

Right Heart Failure in a Patient with Complete Transposition of the Great Arteries and Mustard Operation

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The complete transposition of the great arteries is a severe congenital cyanotic heart disease, which needs surgical repair. The treatment of choice in the past was the atrial switch operation (Mustard or Senning techniques). Although this procedure increases life expectancy, it is implicated in serious delayed complications and tends to be substituted by the arterial switch operation. We present the case of a 33-year-old man with complete transposition of the great arteries and Mustard operation during his first year of life. The patient suffered from refractory ascites and peripheral edema and the echocardiography implicated obstruction of the venous baffle. Due to the presence of bilateral femoral vein and superior vena cava obstruction, diagnostic transhepatic cardiac catheterization through the hepatic veins was performed. The findings were consistent with biventricular failure and the patient was referred for cardiac transplantation. The diagnostic and therapeutic approach of this patient is briefly discussed. The natural history of patients with Mustard operation and the technique of transhepatic cardiac catheterization are also reviewed in this paper.

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Complete transposition of the great arteries is a severe and potentially lethal form of congenital heart disease which consists of the origin of the aorta arising from the morphological right ventricle and that of the pulmonary artery from the morphological left ventricle. The usual clinical presentation is severe cyanosis during the first days of life and requires surgical management. The procedure of choice in the past was the atrial switch operation, which represents a rather palliative approach and is implicated with serious long-term complications. We are presenting a case of a 33-year-old man with complete transposition of the great arteries who had undergone Mustard operation during childhood. He was admitted with refractory ascites and peripheral edema.

Case description

The patient, a 33-year-old male, father of three children, presented severe central cyanosis at birth due to complete transposition of the great arteries. An urgent percutaneous atrial septostomy was performed stabilizing the patients' condition.

During the first year of life he underwent a Mustard operation (atrial switch). The intraatrial septum was resected and with the use of pericardial tissue (pericardial baffle) the flow of superior and inferior vena cava (SVC, IVC) was diverted through the mitral valve into the left ventricle (LV) and thence through pulmonary artery, while the pulmonary venous flow through the rest of the "common" atrium (called "pulmonary baffle") was diverted through the tricuspid valve and the right ventricle (RV) to the aorta.

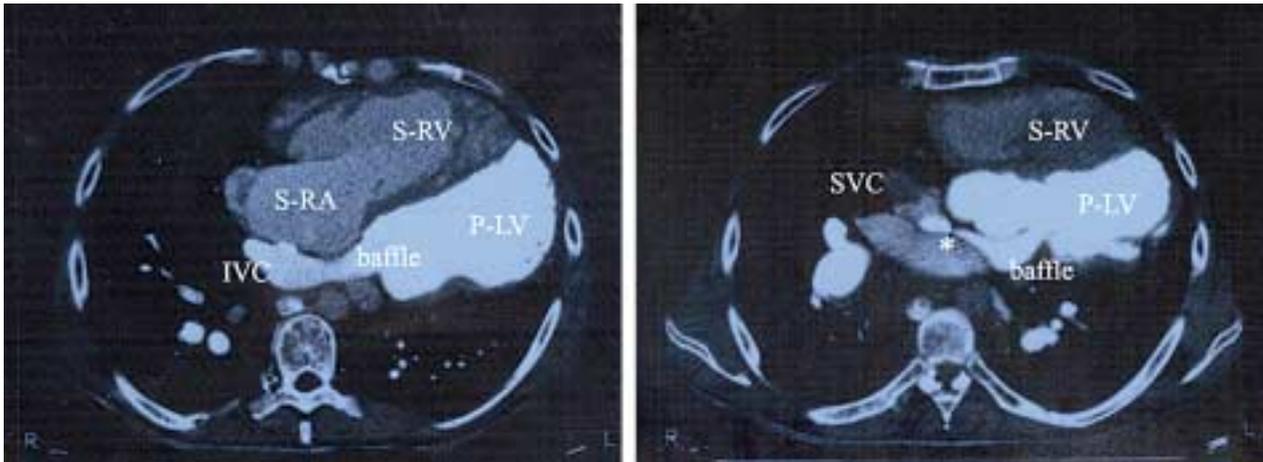


Figure 1. Computerized tomography of the heart. Left: The infusion of contrast agent shows the inferior vena cava (IVC) and the baffle which seem patent. The subpulmonary left ventricle (S-RV) is also showed. At the upper part of the picture is the systemic right atrium (SRA) and the right ventricle which supports the systemic circulation (S-RV). Right: The obstructed superior vena cava (SVC) is showed (*).

As a result, systemic and pulmonary flow were fully separated but right ventricle supported the systemic circulation

His clinical course was good until the age of 29 when routine follow-up echocardiography disclosed severe stenosis at the SVC - pericardial baffle anastomosis. The peak gradient recorded between SVC and the baffle was 19 mmHg. There was also right ventricular dilatation with severe impairment of the systolic function while LV systolic function was good. There was left ventricular outflow tract obstruction with a peak gradient of 35-40mmHg. Cardiac magnetic resonance verified the echocardiographic findings and also disclosed vein flow between SVC and IVC through the azygous vein system.

Radionuclide ventriculography showed a RV ejection fraction of 20% and LV ejection fraction of 46%.

Cardiac catheterization and coronary angiography followed and showed a dilated and hypertrophied systemic RV. There was no coronary artery disease. However, it was not possible to assess pulmonary circulation as both femoral veins were obstructed.

Subsequently, the patient developed complete atrioventricular block (AV block) and a VVI epicardial pacemaker was inserted. However the patient suffered from pacemaker syndrome during VVI pacing and it was decided to programme the pacemaker to a back-up basic rate of 40/min and leave the patient with his underlying rhythm of 60 bpm complete heart block, during which he had less symptoms.

Four years later, and during the six-month period before his current admission he developed recurrent ascites and peripheral edema. A new echocardiogram revealed subpulmonary ventricular dilation (left ventricle), which responded poorly to diuretics. A V/Q scan was consistent revealed pulmonary embolism at this stage. He was put on anticoagulants and had a CT scan (Figure 1) which confirmed SVC obstruction, dilated patent IVC and dilated hepatic veins, implying high venous pressure. The pulmonary arteries were also dilated, a finding consistent with pulmonary hypertension.

Current admission

He was admitted because of shortness of breath on minimal exertion, refractory ascites and peripheral oedema despite medical treatment. His treatment consisted of ACE inhibitor, frusemide, spironolactone and warfarin.

On clinical examination his pulse was regular, 60/min and his blood pressure was 100/60 mmHg. A RV impulse was palpable, whereas the LV impulse was displaced to the left, implying LV dilation. There was an attenuated first heart sound due to RV systolic dysfunction and a single second heart sound (due to the anterior position of aorta and posterior position of pulmonary artery). There was a 2-3/6 ejection systolic murmur radiating to the back (LV outflow tract obstruction), a 2/6 systolic murmur due to atrioventricular valve regurgitation and a soft 1-2/6 diastolic murmur on the left sternal edge, proba-

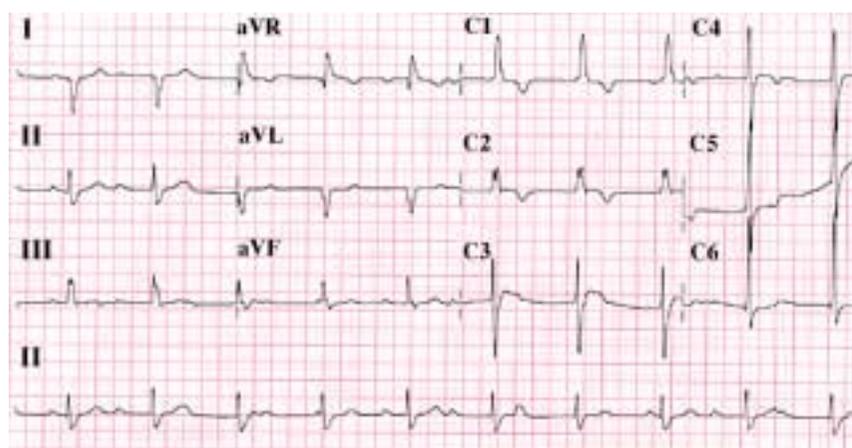


Figure 2. The patient's electrocardiogram. See text for interpretation.

bly due to baffle obstruction. The lungs were clear, the liver was palpable and tender and there was severe ascites.

ECG (Figure 2) revealed complete AV block with a ventricular rate of 57/min, right bundle branch block (QRS duration 125 msec), right axis deviation, and RV hypertrophy (RV hypertrophy is a typical finding of the complete transposition of the great vessels).

Chest X-ray (Figure 3) revealed situs solitus (normal atrial arrangement), levocardia, cardiomegaly, central pulmonary arteries dilation with normal peripheral branches and increased blood flow at the upper lung fields implying venous congestion.



Figure 3. Chest X-ray. See text for interpretation.

There was severe hyponatraemia (114 mEq/l), normal potassium and magnesium, bilirubin of 3.2 mg/dl, urea 54 mg/dl and creatinine 0.7 mg/dl.

Eleven litres of transudate were removed via abdominal puncture.

A new echocardiogram confirmed RV dilation and hypertrophy with severely impaired systolic function and moderate tricuspid regurgitation. The subpulmonary LV was mildly dilated with mildly impaired systolic function. There was dynamic LV outflow tract obstruction with a peak gradient of 25 mmHg and moderate mitral regurgitation. The intraatrial pulmonary vein baffle was patent but there was increased blood velocity (1.8 m/sec) in the intraatrial part of the systemic venous baffle with a peak gradient of 13 mmHg. Transoesophageal echocardiography confirmed that IVC and the part of the baffle near IVC were patent, but revealed intraatrial baffle stenosis at the atrial septum level with a peak velocity of 2.6 m/sec.

As this finding could be clinically significant, cardiac catheterization was decided in order to confirm or exclude IVC baffle stenosis as the reason for patient's clinical deterioration. Transhepatic cardiac catheterization was the only option because of SVC and femoral veins obstruction.

Cardiac catheterization was performed under general anaesthesia. Five attempts were made to insert a catheter in the hepatic veins through the space between lower ribs at first, and through the subxiphoid region consequently. A 5F Cobra catheter was inserted in one hepatic vein using the Seldinger technique and was advanced to the IVC. The angiographic images of IVC revealed that this was patent with satisfactory flow towards the mitral valve and subpulmonary LV (Figure 4). However the SVC baf-

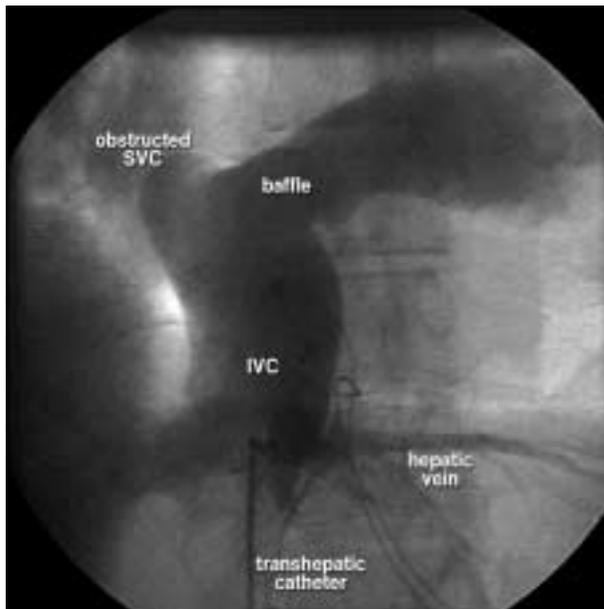


Figure 4. Transhepatic right heart catheterization. Through a hepatic vein with the infusion of contrast agent the inferior vena cava (IVC) the baffle and the subpulmonary left ventricle (S-RV) are showed. The superior vena cava (SVC) is obstructed.

ple was obstructed. LV function was severely impaired and there was moderate mitral regurgitation. The pulmonary valve was morphologically normal. There was mild main pulmonary artery dilation and there was no stenosis of the peripheral pulmonary branches, which were also mildly dilated. There was no gradient between IVC and LV, the end diastolic subpulmonary (LV) pressure was 17mmHg and there was a gradient of 34 mmHg between LV and the pulmonary artery. Pulmonary artery pressure was in the upper normal limits (Figure 5).

The above findings were consistent with biventricular failure. Except for the “expected” right ventricular failure supporting the systemic circulation, the left ventricular function was also poor due to the interdependence between the two chambers.

A twenty-four hour Holter tape revealed multifocal ventricular ectopic beats and short run (4 beats) of non sustained ventricular tachycardia (NSVT). Amiodarone was added to the medical treatment.

A cardiopulmonary exercise test was also performed. The heart rate increased to 69/min and peak oxygen consumption was 15.5 ml/kg/min (40% of predicted). As there were no further medical or sur-

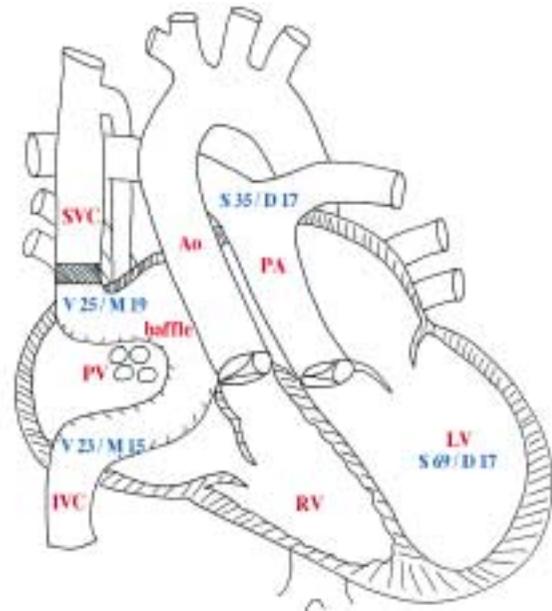


Figure 5. Figure which shows the Mustard operation. The measured pressures during the catheterization are also showed. IVC: inferior vena cava, SVC: superior vena cava, PV: pulmonary veins, RV: right ventricle, LV: left ventricle, Ao: aorta, PA: pulmonary artery, V: V wave, M: mean pressure, S: systolic pressure, D: diastolic pressure.

gical treatment options the patient was referred for cardiac transplantation. Pacemaker upgrade to DDD mode was also decided.

Discussion

Atrial switch operation is a palliative surgical therapy for transposition of the great arteries. The baffle used is made of pericardium or Dacron (Mustard operation) or atrial tissue (Senning operation). The separate circulations are corrected at the atrial level. Most centers do not employ this surgical technique any more due to the long-term complications, which consist of :

1. Systemic RV failure.
2. Moderate or severe tricuspid regurgitation (10%-40%).
3. Arrhythmias and conduction disease (atrial flutter 20% up to the age of 20 and sinus node dysfunction 50% up to the age of 20).
4. Sudden cardiac death which may occur in up to 5% of cases and may be caused by systemic ventricular failure, atrial flutter or pulmonary hypertension.
5. Severe pulmonary vascular disease in patients with non-restrictive ventricular septal defect and de-

layed surgery or patients with haemodynamically significant left-to-right shunt through the baffle.

6. Cyanosis caused by right-to-left shunt through the baffle (not very common).
7. Obstruction of the pulmonary venous pathway which may cause pulmonary venous congestion.
8. Obstruction of the IVC or SVC baffle pathway.

SVC obstruction usually does not present any symptoms as collaterals develop via the azygous vein system. Collaterals do not allow venous pressure to increase dramatically, so SVC obstruction prognosis is good, while IVC obstruction could potentially be life threatening¹. Baffle stenosis is usually diagnosed with Doppler echocardiography as the normal biphasic flow is not present and the velocity through the stenosed baffle area is more than 1m/sec⁷. Baffle balloon dilation and stenting is an option with limited success in adult patients¹.

Most adult patients with Mustard operation are in NYHA class I or II. Two to fifteen percent of these patients presented with symptoms in a 25 year follow-up while systemic ventricular systolic dysfunction was present in 40% of these patients⁶⁻⁷.

Severe biventricular dysfunction makes heart transplantation the only therapeutic option for the patient presented in this report. Heart transplantation in patients with congenital heart disease has a relatively good five- and ten-year survival¹. However, in some patients with systemic right ventricle, life expectancy could be similar to normal subjects³.

As atrial switch operation may cause long-term complications, it has been substituted by arterial switch operation (which was first performed in the late 70s) where the pulmonary artery and aorta are connected to the right and left ventricle respectively and the coronary arteries are attached to the neo-aorta. Since the correction is anatomic, it seems that the above-described operation is advantageous.

Transhepatic catheterization of hepatic veins and right heart is a technique that may be applicable in patients with complex congenital heart disease or children with obstructed systemic veins⁸⁻¹¹. The puncture area^{8,9} is usually situated in the middle of the distance between diaphragm and the lower liver margin at the middle axillary line. The procedure begins with local anaesthesia and subsequently the needle (usually 22G) is advanced under negative pressure with a horizontal or slightly posterior angle towards the liver and up to the middle of the distance to the middle line. Successful catheterization is confirmed with blood aspiration and angiographic im-

ages. Echography may be useful in guiding the needle in cases of difficult access¹¹. The guide wire is advanced to the right atrium and, subsequently, a dilator and a 5-8F sheath are inserted. It is recommended that the activated coagulation time (ACT) should be less than 200sec before sheath removal if heparin has been used. Alternatively, protamine sulfate can be used to reverse heparin action. Haemostasis with collagen based closure devices reduces bleeding complications⁸⁻⁹.

Transhepatic catheterization is a relatively safe alternative option. One patient has been reported to have undergone 11 procedures without any major complications¹², although severe endoperitoneal bleeding cases have also been reported^{10, 13}. The above-mentioned technique has also been used for bundle ablations¹⁴, pacemaker wire insertion¹⁵ and Hickman¹⁶ or haemodialysis¹⁷ catheter insertion.

Conclusions

Although Mustard procedure increases patients' life expectancy, it should be considered as a rather palliative approach that is implicated with severe long-term complications. Failure of the systematic right ventricle is the most serious and often requires heart transplantation. The persistently increasing number of adult patients with complex congenital heart disease necessitates their management in specialized medical centres.

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