Cardiol Young 2005; 15 (Suppl. 1): 68–71
© Cambridge University Press
ISSN 1047-9511

Severe pulmonary insufficiency should be conservatively treated

J. William Gaynor

Daniel M. Tabas Endowed Chair in Pediatric Cardiothoracic Surgery, The Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, United States of America

It's always hard to follow a presentation by Carl Backer. Today, we have heard several presentations about the problem of pulmonary insufficiency. We have been shown multiple images of dilated and poorly functioning right ventricles. The obvious conclusion is that these patients should undergo replacement of the pulmonary valve. I agree with Carl that operative morbidity and mortality for replacing the pulmonary valve is minimal, but we must also consider the outcomes in the long-term. Most physicians would agree that valvar replacement is indicated in a symptomatic patient with right heart failure and exercise intolerance. In order to win this debate, therefore, it will necessary to redirect and alter the question! The question I am going to consider is: "should we replace prophylactically the pulmonary valve in asymptomatic patients with pulmonary insufficiency and right ventricular dilation?

The original title of our debate was "Severe pulmonary insufficiency should be selectively treated". I think that "selectively" is a better term than "conservatively". We should replace regurgitant pulmonary valves in asymptomatic patients only on a selective basis. As you know, the title of Carl's presentation was "Severe pulmonary insufficiency should be *aggressively* treated". I put it to you that we don't necessarily want to be aggressive in treating these patients – we want to be selective. It is important that we are precise in our use of language. In this light, I wanted to know what is meant by "aggressive surgery"? So I went to the ultimate source, the Oxford English Dictionary. Here we find that "Agression" is defined as "an unprovoked attack and assault". "Aggressive" is, "of and pertaining to aggression". An "aggressor" is, "he who sets upon, attacks, or assails another. He who makes the first attack or takes the first step of provoking a quarrel." This is important, since some of you may think that this is just a synonym for a surgeon, but I hope that's not true. So what is "selective" surgery? "To choose or pick out a preference to another or others, having quality of faculty of selecting characterized by choice or selection." And conservative? "Characterized by a tendency to preserve or keep intact". If we choose from the options offered by the Oxford English Dictionary, therefore, I would suggest that, rather then being aggressive, we need to be selective, and we need to be conservative.

There is no prosthesis that will not fail, and will not have complications. The perfect prosthetic valve does not exist. When do the benefits of replacing a valve outweigh the risks? Again the question we must consider is: "when is prophylactic pulmonary valvar replacement indicated?" We must consider the long-term outcome for these patients, incorporating not just the operative mortality of the initial procedure, but also the risks of reintervention, either in the catheter laboratory or in the operating suite. There is little argument about patients with severe symptoms, following an episode of sudden death, or those with severe right ventricular dysfunction and exercise intolerance. Replacement of the pulmonary valve is indicated. The difficult clinical situation is the patient with asymptomatic right heart dilation and pulmonary insufficiency. Does replacement of the pulmonary valve improve their survival, and their quality of life over the long-term?

What are the goals of replacement of the pulmonary valve? Restoration of normal anatomy? Restoration of normal function, of normal life span, normal physiology? Freedom from long-term sequels? Can we achieve any of these goals? Replacing the pulmonary

Correspondence to: J. William Gaynor MD, Division of Cardiothoracic Surgery, The Children's Hospital of Philadelphia, 34th Street and Civic Center Boulevard, Suite 8527, Philadelphia, PA 19104, USA. Tel: +1 215 590 2708; Fax: +1 215 590 2715; E-mail: gaynor@email.chop.edu

https://doi.org/10.1017/S104795110500106X Published online by Cambridge University Press

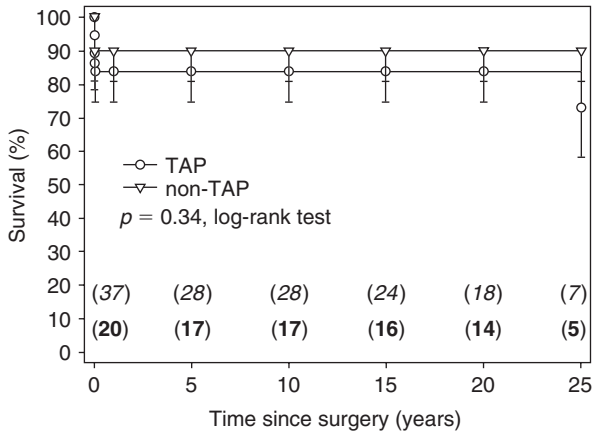


Figure 1.
Kaplan–Meier estimates of overall survival according to type of repair ($p = 0.34$, log-rank test). Error bars depict lower 95 percent confidence limits. TAP: transannular patch. (From Bacha EA, et al. J Thorac Cardiovasc Surg 2001; 122: 154–161, reprinted with permission.) Numbers of patients in follow-up are shown in parentheses in italics (TAP) and boldface (non-TAP).

valve will not make the anatomy normal, and probably will not normalize the ventricular function. By inserting a prosthetic valve, we are trading one disease for another. Hopefully a lesser disease, but it is still a disease. Replacing the pulmonary valve, therefore, does not reconstitute normal anatomy, normal function of the valve, or necessarily normal function of the right ventricle.

If we are to determine if replacement of the pulmonary valve results in restoration of normal life span, it is important to know if use of a so-called transannular patch, in reality a transjunctional patch, adversely affects long-term survival. Investigators from Boston Children's Hospital evaluated patients following repair of tetralogy of Fallot between 1972 and 1977.[1] During this era, it was customary to use large transannular incisions, inserting large transannular patches, with the goal of leaving no residual obstruction. There is a difference in early mortality between patients with and without a transannular patch, but subsequently until 25 years, the survival curves are essentially flat and parallel (Fig. 1). For this cohort, the life expectancy is similar regardless of the type of repair, but worse than for the general population. Thus, restoring a competent pulmonary valve might not alter life expectancy.

So do we restore normal physiology? Hazekamp et al.[2] evaluated replacement of the pulmonary valve in 51 symptomatic patients after repair at a mean age of 26 years. The majority exhibited decreased exercise tolerance. Symptomatic ventricular arrhythmias were present in one-tenth. Operative mortality was low, and follow-up was short, at 1.7 plus or minus 1.4 years, but overall there was improvement in functional class. Preoperative and postoperative right ventricular volumes were examined in only 13 patients, but a decrease in the extent of dilation was documented. There was no improvement in right ventricular ejection fraction, and no decrease in ventricular arrhythmias or duration of the QRS complex. Thus, replacing the valve resulted in improved exercise tolerance, and improved functional class, but it is not clear that the physiology is significantly improved. In addition, there is no evidence that the improvement persists.

Investigators at Emory University[3] evaluated 100 patients following replacement of the pulmonary valve at a median age of 8 years. The majority had symptomatic right ventricular dysfunction. Operative mortality was very low. This study provides evidence that we can do this operation safely; but it does not answer the question "should we"? Nor does it tell us for which patients? The authors do not present functional data, so we really do not know if the patients improved. For the patients less than 3-years old, only two-fifths were free from re-operation at 8 years of follow-up. The outcomes were better for older children. There was a suggestion that the porcine valves had a better outcome than homografts. This study again confirms that we can replace the pulmonary valve safely but, particularly for our smaller patients, there is a significant risk for re-operation.

Therrien et al.[4] evaluated 25 patients using preoperative and postoperative radionuclide angiography after replacement of their pulmonary valves. They found no significant improvement in diastolic volumes, systolic volumes, or right ventricular ejection fraction. They proposed, nevertheless, that the pulmonary valve should be replaced early in the patients with right ventricular dilation, prior to the development of right ventricular dysfunction. In other words, they proposed prophylactically to insert a valve in order to prevent a subsequent decline in right ventricular function. In half of the patients who had "preserved preoperative function", defined as an ejection fraction greater than 40 percent, however, the ejection fraction decreased following valvar replacement. It is difficult to read this report and conclude that replacement of the pulmonary valve is justified in an asymptomatic patient, and will improve right ventricular function.

Replacement of the pulmonary valve has been advocated to prevent ventricular arrhythmias and sudden death. It may be effective, but only if combined with an ablative procedure.[5] The data suggests that, without an ablation, replacement of the valve alone retains a significant incidence of recurrent ventricular arrhythmia. Thus, pulmonary valvar replacement alone is unlikely to prevent symptomatic arrhythmias and decrease the incidence of sudden death.

https://doi.org/10.1017/S104795110500106X Published online by Cambridge University Press

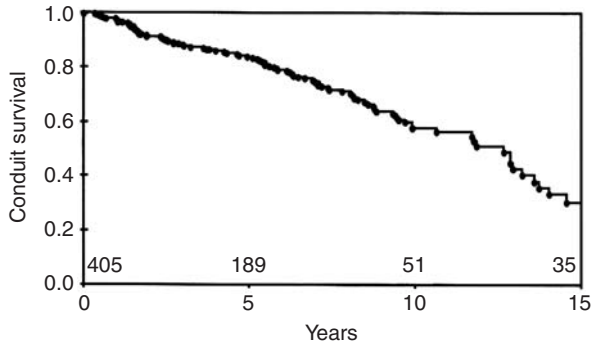


Figure 2.
Survival of homograft conduits at 5, 10, and 15 years. (From Stark J, et al. J Thorac Cardiovasc Surg 1998; 115: 506–516, reprinted with permission.)

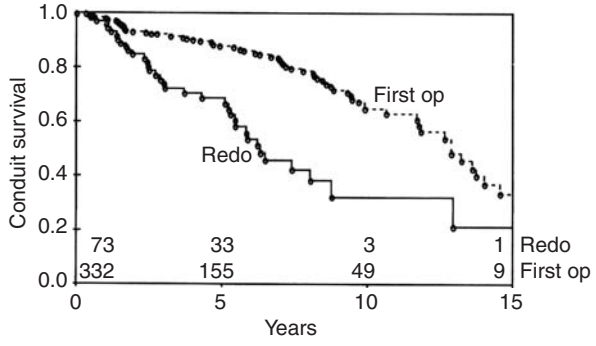


Figure 3.
Survival of first conduits compared with survival of replacement conduits. (From Stark J, et al. J Thorac Cardiovasc Surg 1998; 115: 506–516, reprinted with permission.)

So do we restore normal life span? This is unlikely. Do we restore normal physiology? The data is inconsistent. Some people get better, some people get worse. For those patients who are asymptomatic, an important question is freedom from long-term sequels? Investigators at The Great Ormond Street Hospital for Sick Children[6] evaluated the determinants of failure for homografts placed between the right ventricle and the pulmonary arteries. They evaluated a variety of homografts, preserved in different ways, over a long period of time. They found a fairly consistent rate of increasing failure with increasing duration of time following implantation (Fig. 2). An interesting finding is that the homograft inserted at the first operation was more durable than subsequent conduits (Fig. 3). This is an important issue for young patients, who will require insertion of multiple conduits over their lifetime. In the multivariable analysis, reoperation and later conduits compared to the first conduit were significantly associated with more rapid failure. Similar data is available from investigators in Toronto,[7] who have been advocates of replacing the pulmonary valve. They followed a large number of patients,

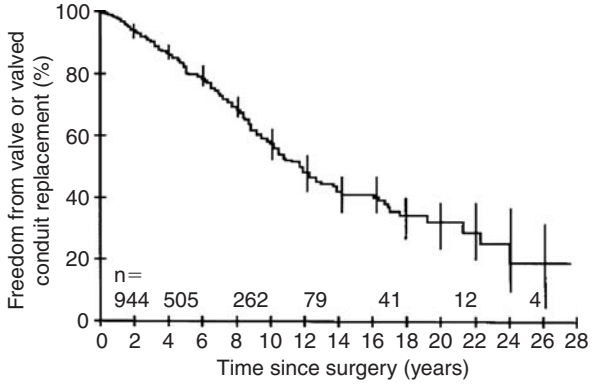


Figure 4.
Kaplan–Meier estimation of freedom from valvar replacement. Vertical lines represent 95 percent confidence intervals. The freedom from valve replacement at 5 years is 82 percent, at 10 years is 58 percent and at 15 years is 41 percent. (From Caldarone CA, et al. J Thorac Cardiovasc Surg 2000; 120: 1022–1031, reprinted with permission.)

finding a significant need for re-operation. By 15 years, freedom from re-operation in their patients is only 41 percent (Fig. 4).

So do we achieve the goals of replacing the pulmonary valve? Restoration of normal anatomy? No. Normal function? No. Normal lifespan? Unlikely. Normal physiology? The data is inconsistent. Freedom from long-term sequels? No. Based on this data, should we replace the valve, or should we just accept the natural history of pulmonary valvar insufficiency. The latter option is also not appropriate. What we need, therefore, is to establish the appropriate indications for pulmonary valvar replacement!

I advocate replacement of the pulmonary valve for symptomatic patients. Patients with exercise intolerance, and heart failure secondary to pulmonary insufficiency, probably should have their valves replaced. A major goal for our patients is to provide as good a quality of life, and as normal a life, as is possible. If patients are unable to exercise, and they are in cardiac failure, then we have not attained this goal. If serial studies show that the right ventricle is pregressively dilitating, and that there is severe tricuspid regurgitation, I find it difficult to believe that such a patient is truly asymptomatic. In a patient who has a dilated but stable right ventricle, with a lot of pulmonary insufficiency, but who is asymptomatic, I don't think we can justify insertion of a valve, either on the basis of what has been proposed by my opponent, or according to the data already published. For the symptomatic patient, who has heart failure, is in a bad functional class, who suffers from exercise intolerance, then yes, we should replace their valves. Ventricular tachycardia with right ventricular dilation associated with syncope or sudden death may be an indication.

https://doi.org/10.1017/S104795110500106X Published online by Cambridge University Press

Replacement must be undertaken, however, in conjunction with ablation. It is a different story when we're considering prophylactic operations hoping to preserve right ventricular function. I can find no evidence that this can be achieved. If it is to be attempted, it should be done as part of a trial that evaluates carefully the function and the durability of the result.

References

1. Bacha EA, Scheule AM, Zurakowski D, et al. Long-term results after early repair of tetralogy of Fallot. J Thorac Cardiovasc Surg 2001; 122: 154–161.
2. Hazekamp MG, Kurvers MMJ, Schoof PH, et al. Pulmonary valve insertion late after repair of Fallot's tetralogy. Eur J Cardiothorac Surg 2001; 19: 667–670.
3. Kanter KR, Budde JM, Parks JW, et al. One hundred pulmonary valve replacements in children after relief of right ventricular outflow tract obstruction. Ann Thorac Surg 2002; 73: 1081–1087.
4. Therrien J, Sui SC, McLaughlin PR, et al. Pulmonary valve replacement in adults late after repair of tetralogy of Fallot: are we operating too late? J Am Coll Cardiol 2000; 36: 1670–1675.
5. Therrien J, Sui SC, Harris L, et al. Impact of pulmonary valve replacement on arrhythmia propensity late after repair of tetralogy of Fallot. Circulation 2001; 103: 2489–2494.
6. Stark J, Bull C, Stajevic M, et al. Fate of subpulmonary homograft conduits: determinants of late homograft failure. J Thorac Cardiovasc Surg 1998; 115: 506–516.
7. Caldarone CA, McCrindle BW, Arsdell GS, et al. Independent factors associated with longevity of prosthetic valves and valved conduits. J Thorac Cardiovasc Surg 2000; 120: 1022–1031.

https://doi.org/10.1017/S104795110500106X Published online by Cambridge University Press