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Arterial Switch for Pulmonary Venous Obstruction Complicating Mustard Procedure

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Two patients underwent an arterial switch procedure for the relief of severe pulmonary venous obstruction complicating a Mustard procedure. Without preparatory pulmonary banding, both patients had adequate left ventricular function due to secondary pulmonary hypertension. At 8 and 4 years after the procedure, both patients are in New York Heart Association functional class I, with echocardiographic evidence of good left and right ventricular function.

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Currently, the arterial switch operation is the operation of choice for transposition of the great arteries [1]. In earlier times, atrial rerouting as in the Mustard procedure, was performed with good early and medium-term results. However, right ventricular dysfunction may be troublesome in the long term [2, 3]. The pulmonary venous obstruction that can arise after a Mustard procedure is a serious complication [1, 4]. Elimination of the obstruction through the placement of a patch carries the risk of restenosis. To overcome the risks of recurrent pulmonary venous obstruction after atrial repair and the long-term complications of the atrial switch procedure, an arterial switch operation should be considered, especially in patients with secondary pulmonary hypertension due to pulmonary venous obstruction. These patients may have a well-trained left ventricle that is capable of supporting the systemic circulation. Two patients underwent a single-stage conversion from a Mustard construction to an arterial switch for the relief of symptomatic pulmonary venous obstruction, without the need for preparatory pulmonary artery banding.

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Table 1. Variables Before Arterial Switch Procedure

Variable	Patient 1	Patient 2
Weight (kg)		
Electrocardiogram	Biventricular hypertrophy	Biventricular hypertrophy
Echocardiography		
Diastolic left ventricular posterior wall thickness (mm)	8 (95th percentile)	6 (75th percentile)
Shortening fraction	0.30	0.29
Cardiac catheterization		
Ejection fraction	0.70	0.65
Left ventricular pressure (mm Hg)	80/5–10	71/1–7
Pulmonary artery pressure (mm Hg)	69/36	68/39
Mean capillary wedge pressure (mm Hg)	29	30
Right ventricular pressure (mm Hg)	91/0–10	95/1–8
Left ventricular-to-right ventricular ratio	0.88	0.75

Case Reports

Patient 1

This patient underwent a Mustard procedure in 1976 at the age of 6 weeks for the repair of transposition of the great arteries with an intact ventricular septum and atrial septum defect, after unsuccessful balloon atrial septostomy. A Mustard procedure with a Teflon baffle was performed using hypothermia and circulatory arrest. The patient was reoperated on twice in the first year of life because of pulmonary venous obstruction. In the first reoperation, a dura mater patch was placed. The second reoperation involved the use of a Teflon patch. In 1985, obstruction recurred with secondary pulmonary hypertension. This caused the patient to be in New York Heart Association functional class IV and ventilator dependent. Preoperative electrocardiography showed biventricular hypertrophy. Echocardiography showed dilatation of the pulmonary veins. The size of the left (pulmonary) ventricle was equal to that of the right (systemic) ventricle, with the left ventricular posterior wall thickness and shortening fraction within normal limits for a systemic ventricle. These findings suggested the existence of severe left ventricular pressure overload (Table 1). Cardiac catheterization showed a high pulmonary capillary wedge pressure, elevated left ventricular and pulmonary arterial pressures, and good left ventricular contractility (see Table 1).

After left femoral artery cannulation and venous cannulation of the right pulmonary artery, because of the

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inaccessibility of the caval veins, extracorporeal circulation was started. Under conditions of deep hypothermia (16°C) and circulatory arrest, an arterial switch using the Lecompte procedure was performed [5]. After excision of the Teflon patch placed during the Mustard procedure, an atrial septum was reconstructed using a USCI patch (C.R. Bard, Billerica, MA). The patient's postoperative course was complicated by a cerebrovascular accident that left her with temporary hemiplegia and pseudobulbar paresis. After a long period of rehabilitation, she recovered completely. Eight years later she is in New York Heart Association functional class I. The electrocardiogram shows a junctional rhythm, and echocardiography shows normal left ventricular function, with still some right ventricular hypertrophy and mild aortic insufficiency.

Patient 2

This patient was admitted with persisting cyanosis after birth, and the diagnosis of transposition of the great arteries, an intact ventricular septum, and open ductus arteriosus was made. A balloon atrial septostomy was performed 2 days after birth. In 1985, at the age of 4 months, the infant underwent a Mustard procedure under circulatory arrest and hypothermia that involved the use of a Teflon baffle. In 1988 exertional dyspnea with a period of pneumonia occurred. Echocardiography revealed distended pulmonary veins and severe narrowing of the communication to the pulmonary venous atrium. The size of the left ventricle was almost equal to that of the right, and both the shortening fraction and the left ventricular posterior wall thickness were within the normal limits for a systemic ventricle (see Table 1). Cardiac catheterization showed pulmonary venous obstruction with elevated left ventricular and pulmonary arterial pressures due to secondary pulmonary hypertension. Left ventricular contractility was good (see Table 1). Subsequently, an arterial switch was performed. With the patient on extracorporeal circulation and under conditions of hypothermia (20°C) and cardioplegic arrest, an arterial switch procedure was done. The Teflon baffle was excised and the atrial septum was reconstructed with a Gore-Tex patch (W. L. Gore, Elkton, MD).

The patient had an uneventful postoperative course, and 4 years later he is in excellent clinical condition (New York Heart Association functional class I) and in sinus rhythm. Echocardiography has confirmed that left and right ventricular contractility is normal, but there is trivial aortic regurgitation.

Comment

Before the era of anatomic repair procedures, children with transposition of the great arteries were treated with a physiologic repair that involved redirecting the systemic and pulmonary flow at the atrial level [1, 6]. In the treatment of transposition with an intact ventricular septum, the atrial switch operation has been found to be associated with a minimal early mortality at many centers, and the actuarial survival results over the first 10 years have been excellent. However, late complications such as rhythm disturbances, right ventricular dysfunction, tricuspid valve incompetence, and sudden death have been reported [2, 3, 7]. This possible long-term outcome after atrial repair is a major argument in favor of the arterial switch procedure. The arterial switch repair can be accomplished with equally good or even better early results and better intermediate results [1, 8]. This is the basis for considering an arterial switch procedure when managing the complications of the atrial switch operation. Cochrane and associates [2] and Mee [3] described the technique for converting from an atrial switch to an arterial switch for the treatment of right ventricular failure. This was done in two stages, with the first stage consisting of banding the pulmonary artery to retrain the left ventricle. Once adequate left ventricular pressure developed, the arterial switch operation was performed.

We describe a one-stage procedure of converting from an atrial switch to an arterial switch for the treatment of pulmonary venous obstruction. Pulmonary venous obstruction is a serious complication after a Mustard atrial switch [1, 4]. Repair using a patch may result in restenosis, as illustrated by patient 1. The pulmonary venous obstruction with secondary pulmonary hypertension may cause the left ventricle to be well trained and thus able to support the systemic circulation without the need for preparatory pulmonary artery banding, as was shown in both our patients (see Table 1). Echocardiography appeared to be a very valuable tool in the selection of candidates for the one-stage approach. After a successful atrial repair for transposition of the great arteries, the left ventricle is flattened due to its low systemic pressure. Echocardiography showed that, in both our patients, the left and right ventricular volumes were comparable. Left ventricular posterior wall thickness had increased to within normal limits for a systemic ventricle. The left ventricular posterior wall shortening fraction was in the lower normal range. However, we believe that the decision to perform a one-stage repair in this setting can only be made adequately with invasively obtained hemodynamic data, using as the cutoff a minimal left ventricular-to-right ventricular pressure ratio of 0.75, as suggested by Mee [3] and Cochrane and colleagues [2].

We conclude that the arterial switch operation is feasible for correction of pulmonary venous obstruction after a Mustard procedure. In this regard, we think that the one-stage arterial switch operation is the operation of choice in patients with adequate left ventricular function.

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INVITED COMMENTARY

This article reports 2 patients who underwent late arterial switch repair after a Mustard procedure performed in infancy, and in whom pulmonary venous obstruction with secondary pulmonary hypertension developed. Because the left ventricular pressure was high, it was not necessary to carry out preliminary pulmonary artery banding.

This is an interesting and novel approach to the management of patients with pulmonary venous obstruction after a Mustard procedure, and fits well with our philosophy of recommending conversion to an arterial switch for all patients who have undergone Senning or Mustard procedures, and who clearly require further revisional operations in whom conversion to an arterial switch is a feasible option—either as a one-stage or two-stage procedure. In these patients, the presence of well-maintained right ventricular function puts them into a different category from that of many patients who become symptomatic and need to be considered for further operation, which, if an arterial switch is not feasible, may well involve transplantation. The fact that the Mustard baffle can be removed in its entirety as part of the conversion to an arterial switch, likely makes it the most satisfactory long-term solution to the problem of pulmonary venous obstruction—which in 1 of the 2 patients described recurred despite two previous attempts at revision.

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Use of the Native Aortic Valve as the Pulmonary Valve in the Ross Procedure

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The placement of a foreign valve in the pulmonary position using the Ross procedure requires reoperation. To circumvent this problem, we devised a method of reimplanting the native aortic valve in the pulmonary position, and successfully performed this procedure in a 12-year-old diabetic boy operated on for the treatment of aortic insufficiency. Although diseased, the reimplanted aortic valve functioned well, with trivial stenosis and insufficiency. This modification offers patients with aortic valve disease a potentially curative operation.

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The Ross procedure requires the implantation of a foreign valve in the pulmonary position using a valve that has longevity. Currently, a cryopreserved homograft is preferred for this purpose, although other materials such as autologous fascia lata and pericardial valves have been used [1]. Valveless conduits also have been used successfully in the pulmonary position, but right ventricular dysfunction is a consequence of this.

Although cryopreserved homografts are preferred and last longer in the pulmonary position, replacement nevertheless will be necessary [2]. Early failures have also been reported and have been attributed to possible immunologic rejection [3].

To circumvent the need for pulmonary valve replacement with the Ross procedure, we devised a method of implanting the native aortic valve in the pulmonary position. This was based on our belief that a diseased aortic valve, especially when the main problem is aortic insufficiency, should function reasonably well in the pulmonary position because of the lower pulmonary vascular pressure and resistance involved. We report on our experience with this method.

A 12-year-old boy was referred to us for the treatment of congenital aortic insufficiency. Cardiac catheterization performed when he was 4 years old had shown a tricuspid aortic valve with mild incompetence. The aortic insufficiency had worsened considerably over the years.

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