anterior oblique position, confirming good mixing between the right and left atria.

DISCUSSION

In this patient with transposition of the great vessels and an intact ventricular septum, clinical deterioration coincided with closure of the ductus arteriosus. Adequate mixing of the systemic and pulmonary circuits became impossible. Although the creation of an atrial septal defect did not cure the underlying condition, the effect of palliative surgery was dramatic and the post-operative cinéangiogram showed a large bidirectional

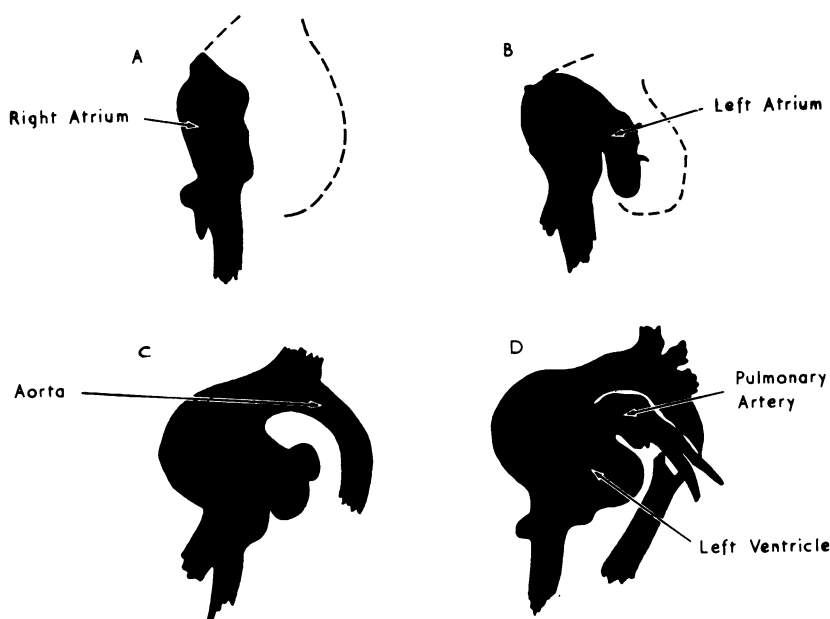


FIG. 3. Venous cinéangiogram after operation—left anterior oblique position. (A) Contrast medium entered the right atrium and refluxed into the hepatic veins. (B) Contrast medium passed across the large atrial septal defect and filled the left atrium. A dilution defect was seen in the right atrium during part of the cardiac cycle, confirming the presence of a bidirectional shunt across the defect. (C) Contrast entered the anteriorly situated aorta which arose from the right ventricle. The left atrium and ventricle were well filled. (D) Opacification of the pulmonary artery from the left ventricle was nearly as dense as opacification of the aorta. There was therefore a large right to left shunt through the newly created atrial septal defect.

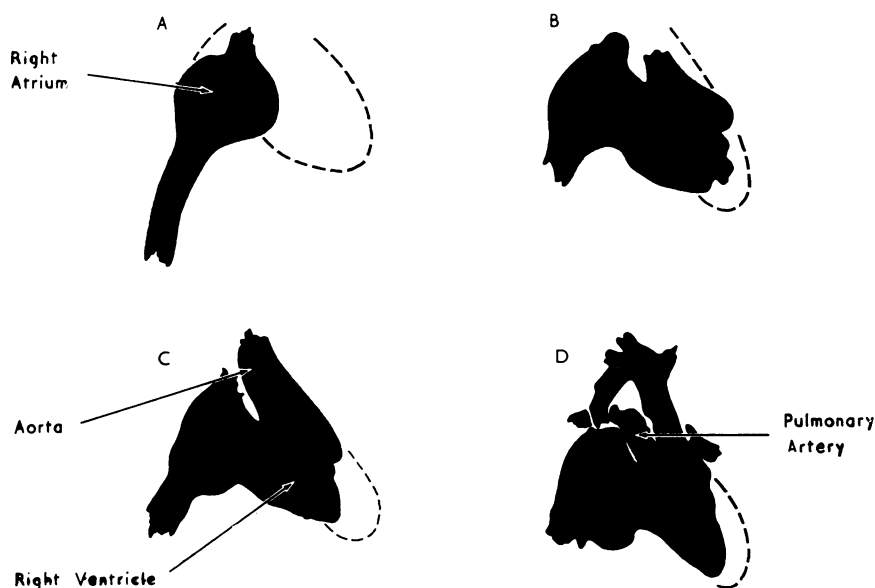


FIG. 4. Venous cinéangiogram after operation—right anterior oblique position. (A) The right atrium filled. (B) The right ventricle filled. Simultaneous opacification of the left atrium and ventricle were not visible in this view. (C) Early opacification of the aorta from the right ventricle. (D) Simultaneous filling of the aorta and pulmonary artery, indicating a good pulmonary blood flow from the systemic venous return.

shunt with adequate mixing through a non-restrictive atrial septal defect. The patient is still alive and well six months after operation.

Ochsner *et al.* (1961) have reported an increase in oxygen saturation and a similar improvement in clinical status after this procedure, and, although Trusler *et al.* (1964) report a high mortality from operation in this group of critically ill infants, the survivors improved. Our patient showed a gratifying and satisfying response to surgery. The operation improves the physiological status of the circulation, and with earlier diagnosis, a more aggressive policy of treatment, and better post-operative care, further improvement can be expected in this difficult group of neonates.

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