

Senning Procedure for Transposition of the Great Arteries in a Patient with Situs Inversus Totalis and Dextrocardia

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ABSTRACT

The arterial switch operation is considered as standard therapy for isolated D-transposition of the great arteries. However, in under-developed countries, patients still present with D-transposition of the great arteries beyond the neonatal age rendering them unsuitable for anatomical repair. These children are often offered physiological repair, i.e. Senning or Mustard procedure. We describe our experience of a successful Senning procedure in a 4.6-year girl, who had atrial and visceral situs inversus totalis, dextrocardia and d-transposition of the great arteries. To our knowledge, this is the first reported case of this operation in a patient with such unique anatomy.

Key Words: *D-transposition of great arteries. Situs inversus. Senning.*

INTRODUCTION

One of the fascinations of congenital cardiac surgery is its complexity and often unexpected findings during surgery. Transposition of great arteries is a condition in which the great arteries arise from the opposite ventricles across the ventricular septum: the aorta from the right ventricle and the pulmonary trunk from the left ventricle. D-transposition of the great arteries affects between 2.6% and 7.8% of infants with congenital heart disease and is the most common cyanotic congenital heart lesion to present in the 1st week of life.¹ D-transposition of the great arteries is considered incompatible with life, unless there is some level of mixing between the two circulations; ventricular septal defects (50%), atrial septal defects (ASDs) (20%), and patent ductus arteriosus (5%).² Transthoracic echocardiography alone is considered sufficient for the diagnosis of this condition as well as to acquire additional information like atrioventricular valve abnormalities (10%), arch obstruction (10%), subaortic or subpulmonary stenosis (5%), and abnormal coronary artery patterns (5%);² this avoids the need for a diagnostic cardiac catheterisation. Prior to balloon atrial septostomy introduced by Rashkind in 1966, those patients without a mixing defect had no survival. He revolutionised the management of these patients; the procedure can be performed in the catheterisation laboratory as well as on the bed side guided by transthoracic echocardiography.³

Visceral situs inversus is characterised by mirror-image location of the thoracic and abdominal organs; there is a higher incidence of cardiac problems than in the general population. Situs inversus has an incidence of about 0.01%.⁴ Frescura and Thiene, after reviewing 177 hearts with discordant ventriculoarterial connection, showed an incidence of situs inversus of 0.7%.⁵

We report a case of a young girl who presented with D-transposition of great arteries and situs inversus and was operated for physiological correction with the Senning procedure.

CASE REPORT

A 4.6-year baby girl was referred from the paediatric cardiology department with the diagnosis of D-transposition of great arteries, situs inversus and dextrocardia. She was born via elective cesarean section and had no family history of cardiac defects. Her parents mentioned that she was unremarkable at the time of birth; however, they later often noticed blue discoloration of her toes and tips of finger as well as her tongue, which were suggested as pneumonic episodes by various doctors. It was on the suspicion of one of her relatives that the parents sought a consultation from a cardiologist; there she was diagnosed with a congenital heart defect and referred to us for further opinion.

On examination, the pulses were normal in volume and character. Palpation of the precordium revealed a ventricular tap at the right lower sternal border and the apex beat at the 5th intercostal space in the right mid-clavicular line. Auscultation revealed a normal 1st heart sound and a single loud 2nd heart sound. No murmurs, clicks, or gallops were noted. The respiratory effort was unlaboured and the lungs were clear to auscultation. The liver was palpable 1 cm below the left costal margin. Her CBC showed an Hb of 13.5, HCT 43.6, and platelets 168,000/ul. X-ray chest showed bilateral lung congestion

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with dextrocardia and a gastric bubble on right side. Echocardiographic report was suggestive of situs inversus with dextrocardia. There was atrioventricular concordance with ventriculoarterial discordance with the aorta anterior to main pulmonary artery. There was a large ASD secundum with bidirectional flow and moderate pulmonary artery hypertension (PAH). Her biventricular function were optimal. She was planned for atrial switch (senning).

Patient underwent intravenous drug induction with maintenance of anesthesia with sevoflurane. Her PO_2 was 37 mmHg at 100% F_2O_2 .

A median sternotomy was performed followed by thymectomy. The pericardium was opened and anatomy assessed; situs inversus with dextrocardia was observed. Standard bicaval cannulation was made and the patient cooled to moderate hypothermia. Del-Nido cardioplegia, at a temperature of 8 to 10°C, was infused at 20 ml per kg over 2 to 3 minutes and repeated at 40 minutes intervals. The surgeon then changed sides to the left of the patient.

A curvilinear right atriotomy incision was made 1 cm away from the sulcus terminalis extending from the base of the right atrium (RA) appendage to the junction of the inferior vena cava (IVC). Another incision was made in the left atrium (LA) parallel to the sulcus terminalis and a vent inserted. The ASD was enlarged by a longitudinal incision over the atrial septum. The posterior flap of the atrial septum augmented with autologous pericardium was used to reconstruct the floor of the systemic venous chamber; stitching was commenced in the floor of the LA separating the pulmonary venous openings from the LA appendage and mitral valve. Next, the anterior lip of the atrial septum was anastomosed with the post margin of the right atriotomy incision, ensuring patency of the openings of the superior vena cava (SVC) and IVC, thus completing the roof of the systemic venous chamber. Finally, the anterior border of the right atriotomy incision was sutured to the posterior lip of the left atriotomy incision, making certain of the patency of the pulmonary venous openings; this completed the pulmonary venous chamber. All anastomoses were performed with 6/0 prolene (8 mm). The vent was readjusted into the RA appendage (now the pul venous chamber).

Once the aortic cross clamp was removed, sinus rythum returned. The patient was rewarmed to 36°C over 20 minutes. Adrenaline at 0.1 ug/kg/min and milrinone at 0.5 ug/kg/min were started. The patient was weaned off-bypass with stable hemodynamics. Total cross-clamp time was 60 minutes and total bypass time was 100 minutes. Post-procedure trans-esophageal echocardiography (TEE) was performed; it showed no significant obstruction in both venous pathways with good biventricular function. Postoperative PO_2 recorded in operating room (OR) was 217 mmHg at 100% F_2O_2 .

Patient was shifted to ICU on mechanical ventilation. She was on adrenaline and milrinone as inotropic agents along with dopamine at renal dose and GTN infusion. She was planned for overnight ventilation with regular monitoring of blood gases. She underwent smooth extubation on the first postoperative day with total ventilator time of 21 hours. She remained in PICU for 4 days and at the fifth postoperative day was shifted. Postoperative echo showed good biventricular function with mild mitral regurgitation and mild tricuspid regurgitation. There was mild turbulence observed in both baffles with no pulmonary regurgitation.

She was discharged from hospital at the ninth postoperative day. Her follow-up visit was unremarkable with SO_2 of 95% at room air.

DISCUSSION

Transposition of great arteries accounts for about 3% of all congenital heart diseases. Patients with absence of mixing lesion are cyanotic at birth and rapidly decompensate due to failure of mixing of the blood from the two parallel circulations. Immediate balloon or surgical septostomy is a pre-requisite for mixing of blood to maintain survival. Definitive intervention involves anatomical correction as described by Adib Dominos Jatene, in the form of an Arterial Switch Operation (ASO), in 1975 at the University of Sao Paulo Heart Institute, Sao Paulo, Brazil.⁶ Nevertheless, there are many patients who present with D-transposition of great arteries unsuitable for ASO due to delayed presentation; such patients require a physiologic correction in the form of atrial switch that results in diverting right atrial blood to mitral valve and left atrial flow towards tricuspid valve; thus the right ventricle becomes the systemic ventricle. The Senning and Mustard baffles remain important techniques for the treatment of congenitally corrected transposition, isolated ventricular inversion, and D-transposition of the great arteries with delayed presentation. In 1957, Ake Senning performed the first successful atrial switch using atrial flaps though the procedure was a failure in the first two patients but his third patient, a 9-year-old boy survived. Senning's operation was technically very complex, resulting in many surgeons abandoning it in preference to the Mustard procedure, a relatively less complex operation.⁷ In 1964, Mustard simplified the procedure by excising the atrial septum and placing a 'single, pantaloons-shaped patch' between the atria to create the 'baffle', again redirecting the blood flow in the same manner as the Senning procedure. Furthermore, use of pericardium rather than atrial septum for baffle creation resulted in a larger atrium with decreased incidence of low cardiac output, associated with small atria as occurred post senning.⁸

Nevertheless, by 1975, interest in the Senning operation was revived. The Senning operation, with its use of

native cardiac tissue, seemed to provide a natural growth potential resulting in less baffle obstruction in growing children.^{9,10}

Currently, we are using the modified Senning operation for patients who present beyond neonatal age or when cardiac status is unsuitable for arterial switch. Considering the potential for baffle obstruction or smaller atrial size, we augment the baffle with pericardium, if required.

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