

HISTORY: , The patient is a 9-year-old born with pulmonary atresia, intact ventricular septum with coronary sinusoids. He also has VACTERL association with hydrocephalus. As an infant, he underwent placement of a right modified central shunt. On 05/26/1999, he underwent placement of a bidirectional Glenn shunt, pulmonary artery angioplasty, takedown of the central shunt, PDA ligation, and placement of a 4 mm left-sided central shunt. On 08/01/2006, he underwent cardiac catheterization and coil embolization of the central shunt. A repeat catheterization on 09/25/2001 demonstrated elevated Glenn pressures and significant collateral vessels for which he underwent embolization. He then underwent repeat catheterization on 11/20/2003 and further embolization of residual collateral vessels. Blood pressures were found to be 13 mmHg with the pulmonary vascular resistance of 2.6-3.1 Wood units. On 03/22/2004, he returned to the operating room and underwent successful 20 mm extracardiac Fontan with placement of an 8-mm fenestration and main pulmonary artery ligation. A repeat catheterization on 09/07/2006, demonstrated mildly elevated Fontan pressures in the context of a widely patent Fontan fenestration and intolerance of Fontan fenestration occlusion. The patient then followed conservatively since that time. The patient is undergoing a repeat evaluation to assess his candidacy for a Fontan fenestration occlusion, as well as consideration for a tricuspid valvuloplasty in attempt to relieve right ventricular hypertension and associated membranous ventricular aneurysm protruding into the left ventricular outflow

tract.,PROCEDURE:, After sedation and local Xylocaine anesthesia, the patient was placed under general endotracheal anesthesia, the patient was prepped and draped. Cardiac catheterization was performed as outlined in the attached continuation sheets. Vascular entry was by percutaneous technique, and the patient was heparinized. Monitoring during the procedure included continuous surface ECG, continuous pulse oximetry, and cycled cuff blood pressures, in addition to intravascular pressures.,Using a 7-French sheath, a 6-French wedge catheter was inserted in the right femoral vein and advanced from the inferior vena cava along the Fontan conduit into the main left pulmonary artery, as well as the superior vena cava. This catheter was then exchanged for a 5-French VS catheter of a distal wire. Apposition of the right pulmonary artery over, which the wedge catheter was advanced. The wedge catheter could then be easily advanced across the Fontan fenestration into the right atrium and guidewire manipulation allowed access across the atrial septal defect to the pulmonary veins, left atrium, and left ventricle.,Using a 5-French sheath, a 5-French pigtail catheter was inserted into the right femoral artery and advanced retrograde to the descending aorta, ascending aorta, and left ventricle. Attempt was then made to cross the tricuspid valve from the right atrium and guidewire persisted to prolapse through the membranous ventricular septum into the left ventricle. The catheter distal wire position was finally achieved across what appeared to be the posterior aspect of the tricuspid valve, both angiographically as well as equal

guidance. Left ventricular pressure was found to be suprasystemic. A balloon valvoplasty was performed using a Ranger 4 x 2 cm balloon catheter with no waste at minimal inflation pressure. Echocardiogram, which showed no significant change in the appearance of a tricuspid valve and persistence of aneurysmal membranous ventricular septum. Further angioplasty was then performed first utilizing a 6 mm cutting balloon directed through 7-French flexor sheath positioned within the right atrium. There was a disappearance of a mild waist prior to spontaneous tear of the balloon. The balloon catheter was then removed in its entirety. Echocardiogram again demonstrated no change in the appearance of the tricuspid valve. A final angioplasty was performed utilizing a 80 mm cutting balloon with the disappearance of a distinctive waist. Echocardiogram; however, demonstrated no change and intact appearing tricuspid valve and no decompression of the right ventricle. Further attempts to cross tricuspid valve were thus abandoned. Attention was then directed to a Fontan fenestration. A balloon occlusion then demonstrated minimal increase in Fontan pressures from 12 mmHg to 15 mmHg. With less than 10% fall in calculated cardiac index. The angiogram in the inferior vena cava demonstrated a large fenestration measuring 6.6 mm in diameter with a length of 8 mm. A 7-French flexor sheath was again advanced cross the fenestration. A 10-mm Amplatzer muscular ventricular septal defect occluder was loaded on delivery catheter and advanced through the sheath where the distal disk was

allowed to be figured in the right atrium. Entire system was then brought into the fenestration and withdrawal of the sheath allowed reconfiguration of the proximal disk. Once the stable device configuration was confirmed, device was released from the delivery catheter. Hemodynamic assessment and the angiograms were then repeated. Flows were calculated by the Fick technique using an assumed oxygen consumption and contents derived from Radiometer Hemoximeter saturations and hemoglobin capacity. Angiograms with injection in the right coronary artery, left coronary artery, superior vena cava, inferior vena cava, and right ventricle. After angiography, two normal-appearing renal collecting systems were visualized. The catheters and sheaths were removed and topical pressure applied for hemostasis. The patient was returned to the recovery room in satisfactory condition. There were no complications.

**DISCUSSION:** , Oxygen consumption was assumed to be normal. Mixed venous saturation was low due to systemic arterial desaturation. There was modest increased saturation of the branch pulmonary arteries due to the presumed aortopulmonary collateral flow. The right pulmonary veins were fully saturated. Left pulmonary veins were not entered. There was a fall in saturation within the left ventricle and descending aorta due to a right to left shunt across the Fontan fenestration. Mean Fontan pressures were 12 mmHg with a 1 mmHg fall in mean pressure into the distal left pulmonary artery. Right and left pulmonary capillary wedge pressures were similar to left atrial phasic pressure with an

A-wave similar to the normal left ventricular end-diastolic pressure of 11 mmHg. Left ventricular systolic pressure was normal with at most 5 mmHg systolic gradient pressure pull-back to the ascending aorta. Phasic ascending and descending aortic pressures were similar and normal. The calculated systemic flow was normal. Pulmonary flow was reduced to the QT-QS ratio of 0.7621. Pulmonary vascular resistance was normal at 1 Wood units. Angiogram with injection in the right coronary artery demonstrated diminutive coronary with an extensive sinusoidal communication to the rudimentary right ventricle. The left coronary angiogram showed a left dominant system with a brisk flow to the left anterior descending and left circumflex coronary arteries. There was communication to the right-sided coronary sinusoidal communication to the rudimentary right ventricle. Angiogram with injection in the superior vena cava showed patent right bidirectional Glenn shunt with mild narrowing of the proximal right pulmonary artery, as well as the central pulmonary artery, diameter of which was augmented by the Glenn anastomosis and the Fontan anastomosis. There was symmetric contrast flow to both pulmonary arteries. A large degree of contrast flowed retrograde into the Fontan and shunting into the right atrium across the fenestration. There is competitive flow to the upper lobes presumably due to aortopulmonary collateral flow. The branch pulmonaries appeared mildly hypoplastic. Levo phase contrast returned into the heart, appeared unobstructed demonstrating good left ventricular contractility. Angiogram with injection in the Fontan

showed a widely patent anastomosis with the inferior vena cava. Majority of the contrast flowing across the fenestration into the right atrium with a positive flow to the branch pulmonary arteries., Following the device occlusion of Fontan fenestration, the Fontan and mean pressure increased to 15 mmHg with a 3 mmHg, a mean gradient in the distal left pulmonary artery and no gradient into the right pulmonary artery. There was an increase in the systemic arterial pressures. Mixed venous saturation increased from the resting state as with increase in systemic arterial saturation to 95%. The calculated systemic flow increased slightly from the resting state and pulmonary flow was similar with a QT-QS ratio of 0.921. Angiogram with injection in the inferior vena cava showed a stable device configuration with a good disk apposition to the anterior surface of the Fontan with no protrusion into the Fontan and no residual shunt and no obstruction to a Fontan flow. An ascending aortogram that showed a left aortic arch with trace aortic insufficiency and multiple small residual aortopulmonary collateral vessels arising from the intercostal arteries. A small degree of contrast returned to the heart., INITIAL DIAGNOSES: ,1. Pulmonary atresia.,2. VACTERL association.,3. Persistent sinusoidal right ventricle to the coronary communications.,4.

Hydrocephalus., PRIOR SURGERIES AND

INTERVENTIONS: ,1. Systemic to pulmonary shunts.,2. Right bidirectional Glenn shunt.,3. Revision of the central shunt.,4. Ligation and division of patent ductus arteriosus.,5. Occlusion of venovenous and arterial aortopulmonary collateral

vessels.,6. Extracardiac Fontan with the fenestration.,CURRENT DIAGNOSES: ,1. Favorable Fontan hemodynamics.,2. Hypertensive right ventricle.,3. Aneurysm membranous ventricular septum with mild left ventricle outflow tract obstruction.,4. Patent Fontan fenestration.,CURRENT INTERVENTION: ,1. Balloon dilation tricuspid valve attempted and failed.,2. Occlusion of a Fontan fenestration.,MANAGEMENT: ,He will be discussed at Combined Cardiology/Cardiothoracic Surgery case conference. A careful monitoring of ventricle outflow tract will be instituted with consideration for a surgical repair. Further cardiologic care will be directed by Dr. X.