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Correction of uncomplicated cases of transposition of the great arteries

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In recent years the surgical treatment of transposed great arteries has become one of the standard operations of cardiac surgery. Though exchange of the great arteries has theoretical attractions, the success of rearrangement of the atrial flow has been so great that this will probably remain the operation of choice for some time yet. The principle of this operation was first described by Albert (1955). Although Senning (1959) was the first to achieve success with this operative approach, its widespread application did not occur until Mustard described his technique in 1964. Other modifications of the principle had been attempted by Creech *et al.* (1958), Merendino *et al.* (1957), Shumacker (1961), and Barnard, Shrire, and Beck (1962), but only Barnard's case had been successful.

Our experience has been with the Mustard operation. Since February 1965 112 cases have had Mustard's operation at The Hospital for Sick Children, Great Ormond Street. The types of cases are shown in Table 1. Seventy-eight anatomically uncomplicated cases form the largest group and discussion will be limited to these.

TABLE 1 *Mustard's operations, February 1965 to May 1970*

Transposition of great arteries (simple)	78
Transposition of great arteries + ventricular septal defect	21
Transposition of great arteries + ventricular septal defect + pulmonary artery unbanding	4
Transposition of great arteries + ventricular septal defect + pulmonary stenosis	7
Transposition of great arteries (complex)	2
Total	112

TABLE 2 *Simple transposition of great arteries: cause of death in hospital in 10 cases*

Case No.	*Cause of death*
9	Superior vena caval obstruction
30	Pulmonary oedema
33	Persistent ductus arteriosus tear
42	? Induced ventricular fibrillation for long period
44	? Induced ventricular fibrillation for long period
60	Post-operative haemorrhage
61	Pulmonary vascular disease ($R_p/R_s = 1{\cdot}2/1$)
68	Dacron patch leak
70	Low output state
111	Pulmonary vascular disease ($R_p/R_s = 2/3$)

Results

Of the 78 simple cases, 10 (13%) died in hospital. The reasons for the deaths are shown in Table 2. The important conclusion is that almost all the causes of death were avoidable. Late deaths have occurred in 7 cases in the entire group of Mustard's operations (112 cases), but only the last two were in cases of simple transposition of the great arteries, the first five also having ventricular septal defects. However, these delayed deaths do emphasize the danger of arrhythmias (especially atrial flutter) and heart block (Table 3).

TABLE 3 *Mustard's operations: late deaths in 7 cases*

Age (yr.)	*Cause of death*	*Interval between operation and death (mth.)*
5	Atrial flutter	6
4	Nodal/atrial flutter	23
1–10/12	3rd degree AV block (late onset)	30
4	3rd degree AV block	32
2	Measles bronchopneumonia	11
1*	Febrile convulsions; inhalation pneumonia	11
16*	Pulmonary vascular disease	7 wk.

* Simple transposition.

Post-operative arrhythmias

The conduction in the first days and weeks after operation is shown in Table 4. The cases are divided into roughly half. In the first 54 cases the incidence of conduction troubles was much greater than subsequently. At this time some minor modifications to Mustard's operation were made in an attempt to reduce the arrhythmias. It can be seen that sinus

TABLE 4 *Mustard's operations: early post-operative rhythm*

Rhythm	*Cases 1–54*	*Cases 55–112*
Sinus	17	40
Nodal	20	2
2nd degree AV block	1	3
Complete AV dissociation	7 (5 s)*	10 (5 s)*
Atrial flutter	5	1 (briefly)

* s = later sinus rhythm.

TABLE 5 *Mustard's operations: long term atrial flutter*

	Cases 1–54	*Cases 55–112*
Post-operative atrial flutter (long term)	14	Nil 1 (briefly)

TABLE 6 *Mustard's operations: variations in technique*

Case No.	*Variations*
2	Atrial septum used as pedicle flap
4	Superior vena cava cannula to right side
23,30	Atrial pedicle flap posterior to coronary sinus
47	Prolonged artificial ventricular fibrillation abandoned
55	Interrupted sutures over atrial conduction paths
60	Coronary sinus not cut back
66	Dacron only used for dividing atria Suture line posterior to coronary sinus
69	Pericardium and Dacron used for dividing atria

rhythm occurred much more frequently in the latter part of the series and nodal rhythm much less commonly. Complete AV dissociation has occurred in a number of cases where sutures have not been placed near the main AV conduction bundle or the AV node, and in a number of these the conduction has later returned to sinus rhythm (see Table 4).

Over a longer term atrial flutter developed in a large proportion of the early part of the series, but since further attempts have been made to avoid damage to either the sinus or the AV nodes or to the interatrial conduction pathways the incidence of atrial flutter has been almost abolished (Table 5).

The modifications made to the operation have been almost all designed to reduce the incidence of arrhythmias and are listed in Table 6. The superior vena cava cannula is now placed as far as possible from the sinus node, and when a suture line crosses the presumed position of the interatrial conduction pathways interrupted sutures are used. To avoid damage to the posterior internodal pathway the coronary sinus is no longer cut back, and since Case 66 the patch has been placed

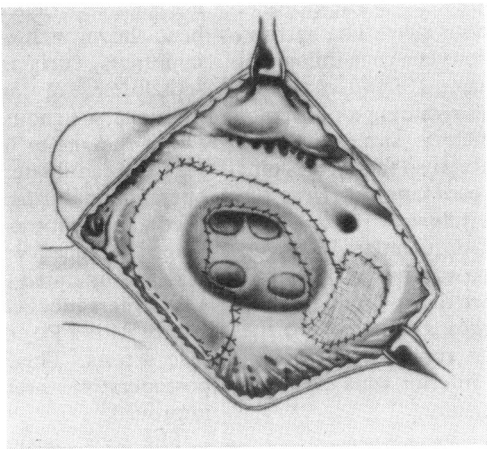
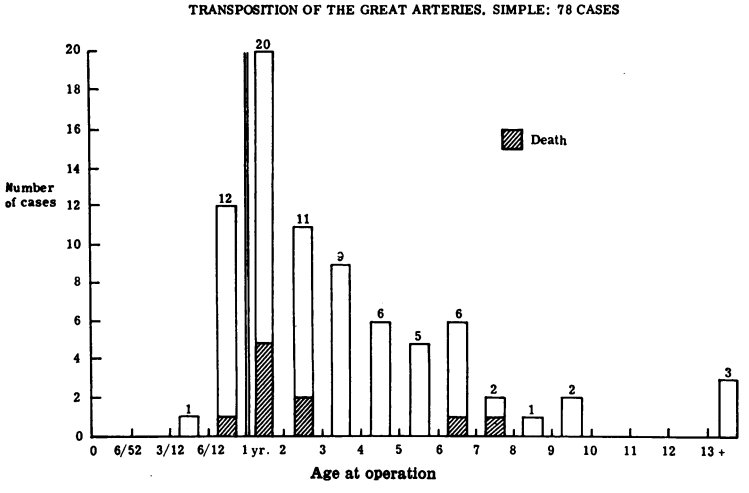


FIG. 1 *Diagram showing completed operation. The pericardial patch surrounds the pulmonary veins, and the 'D' shaped patch of Dacron lies over the inferior vena caval orifice.*

well posterior to the coronary sinus allowing this to drain to the pulmonary atrium, but the effect of this is not significant as a cause of systemic desaturation.

The type of operation which has been used in the last 43 cases is shown in Fig. 1. An attempt was made to use Dacron for the whole interatrial division, but this proved unsuccessful in two of three cases, so the technique used thereafter was to employ pericardium for most of the patch except where a narrow strip of Dacron was required – that is, over the inferior vena caval inflow. (In those cases which we have seen at necropsy some time after operation the pericardium

FIG. 2 *Comparison of hospital death with age at operation.*



appeared to have contracted to one-third or one-quarter of its previous size.) The narrowest portion of the inferior vena cava inflow has therefore been made using Dacron cut to the shape of a 'D', and also retaining a portion of the atrial septum which is sutured to the straight edge of the Dacron so that at no point does the pericardium pass completely across the inferior vena caval inflow.

Superior vena caval obstruction is known to have occurred in two surviving cases after operation, but in neither did it produce signs of any note as the azygos vein accepted the full flow, and thus the superior vena caval flow was routed to the inferior vena cava and back to the systemic atrium.

Optimum age at operation

Effective palliation by the creation of an atrial septal defect has made an immense difference to the natural history of transposition of the great arteries. Whereas 85 per cent of cases were dead by six months of age, the highest mortality being in the uncomplicated cases, now the uncomplicated cases appear to have about a 10 per cent risk if the Rashkind balloon septostomy is made in the early weeks of life (Rashkind and Miller, 1966). There would also seem to be about a 10 per cent risk of death in the next year or so after the septostomy. In addition it has been appreciated that there is quite a high risk of cerebral infarction. One of our cases which died 11 months after Mustard's operation (performed when the child was 1 year old) died with persistent febrile convulsions and inhalation pneumonia. He had been thought to have had a relatively minor stroke as an infant, but post-mortem examination showed a large area of brain scarring.

There is therefore some urgency about performing correction for transposition of the great arteries. If one simply considered the mortality risk it would be reasonable to perform Mustard's operation at six months of age if the mortality rate were no more than 10 per cent greater than the rate if the operation were performed at about 2 years of age. In the past three years we have been operating on children at a progressively younger age. Our experience so far suggests that, as has been shown with other open-heart operations in infants, such as correction of ventricular septal defect or correction of total anomalous pulmonary venous drainage, an infant between the age of 6 and 12 months is a good surgical candidate (Fig. 2). Among 13 cases operated on during infancy there has been one death, and that was possibly the result of prolonged induced ventricular fibrillation. One case was operated on at 3 months of age as a semi-emergency because the child was in severe failure with a Po_2 of 25 and had had two cardiac arrests. This child required prolonged postoperative intensive care but eventually improved.

The series is not yet large enough to be dogmatic, but it seems that, given a good background of intensive care for infants, especially intensive respiratory care, the elective age for performing Mustard's operation in uncomplicated cases of transposition of the great arteries is probably in infancy, and perhaps at about six months of age.

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