REPAIR OF CYANOTIC HEART DISEASE IN INFANCY WITH HYPOTHERMIC PERFUSION AND CIRCULATORY ARREST

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INTRODUCTION

Circulatory arrest with profound hypothermia now permits intracardiac repair of congenital heart disease in infants[1,7]. In this paper we wish to concentrate on the medical and surgical implications of the operations now possible for the treatment of cyanotic congenital heart disease in this age group.

MATERIAL

Until July, 1972, we have used profound hypothermic arrest on eighty two patients, sixty seven of them less than two years of age. Of these infants, fourteen had tetralogy of Fallot and twenty two had transposition of the great arteries. The ranges of age and weight are set out in Table 1.

METHODS

Hypothermia was induced and reversed by extracorporeal circulation with a pump oxygenator and heat exchanger[2]. Details which we believe to be important in obtaining consistently satisfactory results include:—

1. The use of minimum priming volumes for the extracorporeal circuit has led to increasing reliance on a bubble oxygenator with integral heat exchanger* for children below about 8 Kg. body weight, and with an additional heat† exchanger in bigger babies.
2. The use of heparinised blood for preference in priming, particularly for younger infants.
3. Haemodilution, using Hartmann's solution to produce a perfusate haematocrit of approximately thirty per cent.
4. The use of five per cent carbon dioxide in oxygen for the oxygenator at temperatures below thirty degrees centigrade.
5. An arbitrary minimum time of cooling perfusion of twenty minutes, or long enough to allow stabilisation of oesophageal, nasopharyngeal and arterial line temperatures at or below twenty degrees centigrade, in an attempt to minimise temperature gradients between different body areas.
6. Careful left heart venting, and monitoring of pulmonary venous pressure during rewarming to avoid lung damage.

RESULTS

Tetralogy of Fallot

Fourteen corrective operations were attempted. In all cases the ventricular septal defect was repaired with a knitted dacron patch. Usually several interrupted mattress sutures were used for the posteroinferior portion of the repair, and running sutures for the remainder. The right ventricular outflow obstruction was relieved by infundibular resection alone (seven patients) or in combination with pulmonary valvotomy (three patients) in ten patients. In four patients a gusset of pericardium was used to enlarge the pulmonary annulus and main pulmonary artery; in all four the gusset extended distally into one or both of the pulmonary artery branches.

Two patients died after surgery. One, where the right ventricular systolic pressure was greater than systemic, even after the main, right and left pulmonary arteries had been enlarged by gusset angioplasty, died within half an hour; the other died suddenly at twelve hours, from no obvious cause. Respiratory insufficiency with hypoxaemia was not a problem after surgery, since only one case required prolonged orotracheal intubation and artificial ventilation; for twenty four hours.

Neither permanent heart block nor clinically significant residual ventricular septal defect has occurred. The surviving patients are well.

Transposition of the great arteries with intact ventricular septum or small ventricular septal defect

Seventeen patients, all initially palliated by balloon atrial septostomy[6], were treated by intra-atrial baffle repair[5] using a bow-tie shaped patch of fine knitted dacron which was made neither taut nor redundant. In the region of the His bundle and atrioventricular node the suture line was confined to the base of the tricuspid valve leaflet, except in our most recent patient, in whom it ran superiorly and to the left of the coronary sinus ostium.

There were three deaths. In one patient the cardiac septa had reclosed except for a minute ventricular septal defect; he died of respiratory insufficiency secondary to pulmonary infarction and at post mortem the pulmonary arteries and inferior vena cava contained extensive thrombi. In the two others the cause of death is not known.

Four of the fourteen survivors required artificial ventilation for periods of between two and twenty four hours.

Neither permanent heart block nor troublesome atrial arrhythmias have occurred. One surviving patient has significant tricuspid incompetence and needs tricuspid valve replacement. One other patient has mild to moderate tricuspid incompetence.

Transposition of the great arteries with large ventricular septal defect or atrio-ventricularis communis defect

None of our four patients with this complex have survived operation. In two, the pulmonary artery systolic pressure exceeded systemic following repair. In one, ventricular fibrillation occurred while the heart was being exposed and effective cardiac output could not be established either before or after the repair. In the fourth patient, severe tricuspid incompetence was unrelieved by repair of the atrio-ventricular canal defect.

Transposition of the great arteries, large V.S.D., and previous banding of the main pulmonary artery

One such patient treated by repair of the ventricular septal defect (V.S.D.), baffle operation and reconstruction of the main pulmonary artery, at the age of twenty three months, has done well.

DISCUSSION

Tetralogy of Fallot

Intracardiac repair is indicated for most infants with tetralogy of Fallot who are having cyanotic spells or who are severely cyanosed, whatever their age or size. Although we have achieved fair results from ascending aorta to pulmonary artery shunts even in small infants[3], the risk of palliation in infancy is significant, the improvement not always predictable and the secondary intracardiac repair not without significant risk. Nevertheless, when the pulmonary arteries are less than about four millimetres in diameter, a palliative shunt procedure may still be indicated.

In infancy total correction of tetralogy of Fallot is surprisingly easy, perhaps because there is less secondary ventricular hypertrophy and aortic override. In reported series of patients who have been corrected in infancy, gusset reconstruction of the right ventricular outflow tract has been used frequently[1]. However, in our experience this

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TABLE I

RESULTS OF HYPOTHERMIC PERFUSION AND CIRCULATORY ARREST

Diagnosis	Number	Age (mean)	Weight (mean)	C.A.T. (mean)	Deaths	Time and cause of death
Tetralogy of Fallot	14	1 - 23 Mo (11 Mo)	3·3 - 11·6 Kg. (7·3 Kg.)	34 - 66 min. (48 min.)	2	½ Hr. unrelieved R.V. obstruction 12 Hr. sudden—? cause.
T.G.A.—Intact ventricular septum or small V.S.D.	17	2 - 23 Mo (11 Mo)	3·7 - 11·9 Kg. (7·4 Kg.)	37 - 70 min. (48 min.)	3	12 Hr. sudden—? cause 4 days. Pulmonary dysfunction with infarction. 8 days. R.P.A. stenosis—complex metabolic disturbance.
A-V Communis	1	4 days	3·0 Kg.	56 min.	1	½ Hr.—severe tricuspid incompetence.
Large V.S.D.	3	8 days - 8 Mo	3·2 - 10·7 Kg.	65 - 67 min.	3	10 min. severe pulmonary hypertension. ½ Hr. ventricular fibrillation before repair—low cardiac output. ½ Hr. severe pulmonary hypertension.
Large V.S.D. Banded M.P.A.	1	23 Mo	9·4 Kg.	64 min.	0	

C.A.T. = circulatory arrest time.
T.G.A. = transposition of the great arteries.
A-V communis = atrioventricularis communis.
V.S.D. = ventricular septal defect.
M.P.A. = main pulmonary artery.

has only been necessary in about thirty per cent of patients; a proportion comparable to that in older patients[4]. Many of our patients had almost "pure" infundibular stenosis but large distal pulmonary arteries and did not require a gusset. At the other extreme, some of our patients had such severe narrowing of the pulmonary annulus, main pulmonary artery and its branches, that a gusset was clearly necessary: whether any of these patients might, after shunting, have become suitable for less radical repairs is not yet clear.

Transposition of the great arteries

Elective physiological correction in patients with transposition of the great arteries can readily be performed in the second six months of life, thereby minimising the risks of death or serious neurological complications. The time for operation remains a compromise between the cumulative hazards of delay, and the potential technical difficulties of too early correction.

During the past eighteen months it has been our policy to correct well palliated infants between the ages of six and nine months. During this same period two patients have needed earlier surgical repair because of septal closure, with one death. One other patient died of a cerebro-vascular accident while awaiting restudy prior to corrective surgery. On the other hand, none of the survivors of the operation have yet had significant systemic or pulmonary venous obstruction due to an inadequate baffle.

Tricuspid incompetence after repair has been noted by others[8], and may prove to be a serious long term problem. Since we have become aware of this, we give digoxin prophylactically, to try to minimise annular dilatation, thereby possibly delaying the onset, or ameliorating the severity, of tricuspid incompetence. Also, we have recently avoided suturing on the tricuspid leaflet.

Transposition of the great arteries with a large ventricular septal defect remains a formidable problem. Severe pulmonary arteriolar occlusive disease can occur within the first weeks of life and therefore precludes successful repair, as in two of our cases. Although palliation by pulmonary artery banding, with subsequent repair,was successful once in our experience, we intend in the future to attempt early complete physiological correction.

SUMMARY

The results of intracardiac repair of tetralogy of Fallot and transposition of the great arteries in infants, using profound hypothermia and circulatory arrest, are reported. The operations are technically no more difficult then when performed in older children, and offer considerable advantage; without increased mortality.

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