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Severe pulmonary valvar insufficiency should be aggressively treated

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MY ROLE IN THE DEBATE BETWEEN MYSELF AND Bill Gaynor is to substantiate the notion that severe pulmonary valvar insufficiency should be treated in aggressive fashion. This gives me few problems. At Children's Memorial Hospital in Chicago, we have a long tradition of favouring early insertion of new pulmonary valves in patients with significant pulmonary valvar insufficiency following repair of tetralogy of Fallot. Our results with this strategy in the current era were first presented in 1986 at the Western Thoracic Surgical Association.[1] At that time, we had inserted pulmonary valves late following repair of tetralogy of Fallot in 42 patients. We postulated that early control of pulmonary insufficiency may prevent long-term deterioration in right ventricular function. The fact that this is a controversial issue became immediately apparent during the discussion of our presentation. Dr. Frank Spencer, from New York, stated "I would completely disagree with the concept of electively inserting a porcine valve in a child on the basis of haemodynamic data. I fear that the approach recommended in this presentation is probably treating one disease by creating a worse one."

Several questions, therefore, emerge as part of the controversy regarding these patients.

- Why insert a pulmonary valve after repair of tetralogy of Fallot?
- What are the risks of inserting a pulmonary valve?
- What is the benefit to the patient of insertion of the valve?
- What is the long-term result of insertion?
- What is the risk of re-replacement of the pulmonary valve?

In this review, I will attempt to answer all of these questions, which hopefully will substantiate my viewpoint that severe pulmonary valvar insufficiency should be aggressively treated.

Why insert a pulmonary valve after repair of tetralogy of Fallot?

The answer to this starts with the fact that many patients who have undergone repair of tetralogy of Fallot have had a transjunctional, or so-called "transannular", patch. Of necessity, insertion of such a patch across the ventriculo-pulmonary junction creates severe pulmonary valvar insufficiency. This, in many patients, leads to right ventricular dilation. This, in turn, leads to tricuspid valvar insufficiency and, eventually, in some patients, to right ventricular failure. All of the above problems can contribute to postoperative arrhythmias and the potential for sudden death. In the series of patients described from the Mayo Clinic in 2001,[2] valves had been inserted in 42 patients following repair of tetralogy of Fallot. The indications for insertion were decreased exercise tolerance in three-fifths, right heart failure in one-fifth, arrhythmia in one-sixth, syncope in one-tenth, and progressive right ventricular dilation in about one-fifteenth.

In another series of patients from our own hospital,[3] statistical analysis demonstrated a significant relationship between reduced work performance and residual disease after repair of tetralogy of Fallot, including, in particular, pulmonary valvar incompetence. Carvalho et al.[4] had studied the exercise capacity of patients after complete repair of tetralogy of Fallot. They concluded that impaired exercise capacity

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was directly related to the degree of residual pulmonary regurgitation. In that study, patients with an abnormal maximal uptake of oxygen, less than 85 percent of the predicted normal valve, had significantly greater residual pulmonary regurgitation than those in whom uptake of oxygen was normal.

Marie et al.[5] evaluated the influence of right ventricular volume and pressure overload on inducible, sustained ventricular tachycardia in patients who had undergone surgical repair of tetralogy of Fallot. They concluded that the presence of right ventricular systolic overload, be it barometric or volumetric, was an important independent predictor, which suggests that increased right ventricular systolic wall stress may be an essential determinant of the inducibility of sustained ventricular tachycardia. Khairy et al.[6] recently showed that patients with such inducible sustained polymorphic ventricular tachycardia are at increased risk of future clinical ventricular tachycardia and sudden cardiac death.

Gatzoulis et al.[7] have evaluated the relationship of prolongation of the QRS complex to right ventricular size and the ability to predict ventricular arrhythmias. They concluded that chronic right ventricular volume overload due to pulmonary insufficiency after repair of tetralogy of Fallot is related to diastolic function, and correlates with the degree of prolongation of the QRS complex. The risk of symptomatic arrhythmia is high in the setting of marked right ventricular enlargement and marked prolongation of this complex. A duration on the resting electrocardiogram of greater than or equal to 180 milliseconds is one of the most sensitive predictors of life-threatening ventricular arrhythmias yet described.

Recently, magnetic resonance imaging has been used to assess the hemodynamic effect of inserting a pulmonary valve in adult patients after repair of tetralogy of Fallot. Vliegen et al.[8] concluded that, in such adult patients with pulmonary regurgitation and right ventricular dilation, insertion of a new valve produces remarkable haemodynamic improvement of right ventricular function. They therefore advocated a "less restrictive" approach to inserting a valvein these patients. All of the above answer to the question "Why insert a pulmonary valve after tetralogy of Fallot repair?" Now, what are the risks of inserting the valve?

What are the risks of inserting a pulmonary valve?

Table 1 shows a meta-analysis of 202 patients who have undergone replacement of the pulmonary valve late after repair of tetralogy of Fallot.[2,9–14] Mortality was low at 1 percent. This risk must now be compared to the benefit to the patient of inserting the valve.

Table 1. Meta-analysis of 202 patients who underwent replacement of the pulmonary following surgical repair of tetralogy of Fallot.

Institution	Year	#Pts	Mortality
Mayo Clinic[2]	2001	42	1
University of Toronto[9]	2000	55	0
Cliniques Universitaires Saint-Luc-Brussels[10]	1998	15	0
Children's Memorial Hospital-Chicago[11]	1997	70	1
Tufts-Boston[12]	1993	16	0
SUNY Health Science Center at Syracuse[13]	1985	11	0
University of California-San Francisco[14]	1983	12	0
		202	2 (1%)

Abbreviation: SUNY: State University of New York

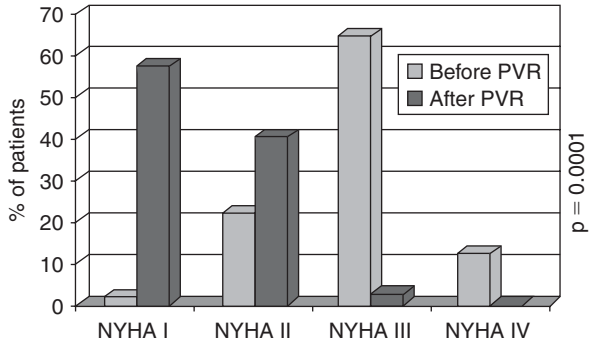

Figure 1.
The New York Heart Association (NYHA) Class before and after insertion of the pulmonary valve (PVR) in the series of 42 patients reported from the Mayo Clinic. From Discigil B, Dearani JA, Puga FJ, et al. Late pulmonary valve replacement after repair of tetralogy of Fallot. J Thorac Cardiovasc Surg 2001; 121: 344–351. Reproduced with permission.

What is the benefit to the patient of insertion of the valve?

The benefits include increased exercise tolerance, decreased risk of supraventricular and ventricular tachycardia, reversal of right ventricular dilation, and preservation of right ventricular function. In this respect, analysis of the series reported from the Mayo Clinic[2] showed that valvar insertion significantly improved right ventricular function, functional class (Fig. 1), and reduced the incidence of atrial arrhythmias, and showed that it can be performed with low mortality. Bove et al.[13] also concluded that such insertion if a valve resulted in objective improvement in right ventricular function. Insertion of the valve produced a dramatic decrease in heart size (Fig. 2). Warner et al.,[12] having replaced the valve with cryopreserved allografts, also demonstrated improved exercise

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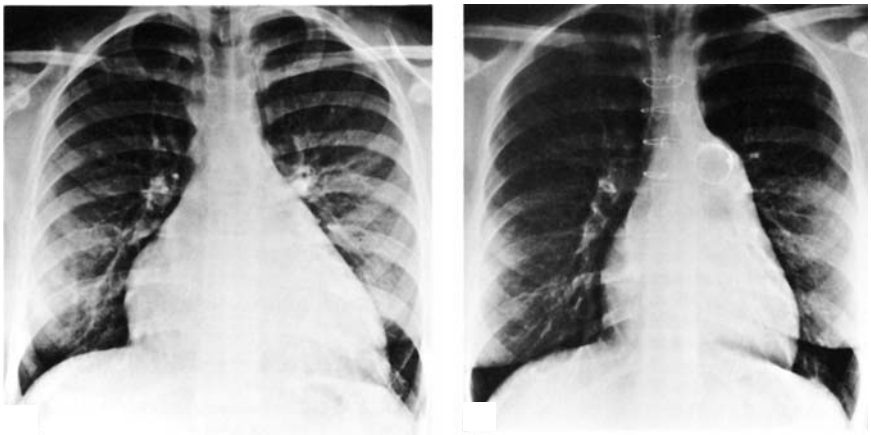

Figure 2.
Significant cardiomegaly on chest X-ray preoperatively. Following insertion of the pulmonary valve, there is a dramatic decrease in the size of the right ventricle and right atrium. From Bove EL, Kavey R-EW, Byrum CJ, et al. Improved right ventricular function following late pulmonary valve replacement for residual pulmonary insufficiency or stenosis. J Thorac Cardiovasc Surg 1985; 90: 50–55. Reproduced with permission.

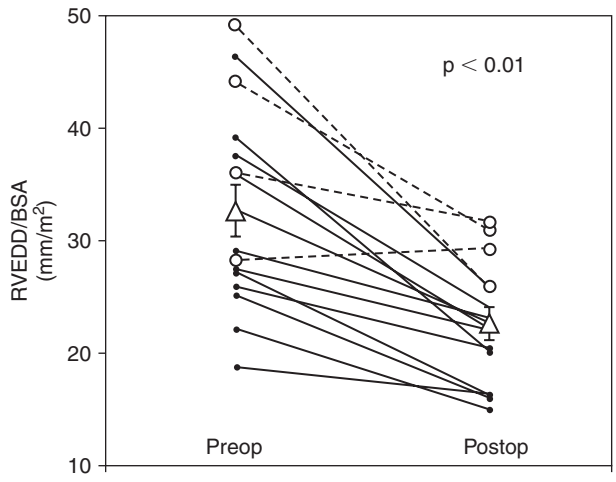

Figure 3.
Graph showing preoperative and postoperative measurements of right ventricular end-diastolic diameter (RVEDD) indexed to the body surface area (BSA) for group 1 (trace or mild regurgitation, solid lines) and group 2 (moderate regurgitation, dashed lines) after insertion of a pulmonary valve. These values decreased in all but one patient. From Warner KG, Anderson JE, Fulton DR, et al. Restoration of the pulmonary valve reduces right ventricular volume overload after previous repair of tetralogy of Fallot. Circulation 1993; 88: 189–197. Reproduced with permission.

tolerance and diminished right ventricular volume overload (Fig. 3). Having summarized the reasons for inserting the valve, and stressing the low mortality insertion, as well as the benefits of insertion, we can turn our attention to the long-term results.

What is the long-term result of insertion?

In the series reported from the Mayo Clinic, survival at 10 years was 76 percent.[2] In that same series, freedom from valvar replacement was 93 percent at 5 years, and 70 percent at 10 years. In our own experience at Children's Memorial Hospital in Chicago,[11] the freedom from replacement was 84 percent at 10 years.[1] A study from Sapporo, in Japan, showed a 90 percent freedom from valvar replacement at 5 years.[15]

What is the risk of re-replacement of the pulmonary valve?

In the series from the Mayo Clinic,[2] re-replacement was needed in 8 patients, with no mortality. At our institution,[11] the valve has needed to be replaced again in 10 patients, but again with no mortality. Thus, replacing the valve is an operation carrying low risk, even on the second occasion, despite requiring a repeat sternotomy.

Conclusions

Why should we insert a pulmonary valve after repair of tetralogy of Fallot? As I have shown, the insertion of a new valve improves exercise tolerance, decreases the risk of arrhythmias, decreases the duration of the QRS complex, preserves right ventricular function, has a very low operative risk, has excellent long-term valve durability, and carries a very low risk should the valve need to be replaced again on further occasions. Insertion of a new valve, therefore, is definitely beneficial for patients with pulmonary insufficiency after surgical repair of tetralogy of Fallot, the more so for those patients with decreased exercise tolerance, dilation or failure of the right heart, and arrhythmias. I find it hard to believe that my colleague, Bill Gaynor, would be able to sway you from the opinion that severe pulmonary valvar insufficiency following repair of tetralogy of Fallot is deserving of aggressive treatment.

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