Blade Atrial Septostomy: Collaborative Study

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SUMMARY During the past 4 years, five institutions have collaborated in evaluating the efficacy of blade atrial septostomy. The procedure was performed in 52 patients, including 31 with transposition of the great arteries, 10 with mitral atresia, five with tricuspid atresia and six with miscellaneous anomalies. The patient’s ages ranged from 1 day to 12 years (mean 13 months). Improvement occurred in 41 of 52 patients (79%). Four patients had an intact interatrial septum, and blade atrial septostomy was successfully performed by a transseptal technique. One patient died from a lacerated left atrial wall; other complications occurred in four patients. Blade atrial septostomy is an effective palliative procedure, even when the interatrial septum is thickened or intact.

IN PATIENTS with certain varieties of congenital heart malformations, an adequate interatrial opening (IAO) is essential for survival. An IAO is important in transposition of the great arteries (TGA), as well as for obligatory interatrial shunt lesions such as mitral atresia, tricuspid atresia and total anomalous pulmonary venous return. An adequate IAO is hemodynamically beneficial to patients with double-outlet right ventricle and a restrictive ventricular septal defect (VSD) and those with a univentricular heart and a unilateral restrictive atrioventricular valve. Balloon atrial septostomy, introduced by Rashkind and Miller in 1966,1 was the first innovative nonsurgical therapeutic modality in pediatric cardiac catheterization. Palliation has been achieved in the majority of patients who have undergone this procedure. However, in some patients, particularly older infants and children with a thickened interatrial septum, this procedure is usually not successful. In patients with an inadequate IAO after balloon atrial septostomy in infancy, the interatrial septum is often thickened, and a repeat balloon atrial septostomy is often ineffective.2

In 1973, a catheter with a built-in surgical blade was developed to enlarge the IAO in such patients. Experiments in animals substantiated its efficacy and safety,3 and clinical use of this technique has been successful in a limited number of patients.4 To extend the clinical trial of the technique, a collaborative study was organized in 1977.

In this report, we review the overall result of a collaborative study of blade atrial septostomy. The procedure is described in detail because the catheter

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TABLE 1. Participating Institutions and Principal Investigators

<table>
<thead>
<tr>
<th>Participating institution</th>
<th>Principal investigator</th>
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</thead>
<tbody>
<tr>
<td>Children's Hospital, Pittsburgh, Pennsylvania</td>
<td>S.C. Park</td>
</tr>
<tr>
<td>Texas Children's Hospital, Houston, Texas</td>
<td>C.E. Mullins</td>
</tr>
<tr>
<td>Indiana Univ. Hospital, Indianapolis, Indiana</td>
<td>D.A. Girod</td>
</tr>
<tr>
<td>Hospital for Sick Children, Toronto, Canada</td>
<td>P.M. Olley</td>
</tr>
<tr>
<td>Bakulev Institute, Moscow, USSR</td>
<td>G. Falkowski, V.A. Garibjan</td>
</tr>
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</table>

and the technique have been modified since the initial report.4

Materials and Methods

Five institutions participated in the clinical evaluation of the blade atrial septostomy catheter (table 1). Each investigator obtained appropriate approval from the respective institutional review board. Informed consent was obtained from each patient's parents before blade atrial septostomy.

Each patient selected for the procedure had a cardiac anomaly in which a sizeable IAO would be beneficial. Group 1 included 30 patients in whom balloon atrial septostomy did not create an adequate IAO; group 2 included 10 patients older than 1 month of age in whom the interatrial septum was expected to be too thick for effective balloon atrial septostomy; and group 3 included four patients with an intact interatrial septum.

Fifty-two patients were included in this study (table 2). The most common cardiac anomaly was TGA (31 patients). Of the TGA patients, 20 had no associated cardiac lesion or only a small VSD, while 11 patients had associated cardiac lesions. Of these 11 patients, seven had a large VSD (two of whom had pulmonary artery banding in early infancy), two had both a VSD and pulmonary stenosis, and one patient each had either pulmonic stenosis or a large patent ductus arteriosus. Other common indications for blade septostomy were mitral atresia complex (10 patients) and tricuspid atresia (five patients). A miscellaneous group included two patients with double-outlet right ventricle and a restrictive VSD, two patients with pulmonary valve atresia and an intact ventricular septum, one patient with total anomalous pulmonary venous return to the coronary sinus and one patient with Taussig-Bing anomaly and inadequate interatrial mixing.

The patients were 1 day to 12 years old (mean 13 months). Two-thirds of the patients were younger than 6 months of age at the time of the procedure; eight (15%) were 1 year of age or older and three were 5 years of age or older.

This study was designed to evaluate the immediate result of blade atrial septostomy; follow-up information is available in only 38 of the 52 patients (73%). The follow-up period ranged from 2 months to 4 years.

Blade Atrial Septostomy Catheter

The catheter is 65 cm long and is made of #6F radiopaque polyethylene tubing (Cook Incorporated). The catheter tip consists of a 3.5-cm section of stainless steel tubing with a 2.5-cm slit on its long axis. The metal tubing contains a small blade that is linked to a lever whose distal portion pivots at the catheter tip. The proximal portion of the blade is linked to a solid guide wire that passes through the entire catheter and exits at the hub. Advancing the wire extends the blade and the lever through the slit to form a triangle (fig. 1). There is a Y connector at the proximal portion of the catheter; one side branch is for fluid infusion and pressure measurement and the other for the wire that controls the blade.

Preparation

The procedure was performed by cutdown or percutaneous technique through either the saphenous or femoral vein. Biplane or movable "C" arm image intensifiers for rapid access to lateral fluoroscopy without moving the patient were used. The patients in this study usually received standard premedication for cardiac catheterization, but no patient received general anesthesia. When the procedure was done percutaneously, it was necessary to use a sheath that was large enough to accommodate either the blade septostomy or the balloon septostomy catheter. Although the blade septostomy catheter is #6F, a #7F introducer should be used because most balloon septostomy catheters cannot be introduced through a smaller sheath. A special sheath with a device to control back-bleeding is recommended to prevent blood loss due to mismatch between the size of the introducer and the smaller size

TABLE 2. Type of Cardiac Anomaly and Age Distribution of Patients

<table>
<thead>
<tr>
<th>Anomaly</th>
<th>No. of pts</th>
<th>Age distribution</th>
<th>Age range</th>
<th>Mean (months)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>&lt; 1 mo</td>
<td>1-3 mos</td>
<td>4-6 mos</td>
</tr>
<tr>
<td>TGA</td>
<td>31 (60%)</td>
<td>6</td>
<td>10</td>
<td>9</td>
</tr>
<tr>
<td>Mitral atresia</td>
<td>10 (19%)</td>
<td>1</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>Tricuspid atresia</td>
<td>5 (10%)</td>
<td>0</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>Miscellaneous</td>
<td>6 (11%)</td>
<td>1</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>52 (100%)</td>
<td>8</td>
<td>14</td>
<td>13</td>
</tr>
</tbody>
</table>

Abbreviation: TGA = transposition of the great arteries.
catheter during the procedure. Selective left atrial angiography is recommended before blade septostomy, to evaluate the size of the left atrium and the location of the IAO, particularly when the catheter passage across the IAO is unusual. In patients with mitral atresia, angiographic evaluation of left atrial size is essential because the left atrium is often small in infants.

Prerequisites

Because blade atrial septostomy is performed from the femoral vein approach, bilaterally thrombosed femoral or iliac veins, an absent hepatic segment of the inferior vena cava, or other conditions that prohibit the femoral approach preclude using this technique. A superior vena caval approach has not been attempted. The maximal height of the extended blade is 12–13 mm. Thus, when the diameter of the IAO is greater than 10 mm, it is unlikely that the blade could effectively engage the interatrial septum and the procedure is not recommended. To estimate the size of the IAO, a balloon catheter is inflated in the left atrium to a diameter larger than that of the IAO and it is gently pulled against the interatrial septum. The balloon is then gradually deflated until it passes through the defect. The passage of the balloon through the IAO is recorded cineangiographically at approximately 30 frames/sec. A calibrated measuring grid is recorded at the level of the right atrium to calculate the magnification factor. Patients with an IAO 8–10 mm in diameter are in the borderline group. In patients with a borderline-sized IAO, repeated withdrawal with variable angulation of the blade across the IAO successfully incises the atrial septum.

Procedure

1. The catheter system is checked before introduction (fig. 1). The locking device (D) is loosened from the wire holder (E) and the locking device and wire holder are moved toward the gasket (B) so that no gap is present between the locking device (D) and the gasket (B). The locking device is then tightened to prevent inadvertent protrusion of the blade during the manipulation of the catheter. The flexible plastic tubing is connected between the port (C) and a pressure transducer or an i.v. infusion line. The catheter system is flushed with heparinized solution.

2. The catheter is introduced into the left atrium (fig. 2-1). Because the side arm (C) is in the same plane as the curve of the catheter tip, passage of the catheter into the left atrium can be facilitated by maintaining the side arm in a posterior and leftward orientation. The location of the catheter in the left atrium is confirmed by lateral fluoroscopic examination and pressure measurement.

3. After the position of the catheter tip in the left atrium is confirmed, the locking device (D) is loosened from the control wire (F) and pulled backward until the gap between the gasket (B) and the locking device (D) is 12 mm; the holder (E) is then tightened. Next, the blade is extended by advancing the blade control wire holder (E) gently toward the catheter tip under fluoroscopic control (fig. 2-2). If resistance is met or the blade cannot be fully extended, it should be suspected that the catheter tip is positioned in the left atrial appendage or in a pulmonary vein. In this situation, the control wire holder (E) is withdrawn to fold the blade back into the catheter. Then the entire catheter system
is slightly withdrawn and the same maneuver repeated. Once the blade has been extended, the gasket (B) and the locking device (D) are held together with the thumb and index finger. The catheter is then slightly rotated counterclockwise until the blade is facing somewhat anteriorly as seen by lateral fluoroscopy (fig. 2–3).

(4) The entire catheter system is slowly withdrawn to the right atrium using both hands to maintain the same catheter orientation (fig. 2–4). Resistance of the interatrial septum is usually encountered in the middle or lower portion of the cardiac silhouette. Gentle but firm force is maintained to withdraw the catheter from the left atrium to the right atrium until a sudden decrease in resistance is felt. Continued resistance may be felt despite withdrawal of the catheter to the level of the diaphragm or even lower, especially if the left atrium is large or if the interatrial septum is quite stiff. Under no circumstances should rapid withdrawal be attempted as required for balloon atrial septostomy.

(5) Once the blade septostomy catheter has been withdrawn across the interatrial septum, the catheter is advanced to the middle right atrial position and the blade is folded back into the catheter lumen by withdrawing the locking device (D) and blade control wire holder (E). Although one passage of the blade across the interatrial septum usually suffices, the procedure may be repeated if little resistance was encountered during initial withdrawal. The angle of the blade should be changed slightly on subsequent withdrawals to ensure adequate incision of the interatrial septum. When the interatrial septum is unusually thick and the IAO is very small (less than 4 mm in diameter or in a transseptal approach) the first withdrawal of the catheter is done with only a partially extended blade. This is followed by withdrawal with a fully extended blade. This stepwise manner facilitates initial withdrawal of the blade across the interatrial septum and causes less stress on the delicate blade assembly.

(6) Using a Rashkind septostomy catheter (USCI) or a Fogarty septostomy catheter (Edwards Laboratories), balloon atrial septostomy is performed to further enlarge the IAO.

Blade atrial septostomy has been successfully performed with a transseptal technique in four patients with an intact interatrial septum. A long percutaneous transseptal sheath (Mullins transseptal catheter introducer, USCI) is used to introduce the blade catheter into the left atrium. Initially, the long sheath is passed into the left atrium with a Brockenbrough needle and the Mullins introducer set. The introducer catheter is replaced with the blade catheter and later with a balloon catheter. The long introducer sheath has also been successfully used in eight patients with a restrictive interatrial communication in whom it was difficult to maneuver the stiff blade septostomy catheter into the left atrium. The long sheath is initially introduced into the left atrium over a regular catheter, which can be more easily maneuvered. Once the sheath is properly positioned in the left atrium, the catheter is replaced with the blade catheter. Before the blade septostomy procedure, the tip of the sheath is withdrawn to the level of the middle inferior vena cava. When the blade catheter is withdrawn for septostomy, the sheath must be withdrawn simultaneously to prevent possible cutting of the sheath.

Assessment of the Procedure

To assess the effectiveness of the technique objectively, the following data were obtained before and after the procedure: (1) phasic and mean pressure measurement during catheter withdrawal from the left to the right atrium; (2) systemic arterial oxygen saturation; and (3) the estimated diameter of the IAO as described above. The actual diameter of the IAO was measured with a ruler at open heart surgery or a postmortem examination. If the IAO was oval or elliptical, maximal and minimal diameters were obtained, but only the maximal diameter was used.

Criteria for Improvement

Criteria for a successful blade atrial septostomy were arbitrarily established depending on the type of cardiac lesion (table 3). Equalization or reduction of pressure gradient between the two atria and measured size of the IAO were essential variables. In the TGA group, a change in systemic oxygen saturation was the most important indicator of success. Therefore, some patients had successful enlargement of the IAO (over 1 cm in diameter) and complete abolition of the inter-

<table>
<thead>
<tr>
<th>Indication</th>
<th>Systemic oxygen saturation</th>
<th>Interatrial pressure gradient</th>
</tr>
</thead>
<tbody>
<tr>
<td>Interal atrial mixing</td>
<td>&gt; 55%</td>
<td>&lt; 2 mm Hg (LA &gt; RA)</td>
</tr>
<tr>
<td>TGA</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Taussig-Bing anomaly</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Interal L→R shunt</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Obligatory</td>
<td>NA</td>
<td>&lt; 10 mm Hg &gt; 10 mm (LA &gt; RA)</td>
</tr>
<tr>
<td>Mitral atresia</td>
<td>NA</td>
<td>&lt; 5 mm Hg &gt; 10 mm (LA &gt; RA)</td>
</tr>
<tr>
<td>Facilitative</td>
<td></td>
<td></td>
</tr>
<tr>
<td>DORV with restrictive VSD</td>
<td>NA</td>
<td></td>
</tr>
<tr>
<td>Univentricle with restrictive left AV valve</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Interal R→L shunt</td>
<td>NA</td>
<td>&lt; 2 mm Hg &gt; 10 mm (RA &gt; LA)</td>
</tr>
<tr>
<td>Tricuspid atresia</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pulmonary valve atresia</td>
<td></td>
<td></td>
</tr>
<tr>
<td>TAVR</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Abbreviations: DORV = double-outlet right ventricle; IAO = interatrial opening measured by balloon or angiography; NA = not applied; TAVR = total anomalous pulmonary venous return; TGA = transposition of the great arteries; VSD = ventricular septal defect; LA = left atrium; RA = right atrium; AV = atrioventricular R→L = right to left; L→R = left to right.
atrial pressure gradient, but if the systemic oxygen saturation did not increase to more than 55%, they were regarded as having had no improvement. On the other hand, some patients with TGA and VSD had systemic oxygen saturations greater than 55% before the procedure. In these patients the systemic oxygen saturation may not improve. However, if the IAO was enlarged by more than 10 mm and the interatrial pressure gradient was abolished or reduced to less than 2 mm Hg, these patients were regarded as having improvement, even though the systemic oxygen saturation did not increase.

In patients with left-sided obstructive lesions, left atrial hypertension is usually present, particularly when pulmonary blood flow is increased. Since mitral atresia necessitates an obligatory interatrial left-to-right shunt, a left-to-right atrial pressure gradient may persist even with an adequate IAO. Therefore, the combination of an interatrial pressure gradient less than 10 mm Hg and an IAO larger than 10 mm was regarded as an improvement. In other relative left-sided obstructive lesions (e.g., double-outlet right ventricle with a restrictive VSD), an interatrial pressure gradient less than 5 mm Hg and an IAO larger than 10 mm were considered an improvement.

In contrast, in patients with interatrial right-to-left shunt lesions (e.g., tricuspid atresia, pulmonary valve atresia and total anomalous pulmonary venous return), right atrial pressure equals or exceeds left atrial pressure. In these lesions, a residual interatrial pressure gradient of less than 2 mm Hg and an adequate IAO larger than 10 mm were considered an improvement.

Statistical analysis of the results was performed using the t test for paired observations. All values are expressed as mean ± sd.

Results

Transposition of the Great Arteries

Patients with TGA were the largest group to undergo blade atrial septostomy (31 patients [60%]). Twenty-five patients (81%) were younger than 6 months of age at the time of the procedure. Four patients, ages 1–18 days, did not improve after balloon atrial septostomy and therefore underwent blade atrial septostomy. The remaining 27 patients had previously undergone balloon atrial septostomy but had become more cyanotic and underwent repeat cardiac catheterization. Two patients, ages 4 months and 3 years, had an intact interatrial septum, and blade atrial septostomy was successfully performed by the transseptal technique.

Overall changes in the interatrial pressure gradient, oxygen saturation and size of the interatrial opening before and after the procedure are summarized in figure 3. Although overall improvements in those variables were statistically significant in each category, only 23 of the 31 patients (74%) met the criteria for improvement after the procedure. The mean systemic arterial oxygen saturation increased from 47.3 ± 15.9% to 63.2 ± 13.1% (p < 0.001). The interatrial pressure gradient was reduced from 3.1 ± 2.7 mm Hg to 0.6 ± 1.3 mm Hg (p < 0.001) and the IAO increased from 6.4 ± 1.6 mm to 12.1 ± 2.2 mm (p < 0.001). In one patient, the right ventricular outflow tract was perforated by the blade catheter tip. The remaining seven patients did not improve. Four of these seven patients had a borderline-size IAO and the blade did not engage the interatrial septum. (Two underwent Blalock-Hanlon surgical septostomy 1 day after the procedure, with one surgical mortality. Another two had spontaneous improvement in arterial oxygen saturation after the procedure and required no further treatment.) The remaining three patients had no improvement in systemic arterial oxygen saturation despite apparently successful enlargement of the IAO.

The third patient subsequently underwent early Mustard operation 8 days after the procedure. After blade atrial septostomy, two additional patients improved at first but then deteriorated and underwent Mustard operation 14 days and 6 weeks after the procedure. Sizeable interatrial defects measuring 12–15 mm were found at operation. These patients apparently had inadequate interatrial mixing despite the presence of an adequate IAO.

One patient died of a gastrointestinal disorder and possible digitalis toxicity 2 weeks after the procedure. At autopsy, a 10 mm cut in the interatrial septum was found.

Fifteen patients subsequently underwent elective Mustard operation 6–16 months (mean 11 months) after blade atrial septostomy. At operation, 11 of these 15 patients had an IAO greater than 8 mm in diameter. The remaining three patients had a restrictive IAO less than 7 mm in diameter.

Mitral Atresia Complex

Blade atrial septostomy was performed in 10 patients with mitral atresia complex and a restrictive IAO. Their ages ranged from 1 month to 12 years, and six were younger than 1 year of age. One patient had undergone balloon atrial septostomy, but the IAO had become inadequate.

Seven of 10 patients improved promptly after blade atrial septostomy. Systemic oxygen saturation did not change significantly after the procedure (fig. 4). The mean pressure gradient between right atrium and left atrium was 14–33 mm Hg (average 24 mm Hg) before the procedure in these seven patients. After blade atrial septostomy, the mean gradient decreased significantly, to 3–9 mm Hg (average 5 mm Hg). The mean IAO increased from 5.6 mm to 14.2 mm (p < 0.001). In two of the remaining three patients, ages 1 month and 4 months, the blade could not be fully extended because the left atrium was exceedingly small (10–13 mm in maximal transverse diameter on the angiocardigram.) One of these two patients underwent open surgical septectomy 8 days after the procedure and a 3-mm IAO was found. The other patient died suddenly while awaiting operation; an IAO less than 2 mm and a thickened interatrial septum were found at postmortem...
examination. The last patient, a 12-year-old, suffered a laceration of the left atrial wall during the procedure. Of the seven patients who improved initially, one patient died of sepsis 2 months after the procedure. At postmortem examination, a 12 × 14-mm atrial septal defect was found. Three patients who had the procedure at 3, 3.5 and 9 years of age are doing well 3–4 years later. One of these patients subsequently had a cardiac catheterization, which confirmed the adequacy of the IAO. Three other patients who underwent the procedure in early infancy were found to have a restrictive IAO. These three patients underwent open surgical septectomy 5, 14 and 18 months after the blade septostomy and were found to have 6–7-mm IAOs.

Tricuspid Atresia Complex

All five patients with tricuspid atresia had hemodynamic improvement from the procedure. They were 2–9 months old (mean 7 months) at the time of the procedure. The mean interatrial pressure gradient was 5–9 mm Hg before the procedure and 0–2 mm Hg afterward ($p < 0.001$) (fig. 4). The IAO increased significantly after the procedure, but the change in systemic oxygen saturation was not significant.

Of five patients with tricuspid atresia, four are doing well 2–24 months (mean 13 months) after the procedure. Another patient with tricuspid atresia died at operation for pulmonary artery banding 2 days after blade septostomy. At postmortem examination there was a large IAO (15 mm in diameter).

Miscellaneous Group

Two patients, ages 15 months and 7 years, had double-outlet right ventricle and a restrictive VSD. Left ventricular systolic and end-diastolic pressures were elevated, as was left atrial pressure. The interatrial pressure gradients in these two patients were 14 and 22 mm Hg, respectively. Transseptal blade atrial septostomy was successfully performed using the long percutaneous sheath. The interatrial gradient was reduced to 5 mm Hg in each patient and both also had symptomatic improvement after the procedure (fig. 4). Two patients with pulmonary valve atresia, an intact ventricular septum and a restrictive IAO underwent blade atrial septostomy at 4 days and 12 months of age. Each improved after enlargement of the IAO. A 3-month-old infant with total anomalous pulmonary venous return to the coronary sinus was found to have severe congestive failure and pulmonary hypertension in association with a restrictive IAO. After blade septostomy, the interatrial pressure gradient was completely abolished and there was striking clinical improvement.
The last patient in this group was a 5-month-old infant with the Taussig-Bing complex and a restrictive IAO. Blade atrial septostomy enlarged the interatrial communication from 3 mm to 12 mm in diameter and resulted in clinical improvement as well.

Complications

Major complications occurred in five patients (table 4). One patient died, a 12-year-old boy with mitral atresia complex and a restrictive IAO. His IAO was located in an unusually superior and posterior position and it was very difficult to maneuver even a standard catheter into the left atrium. The procedure resulted in a laceration of the left atrial posterior wall and massive hemopericardium. Emergency surgical repair of the laceration and a surgical septostomy were performed. The patient died of neurologic sequelae.

A 3-month-old patient with TGA underwent blade atrial septostomy because of severe hypoxemia. Attempts to place the blade catheter in the left atrium resulted in a perforation of the right ventricular outflow tract by the catheter tip; the perforation was confirmed at operation. Since single-plane fluoroscopy with a moveable “C” arm was being used, the position of the catheter tip in the lateral view could not be constantly monitored. A Blalock-Hanlon atrial septectomy was done, followed 8 months later by a Mustard operation. This patient had a neurologic insult at the time of the complication and some neurologic deficit persists.

Three other patients with TGA had neurologic problems after blade atrial septostomy. Two patients had generalized seizures within 12 hours and required anticonvulsant medication. Another patient developed hemiparesis, presumably due to a cerebrovascular accident, shortly after the procedure. These three patients have improved, and follow-up examination showed no permanent sequelae.

Four patients had excessive blood loss during the procedure and required blood transfusion. The blood loss was mostly related to leakage of blood between

<table>
<thead>
<tr>
<th>Anomaly</th>
<th>No. of procedures</th>
<th>Improvement</th>
<th>Complications</th>
</tr>
</thead>
<tbody>
<tr>
<td>TGA</td>
<td>31</td>
<td>23 (74%)</td>
<td>RVOT perforation (n = 1)</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>CNS insult (n = 3)</td>
</tr>
<tr>
<td>Mitral atresia</td>
<td>10</td>
<td>7 (70%)</td>
<td>LA laceration (n = 1)</td>
</tr>
<tr>
<td>Tricuspid atresia</td>
<td>5</td>
<td>5 (100%)</td>
<td>None</td>
</tr>
<tr>
<td>Miscellaneous</td>
<td>6</td>
<td>6 (100%)</td>
<td>None</td>
</tr>
<tr>
<td>Total</td>
<td>52</td>
<td>41 (79%)</td>
<td>5 (10%)</td>
</tr>
</tbody>
</table>

Abbreviations: CNS = central nervous system; LA = left atrium; RVOT = right ventricular outflow; TGA = transposition of the great arteries.
the percutaneous sheath and the smaller balloon septostomy catheter.

Discussion

This collaborative study has shown blade atrial septostomy to be a relatively safe and effective procedure compared with surgical atrial septectomy. However, some serious complications have occurred. Most patients who undergo this procedure are cyanotic or clinically unstable, and are therefore potentially prone to certain complications. An insult to the central nervous system after routine cardiac catheterization in cyanotic patients is known to occur. Therefore, the blade atrial septostomy procedure may not be the sole reason for such complication.

Blood loss has not been a problem since the introduction of special sheaths with a bleeding control device. These devices also minimize the risk of air embolism, which is a potential hazard during cardiac catheterization, particularly in patients with cyanotic cardiac malformations. The insult to the central nervous system in one patient in this series was probably related to thromboembolic phenomena or hypoxemia, but air embolism cannot be ruled out. Improved construction of balloon catheters in recent years has virtually eliminated embolization of broken balloon fragments.

The fatal complication of left atrial wall laceration was related to the unusual location of the IAO and the orientation of the blade of the prototype catheter used in this patient. The catheter system was subsequently modified and the blade component design now protrudes inferiorly instead of superiorly. This modification has eliminated the need for as much rotation of the blade tip in the left atrium and should lessen the danger of inadvertent trauma or laceration of the adjoining cardiac tissue.

Perforation of the right ventricular outflow in another patient was related to the lack of a lateral fluoroscopy unit. A constant lateral fluoroscopic capability without moving the patient is essential for this procedure. Also, pressure should be carefully monitored through the catheter system during catheter manipulation to avoid inadvertent passage of the catheter into the right ventricle instead of the left atrium.

The introduction of the long percutaneous sheath has made transseptal blade atrial septostomy possible in a patient with an intact interatrial septum. It also facilitates quick introduction of the blade catheter into the left atrium in other patients. Thus, use of the long sheath technique eliminates prolonged and potentially traumatic maneuvering of the blade catheter and should minimize the incidence of major complications.

Major impediments to the blade septostomy procedure are an excessively small left atrium and inability to approach from the femoral vein because of a thrombosed femoral venous system or absence of the hepatic segment of the inferior vena cava.

The long-term fate of the IAO created by blade septostomy is not known. Our preliminary study with up to 4 years of follow-up of 38 patients indicates that the IAO has remained stable in 32 patients (84%), while in six patients (16%) it was found to be reduced in size several months to years later. On review, the blade atrial septostomy was not satisfactory in three patients whose IAO was subsequently found to be inadequate. In two of these three patients, the blade was poorly positioned and in the other, withdrawal of the blade across the interatrial septum was incomplete. Although we suggested one pullback of the blade across the interatrial septum in a previous communication, we now believe that several passages of the blade across the interatrial septum, with a slight change in the angulation of the blade, should minimize the possibility of an inadequate blade septostomy. This is particularly true in patients with an unusually small IAO or in the case of a transseptal approach, in which a stepwise procedure is also recommended. The cause of an inadequate IAO after balloon or blade atrial septostomy may be related to stretching of the IAO instead of a true tear of the interatrial septum. This can be due to an unusually thick interatrial septum or to inadequate technique. Despite this failure, some of these patients may show clinical improvement, such as increased oxygen saturation and a larger IAO as measured by balloon withdrawal. Later, these patients may show clinical deterioration and repeat study may show reduction of the IAO. Most of these patients likely had stretching of the IAO rather than true tear of the interatrial septum at the initial procedure.

No patient with long-term failure of the procedure has undergone a repeat blade atrial septostomy in the present series. A repeat procedure should be considered in patients who have undergone blade atrial septostomy in early infancy and are later found to have an inadequate IAO. Although only a few patients in our series have been followed by two-dimensional echocardiography to evaluate changes in size of the IAO, such serial studies may be valuable. Most neonates with mitral atresia do not have a significant interatrial pressure gradient despite the presence of a small IAO. In this series, balloon atrial septostomy was not performed in the majority of these patients, as they had no significant pressure gradient. In retrospect, all patients with mitral atresia should have balloon or blade atrial septostomy if technically possible, regardless of the interatrial pressure gradient, when the actual IAO is small by balloon measurement.

In conclusion, blade atrial septostomy is an effective means of enlarging or creating an IAO without thoracotomy. The procedure is particularly useful in patients who are critically ill because of an inadequate IAO and when the surgical risk is likely to be substantial.

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Pulmonary Atresia and Intact Ventricular Septum: A Revised Classification


SUMMARY The dismal outlook for patients with pulmonary atresia with intact ventricular septum may be related to associated right ventricular hypoplasia. Study of 32 autopsy specimens and 46 angiograms of neonates with this lesion suggested that the cavitary hypoplasia was related to massive hypertrophy of the right ventricular wall. This hypertrophy was sufficient to obliterate the trabecular and/or infundibular portion of the ventricular cavity entirely in one-third of the cases; this observation forms the basis for a revised classification of these hearts. Three autopsies and 14 angiograms of neonates with critical pulmonary stenosis were examined. Hearts with obliterated infundibular and trabecular cavities had thicker walls and smaller tricuspid valves, as estimated angiographically or at autopsy, than those in which the normal three portions of the ventricular cavity were represented.

PULMONARY ATRESIA with intact ventricular septum (PA:IVS) has a high early and late mortality. The pathologic classification of Greenwold et al. into commoner type 1 cases with a small right ventricular cavity and type 2 with a normal or dilated cavity has been the key to the description and management of the disease. The implication at that time was that patients with smaller cavities were less likely to survive and that the abnormality was not ultimately correctable. However, morphologic studies of PA:IVS have revealed a continuum of right ventricular cavity size; and, as clinical series were increasingly reported, it emerged that some cavities can grow.

We propose a revised classification of the right ventricular appearances in this condition, based on the tripartite approach to right ventricular morphology. This new classification has relevance for the surgical management of PA:IVS. Ventricular size is important, but there are limitations to the angiographic determination of the volume of these bizarrely shaped cavities.

We therefore prefer to measure the tricuspid annulus diameter and relate tricuspid valve size to our morphologic classification.

In the neonatal period, patients may present with a hemodynamic disturbance indistinguishable from PA:IVS (suprasystemic right ventricular pressure, obligatory right-to-left atrial shunt and a persistent ductus arteriosus) but with a pinhole patency of the pulmonary valve visible on angiography. The ventricular morphology of these patients was examined for comparison.

Materials and Methods

Thirty-two specimens from patients who died with PA:IVS during the neonatal period were available from the pathologic collection at the Hospital for Sick Children. Three specimens from patients with critical pulmonary stenosis from the same age group were also studied. Each specimen was examined to determine the presence or absence of the inlet, trabecular and infundibular portions of the right ventricular cavity. The inlet portion is defined as the part of the ventricle that incorporates the tricuspid valve apparatus. The trabecular portion lies beyond the insertion of the papillary muscles of the tricuspid valve toward the apex, while the infundibulum or outlet portion leads to the atretic pulmonary valve. The tricuspid diameter was measured using Hegar dilators, as this appeared more clinically meaningful than the conventional pathologic measurement of tricuspid annulus circumference. Maximal wall thickness, usually toward the apex of the cavity, was also measured.

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