PAPERS AND ORIGINALS

Transposition of the great arteries: logical anatomical arterial correction

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Summary

In a 20-month-old child with classic transposition of the great arteries the pulmonary artery, coronary arteries, and aorta were successfully retransposed at arterial level. It was difficult to reimplant the coronary arteries in the dilated pulmonary artery root without damaging the attachments of the valve cusps, and it may be better to divide the pulmonary artery first to avoid this hazard. Retransposition at arterial level will not be suitable for all patients with transposition of the great arteries, and many questions about long-term outcome remain to be answered.

Introduction

When a child is born with transposition of the great arteries the aorta arises directly from the morphological and functional right ventricle and receives systemic venous blood, while the pulmonary artery arises from the left ventricle and receives oxygenated pulmonary venous blood. This transposition is life-threatening unless there is a sizeable communication between the two circulations. Creation of an atrial septal defect by Rashkind septostomy[1] has saved many infants in the first few weeks of life and dramatic surgical sucess has been achieved by physiological interatrial correction—namely, Mustard's operation.[2-3] Those with an added large ventricular septal defect and pulmonary stenosis have been helped by the use of a valved conduit.[4]

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The logical method of treating true classic transposition of the great arteries, however, is to retranspose the aorta and pulmonary artery at arterial level. The main problem has been in transferring the coronary arteries. Several pioneer attempts were unsuccessful in the early days of open heart surgery,[5-7] but experiments have shown the practical possibility of retransposition.[8-11] Recently Jatene *et al*[12] reported the first successful correction of transposition of the great arteries at arterial level. This achievement opens up a new future for sick children with this congenital cardiac malformation, and we report here on another child who was successfully treated by returning the great arteries to their normal physiological and anatomical positions.

Case report

A 20-month-old girl was referred for treatment to the National Heart Hospital because of increasing cyanosis obvious since the age of 3 months and recurrent heart failure, thriving problems, and respiratory infections since the neonatal period.

She was a blue, small child with bulged chest, a large heart and overactive right ventricle, a long systolic murmur conducted to the right of the sternum, a single loud second heart sound, and slightly jerky pulse. The chest radiograph showed cardiomegaly, intense pulmonary plethora, and a large right pulmonary artery to the right of the sternum. The electrocardiogram confirmed biventricular hypertrophy. Cardiac catheterisation confirmed the diagnosis of transposition, atrial septal defect and a large duct with slight pulmonary stenosis (gradient 20 mm Hg), and important pulmonary hypertension (see table). Angiography showed the anterior aorta lying to the left and arising from the right ventricle and the posterior medial large main pulmonary artery (fig 1). In view of the absence of a ventricular septal defect and the presence of hyperkinetic pulmonary hypertension with a left ventricle contracting at systemic pressure we considered that this would be the ideal case in which to retranspose the aorta and pulmonary artery.

On October 30 1975 DR, using surface cooling with cardiopulmonary bypass with reduced flow and further cooling to 17°C, exposed the heart through a median sternotomy. The external appearances were those of a D-transposition but with juxtaposition of the atrial appendages on the left side. There was a large tense pulmonary artery lying behind the moderately well-developed anterior ascending aorta, and the enlarged left and right ventricles were equal in size and morphologically normal. The pulmonary arteries were large.

Haemodynamic data in child aged 20 months with transposition of great vessels and persistent duct and atrial septal defects before arterial correction and five weeks after operation. Both were studies performed under local anaesthesia, breathing air. Pressures are given as systolic/diastolic or mean.

	Pressures (mm Hg)						O_2 Saturations (%)					
	RA	RV	PA	LA	LV	AO	RA	RV	PA	LA	LV	AO
Before operation	4	74/5	54/42	5	78/7	73/49	55	63	87	90	88	62
After operation	2	40/3	38/11		97/7	76/39	67	63	64		89	90

RA = Right atrium. RV = Anatomical right ventricle. PA = Pulmonary artery. LA = Left atrium. LV = Anatomical left ventricle. AO = Aorta.

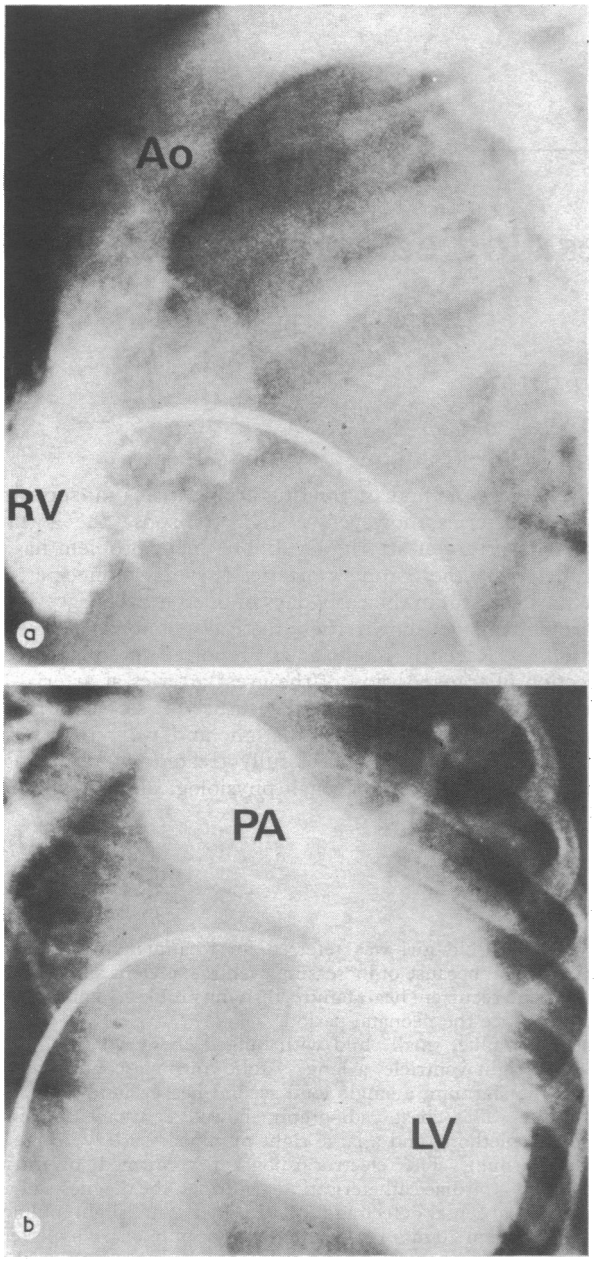

FIG 1—Preoperative angiocardiograms. (*a*) Lateral view showing aorta (Ao) arising anteriorly from right ventricle (RV). (*b*) anteroposterior view showing pulmonary artery (PA) arising from left ventricle (LV).

The coronary arteries did not appear to arise higher than usual from the root of the aorta. The large ductus was dissected out and surrounded by a loose ligature.

The ductus was closed and during this time right and left coronary arteries were dissected close to the aorta, and, at a temperature of about 20°C, the aorta was cross-clamped. The right and then the left coronary artery were excised with a disc of aorta and the holes in the aorta were closed with a sheet of homologous dura mater sewn in place with continuous 6/0 Prolene. The aorta was then transected to give good access to the root of the pulmonary artery and the coronary arteries were implanted into the sides of the pulmonary artery root using 6/0 Prolene. It was difficult to do this without damaging the attachments of the valve cusps. The pulmonary valve, arising from the left ventricle, was normal and non-stenotic but immediately below it—that is, within the left ventricle—there was a fibrous sub-valvar stenosis, which was excised.

The huge pulmonary artery was sewn across half its diameter and anastomosed to the ascending aorta with 4/0 Tycron sutures. The transsected main pulmonary artery could not be approximated to the remains of the aortic root emerging anteriorly from the right ventricular outflow vessel, so a length of aortic homograft about 2 cm long had to be interposed to achieve this connection between the main pulmonary artery and the "aortic" root arising from the right ventricle.

Rewarming was carried out during this anastomatic procedure, and at this stage the right atrium was opened. A full-sized fossa ovalis septal defect, but no ventricular septal defect, could be seen or felt through the tricuspid valve. The atrial septal defect was closed with a patch of autogenous pericardium, and on closure of the right atrium the heart came off bypass without difficulty to sinus rhythm. The child was ventilated over the first night after operation and kept on isoprenaline. There were no special postoperative problems except for some right heart failure and pneumonitis. Five weeks after operation she was readmitted for cardiac catheterisation (table I). There was a persistent subvalvar gradient of 20 mm Hg across the left-sided outflow tract. Three months after the operation she was leading a normal life without symptoms or medication.

Discussion

This case illustrates the logical form of correction for patients born with transposition of the great arteries. Not all patients will be suitable for this procedure, however, because a left ventricle which is used to beating at systemic pressure is needed to cope properly with systemic arterial resistance and pressures. This may explain some of the earlier failures with the more simple forms of transposition where only an interatrial communication is present and left ventricular pressure is low. Patients who may be suitable for arterial correction will thus be those with a large ventricular septal defect, such as Jatene's patient, or with a large duct or those who have important pulmonary stenosis which raises the left ventricular pressure.

At present we would not be happy to switch the vessels in those with pulmonary valve stenosis, however easy this may be, because of the obvious additional problems of an "aortic" valve replacement either at the time or subsequently. Ideally, patients with transposition who are suitable for a "switch" should have either a banded pulmonary artery, subpulmonary stenosis not due to mitral cusp tissue, or hyperkinetic pulmonary hypertension. Unfortunately children with these added large lesions tend to develop early important pulmonary vascular disease, which may prevent a good lasting result of any type of corrective surgery. The child we treated was unusual in having a large natural atrial septal defect and persistent duct which permitted survival and created favourable haemodynamics for this procedure. The subpulmonary obstruction, common but unpredictable in true transposition, was probably acquired from turbulent fibrosis. If present in association with ventricular septal defect it would offer some protection from pulmonary vascular disease but with a duct would merely encourage more left-to-right shunting through the duct and possibly aggravate the development of peripheral pulmonary vascular changes. The

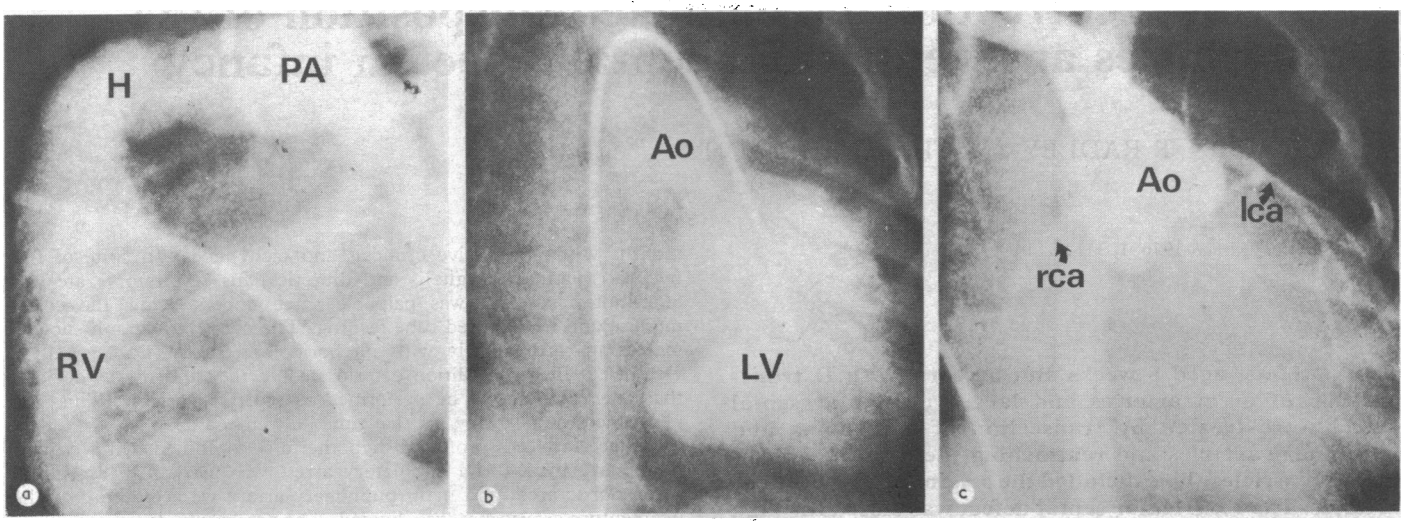

FIG 2—Postoperative angiocardiograms. (*a*) Lateral view showing connec tion of right ventricle (RV) to pulmonary artery (PA) with homograft (H) segment. (*b*) Anteroposterior view showing connection of aorta (Ao) to left ventricle (LV). (*c*) Anteroposterior view of aorta (Ao) showing origins of left (lca) and right (rca) coronary arteries.

postoperative systolic pulmonary artery pressure of 38 mm Hg suggests that there was already some pulmonary vascular damage. Only time will show if this is reversible but the fall in pulmonary artery pressure from the high levels before operation suggests that much of the pulmonary hypertension was in fact hyperkinetic.

Most children with classic transposition who are suitable for arterial correction—that is, those with a high left ventricular pressure—will have an added ventricular septal defect. Closing this adds the risk of heart block and a longer period on cardiopulmonary bypass, thus increasing mortality and morbidity. The risks of this new procedure must be carefully balanced against the problems of the Rastelli procedure and the valve conduit used.

Our patient had unusual associated natural anomalies which cannot be expected in more than 5-8% of children with transposition. Only time will tell if this logical operation will have better results than an interatrial correction, which adds a further structural abnormality to the malformed heart. What the evolution of the subaortic gradient (once subpulmonary) will be is unknown. Since the septum no longer moved paradoxically on the echo cardiogram after correction, there may be no continued left subvalvar turbulence or fibrous deposition so, we hope, it should not progress.

Technically, there were several problems. One of them was the difficulty in implanting the coronary arteries into the dilated root without damaging the valve cusps: it is probably preferable to divide the pulmonary artery first to avoid this possible hazard. Another problem was the huge pulmonary artery which had to be pleated to fit the smaller aortic root.

Nothing is known of the future of such a procedure. There are various questions to be answered: will there be growth of the suture lines? Will the coronary arteries remain patent? Will the anatomically abnormal right ventricle function into adult life? What is the ideal time to do this physiologically correct procedure? The reason for this report is to stimulate further thought and work in the management of this complex and not uncommon form of congenital heart disease.

We thank Dr Richard Emanuel who referred the child for treatment and has given us permission to report these details and Dr Keith Jefferson for his radiological assistance.

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ADDENDUM—Since submitting this paper for publication another child aged 4 years with severe asthma has had successful arterial correction for transposition of the great arteries, large ventricular septal defect, and subpulmonary obstruction. The child had been breathless at the age of 1 week and was helped by a Rashkind septostomy. Progressive cyanosis and disability continued, reflected by a preoperative haemoglobin level of 21 g/dl. The pulmonary artery pressure was 52/4; mm Hg with left ventricular pressure 82 mm Hg. The operative technique was the same as in the case reported. The holes in the root of the aorta where the coronaries had been removed were patched with dura mater and the long distance between the main pulmonary artery and the aortic stump was joined with a length of preclotted Dacron. The postoperative right ventricular pressure was 40 mm Hg when the aortic pressure was 100 mm Hg. The first weeks after operation have been uneventful.

References

[1] Rashkind, W I, and Miller, W, *Journal of the American Medical Association*, 1966, **196**, 991.
[2] Mustard, W T, *Surgery*, 1964, **55**, 469.
[3] Stark, J, *et al*, *Journal of Thoracic and Cardiovascular Surgery*, 1974, **67**, 673.
[4] Rastelli, G C, Wallace, R B, and Ongley, P A, *Circulation*, 1969, **39**, 83.
[5] Mustard, W T, *et al*, *Surgery*, 1954, **36**, 39.
[6] Bailey, C P, *et al*, *Journal of Thoracic Surgery*, 1954, **27**, 73.
[7] Kay, E B, and Cross, F S, *Surgery*, 1955, **38**, 712.
[8] Idriss, F S, *et al*, *Circulation*, 1961, **24**, 5.
[9] Baffes, T G, Ketola, F H, and Tatooles, C J, *Diseases of the Chest*, 1961, **39**, 648.
[10] Anagnostopoulos, C E, *Annals of Thoracic Surgery*, 1973, **15**, 565.
[11] Anagnostopoulos, C E, *Annals of Thoracic Surgery*, 1973, **16**, 458.
[12] Jatene, A D, *et al*, *Arquivos Brasileiros de Cardiologica*, 1975, **28**, 461.