SURGERY FOR CONGENITAL HEART DISEASE

IMPROVED RESULTS WITH SELECTIVE MANAGEMENT IN PULMONARY ATRESIA WITH INTACT VENTRICULAR SEPTUM

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Objective: Late outcome of neonatal pulmonary atresia with intact ventricular septum remains poor in most reported series. We have followed a selective approach toward either single ventricle repair versus complete or partial biventricular repair based on the presence of right ventricle-dependent coronary circulation and growth of the right ventricle. **Methods:** A retrospective chart review was conducted of 47 patients who underwent surgery between January 1991 and September 1998. **Results:** Sixteen (34%) patients had a right ventricle-dependent coronary circulation, with a tricuspid valve Z-score of -3.0 ± 0.66 versus -2.0 ± 0.95 ($P = .002$) for those without a right ventricle-dependent coronary circulation. A systemic-pulmonary artery shunt only was performed in all patients with a right ventricle-dependent coronary circulation, with 1 death. Fourteen of 16 patients with a right ventricle-dependent coronary circulation underwent a bidirectional Glenn shunt at a median of 9 months after their first operation, 9 of whom have had a Fontan procedure (no deaths). In the 31 (66%) patients without a right ventricle-dependent coronary circulation, 6 patients underwent only a systemic-pulmonary artery shunt, 23 had a shunt and right ventricular decompression, and 2 had only a transannular patch. In this group, 10 patients received a 2-ventricle repair, 6 a 1.5-ventricle repair, and 8 patients had a Fontan procedure. There was 1 early death and the overall survival was 98% at 1 year, 5 years, and 7 years. **Conclusions:** If patients are stratified well, excellent survival can be achieved in the treatment of pulmonary atresia with intact ventricular septum. This result may be at the price of achieving a 1-ventricle as opposed to a 2-ventricle repair. (J Thorac Cardiovasc Surg 1999;118:1046-55)

The outcome of neonatal pulmonary atresia with intact ventricular septum (PA/IVS) remains poor in most reported series.^{1,2} A recent multi-institutional study reported a survival of 60% at 4 years.³ In the current era, these results do not match the surgical success of other complex congenital heart defects such as trans-

position of the great arteries. This may reflect partly the morphologic heterogeneity of this condition and partly the interpretation of this morphology by different institutions. Furthermore, the numerous physiologically based management protocols and algorithms are not supported by surgical outcome.

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The morphologic heterogeneity in PA/IVS encompasses a spectrum of lesions including hypoplasia of the right ventricle (RV) and the tricuspid valve, abnormal coronary circulation, and pulmonary atresia itself.⁴ Ideally, surgery should optimize growth of the RV and the tricuspid valve without compromising the coronary circulation and ventricular function. If all measures are favorable, a 2-ventricle repair will be achieved; if not, a 1- or 1.5-ventricle repair will be done. We have followed a selective approach toward either single ventricle repair versus complete or partial biventricular repair based on the presence of RV-dependent coronary circulation and growth of the RV after decompression with an RV outflow patch.

The aim of this study was to examine the neonatal strategies, surgical management, and midterm survival for patients with PA/IVS.

Methods

Patients who underwent surgery for PA/IVS between January 1991 and September 1998 were studied retrospectively. We chose January 1991, since patients with PA/IVS up to this date have previously been included in the Congenital Heart Surgeons Society data. A total of 47 patients were identified. Patients with Ebstein's anomaly were excluded. The median age at the first operation was 4 days (range 2 days–6 weeks). All patients were younger than 1 month of age at the time of initial presentation, except 1 patient who was 6 weeks old. The median weight was 3.4 kg (range 2.8–4.3 kg). Initial diagnosis was made by transthoracic echocardiography in all patients.

Tricuspid valve and RV cavity size. The tricuspid valve diameter was measured by 2-dimensional transthoracic echocardiography and is expressed as the Z-value.⁵ The RV cavity size is expressed as 3° of hypoplasia, mild, moderate, or severe as measured by transthoracic echocardiography. In this study, angiographically derived RV volumes calculated by the Simpson technique⁶ or methods relying on inlet and outlet indices are avoided because of the unusual shape of the morphologic RV in patients with PA/IVS.

Coronary circulation. All patients underwent coronary angiography. Coronary artery-to-RV fistulas, coronary sinusoids, and coronary stenoses were identified. We define patients with an RV-dependent coronary circulation as those with RV-to-coronary artery fistulas plus stenoses of the right and/or left coronary arteries. Patients with an RV-dependent coronary circulation have a major portion of their left ventricular myocardium dependent on supply from the RV and therefore at risk for ischemia when the RV is decompressed. Death after RV decompression depends on the amount of left ventricular myocardium at risk for ischemia, that is, the amount of myocardium that is distal to coronary artery stenoses in the presence of stenoses of other coronary vessels that prevent collateralization. Therefore, RV decompression is not necessarily contraindicated in patients with fistulas

without associated coronary stenoses or in patients with fistulas with stenosis of a single coronary artery.

Surgical intervention. All modified Blalock-Taussig shunts were performed on the right side through a median sternotomy. A 3.5-mm polytetrafluoroethylene tube graft was used to construct the shunt in almost all patients, with the proximal anastomosis being placed at the origin of the right subclavian artery. RV obstruction was relieved by using glutaraldehyde-treated autologous pericardium as a transannular patch for patients in whom RV-dependent coronary circulation was not present. The atretic pulmonary valve was usually excised. Subsequent interventions were a bidirectional Glenn procedure with or without closure of an atrial septal defect and the fenestrated Fontan operation, either the lateral tunnel or the extracardiac conduit technique.

Outcome measures and eventual ventricular repair

Two-ventricle repair. The group undergoing 2-ventricle repair is defined as those in whom residual RV outflow tract obstruction was relieved, all intracardiac and extracardiac shunts were closed, and each ventricle was connected to its respective great vessel.

One-ventricle repair. The patients undergoing 1-ventricle repair are defined as those who underwent a Fontan operation.

One and a half-ventricle repair. The 1.5-ventricle repair group comprises those in whom RV-pulmonary artery continuity was present, some of the systemic venous return was pumped to the pulmonary circulation by the RV, and the rest of the systemic return was directed to the pulmonary circulation by a cavopulmonary anastomosis.

Intermediate stage. This group comprises those patients who have undergone a Blalock-Taussig shunt with or without RV decompression or a bidirectional Glenn procedure with a follow-up of 1 year or less. The ultimate fate of the ventricular repair has not yet been determined.

Statistical analysis. Z-scores for the tricuspid valve for patients with and without an RV-dependent coronary circulation were compared by means of the 2-sample Student *t* test. Actuarial survival with a 95% confidence interval was estimated by means of the Kaplan-Meier product-limit method.⁷ The Lifetest procedure in the SAS software package (version 6.12, SAS Institute, Inc, Cary, NC) was used for data analysis.

Results

Tricuspid valve and RV cavity size. The Z-value of the diameter of the tricuspid valve was between –2 and –3 in 32 (68%) patients and less than –3 in 15 (32%) patients. The RV was severely hypoplastic in 14 (30%) patients, moderately hypoplastic in 32 (68%), and mildly hypoplastic in 1 (2%) patient. The mean Z-value for tricuspid valve diameter for patients with a severely hypoplastic RV was -2.9 ± 0.62 and for those with a moderately hypoplastic RV, -2.2 ± 0.67 . This difference was statistically significant ($P = .03$).

Coronary circulation. Sixteen (34%) of the 47

Fig 1. Flow diagram to show outcome measure and eventual ventricular repair in patients with RV-dependent coronary circulation (RVDCC). PA, Pulmonary artery; BDG, bidirectional Glenn shunt.

patients had an RV-dependent coronary circulation. None of these patients underwent RV decompression. Of the 14 patients with severe RV hypoplasia, 7 had an RV-dependent coronary circulation, and of the 32 patients with moderate RV hypoplasia, 9 had an RV-dependent coronary circulation. This difference did not reach statistical significance ($P = .18$, Fisher exact test).

Surgical intervention. Systemic-pulmonary artery shunt was performed in the 16 (34%) patients with an RV-dependent coronary circulation in the neonatal period (Fig 1). There was 1 death in this group. A neonate weighing 2.8 kg who had metabolic acidosis despite alprostadil (prostaglandin E_1) and inotropic support was taken to the operating room for an emergency Blalock-Taussig shunt. When the chest was opened, the patient had an episode of ventricular fibrillation that proved resistant to all treatment with eventual death. Fourteen of the 16 patients with an RV-dependent coronary circulation underwent a bidirectional Glenn procedure at a median of 9 months after the first operation. Of these patients, 9 have subsequently undergone the Fontan procedure, and 6 are in the intermediate stage. One patient underwent the Fontan procedure without a preliminary bidirectional Glenn shunt.

Among the 31 (66%) patients without an RV-dependent coronary circulation (Fig 2), 6 patients underwent an initial systemic-pulmonary artery shunt alone because of very small RV size, usually with minimal or no infundibular development, and 23 had a shunt and RV decompression performed. In 2 patients in this latter group, the shunt and RV decompression were staged. In 1 patient RV decompression was performed at 2 weeks and in another at 2 months after the shunt. This was due to the development of necrotizing enterocolitis in both patients. Two patients had a transannular patch without a shunt. In the group without an RV-

dependent coronary circulation, 10 patients received a 2-ventricle repair, 8 patients a 1-ventricle repair, and 6 a 1.5-ventricle repair.

Of the 17 patients who underwent the Fontan procedure, 12 patients had a lateral tunnel and 5 had an extracardiac conduit performed.

Follow-up. The median follow-up from the last procedure was 33 months (range 2 months–7.5 years) for all patients. Of the 47 patients who underwent surgical intervention, only 1 patient died. Follow-up is complete and up-to-date in all patients except for 1 patient who lives outside the United States. The actuarial survival at 1 year, 5 years, and 7.5 years was 98% (Fig 3).

Discussion

The goals of treatment for PA/IVS are to separate the pulmonary and systemic circulations to eliminate cyanosis, without compromising cardiac output and without inducing excessive systemic venous hypertension. These goals can be achieved by univentricular, biventricular, or 1.5-ventricular strategies, dictated by indices that are predictive of success or failure of each approach. A particular challenge in planning a strategy for the treatment of PA/IVS is the fact that RV growth and contribution to pulmonary flow is difficult to predict, yet critical to the question of whether the patient will benefit from efforts to effect a biventricular repair. Various authors have proposed factors predicting RV growth. Bull and colleagues⁸ have proposed the tripartite classification of the RV and the size of the tricuspid valve as determinants of RV growth. In an angiographic and autopsy study, they demonstrated that hearts with obliterated infundibular and trabecular cavities had thicker walls and smaller tricuspid valves than those in which the normal 3 portions of the RV cavity were represented. Similarly, others have suggested that the tricuspid valve size is a major prognostic determinant of the RV growth and eventual achievement of a 2-ventricle repair.³ Presumably, achieving a 2-ventricle repair is ideal. Earlier attempts at RV decompression and allowing for the growth of the RV and tricuspid valve had resulted in a high early and late surgical mortality in single-center and multicenter reports.^{2,3} This may have been due partly to the presence of RV-dependent coronary circulation. Because the suprasystemic RV may supply part of the myocardial blood flow in the presence of fistulas with or without coronary stenoses, surgical procedures that result in RV decompression may compromise perfusion of both the RV and the left ventricle. The potential mechanisms for adverse outcome after RV decompression may be due to the RV-to-coronary fistulas without coronary stenoses, resulting in the runoff from the aorta into the

Fig 2. Flow diagram to show outcome measure and eventual ventricular repair in patients without RV-dependent coronary circulation (RVDCC). PA, Pulmonary artery; BDG, bidirectional Glenn shunt; RVD, RV decompression; V, Ventricle.

RV during diastole (steal phenomenon). If stenosis is present, either proximal or distal to the fistula, both ischemia and steal may account for poor performance. Giglia and colleagues⁹ demonstrated that potential RV steal without coronary stenoses does not preclude RV decompression; however, RV decompression is contraindicated in the presence of stenosis and/or occlusion of the coronary arteries. Death in patients with an RV-dependent coronary circulation who undergo RV decompression is most probably related to the amount of left ventricular myocardium at risk of ischemia. Regional left ventricular dysfunction was shown to be rare in patients without coronary artery abnormalities but common after RV decompression in those with multiple stenoses or obstruction.¹⁰

In a study by Coles and associates,² 39 of 50 patients with an RV-dependent coronary circulation underwent RV decompression. The 30-day mortality in this group was 41.7%. In patients with an RV-dependent coronary circulation, some have proposed the placement of a conduit between the aorta and the RV.¹¹ de Leval¹² demonstrated that such a procedure could actually equilibrate the systolic and diastolic pressures in the RV and the aorta, thus using the RV as a conduit to perfuse the distal coronary arteries.

During the late 1980s we gradually adopted a policy of RV decompression according to the presence or absence of RV-dependent coronary circulation. By 1991 this had become a standard institutional protocol. As is shown in this study as well as previous reports,³ there is a direct correlation between the size of the RV and tricuspid valve and the presence of an RV-dependent coronary circulation. Those patients with an RV-

Fig 3. Kaplan-Meier estimated survival after surgery for PA/IVS. The 95% confidence intervals are provided around the survival curve and the numbers of patients on whom the estimates were based are shown in parentheses.

dependent coronary circulation tend to have smaller RVs and tricuspid valves.

A 1-ventricle repair is planned in patients in whom RV decompression cannot be achieved because of RV-dependent coronary circulation or in whom tricuspid valve or RV hypoplasia is severe. In the current study, there was no death after the Fontan procedure with a median follow-up of 2.8 years. Delius and coworkers¹³ reported that a low-risk Fontan procedure may produce better long-term results than a higher-risk biventricular repair for some patients with moderately functioning ventricles with complex disease. Similarly, others have

reported actuarial survivals of 85% and 80% at 6 and 10 years' follow-up with an operative mortality of 8%.^{14,15} Longer follow-up is needed in this group to assess the rise in hazard function.

Although there is consensus in the strategy of treatment for the extremes of anatomic presentations, opinions vary concerning how to optimize outcome between these extremes of PA/IVS. The 1.5-ventricle repair provides a means of allowing some pulsatile blood flow when the RV is insufficient to conduct an entire cardiac output. There are appealing theoretical advantages to the 1.5-ventricle repair as an end point in itself or as a strategy to encourage RV and pulmonary artery growth before a 2-ventricle repair. The strategy is to relieve RV outflow tract obstruction, to encourage forward flow, and to minimize RV volume loading. Recent modifications of the 1.5-ventricle repair to manage the deficient RV have been to combine an RV outflow procedure with a bidirectional Glenn,^{16,17} atrial decompression by septostomy,¹⁸ or double cavopulmonary anastomosis,¹⁹ with or without a systemic-pulmonary shunt to augment pulmonary blood flow before the bidirectional Glenn procedure.²⁰ Angiography and echocardiography after these procedures have demonstrated antegrade, pulsatile pulmonary blood flow, without excessive systemic venous hypertension, despite potentially competitive sources. Measures such as proximal right pulmonary artery ligation may be carried out to separate bidirectional Glenn and RV flow in some cases in which circular shunting is anticipated.¹⁹

Controversy remains as to the significance and predictability of competition from multiple sources of pulmonary blood in the 1.5-ventricle repair. The theoretical advantages to additional pulmonary blood flow are (1) increased arterial oxygen saturation reducing cyanosis, (2) providing hepatic blood to the pulmonary circulation, which may have an anti-angiogenesis factor reducing the development of pulmonary arteriovenous malformations, (3) providing an element of pulsatility, and (4) encouraging the growth of the pulmonary arteries. However, there are no strong data to support these theoretical advantages. Webber and associates²¹ reported results of a multi-institutional study of patients undergoing a bidirectional Glenn shunt with or without competitive sources of pulmonary blood flow and demonstrated improved early postoperative recovery with competitive flow.

This study demonstrates the limitations of RV growth after decompression in terms of achieving suitability for a 2-ventricle repair. In the majority of the patients, a 1.5-ventricle or a 1-ventricle repair was required despite successful decompression of the RV. Never-

theless, if stratified well on the basis of the presence or absence of RV-dependent coronary circulation, excellent results and survival can be achieved in patients with PA/IVS.

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Discussion

Dr Frank L. Hanley (*San Francisco, Calif*). Dr Jahangiri, I thoroughly enjoyed your excellent presentation. Your outstanding results set a new benchmark for outcomes in this very complex and difficult problem.

The publication in 1993 from the Congenital Heart Surgeons Society on PA/IVS emphasized a very important dilemma in this lesion, pitting survival against a desire to create as many 2-ventricle repairs as possible. Maximizing survival seemed to require that fewer 2-ventricle repairs would have to be performed by a stratification of management. On the other hand, if one attempted to maximize the number of successful 2-ventricle repairs, it seemed that this could be achieved only at the expense of a higher overall mortality.

Most would agree that the philosophy of maximizing survival would be the appropriate one. Your experience in this study confirms that moving away from a strategy that is determined to achieve a 2-ventricle repair at all costs does not maximize survival. This concept is supported by your data showing that more than 50%, 24 of the 47 patients in the study, received a shunt alone at the very first operation. In the entire population, of those achieving a definitive operation to date, 29% (10/34) have received 2-ventricle repair and the remainder have received something else. Excluding those patients with an RV-dependent coronary circulation, fewer than 45% have received a 2-ventricle repair. The benefit of this stratification can be seen in that you have only 1 death at midterm follow-up in your series of 47 patients.

I would like to address several areas. Several have to do with definitions in the manuscript and in your presentation.

First, the concept of RV-dependent coronary circulation has some intrinsic subjectivity. Exactly what do you mean by RV-dependent coronary circulation? Does dependency of just the circumflex distribution, just the left anterior descending, or just the distal half of the left anterior descending after the first septal perforator qualify? Where do you draw the line in determining the definition?

Of the 31 patients who did not, by your definition, have RV-dependent coronary circulation, how many actually had fistulas?

How did you define mild, moderate, and severe RV hypoplasia?

Additionally, as your study and previous studies have shown, there seems to be a fairly good correlation between RV size and tricuspid valve size. In the analysis of your patients, did you find that looking at the RV per se in addition to or separate from the tricuspid valve size helped you in your stratification process?

I noted that 30% of the patients who received a Fontan operation in the group with an RV-dependent coronary circulation received an extracardiac Fontan conduit. Do you believe that there is a particular benefit for the extracardiac Fontan conduit in this subset of patients, or does this represent a more general change in the philosophy of how the Fontan conduit is constructed at your institution?

Could you also define more precisely your 1.5-ventricle repair operation? You state that it involves forward flow across the RV outflow tract with a bidirectional Glenn shunt. Does this mean that the atrial septal defect is always closed, or do you include in this group some patients who still have an open atrial communication?

Dr Jahangiri. Dr Hanley, thank you for your comments and your questions.

Regarding the definition of RV-dependent coronary circulation, we do not believe that a single coronary stenosis precludes RV decompression. I cannot give you exact information as to the anatomy of all the RV circulations, but they were labeled as dependent circulations from the echocardiograms and the angiograms; the majority of these patients had both a left anterior descending and a right coronary stenosis. Almost none of the patients had a single stenosis of the circumflex coronary artery. Thus we do not believe that single coronary stenosis should preclude RV decompression. The decision whether a patient does or does not have an RV-dependent coronary circulation has generally been made by the team. Although I spent a very short period at the Children's Hospital, I do not believe there have ever been discussions and discrepancies about whether a patient had RV dependence or not. The diagnosis usually is pretty clear-cut.

Regarding your second question, none of the patients without an RV-dependent coronary circulation had any fistulas.

Your third question concerned the definition of RV hypoplasia. We are aware of the tripartite classification of the RV, but in this study we were more concerned with the overall size of the RV cavity. Similarly, we did not use the Simpson method or volumetric methods. We used echocardiography to judge RV hypoplasia.

As you previously demonstrated in the Congenital Heart Surgeons Society report, the tricuspid valve size has correlated with the presence of RV-dependent coronary circulation. Thus patients with RV-dependent coronary circulations do have smaller tricuspid valves and smaller RVs.

Performance of an extracardiac Fontan operation was purely the choice of the surgeon. I cannot distinguish any difference between functional status or well-being when compared with intracardiac cavopulmonary connections.

Regarding the 1.5-ventricle repair, some patients in that cohort still have a communication in their atrial septum.

Dr Hanley. Just to clarify one point: If a fistula was present, was the coronary circulation always RV-dependent?

Dr Jahangiri. No. Of the patients without RV-dependent coronary circulations, 6 had a single coronary stenosis with fistulas.

Dr Hanley. Of the 31 patients who were not RV-dependent, some had fistulas?

Dr Jahangiri. Absolutely. With single coronary stenosis.

Dr Hillel Laks (*Los Angeles, Calif*). We also have had a selective approach to the treatment of pulmonary atresia, also dividing the patients into those with mild, moderate, and severe hypoplasia. In the neonate, the tricuspid valve size does generally correlate with the RV size. We have defined mild as Z-scores of 0 to -2, moderate as -2 to -4, and severe as -4 onward.

We have had patients who have had RV-dependent coronary circulations of varying degrees that we have converted to non-RV-dependent coronary circulations. Again, this is hard to define. But we have performed RV decompression at the time of the Glenn or Blalock-Taussig shunt in patients who have had multiple tortuous sinusoids. In the case of severe hypoplasia, we have done it by tearing the tricuspid valve through the right atrium with a rhizotomy knife; in the case of moderate or mild hypoplasia, we have inserted a transannular RV outflow tract patch. We have seen those tortuous sinusoids that may fill the right and the left coronary arteries to disappear with time with that kind of decompression. Large fistulas that could be identified on the surface, we have ligated.

I am wondering whether some of your cases could have been converted to non-RV-dependent coronary circulations. You had quite a large number in the moderate hypoplasia group who had RV coronary abnormalities which, if they had been converted, might have been good candidates for a biventricular repair. Thus, my first question is this: Do you think that some patients can be converted from an RV-dependent to a non-RV-dependent coronary circulation?

My second question concerns 1.5-ventricle repairs. We have done procedures to increase the size of RVs that have been as small as between one third to one half of normal, which we would consider the more extreme form of the moderate hypoplasia. We put in an adjustable atrial septal defect, do an RV myectomy to enlarge the RV, and insert a competent pulmonary valve. Those procedures can yield quite dramatic growth of both the tricuspid valve and the RV. If you use a transannular patch and do not have a restrictive atrial septal defect, the tricuspid valve will not grow. We have seen some patients lost to follow-up who have come back with large RVs but without growth of the tricuspid valve. I wonder whether you have seen anything like that.

Dr Jahangiri. Dr Laks, thank you for your comments and your questions.

Your first question concerned converting dependent coronary circulations to nondependent coronary circulations. As I mentioned earlier in answer to Dr Hanley, we define RV-dependent

coronary circulations as those with more than just a single coronary stenosis. I believe this is a more severe end of the spectrum. Therefore, we do not believe that any of this cohort of patients can be converted to a nondependent circulation.

With regard to the 1.5-ventricle repair, because of the pathology of this condition, after we find that the atrial septal defect is not restrictive, we do not need to interfere with its size.

Dr Laks. You say it is nonrestrictive?

Dr Jahangiri. Yes.

Dr Laks. That is the reason I am suggesting that a controllable atrial septal defect, which will raise the right atrial pressure, would encourage forward flow through the hypoplastic RV and develop growth provided the pulmonary valve is competent as well.

Dr Jahangiri. I'm sorry, I misunderstood your question. That is a very good idea. We have not done it in practice.

Dr Hani K. Najm (*Riyadh, Saudi Arabia*). Do you think the RV-dependent coronary circulation is always congenital, or do you think it could be acquired? On the basis of the nature of blood flow from the RV through the fistula to the coronary artery, which is faced with opposing antegrade blood flow from the coronary artery, the stenosis may develop. If you think it could be acquired, could you prevent it?

Dr Jahangiri. I really do not know.

Dr Najm. By early decompression of the RV, even in the presence of RV-coronary artery fistulas, you could avoid postnatal development of coronary artery stenoses, thereby avoiding RV dependency. Otherwise, this opposing flow of blood in the coronary arteries produces intimal changes leading to development of stenoses or even interruptions of the coronary arteries.

Dr Dominique R. Metras (*Marseille, France*). I rise to compliment Dr Jahangiri and the Boston group for an outstanding and certainly difficult-to-reproduce series of results. I have a question concerning the selection of patients. Was this was a consecutive series of patients presenting at Boston Children's Hospital with pulmonary atresia or a consecutive series of patients escaping interventional catheterization with perforation and dilatation of the pulmonary orifice? In other words, what is the policy of your center regarding interventional catheterization in this disease?

Dr Jahangiri. This is a series of consecutive patients with the diagnosis of PA/IVS who underwent surgery. We learned a long time ago that an isolated pulmonary valvotomy, either by catheter or surgery, or even a valvectomy does not relieve the RV outflow tract obstruction adequately. Patients often return with some element of residual obstruction. This is a consecutive, genuine series of patients with this diagnosis.