Surgical Treatment of Pulmonary Hypertension, A Proposal for A Novel Procedure: The Khouqeer Shunt

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Citation

Abstract
Pulmonary hypertension is a dreadful pathological state that carries very high mortality. It has an average survival of 2 - 3 years from the onset of heart failure. Unfortunately, this holds true in spite of major progress in therapy. We here put forward a novel procedure to palliate this condition at its late stages (or even may help to halt its progression in earlier stages) in hope to improve the natural history, of patients suffering from pulmonary hypertension. The proposed connection shunts blood from the pulmonary artery or one of its branches, away from the lung, to the left atrium or one of the pulmonary veins. This maneuver will maintain the cardiac output at the expense of arterial saturation.

INTRODUCTION
Pulmonary hypertension is a dreadful pathological state that carries very high mortality, in spite of progress in suggested treatments. We here put forward a concept for a novel procedure to palliate this condition which may improve the natural history, of patients suffering from pulmonary hypertension.

PATHOLOGY OF PULMONARY HYPERTENSION
There are seven different entities; plexogenic arteriopathy, embolic and thrombotic pulmonary hypertension, pulmonary venous hypertension, pulmonary veno-occlusive disease, hypoxic pulmonary hypertension, pulmonary hypertension in lung fibrosis and primary pulmonary hypertension. The pathophysiological disturbances associated with all these entities are uniquely similar and a new classification is now proposed by the World Health Organization. Cyanosis, arrhythmia, increased pulmonary artery pressure, right side heart failure and premature death. Increased pulmonary artery pressure will lead to stretching a patent foramen ovale with resultant right to left shunt and contribute to cyanosis. Developing pulmonary arterial venous connections may result in hemoptysis and contribute to cyanosis. Distension, dilation and hypertrophy of the pulmonary arteries, the right ventricle and right atrium will result eventually in failure; and that is the most common cause of death in pulmonary hypertension. In addition, this may lead to more arrhythmia and sudden death. Erythropoietic response to cyanosis will promote erythrocytosis, and with phlebotomy, microcytosis will increase hyperviscosity and its likely complications such as cerebrovascular and thromboembolic events.

SURVIVAL
Once the patient develops irreversible pulmonary vasculopathy and heart failure, death usually occurs with a mean survival of only 2 - 3 years. Some patients with pulmonary hypertension secondary to severe chronic mitral stenosis may show regression post operatively, but in general, they die prematurely if they develop frank right ventricular failure. Different modalities of therapy have changed the course of the disease significantly. Most of the reports implicate right ventricular failure to be the number one cause of death followed by the arrhythmogenic sudden death. Pneumonia is the next but far less cause of death. The main reasons for the clinical neglect of primary pulmonary hypertension have been its dismal prognosis when untreated, the absence of effective treatments. However, the management of primary pulmonary hypertension has been revolutionized by the demonstration that long-term intravenous epoprostenol (Prostacyclin) improves survival and quality of life for patients with this disorder, but it still dismal by any standard. In this group median survival to only few months if mean right atrial pressure exceeds 10mmHg, the mean pulmonary artery pressure exceeds 85 mmHg, or the cardiac index is less than 2 L/min/m².
SURGICAL RATIONAL

The best surgical procedure would be a treatment modality that alleviates the pulmonary hypertension to improve the cardiac output and reduce the chances of arrhythmogenic irritable right ventricle and in this day in age be cost effective. Theoretically, this should improve survival. In spite of treatment of primary pulmonary hypertension with continuous, intravenous Prostacyclin (PGI2), lung transplantation and vasodilators including Calcium channel blockers, the natural history reveals appalling premature death (Fig.1).3,5,6,7,8

There are several observations suggesting that atrial shunting in primary pulmonary hypertension seems to give better survival. It has been shown that patient with congenital heart disease, pulmonary hypertension and Eisenmenger syndrome live longer. Patients with primary pulmonary hypertension and a patent foramen ovale live longer.5,11 The presence of an atrial septal communication in this setting will increase the left ventricular output. The worry, being of uncontrolled size, would be the development of too much or too little shunt, with subsequent severe desaturation or inadequate relieve of the right sided pressures respectively. Other surgical techniques include a fenestrated Fontan, and 2 patches ventricular septal defect closure.12,13,14 Both these techniques allow shunting away from the pulmonary circulation and result in great reduction of the postoperative morbidity associated with these high-risk procedures. There is one report of interest that describes a temporary aorto-pulmonary shunt post truncus repair, but it is constructed between two high-pressure systems. Therefore, it will not lower the pulmonary pressure below that of the systemic circulation.

The new proposed technique is a tube graft shunt to be placed between the high-pressure system (pulmonary artery or one of its main branches) and the low-pressure system (the left atrium or one of the pulmonary veins). This would have many advantages such as:

1. Shunting at level distal to the right ventricle would decrease the wall tension and reduce systolic and diastolic pressures of the right ventricle, compared to shunting at the atrial level. This is more likely to reduce arrhythmic death and help to alleviate the right sided failure

2. The relatively high gradient across the shunt will keep it open longer compared to the balloon atrial septostomy, 5 with less need for reintervention.

3. The length and the size of the tube graft can be estimated mathematically preoperatively according to the blood flow characteristics, the current and the desired postop hemodynamic.

4. By lowering the PA pressure and maintaining the cardiac output, some regression of the vascular pathology may be noticed.16 Though desaturation may elicit other mechanisms for pulmonary vasculopathy.

5. This shunt can be preformed in conjunction with primary repair of a cardiac defect (ventricular or atrial septal defects) or even in cases for pulmonary endarterectomy associated with systemic values of pulmonary hypertension. There is of course the possibility of transcatheter occlusion if no longer needed later.

6. Much less cost will be expected as compared with the long term medical treatment , lung or heart-lung transplantation.

CONCERNS

The application of this new surgery has to be in accord with the standard set forward by the American College of Surgeons.18 The possibility of endocarditis, the incidence of neurological complications, left-sided heart failure will be acceptable, perioperative mortality as the post-cardiac catheterization mortality are recognized risks. Acceptable saturation post operatively should be in the range of 60-70%, in order to minimize this risk.4 This may be a significant factor to assure that a minimum percentage of the hepatic venous drainage perfuse the pulmonary bed, to reduce the formation of arterio-venous malformation in the lung. A sound experimental protocol for randomized multicenter study will be highly recognized and appreciated in the medical community.

References

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