Transposition of the Great Arteries

Results of Palliation by Balloon Atrioseptostomy in Thirty-one Infants

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SUMMARY

A nonsurgical method of creating atrial septal defects using a balloon catheter has been reviewed, and important procedural and technical details have been presented. The results of balloon atrioseptostomy in 31 infants with transposition of the great arteries have been reported and compared with those from the surgical creation of atrial septal defects. All 31 patients survived balloon atrioseptostomy without complications; 26 of them (84%) had effective immediate palliation, and 22 (71%) are long-term survivors. These results demonstrate that effective palliation of transposition of the great arteries, with or without associated ventricular septal defect, can be provided rapidly and safely by balloon atrioseptostomy. All patients with transposition and a large ventricular septal defect should have balloon atrioseptostomy and may also need pulmonary artery banding. For some of these patients banding may be postponed safely for many months, may occur spontaneously, or may be avoided completely.

Atrioseptostomy by the balloon catheter technique requires neither sedation nor general anesthesia, and obviates the need for thoracotomy and cardiectomy. The rapidity with which it can be performed makes it particularly advantageous for the desperately hypoxic newborn with transposition of the great arteries who may be considered too sick to withstand major surgery.

Additional Indexing Words:
Patent ductus arteriosus
Nonsurgical creation of atrial septal defects
Cyanotic congenital heart disease

Transposition (TGA) of the great arteries is a common and frequently lethal congenital cardiac malformation. Among 100 infants with congenital heart disease who died within the first month of life, Ober and Moore found that 27 had TGA. Nearly all untreated infants with this entity die before 6 months of age, and most of them before 3 months. In the past few years a variety of surgical procedures for correction of this defect have been devised. In patients beyond 1 year of age and without other associated surgical defects, correction of transposition has been attempted with a mortality of 17 to 29%, when the method perfected by Mustard and his associates has been used. However, since most patients with TGA die in infancy before definitive surgery is feasible, early palliation is mandatory. Several techniques have been effective in providing adequate mixing of the pulmonary and systemic circulations. In the past the approach used most often has been the surgical creation of an interatrial communication by the Blalock-Hanlon method or a modification of it. Successful palliation has also been accomplished by operations developed by Baffes and by Edwards and Barger.
In 1966 the authors described a nonsurgical method of creating an atrial septal defect for palliation of TGA, using a balloon-tipped intracardiac catheter. Since that time additional instances of successful palliation by this technique have been documented by others. The purpose of this report is to review the method of balloon atrioseptostomy (BAS), to present the results of this treatment in 31 infants with TGA, and to compare these results with those following surgical creation of atrial septal defects using thoracotomy.

Methods

From May 1965 to October 1967, thirty-two infants have been admitted to The Children's Hospital of Philadelphia with TGA (dextro) with or without a ventricular septal defect and a patent ductus arteriosus. One of these patients died before BAS could be performed. In each of the other 31 infants, who form the basis of this report, BAS was carried out as an integral part of the cardiac catheterization procedure immediately after the anatomic diagnosis was established. In 21 patients no other defects of hemodynamic significance were found at the time of the original diagnostic study, and this group is hereafter identified as "isolated" TGA (table 1). Among the remaining 10 patients, significant ventricular septal defects were present in seven, and large patent ductus arteriosus were demonstrated in three. Excluded from this report are patients with TGA in association with coarctation or atresia of the aorta, stenosis or atresia of intracardiac valves, single ventricle, or total anomalous drainage of systemic or pulmonary veins.

At the time of BAS the ages of the patients ranged from 4 hours to 3½ months (average, 28 days), and all but two weighed less than 9 pounds. All 31 infants had obvious physical and roentgenographic signs of congestive heart failure and at least moderate degrees of cyanosis. Among 18 with severe heart failure or life-threatening hypoxemia, 11 were moribund at the time of admission to the hospital. Cardiac catheterization and BAS were performed from a femoral vein with local anesthetization of the femoral area, without patient sedation. Diagnostic procedures were completed as rapidly as possible with careful monitoring and treatment of metabolic acidosis and hypothermia. Systemic arterial oxygen saturation was measured before and immediately after BAS in 20 of the 31 patients. In small infants the primary skin incision was made at, or slightly above, the level of the inguinal crease to expose the common femoral vein. After definitive diagnostic study, a double-lumen, balloon-tipped 6.5F catheter was introduced and advanced to the right atrium and across the patent foramen ovale to the left atrium or a pulmonary vein. The location of the catheter in the left atrium was verified by angiography, sampling of arterial blood from a chamber with atrial pressure, from a posterior position at the upper left cardiac border, or on entry into a pulmonary vein. The balloon was inflated with diluted radiopaque solution to a diameter of 1.0 to 1.5 cm and then rapidly withdrawn across the atrial septum with a sudden, sharp, short pull. During withdrawal the septum was displaced toward the inferior vena cava. Pullback was made

![Figure 1](attachment://image.png)

**Figure 1**

Double lumen 6.5F catheters with the balloon empty and inflated.

<table>
<thead>
<tr>
<th>Table 1</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Results of Balloon Atrioseptostomy in 31 Infants with Transposition of the Great Arteries</strong></td>
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<tr>
<td>Associated defects</td>
</tr>
<tr>
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<td>&quot;Isolated&quot;*</td>
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<tr>
<td>Ventricular septal defects</td>
</tr>
<tr>
<td>Patent ductus arteriosus</td>
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<tr>
<td>Total</td>
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</tbody>
</table>

*This category includes patients with small ventricular septal defects and patent ductus arteriosus of little or no hemodynamic significance.
TRANSPOSITION OF GREAT ARTERIES

Figure 2

Isolated, sequential frames of cineangiograms at 30 frames per second of a BAS illustrating the direction and rapidity of catheter withdrawal from left to right atrium.

to carry the balloon in a single motion from the left atrium to the junction of the inferior vena cava and right atrium. Then the balloon was advanced rapidly from the vena cava to the right atrial cavity and deflated. Using gradually increasing balloon volumes, BAS was repeated five to ten times until no resistance was felt during withdrawal of a balloon inflated to a diameter of 1.5 to 2.0 cm. Figure 1 illustrates the balloon catheters, and figure 2 shows cineangiograms of one septostomy.

Results

All 31 infants survived BAS without complications from the procedure itself. Transient arrhythmias during BAS were uncommon, and no patient had permanent abnormalities of conduction. Figure 3 illustrates the minor changes in the ECG and femoral arterial pressure during typical septostomy. Resting left atrial-to-right atrial pressure gradients were measured before and after BAS in 11 patients. A decrease in the a-wave gradient was recorded in nine of them and a reduction in mean pressure gradient was demonstrated in 10 (fig. 4). An increase in arterial oxygen saturation was found immediately after BAS. In 20 patients in whom saturation changes were measured (fig. 5), the mean increase was 23% (range, +7 to +42%). In 15 patients with “isolated” TGA, mean saturations of 41% (range, 12 to 86%) increased to 64% (range, 32 to 94%) after BAS. In five patients with TGA and large ventricular septal defects mean saturation of 53% (range, 40 to 76%) increased to
Figure 3

(Upper tracing) Electrocardiogram of one patient during a BAS. (Lower tracing) Arterial blood pressure record of another patient during BAS.

Figure 4

Changes in the interatrial pressure gradients (LA/RA). The left column shows the a-wave changes, and the right column the mean changes in 11 patients with TGA before, and immediately after, BAS.

Figure 5

Systemic arterial oxygen saturations before, and immediately after, BAS in 15 patients with "isolated" TGA (left column), and in five patients with TGA and ventricular septal defects (right column).

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74% (range, 62 to 92%). In each of the five infants with oxygen saturations below 50% immediately after BAS, further reduction in cyanosis was evident 1 to 3 days later. In one a saturation of 32% after the procedure increased to 75% within 2 days. Twenty-six of the 31 patients (84%) had clinical evidence of marked reduction in cyanosis and elimination of congestive heart failure after BAS, and these infants were discharged from the hospital within 4 to 14 days. Seventeen (81%) of the 21 patients with “isolated” TGA and five (71%) of the seven with associated ventricular septal defects are long-term survivors (table 1). These 22 living patients have been followed for 8 to 33 months after BAS (fig. 6). Each has only mild or moderate cyanosis. None demonstrates clinical or roentgenographic signs of congestive heart failure (fig. 7); 12 receive maintenance doses of digoxin. Strokes have occurred in two survivors during episodes of severe gastroenteritis with fever and dehydration.

A second BAS was performed in three patients with “isolated” TGA at 1½, 2, and 5 months after the first procedure. In each patient the development of cardiomegaly, plus some increase in cyanosis, prompted the second diagnostic catheterization, which confirmed the need for greater interatrial mixing. After the second BAS each patient had prompt and sustained diminution in
cyanosis and elimination of the signs of heart failure.

Nine patients have died (table 1). Early deaths, before discharge from the hospital, occurred in five infants. Four of them when admitted were moribund with severe congestive heart failure, intense cyanosis, and marked metabolic acidosis and hypothermia. Emergency BAS, performed within 1 to 12 hours after admission, resulted in significant improvement in cyanosis in each one, but persistent heart failure and acidosis resulted in their deaths from 2 to 24 hours later. Postmortem examinations demonstrated large associated patent ductus arteriosus in three and isolated TGA in one. In a fifth infant with an accompanying ventricular septal defect, BAS produced moderate improvement in severe congestive heart failure and reduction in cyanosis for 9 days. Death occurred on the tenth day from acute pulmonary edema before pulmonary artery banding could be accomplished. Postmortem examination of this infant demonstrated a large ventricular septal defect of the endocardial cushion variety with deep posterior extension.

Late deaths occurred in four patients. One infant with an associated ventricular septal defect died 2 months after BAS, and postmortem examination revealed a large cerebellar cyst. The death of another with isolated TGA, 1 day after a second BAS, could not be explained by thorough postmortem studies. Autopsy was not performed on two other patients, each with "isolated" TGA. One died from severe diarrhea and dehydration 8 months after effective BAS. The other died 2 months after a second BAS had resulted in significant diminution in cyanosis and disappearance of the signs of heart failure. In all seven of the autopsied cases, a large interatrial communication was found.

Discussion

These results indicate that BAS is a safe and effective means of palliation of severe congestive heart failure and hypoxemia from TGA. None of these patients died as a result of the procedure and none experienced complications from it. Because of the rapidity with which it can be performed, it is particularly applicable as an emergency procedure for those infants with "isolated" TGA who develop critical hypoxemia and heart failure soon after birth. In this series BAS resulted in effective long-term palliation in 81% of such infants.

Even when a large ventricular septal defect is present, BAS alone can provide effective palliation throughout infancy, as demonstrated by five of the seven such patients in this study. In this group of patients, banding of the pulmonary artery will usually be necessary to prevent the development of pulmonary vascular obstruction, but in most instances it may be deferred for 6 to 12 months. Postponement of banding beyond early infancy is not justified, however, if the signs of congestive heart failure are not eliminated within a week after BAS. Evaluation of the optimal time for banding may require serial cardiac catheterizations to measure pulmonary arterial pressure, flow, and resistance.

Patency of the ductus arteriosus is relatively common in infants with TGA, but it is rarely of hemodynamic significance. However, in those patients with TGA and patent ductus arteriosus in whom aortography illustrates a large systemic-to-pulmonary arterial shunt, an ominous situation may exist. In this series, each of three infants with this degree of ductal patency developed severe congestive heart failure at an early age and died within hours after emergency BAS. In patients such as these, if congestive failure persists after BAS has provided adequate mixing, the ductus should be ligated.

The overall long-term mortality in these 31 patients was 28%, and in those with "isolated" TGA it was 19%. Among the nine infants who died, four were moribund before BAS, and two died of serious, noncardiac abnormalities. From three large medical centers where surgical creation of atrial defects has been used for palliation of TGA, mortality rates of 30 to 57% have been reported.13, 15, 26, 27 The number of infants with TGA who die before surgical palliation can be accomplished is generally not known. But
a recent report from one center indicates that among 52 infants hospitalized with TGA, 22 (42%) died before surgery. It is important to emphasize that since 1965, when BAS was first introduced, only one infant with TGA, with or without ventricular septal defect or patent ductus arteriosus, has died at this institution before this nonsurgical method of palliation could be performed.

Irrespective of the method of creating the atrial defect, further reduction in the mortality from TGA to levels below 20% will depend primarily on the earlier referral of cyanotic infants, especially newborns, to major treatment centers. These infants, particularly if they show signs of congestive heart failure, should be handled as emergencies. The details of general management before and during cardiac catheterization are critical. Effective digitalization and prompt diuresis are imperative if time permits. Hypothermia and metabolic acidosis from severe hypoxemia and heart failure are ominous signs for which high oxygen environment, effective body warming, and prompt administration of intravenous buffers are indicated. However, these temporary measures become increasingly ineffective, and a rapidly executed diagnostic cardiac catheterization, followed by adequate septostomy, is mandatory. A dramatic diminution in the signs of respiratory distress is usually seen after the first catheter septostomy. This immediate improvement is probably the result of decompression of the left atrium with reduction in pulmonary venous hypertension and intrapulmonary-left heart blood volume. Left atrial-to-right atrial pressure gradients are decreased by BAS (fig. 8). Measurement of this reduction, particularly the left atrial a-wave pressure, is a useful guide to the adequacy of septostomy, as described by Muster and his associates.

All patients with TGA require careful management after successful palliation, whether by surgical or nonsurgical means. Many continue to demonstrate moderate cyanosis. All of the patients in this study showed an early increase in systemic arterial oxygen saturation. In the 20 in whom the change was measured, a mean increase of 23% was found immediately after BAS. The increase in systemic arterial oxygen saturation after BAS is comparable to that found after surgical creation of atrial septal defects. The increase in systemic arterial oxygen saturation several months after Blalock-Hanlon operation has recently been reported in 27 patients with TGA. The mean change was 18%; seven of the 27 had values below, or equal to, those before surgical palliation, and 24 had saturations of 75% or less. Regardless of the degree of palliation, all patients with TGA remain highly susceptible to infection and subject to the risk of cerebrovascular thrombosis from paradoxical embolus or episodes of dehydration.

Following palliation, a moderate increase in cyanosis alone is not sufficient indication for restudy. Progressive cardiomegaly is the

Figure 8
Records of interatrial pressure gradients measured before and immediately after BAS in a patient with isolated TGA.
most reliable sign of inadequate mixing, and its development should lead to repeat cardiac catheterization to evaluate the need for further enlargement of the interatrial communication. If a second BAS, performed in conjunction with recatheterization, does not result in significant diminution in cyanosis and in the signs of heart failure within 1 or 2 weeks, surgical enlargement of the atrial defect seems justified in these older and more stable patients. Although none of our patients has required surgical septostomy, three have undergone a second BAS.

Several procedural details deserve emphasis. Although inability to insert the balloon catheter into the femoral vein has been reported to us, we have not encountered that problem. The skin incision must be made high enough to obtain a vein of large diameter. Gentle dilatation of the vein by a vessel dilator or smooth-tipped hemostat is often helpful. If the vein is too small to accept a 6.5F catheter, double-lumen 5F and 6F catheters or single-lumen 4F to 6F catheters are available and may be used. Catheter withdrawal from either ventricle can produce serious damage to the atrioventricular valves. When a double-lumen catheter is being used, this hazard is easily prevented by a combination of the methods described earlier to detect that the catheter tip is in a ventricle. If a single-lumen catheter is necessary, left atrial position is verified by catheterization of a pulmonary vein or by measurement of gross pressure through a partly inflated balloon, which distinguishes between atrial and ventricular pressures. Damage to a pulmonary vein from balloon distention is avoided by slow inflation which results in gentle extrusion of the entire tip into the left atrial cavity. The correct placement of the balloon in the left atrium has not been a problem when one or more of these methods of identification has been used.

Inadequate enlargement of the interatrial opening may result from improper pullback technique or inadequate balloon diameter. The valve of the foramen ovale may be stretched, but not ruptured, if the catheter is withdrawn slowly, or if the inflated balloon has a small diameter or is too long. Rapid pullback, using a short, wide balloon, should overcome this difficulty.

During the development of satisfactory balloon catheters, several design problems have been encountered: namely, balloons with inadequate diameters or with long sausage shapes, catheters with redundant balloons, too large to permit insertion, and inadequate balloon ties or thin-walled balloons, resulting in easy rupture. All of these equipment deficiencies have been eliminated or minimized in the commercially produced balloon catheters presently available. Sterilization by heat or gas damages both the balloon and the tie, and only chemical sterilization should be employed.

BAS has been used, alone or in conjunction with surgical procedures, in the palliative treatment of infants with a variety of other congenital cardiac defects: tricuspid atresia, total anomalous pulmonary venous drainage, pulmonary atresia with intact ventricular septum, mitral atresia, and critical Ebstein's malformation. Moreover, it has been successfully performed in several older children: a 2-year-old child and a 6-year-old child with TGA and pulmonic stenosis; a 3-year-old child

*United States Catheter & Instrument Corporation, Glens Falls, N. Y.

Figure 9
In situ photograph taken at time of Mustard correction, demonstrating the atrial septal defect produced by BAS 2 years earlier. The defect is 1.5 by 1.0 cm.
and an 8-year-old child with TGA, ventricular septal defect, and pulmonary vascular obstruction; and a 12-year-old patient with tricuspid atresia and a Potts' anastomosis. In all patients who have had BAS, an adequate interatrial communication has been produced, as verified either by clinical survival, surgical inspection (fig. 9), or postmortem examination.

There are several ancillary, but important, advantages of this technique. General anesthesia is avoided, and in infants even sedation is not indicated. Thoracotomy and cardiectomy are obviated, thereby eliminating the difficulties of adhesions at the time of subsequent surgery and providing adequate, unscarred pericardial tissue for use as the corrective baffle for the Mustard procedure. Patients treated by BAS are returned to the general medical ward and do not require specialized or intensive postoperative care.

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