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**Congenital Heart Disease**

# Perforation of the Atretic Pulmonary Valve

## Long-Term Follow-Up

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**OBJECTIVES** We evaluated the long-term results of perforation of the pulmonary valve in patients with pulmonary atresia with an intact ventricular septum (PA-IVS).

**BACKGROUND** Interventional perforation of the pulmonary valve is considered the elective first stage treatment for PA-IVS, particularly in patients with a tripartite right ventricle (RV) and normal coronary circulation. However, the long-term results of this procedure are lacking.

**METHODS** Between January 1991 and December 2001, 39 newborns with a favorable form of PA-IVS underwent attempted perforation of the pulmonary valve. We evaluated the early and long-term outcomes.

**RESULTS** Median tricuspid and pulmonary *z* values were −1.2 and −2.4, respectively. Perforation was successful in 33 patients. Among them, 17 needed neonatal surgery, 13 did not need any surgery, and 3 had elective surgery after the first month of life. There were two procedure-related deaths, seven nonfatal procedural complications, and four postsurgical deaths. Compared with patients needing neonatal surgery, those having no or elective surgery had a higher incidence of a tripartite RV and a higher median tricuspid *z* value (92% vs. 53%, $p = 0.04$ and −1.7 vs. −0.5, $p = 0.03$). At a median follow-up of 5.5 years (range 0.5 to 11.5), survival was 85% and freedom from surgery was 35%. Five patients, four of whom had neonatal surgery, underwent a partial cavo-pulmonary connection.

**CONCLUSIONS** Our results show that this technique, although burdened by non-negligible mortality and morbidity, is effective in selected patients with a normal-sized RV. Preselection of patients allows interventional or surgical biventricular correction in the majority of cases. (J Am Coll Cardiol 2003;41:1399–403) © 2003 by the American College of Cardiology Foundation

Pulmonary atresia with an intact ventricular septum (PA-IVS) is a complex congenital heart disease that has a wide morphologic diversity. The degree of right ventricular (RV) hypoplasia, the presence of a patent infundibulum, and the occurrence of right ventriculo-coronary connections strongly influence the therapeutic approach (1–3). In the forms with moderate RV hypoplasia, a patent infundibulum, and an atretic or imperforate pulmonary valve, RV decompression has been advocated as the initial palliative procedure (4). This goal can be achieved surgically or by interventional perforation of the pulmonary valve (5). During the last 10 years, interventional catheterization has been chosen as the elective primary treatment for favorable forms of PA-IVS (6–11). This procedure is feasible either by a guide wire, radiofrequency, or laser, with a relatively low periprocedural risk (6,8,10). However, most investigators have shown that, at early follow-up, the large majority of patients still needs palliative cardiac surgery (10,12). In addition, small series of patients primarily treated by perforation of the pulmonary valve have been examined so far (5,6,8,12), and data on long-term follow-up are lacking.

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In our institution, this technique was introduced in 1991. The purpose of this study was to review our experience, focusing attention on early and long-term results. We also tried to identify criteria that could predict the need for neonatal surgery, despite successful perforation of the pulmonary valve.

## METHODS

**Patients.** Between January 1991 and December 2001, 39 newborns with favorable forms of PA-IVS underwent attempted perforation of the pulmonary valve. Inclusion criteria did not change during the study period. They included neonates and infants with an unoperated PA-IVS, bipartite or tripartite RV, tricuspid *z* value not less than −3.5, pulmonary *z* value not less than −5, absent or mild Ebstein anomaly, and normal coronary anatomy. The exclusion criteria included a monopartite RV, coronary fistulae with RV-dependent coronary circulation, and moderate to severe Ebstein anomaly. The *z* value was defined as follows: (measured diameter − mean normal diameter)/(standard deviation of mean normal diameter). The population included 18 males and 21 females. The median age and weight at perforation were three days (range 1 to 90) and 3 kg (range 2 to 4), respectively. One patient had quadrisomy of chromosome X. Two patients had a mild Ebstein

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**Abbreviations and Acronyms**

ASD = atrial septal defect
BT = Blalock-Taussig
NYHA = New York Heart Association
PA-IVS = pulmonary atresia with intact ventricular septum
PCPC = partial cavo-pulmonary connection
PG = prostaglandin
RV = right ventricle/ventricular
RVOT = right ventricular outflow tract

anomaly of the tricuspid valve. The RV was bipartite in 12 and tripartite in 27 patients. The median tricuspid diameter was 11 mm (range 7 to 18), and the median tricuspid *z* value was −1.2 (range −3.5 to 1.5). The median diameter of the atretic pulmonary annulus was 7 mm (range 5 to 9), and the median pulmonary *z* value was −2.4 (range −4.5 to 1.5). All but three patients, who had a patent ductus arteriosus and underwent a late diagnosis, were under prostaglandin (PG) infusion.

**Transcatheter procedure.** The femoral vein was cannulated using a 4F or 5F sheath. All patients underwent RV angiography in the lateral and/or four-chamber view to exclude RV-dependent coronary circulation and to visualize the pulmonary infundibulum. Guide-wire perforation was obtained by pushing the rigid extremity of a variety of guide wires against the pulmonary valvular plane through a 4F or 5F Judkins right catheter. Radiofrequency valvotomy was performed using the Medtronic Cardiorhythm generator (Rueil Mailmaison, France) connected to a Cerablate PA 120 unipolar catheter (Osypka, Rheinfelden-Herten, Germany) through a 5F Judkins right catheter. After successful valvotomy, a 0.014-in. or 0.018-in. coronary guide wire was passed into the descending aorta through the pulmonary artery and patent ductus arteriosus. Sequential dilations of the pulmonary valve were then performed, until obtaining a full opening of the pulmonary annulus. The RV to aortic pressure ratio was measured before and after successful dilation of the pulmonary valve. Procedural failure was defined as the inability to perforate the atretic valve.

**Follow-up.** Patients were gradually weaned from PG infusion. Patients who, after successful perforation, continued to have inadequate anterograde pulmonary flow and hypoxemia, underwent a Blalock-Taussig (B/T) shunt procedure. The B/T shunt was associated with the right ventricular outflow tract (RVOT) patch when the outflow tract was hypoplastic or stenotic.

Persistent infundibular stenosis with or without pulmonary valve stenosis was an indication to use a late, elective RVOT patch. Patients with persistent systemic desaturation at rest and a right to left shunt through an atrial septal defect (ASD) or patent foramen ovale underwent a partial cavo-pulmonary connection (PCPC).

Patients with mild desaturation on effort (≤95%) and unchanged right atrial pressure during test occlusion of ASD underwent ASD interventional closure. Patients with normal saturation at rest and during test occlusion of the B/T shunt underwent interventional embolization.

Hospital records were reviewed for any surgical or interventional procedure and for the most recent clinical and echocardiographic evaluation. We took into account the arterial oxygen saturation at rest and on effort, the right to left shunt through an ASD or foramen ovale, and the residual pulmonary stenosis.

On the basis of follow-up data, the patients were classified into two outcome groups: the "no neonatal surgery" group (n = 16) comprised patients who, after successful perforation of the pulmonary valve, did not need any surgical procedure, or needed elective surgery after the neonatal period and had no or mild cyanosis on effort at late follow-up. The "neonatal surgery" group (n = 17) comprised patients who, despite successful perforation of the pulmonary valve, needed neonatal surgery.

**Statistical analysis.** For analysis of data, we used the statistical package SAS version 6.12. The Kaplan-Meier test was employed for analysis of survival and freedom from surgery. The paired Student *t* test was used to compare hemodynamic values before and after interventional catheterization. The Wilcoxon and Fisher exact tests were employed to compare variables between the "no neonatal surgery" and "neonatal surgery" groups. For statistical analysis, patients with a patent ductus arteriosus and late diagnosis were excluded. Age, weight, and anatomic values were expressed as the median value and range. A p value $<0.05$ was considered statistically significant.

## RESULTS

Figure 1 summarizes the results of interventional and surgical procedures. Perforation, obtained by a guide wire in 20 and by radiofrequency in 19 patients, was successful in 33 (85%). Right ventricular pressure fell from 112 ± 21 to 49.6 ± 9.7 mm Hg ($p < 0.001$). There were six (15%) procedural complications (2 deaths due to infundibular perforation, 1 pericardial effusion, 3 cases of atrial flutter) and three (8%) postprocedural complications (3 cases of necrotizing enterocolitis). Infusion of PG was continued for a median period of 12 days (range 4 to 40). At a median follow-up of 15 days (range 1 to 30), 21 patients, all under PG therapy, needed cardiac surgery (12 B/T shunts, 8 B/T shunt plus RVOT patch, 1 RVOT patch). There were four (19%) postsurgical deaths. Two more patients had an elective RVOT patch after the neonatal period. Four patients had redilation of the pulmonary valve at 13, 11, 20, and 1.5 months; three had interventional shunt embolization at 12 and 29 months and 6 years, and three had interventional closure of their ASD at 5, 6, and 4 years. In the "no neonatal surgery" group, no patient underwent interventional catheterization after the first perforation of the pulmonary valve. Four patients who had previous neonatal surgery and one unoperated patient underwent PCPC. None had the Fontan operation.

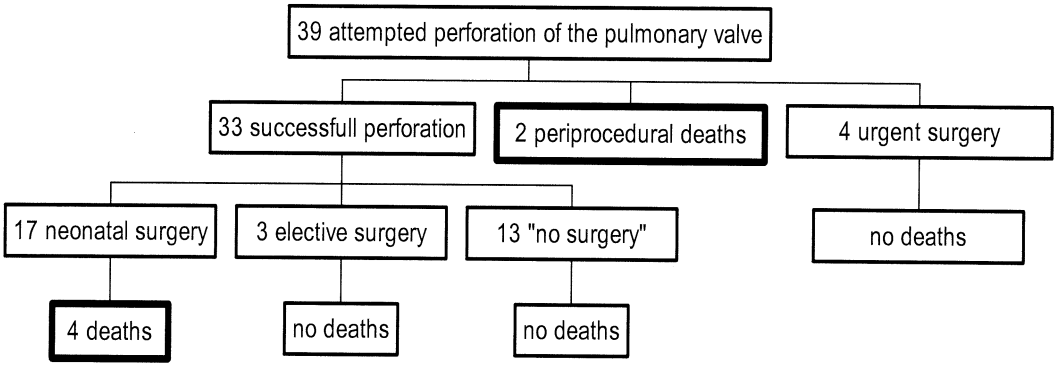


**Figure 1.** Flow chart summarizing the procedures and outcomes (death or no death) of patients.

Thus, at a median follow-up of 5.5 years (range 0.5 to 11.5), 13 patients did not need any surgery, and three had elective surgery after the neonatal period. The remaining patients needed neonatal surgery, four due to failed perforation and 17 despite successful perforation of the pulmonary valve. Table 1 illustrates the anatomic "predictors of outcome" in the "no neonatal surgery" and "neonatal surgery" groups. Patients who needed neonatal surgery had a tricuspid $z$ value significantly smaller than that of patients who did not need neonatal surgery; 47% of them had a bipartite RV. In this group, the pulmonary annulus was smaller than that in the "no neonatal surgery" group, but not significantly so. No patient in the "no neonatal surgery" group had a tricuspid $z$ value less than $-2$ and a pulmonary $z$ value less than $-3$; all but one had a tripartite RV. Nevertheless, two patients with tricuspid and pulmonary $z$ values $>0$ and a tripartite RV needed neonatal surgery.

The neonatal mortality rate was 15%. No late deaths occurred. Figure 2 illustrates the freedom from surgery. Neonatal surgery was needed in 52% of patients with a successful pulmonary perforation. Overall late freedom from surgery was 35%. Nine patients needed multiple operations.

At the last clinical and echocardiographic examination, all patients except one, who had a chromosomal anomaly, were in New York Heart Association (NYHA) functional class I. Cyanosis at rest was present in seven patients: two are waiting for PCPC; two have PCPC with a mild right to left shunt at the atrial level; and three have a mild right to left shunt through a patent foramen ovale and are waiting for its interventional closure. One patient had mild cyanosis on effort. No patient has residual pulmonary stenosis. Mild to moderate pulmonary incompetence was present in five patients and tricuspid incompetence in one. All patients with PCPC have nonobstructed flow in the anastomosis.

## DISCUSSION

Perforation of the pulmonary valve, introduced for the first time in 1991 by Qureshi et al. (13) has been validated by various investigators (6,8–10). The successful perforation rate ranges between 75% and 100% (6,8,10,12); however, a perioperative mortality rate up to 17% has been reported (12). When selecting patients for perforation, choosing only those with favorable forms has worked to diminish procedure-related death and improve outcome (6). Nevertheless, perforation of the infundibulum, sepsis, and multiorgan failure are not occasional complications (5,7,10,12). In our series, perforation was obtained in 85% of cases. The procedural mortality rate was 5% and procedural morbidity was 12%, due to either pericardial effusion or arrhythmias. We confirmed a non-negligible incidence of necrotizing enterocolitis in neonates who remained duct-dependent after successful pulmonary valve dilation (14).

After a successful procedure, the impaired compliance of the RV often prevents the surgeon from avoiding neonatal surgery. The reported need of a B/T shunt and/or RVOT patch ranges from 0% to 75% (6,10). This large variability is likely linked to anatomic differences among the treated patients or to different attitudes regarding the maximal accepted duration of PG infusion. In our series, neonatal surgery was needed in more than 50% of infants with successful valve perforation. Various authors have highlighted the increased risk of surgery in duct-dependent patients (12). This could be explained by prolonged hypoxemia, chronic vasodilation, poor trophism, immunodepression, and tissue edema, all generally found in these patients (14). In this setting, we reported a 24% postsurgical mortality rate, due to either sepsis or shunt thrombosis, whereas no deaths occurred in patients undergoing immediate sur-

**Table 1.** Anatomic Characteristics of Patients Who Did Not Need or Who Needed Neonatal Surgery

| | No Neonatal Surgery Group (n = 13) | Neonatal Surgery Group (n = 17) | p Value |
|---|---|---|---|
| Tripartite right ventricle (%) | 92 | 53 | 0.04 |
| Median tricuspid $z$ value | −0.5 | −1.7 | 0.03 |
| Median pulmonary $z$ value | 0.5 | −1.5 | 0.14 |

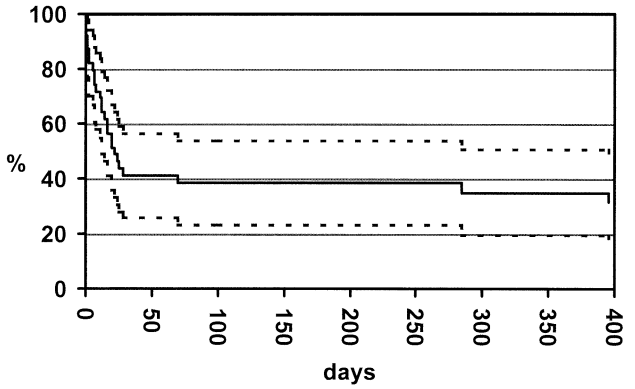


**Figure 2.** Actuarial freedom from surgery for the entire study group of 36 surviving patients. **Dotted lines** enclose a 95% confidence interval.

gery after failed perforation. Nevertheless, other investigators have reported a high mortality in patients undergoing urgent surgery after failed perforation (12).

The morphologic characteristics of the RV are considered to be the main determinant of outcome; thus, the main concern is the preselection of patients. Even if it has been shown that even very hypoplastic RVs can grow (12,15–17), most authors agree that biventricular repair is unlikely to be attained when the tricuspid $z$ value is less than $-4$ (18). In addition, it has been demonstrated that patients needing interventional catheterization only must have a tricuspid $z$ value greater than $-0.5$ and a pulmonary $z$ value greater than $-4$ (7,19).

Our data show that biventricular correction was attained in 87% of patients. All but one patient who did not need neonatal surgery had a tripartite RV. None of them had a tricuspid $z$ value less than $-2$ and a pulmonary $z$ value less than $-2.8$. Nevertheless, although patients needing neonatal surgery had a tricuspid $z$ value significantly smaller than that of patients who did not need any surgery, three patients with tricuspid and pulmonary $z$ values $>0$ needed neonatal surgery. Other investigators have highlighted some morphologic overlapping between groups of patients with different outcomes, suggesting that factors promoting RV growth have not been completely identified (12,15).

Thus, if an accurate preselection of patients is mandatory for obtaining biventricular repair, it is probably insufficient to identify patients who will not need any surgery.

Although an elective surgical approach for patients with PA-IVS is known to have an elevated mortality (20), recently, some authors have reconsidered the surgical treatment as an alternative to pulmonary valve perforation (5,8). The results are discordant due to different populations examined and a different surgical approach. As no randomized studies have compared interventional versus surgical treatment for unselected or selected patients with PA-IVS, it is difficult to establish the best first-stage treatment.

Mid-term survival in our series was similar to that previously reported (5,10,12). Publications on mid-term and late outcomes are scanty. Our data show that, after the neonatal period, mortality was zero and freedom from surgery decreased only slightly, confirming that neonatal strategy and outcome are strictly linked.

The results at mid-term follow-up are excellent for patients treated either by surgery or catheterization in terms of NYHA functional class, incidence of systemic desaturation, and pulmonary and tricuspid valve function. Repeated interventional catheterization was observed in the large majority (59%) of patients who needed neonatal surgery, but not in any patient who did not need surgery, confirming that different populations of patients are treated and that, again, a strict preselection of patients is mandatory before choosing either the surgical or interventional approach.

**Conclusions.** Our results show that perforation of the pulmonary valve, although carrying non-negligible mortality and morbidity, is effective in selected patients with a normal-sized RV. Preselection of patients allows biventricular correction in the majority of them. Limiting the interventional approach to forms of PA-IVS with a tripartite RV and a tricuspid $z$ value no less than $-2$ will probably allow us to identify more precisely patients in whom biventricular circulation will be restored by pulmonary valve perforation only.

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