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Case Report

Anatomical repair of congenitally corrected transposition in the fifth decade of life



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ABSTRACT

Successful repair of congenitally corrected transposition with ventricular septal defect and pulmonary stenosis presenting with heart failure in the fifth decade of life is described. This is the oldest patient to undergo this surgery, as per existing literature.

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1. Introduction

Congenitally corrected transposition (CCTGA) is a rare congenital heart defect with 0.5–1% incidence of all congenital heart disease.¹ Instances of survival without intervention are well known as late as the 8th decade of life. Adult patients of unoperated CCTGA without associated lesions, who present with symptoms usually do so secondary to worsening systemic ventricular (right ventricular (RV)) function along with systemic atrioventricular (AV) valve incompetence. The other presentation is due to spontaneous AV block that is known to develop.¹

The other category of patients, who would be associated with longevity without surgical intervention are those with associated large ventricular septal defect (VSD) and significant pulmonary stenosis (PS) not tight enough to cause excessive cyanosis but of enough severity to have protected the pulmonary circulation from overflow.

2. Case report

We report a 51-year-old lady, who had been diagnosed with CCTGA, VSD, and PS in childhood, but had a balanced shunt without clinical cyanosis. She was on irregular follow-up, had had a normal pregnancy 25 years ago and was not on any medications till 2 months before she presented to her cardiologist. The presenting complaints were breathlessness and visible cyanosis on even mild exertion, orthopnoea, and palpitations. On examination, she was found to be in atrial fibrillation (AF) with prominent neck veins, hepatomegaly and with a systolic murmur.

Echocardiography revealed situs solitus, mesocardia, AV and ventriculoarterial discordance, and large VSD with severe PS. In addition, she had severe left AV valve regurgitation with a dilated tricuspid annulus but no displacement. The right AV (mitral) valve was normal. She had good biventricular function.

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2.1. Stabilization

She was started on beta blockers, diuretics, and ace inhibitor and when she presented to us, she was in normal sinus rhythm (NSR) and not in obvious congestive heart failure (CHF), but still had exertional intolerance with aggravation of cyanosis on slight exertion.

Cardiac catheterization and angiography revealed the ventricular end diastolic pressure was a mean of 12 mmHg. Ventriculogram revealed routability of the left ventricle (LV) to the aorta, tight PS with PA pressures of 30/12 mmHg and a normal sized left and a dilated right pulmonary artery (Fig. 1). There was severe left AV valve regurgitation. Coronary angiogram revealed single coronary artery but no coronary artery disease.

3. Treatment options

Options considered were (a) continuation of medical therapy alone, or (b) corrective surgery in the form of anatomical repair (Rastelli + Senning procedure) with tricuspid valve repair. Cardiac transplant was not considered in view of the normal ventricular function and presence of correctable anatomy. Conventional repair of CCTGA was not considered due to the known poor long-term results.⁵

As the lady was sufficiently disabled despite optimal medical treatment, the family opted for corrective surgery.

4. Surgery

A Rastelli + Senning procedure was performed as per our published technique.² The LV was routed to the aorta by an intraventricular tunnel working through the right ventriculotomy and a 24 mm hand made bovine pericardial conduit with a trileaflet valve of 0.1 mm Goretex pericardial membrane

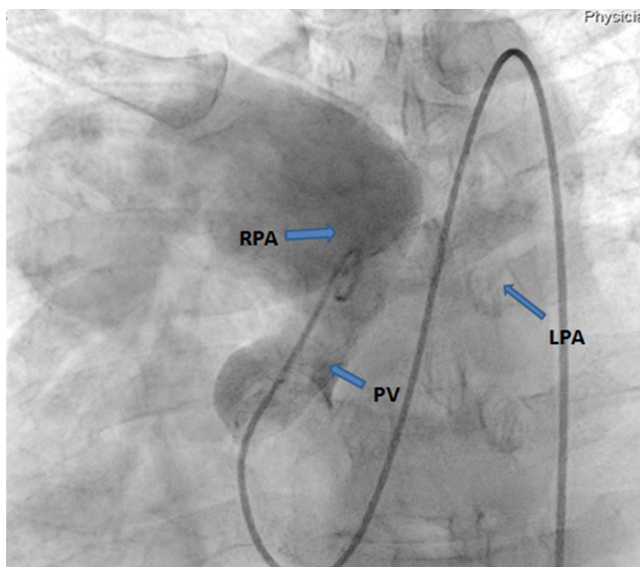


Fig. 1 – Angio showing the dilated RPA, LPA and pulmonary valve.

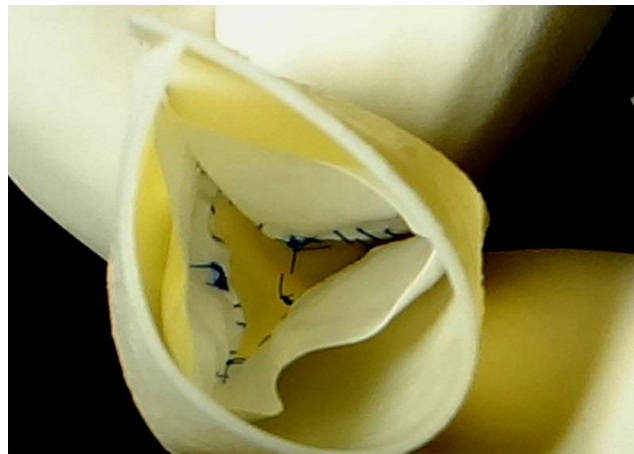


Fig. 2 – Hand made valved conduit.

was inserted between the right ventricle (RV) and pulmonary artery to the left of the ascending aorta (Fig. 2). Venous switch was achieved with a Senning procedure. The tricuspid valve annulus was found to be severely dilated and a reduction posterior semicircular DeVega type annuloplasty was performed using 3-0 prolene double strand suture.

The cardiopulmonary bypass (CPB) and aortic cross clamp times were 285 and 155 min respectively.

The patient could be easily weaned off CPB on low dose dobutamine. TEE revealed satisfactory repair with no TR.

She initially required AV sequential pacing though epicardial temporary pacing wires but reverted to normal AV conduction after 48 h.

At discharge on the 8th post operative day, she was on beta blockers for AF prophylaxis and diuretics. Both ventricles showed good functions and there was no residual anatomical defect. Her O₂ saturation was 95% on room air.

At 3-month follow-up, she had resumed normal activity, was NYHA Class I, and was off all medicines except beta blockers. Echocardiography demonstrated good biventricular function with unobstructed outflow tract (Fig. 3).

5. Comment

The Rastelli + Senning operation involves long intraoperative support times and multiple redirections of the blood stream with multiple potential sites of residual problems. Hence, it is not surprising that even today, when anatomical repair for CCTGA with or without its associations is established; there is reluctance among the cardiology fraternity in referring a patient for such an operation. Especially if a patient has balanced pulmonary and systemic blood flows, and is relatively asymptomatic, it is more than likely that such patients would be followed up conservatively. Truly long-term outcomes of repaired CCTGA are not entirely clear, and this also contributes to this tendency.

The present case is the oldest described patient known to have undergone a Rastelli + Senning operation in the world literature, the oldest to date being 40-year-old,⁴ to the best of our knowledge.

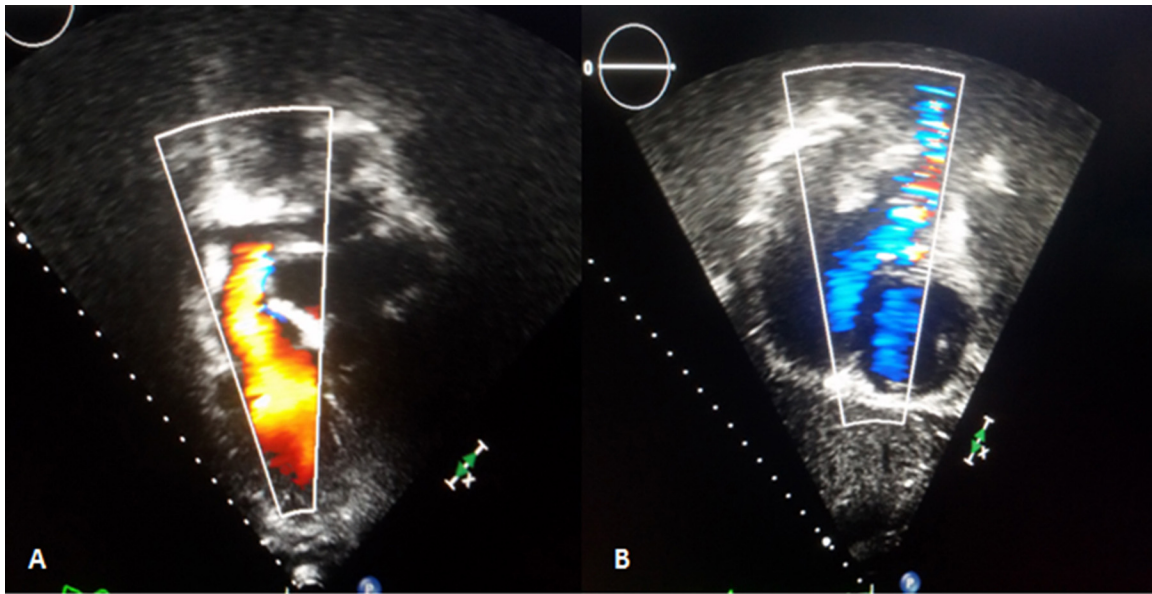


Fig. 3 – Echo demonstrating (A) unobstructed pulmonary venous baffle. (B) Laminar flow in LV to aorta tunnel.

The Rastelli + Senning operation was first described by Ilbawi et al.³ At that time our patient would have been in her twenties. The operation was first performed in India in 1993.² It is therefore understandable that she was managed conservatively till well into her adulthood.

It is unclear when she developed AF but her relief in symptomatology after reverting to NSR points to this having been the precipitating cause. With loss of atrial kick, she lapsed into CHF and with the reduction in forward output, her pulmonary blood flow probably suffered, making her clinically cyanotic.

The left AV valve leak would obviously have been a long standing development but was not documented in any of her previous records.

We were concerned about the reported low compliance of the hypertrophied adult ventricle, and how it would behave after a long ischemic time. However, the postoperative recovery was exceptionally smooth.

As is quite common after the Rastelli + Senning operation, temporary AV block ensued. As is also so common after this operation in our experience, NSR resumed within 48 h of the operation.²

One of the important steps in the operation was to achieve a competent TV. Reduction of TR is reported after the TV becomes the pulmonary AV valve following the anatomic repair of CCTGA secondary to drop in RV pressures. However, in this lady this was unlikely to resolve spontaneously in view of the dilated tricuspid annulus. We were fortunate that there was no Ebsteinoid displacement of the TV annulus into the RV and a simple posterior semicircular annuloplasty could achieve the desired goal.

6. Compliance with ethical standards

Ethical committee clearance from hospital and patient's informed consent was taken for study and research purpose.

Conflicts of interest

The authors have none to declare.

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