Palliative procedures for transposition of the great arteries[1]

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Palliation in uncomplicated transposition of the great arteries is a matter of achieving increased intracardiac mixing. This can be done by surgical atrioseptectomy or by balloon atrioseptostomy. The latter technique has been proved to be a safe and effective procedure for long-term palliation. In the presence of complicating lesions adjustments of the pulmonary blood flow must be made. When the complicating lesion is a ventricular septal defect with increased pulmonary blood flow, pulmonary artery banding is often required. In the presence of severe pulmonic stenosis with a ventricular septal defect, aorticopulmonary shunting may be required. Judicious application of these palliative procedures has completely altered the outlook of the infant born with transposed great arteries, with or without complicating lesions.

Procedures for palliative treatment of complete transposition of the great arteries are directed toward delivering the maximum amount of desaturated (systemic venous) blood to the pulmonary circulation and minimizing the delivery of saturated (pulmonary venous) blood to the pulmonary circulation. This is accomplished by two general techniques: first, provision of adequate intracardiac mixing, and second, alteration of pulmonary blood flow, either decreasing it by pulmonary artery banding or ductus ligation, or increasing it by anastomosing a systemic artery to a pulmonary artery. When transposition of the great arteries is not accompanied by a large patent ductus arteriosus or by a large ventricular septal defect, with or without pulmonic stenosis, provision of adequate intracardiac mixing is sufficient to provide long-term palliation. In practical usage increased intracardiac mixing is achieved by producing or enlarging an atrial septal defect.

Blalock and Hanlon (1950) first introduced a surgical technique for atrioseptectomy. The results of the experience of the Hopkins group were reported by Cornell *et al.* in 1966. Subsequent experience in other clinics shows that septectomy can be provided for many sick infants with transposition of the great arteries with increasingly better results and lower mortality and morbidity than achieved in the earlier studies. Improvements in surgical techniques and in ancillary management

details – namely, close regulation of body temperature, acid-base equilibrium (Shaher and Kidd, 1967), and postoperative respiratory support – are mandatory adjuncts to achieving good results in these infants regardless of the technique of palliation used. Since the introduction of the balloon atrioseptostomy technique by Rashkind and Miller in 1966 this technique has achieved wide usage as a rapid, effective means of palliating these very sick infants. The primary purpose of this report is to describe the long term results of palliation in the first 45 patients treated by balloon atrioseptostomy at the Children's Hospital of Philadelphia. The first 45 patients were selected, since at least one year's follow-up data were available on all survivors.

Technique of balloon atrioseptostomy

Insertion of catheter It is mandatory to use a catheter with a balloon of sufficient size to be inflated to a diameter of at least 1·0 cm., and preferably to a diameter of 1·5 cm. This necessitates the use of a catheter of at least No. 5·5 French gauge, and preferably No. 6·5 French gauge. To ensure access to a vein of adequate size to permit insertion of such a catheter it is important to make the initial incision much higher than that usually used for cardiac catheterization in infants. In the young infant, in whom the bulk of these procedures is performed, the saphenous bulb is nearly at the level of the inguinal crease. The inguinal ligament lies cephalad to that crease by approximately 1 cm. Therefore, it is advisable to make the initial incision approxim-

ately 1 cm. cephalad to the inguinal crease. This will permit insertion of a No. 6·5 French catheter in almost all infants over 2·5 kg. in weight. It is sometimes useful to use a small mosquito haemostat with a very fine point to dilate the vein, after it has been incised, to facilitate initial introduction of the catheter tip.

Localization of balloon in left atrium

When a double-lumen catheter can be used – and this is always preferable to the use of a single-lumen catheter – presence of the balloon tip of the catheter in the left atrium can be verified by angiography or by sampling arterialized blood from a chamber with atrial pressure. If it is necessary to use a single-lumen catheter, left atrial position is verified by catheterization of the pulmonary vein, by measurement of pressure wave transmission through the partly inflated balloon (which can distinguish between atrial and ventricular pressure pulses), or by posterior position of the catheter tip at the upper left cardiac border.

Achieving adequate septostomy   Failures can occur either as a result of using inadequate or improper equipment or as a consequence of performing the procedural portion of the septostomy inadequately. Previous mention has been made of the need to use a balloon that is capable of dilating to a diameter of 1·0 to 1·5 cm. The volume required to produce this diameter is not significant. In most of the modern balloon-tipped catheters available the desired diameter can be achieved with 2 to 3 ml. of solution. In order to prevent gradual dilatation, without rupture, of the foramen ovale it is important to do the initial pullback with the balloon inflated to nearly 1·0 cm. in diameter. Subsequent pullbacks can then be done until the balloon inflated to 1·5 cm. diameter can be withdrawn from the left to the right atrium without detecting any resistance. The technique of the pullback is all-important. One must not be too gentle, since slow withdrawal of the balloon from the left to the right atrium is apt to produce dilatation of the foramen ovale without actual tearing of the fossa ovalis tissue. When the position in the left atrium has been verified, and the balloon has been inflated, a sharp rapid tug is made to jerk the balloon from the left atrium to the junction of the right atrium and inferior vena cava in a single motion. When that point has been reached the catheter is rapidly advanced cephalad. It is important to repeat this procedure several times until no further resistance is met on pullback, until a satisfactory arterial oxygen saturation is obtained, or

until the left atrial to right atrial 'a' wave pressure gradient has been abolished.

Procedural problems and their prevention   Damage to a pulmonary vein from balloon distension can be avoided by slow inflation of the balloon. This will result in gentle extrusion of the entire catheter tip back into the left atrial cavity. Injury to either the right or the left atrioventricular valves can be avoided by being absolutely certain that the catheter tip is not in either ventricle. Verification of left atrial positioning has been described above. There has been a report of injury to the inferior vena cava. It is suggested that such injury can be prevented by inflating the balloon adequately for each pullback, so that it cannot be pulled into the inferior vena cava. Moreover, the rapid caudad to cephalad movement tends to minimize this possibility. Rupture of the balloon is a rather common phenomenon. One must be extremely careful to remove all air bubbles from the balloon catheter to prevent air embolization. Though there has been a report of embolization by a piece of the rubber balloon the use of higher grade rubber material for the fabrication of these catheters has virtually abolished this hazard.

On occasion the distal balloon ties will come loose and the balloon will retract above the balloon lumen and be impossible to deflate. This can be managed by inserting a sharp stylet wire inside a very fine catheter to the inferior vena cava/right atrial junction. The wire tip then can be advanced just beyond the catheter, and the balloon tipped catheter pulled against the wire tip until the balloon ruptures. Then both catheter and stylet can be removed.[2]

Results of balloon atrioseptostomy

Forty-five patients with uncomplicated transposition of the great arteries, or with transposition of the great arteries and a haemodynamically significant ventricular septal defect, have had balloon atrioseptostomy. The survivors have been followed for a period of one year or more. The breakdown of the patient groups, the effective palliation rate, and the long-term effects are described in Table I.

The effective palliation rate is approximately 90 per cent, and the long-term survival rate approximately 80 per cent. The long-term survival rate does not include two patients who died after total surgical correction.

[2] A new balloon catheter system is being designed and tested to obviate the possibility of this complication occurring.

TABLE I *Effective palliation and long-term survival rate*

	No.	Effective palliation	Long-term survival
Uncomplicated	31	27	23
Ventricular septal defect	14	13	12

In 5 patients successful palliation was not achieved. Two of these failures occurred in critically ill infants with such profound acidosis and hypothermia at the time of the procedure that they did not recover. Necropsy confirmed that a large atrial septal defect had been made in both. One additional patient died of sepsis, probably present at the time of the procedure. One died of aspiration and pneumonia just before scheduled discharge from the hospital. Necropsy confirmed the presence of a large atrial septal defect. The fifth patient died owing to perforation of the right atrium by the balloon catheter. There were five late deaths in the series. One was due to congestive heart failure, although neither clinical nor necropsy follow-up was available. One died of pneumonia, and three had severe cerebral vascular accidents, probably secondary to sepsis and dehydration.

Post-balloon atrioseptostomy course

Table 2 summarizes the long-term follow-up on the surviving patients. There is some overlap in Table 2, since some of the patients fall into more than one of the listed categories.

TABLE 2 *Long-term follow-up*

Repeat balloon atrioseptostomy	4
Complete correction	22
Pulmonary artery banding	5
Aortico-pulmonic shunting	3
Stroke	5
Acquired pulmonic stenosis	6

Four of the patients required a repeat balloon atrioseptostomy. Twenty-two of these patients have now had total correction.[3] Twenty of them are alive. One death was due to contraction of the baffle that occurred approximately one year after correction. The other death, which occurred on the second postoperative day, was the only patient in the series with transposition of the great arteries, ventricular septal defect, and severe pulmonic stenosis. Five of the patients with ventricular septal defect required pulmonary artery banding. One of these was performed

[3] All performed by Dr. John A. Waldhausen at the Children's Hospital of Philadelphia.

during the initial hospital stay, and four at intervals from 12 to 18 months postseptostomy. Three of these have had total correction with debanding of the pulmonary artery, transatrial closure of the ventricular septal defect, and interatrial rebaffling of the Mustard type. Three of the patients required aorticopulmonary shunting. One of these was a patient who developed pulmonic stenosis with a systolic pressure gradient between the left ventricular and pulmonary artery of 70 mm. Hg. In infancy he had no gradient demonstrable across the left ventricular outflow tract. In the six patients in the group who developed some form of pulmonic stenosis, four of the gradients were between 15 and 25 mm. Hg systolic, and two were over 60 mm. Hg systolic. Five of the patients have had strokes.

Long-term effect of balloon atrioseptostomy on oxygen saturation

The mean preseptostomy arterial oxygen saturation in all patients was 41 per cent, with a range of 12 to 60 per cent. Immediately after septostomy the mean saturation was 64 per cent, with a range of 32 to 82 per cent. Some of the patients had extremely low saturations after septostomy. Subsequent studies showed that their saturations rose to acceptable levels within several days after septostomy. Therefore, when one is certain that septostomy has been performed adequately, failure to achieve a conspicuous increase in arterial saturation immediately after the procedure does not mean that the procedure was unsuccessful. Many of these patients will continue to improve for several days after septostomy. The patient with the postseptostomy saturation of 32 per cent improved to 75 per cent saturation 48 hours later. Just before total correction the mean arterial oxygen saturation in the entire group was 68 per cent, with a range of 60 to 89 per cent.

All patients who have had total surgical correction are clinically pink with arterial oxygen saturations ranging from 80 to 90 per cent. The mean arterial partial pressure of oxygen in this group, however, is 67 mm. Hg. The explanation for this continued desaturation is entirely conjectural. Probably a combination of persistent Thebesian vein drainage into the new 'left' atrium and the persistence of pulmonary arteriovenous communications are factors in this phenomenon.

References

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