

## Modified Mustard Procedure for D-Tga with Left Superior Venacava : a Case Report

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**Abstract:** Transposition of the great arteries refers to ventriculoarterial discordance. Complete transposition is also known as d-TGA and is commonly seen in newborns. Cyanotic babies may be treated percutaneously with a Rashkind atrial balloon septostomy. However, definitive surgical procedures can be an atrial switch or an arterial switch. We present a case of a 10-month-old infant with d-TGA with left SVC, who underwent a modified Mustard and a Glenn procedure for palliation.

**Key Words:** D-TGA (Transposition of great vessels), Mustard, Glenn procedure

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**Introduction:** Transposition of the great arteries refers to ventriculoarterial discordance. The aorta arises from a morphological right ventricle, and the pulmonary artery arises from a morphological left ventricle. Complete transposition of the great arteries is also known as d-TGA; the "d-" refers to the dextroposition of the bulboventricular loop.[1]

D-TGA is commonly seen in newborns, and when the ventricular septum is intact, it is usually cyanotic in the first day of life.[2] If circulatory mixing occurs via a patent ductus arteriosus, physiological closure of the ductus causes abrupt cyanosis and clinical deterioration. Cyanotic babies may be treated percutaneously with a Rashkind atrial balloon septostomy to create a more sizable atrial septal defect, which may dramatically improve their oxygenation until definitive surgery can be performed.[3]

Definitive surgical procedures can be an atrial switch or an arterial switch. The first atrial switch procedure was performed by Senning in 1958, which involves the creation of an intra-atrial baffle from autologous pericardial tissue to direct the venous return to the contralateral atrioventricular (AV) valve and ventricle.[4] An alternative operation was subsequently developed by Mustard, who excised the atrial septum and used synthetic material to create the baffle. Arterial switch operation restores the normal anatomic arrangement of the circulation and, as such, is a more striking physiological long-term option. From its first description by Jatene in 1976 [5], it is now considered as a procedure of choice when the anatomy is appropriate and is usually performed in the first month of life.

We present a case of a 10-month-old infant with d-TGA with left SVC, who underwent a modified Mustard and a Glenn procedure for palliation.

**Case:** 2-month-old baby boy presented to Emergency Room at The Aga Khan University Hospital with a 15-day history of cyanosis. On examination, he was hypoxic with oxygen saturations of 45%, heart rate of 170 beats/minute, and blood pressure of 72/46 mmHg. He was resuscitated with prostaglandin, dopamine, and was ventilated. Transthoracic echocardiography (echo) revealed d-TGA, a small patent foramen ovale, and a tiny PDA.

Emergency atrial septectomy was done on cardiopulmonary bypass; post-procedure echo showed an atrial septal defect (ASD) of 9-10 mm with good flow across it. He was successfully extubated on 3rd postoperative day and was discharged on 10th postoperative day with oxygen saturations of 65%. During follow-up in clinic, he remained well.

At 10-months of age, the patient was admitted for elective Atrial Switch procedure. Pre-operative findings were d-TGA, single left superior vena cava opening into the coronary sinus, hypoplastic left ventricle, and an ASD. Owing to his left superior vena cava (SVC), we did a hemi-Mustard by diverting the IVC to the left atrium using autologous pericardium and directed the left SVC into the pulmonary artery (Glenn shunt). Pulmonary veins were baffled into the right atrium using autologous pericardium. Patient recovered uneventfully and was successfully extubated on 2nd post-operation day. His postoperative echo showed non-obstructive systemic and pulmonary blood flow. Patient was discharged on 6th postoperative day; on his follow-up at 2 years, he was growing well with O<sub>2</sub> saturations of 74% and achieving his expected milestones.

**Operation:** Glenn Shunt (left SVC---Left PA) Hemi mustard operation (IVC to Mitral valve)

d-Tga was palliated by performing a Hemi Mustard procedure and then creating a Glenn Shunt. This was done using a bovine pericardial baffle, thus separating the pulmonary venous and Inferior Vena Caval blood. And then the pulmonary veins were directed to the Right atrial and tricuspid valve using a broad based patch of bovine pericardium.

The Left Superior Vena Cava was disconnected from left atrium. Left atrium was closed, and then the left Superior vena cava was anastomosed to the Left Pulmonary artery to create a left Glenn Shunt.

**Discussion:** d-TGA is one of the most common cyanotic congenital heart disease ,with an incidence of 1 in 3500 -5000 live birth and a male to female ratio of 1.5 to 3.2:1.[6] If not treated it has a mortality rate of 30% in first week, 50%in first month and 90 % by the end of one year.

In developing countries like Pakistan and India , a major issue is that the congenital heart disease are diagnosed late hence about 95% of infants remain untreated.[7]

Initial management of patients with TGA is IV prostaglandin E1, which causes patency of ductus arteriosus thus maintaining the pulmonary flow of blood. If patient is extremely hypoxemic with no or minimum atrial level communication and very little mixing of blood then balloon septostomy, is done . Its limitation includes a rigid interatrial septum. However the ideal procedure is arterial switch procedure, which results in left ventricle to aorta continuity . In patients with aberrant coronary artery anatomy Senning or Mustard procedure ,requiring excision of the atrial septum and formation of a baffle using synthetic material, is seen to be more successful.[1]In Mustard procedure inter atrial septum is excised and a PTFE patch for the SVC and IVC.is then used as a pentaloop shapped baffle. While in Senning the atria are converted into a Y – shaped systemic venous pathway and a C- shaped pulmonary venous pathway using autologous tissue.

In our patient atrial septectomy was done as there was non-availability of the rotar blade for atrial septostomy. Since there was a left SVC Glenn Shunt

was fashioned as it reduces the volume load on the heart and the inferior hemi mustard completed the procedure with excellent outcome.

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