

**HISTORY:** The patient is a 10-1/2-year-old born with asplenia syndrome with a complex cyanotic congenital heart disease characterized by dextrocardia bilateral superior vena cava, complete atrioventricular septal defect, a total anomalous pulmonary venous return to the right-sided atrium, and double-outlet to the right ventricle with malposed great vessels, the aorta being anterior with a severe pulmonary stenosis. He had undergone staged repair beginning on 04/21/1997 with a right modified Blalock-Taussig shunt followed on 09/02/1999 with a bilateral bidirectional Glenn shunt, and left pulmonary artery to main pulmonary artery pericardial patch augmentation. These procedures were performed at Medical College Hospital. Family states that they moved to the United States. Evaluation at the Children's Hospital earlier this year demonstrated complete occlusion of the right bidirectional Glenn shunt as well as occlusion of the proximal right pulmonary artery. He was also found to have elevated Glenn pressures at 22 mmHg, transpulmonary gradient axis of 14 mmHg. The QP:QS ratio of 0.6:1. A large decompressing venous collateral was also appreciated. The patient was brought back to cardiac catheterization in an attempt to reconstitute the right caval pulmonary anastomosis and to occlude the venous collateral vessel.

**DESCRIPTION OF PROCEDURE:** , After sedation and local Xylocaine 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 6-French sheath, a 6-French wedge catheter was inserted in the right femoral vein and advanced from the inferior vena cava into the right-sided atrium pulmonary veins and the right ventricle. Using a 6-French sheath, a 5-French pigtail catheter was inserted into the right femoral artery and advanced retrograde to the descending aorta and ascending aorta. A separate port of arterial access was obtained in the left femoral artery utilizing a 5-French sheath. Percutaneous access into the right jugular vein was attempted, but unsuccessful. Ultrasound on the right neck demonstrated a complete thrombosis of the right internal jugular vein. Using percutaneous technique and a 5-French sheath, 5-French wedge catheter was inserted into the left internal jugular vein and advanced along the left superior vena cava across the left caval-pulmonary anastomosis into the main pulmonary artery and left pulmonary artery with aid of guidewire. This catheter then also advanced into the bridging innominate vein. The catheter was then exchanged over wire for a 4-French Bernstein catheter, which was advanced to the blind end of the right superior vena cava. A balloon wedge angiogram of the right lower pulmonary vein demonstrated back filling of a small right lower pulmonary artery. There was no vascular continuity to the stump of the right Glenn. The jugular venous catheter and sheaths were exchanged over a wire for a 6-French flexor sheath, which was advanced to the proximal right superior vena cava. The

Bernstein catheter was then reintroduced using a Terumo guidewire. Probing of the superior vena cava facilitated access into the right lower pulmonary artery. The angiogram in the right pulmonary artery showed a diminutive right lower pulmonary artery and severe long segment proximal stenosis. The distal pulmonary measured approximately 5.5 to 60 mm in diameter with a long segment stenosis measuring approximately 31 mm in length. The length of the obstruction was balloon dilated using ultra-thin SD 4 x 2 cm balloon catheter with complete disappearance of the waist. This facilitated advancement of a flexor sheath into the proximal portion of the stenosis. A PG 2960 BPX Genesis stent premounted on a 6 mm OptiProbe. A balloon catheter was advanced across the area of narrowing and inflated with a near-complete disappearance of proximal waist. Angiogram demonstrated a good stent apposition to the caval wall. Further angioplasty was then performed utilizing an ultra-thin SDS 8 x 3 cm balloon catheter inflated to 19 atmospheres pressure with complete disappearance of a distinct proximal waist. Angiogram demonstrated wide patency of reconstituted right caval pulmonary anastomosis though there was no flow seen to the right upper pulmonary artery. The balloon wedge angiograms were then obtained in the right upper pulmonary veins suggesting the presence of right upper pulmonary artery and not contiguous with the right lower pulmonary artery. Bernstein catheter was advanced into the main pulmonary artery where a wire probing of the stump of the proximal right pulmonary artery facilitated access to the right upper

pulmonary artery. Angiogram demonstrated severe long segment stenosis of the proximal right pulmonary artery. Angioplasty of the right pulmonary was then performed using the OptiProbe 6-mm balloon catheter inflated to 16 atmospheres pressure with disappearance of a distinct waist. Repeat angiogram showed improvement in caliber of right upper pulmonary artery with filling defect of the proximal right pulmonary artery. The proximal right pulmonary artery was then dilated and stent implanted using a PG 2980 BPX Genesis stent premounted on 8-mm OptiProbe balloon catheter and implanted with complete disappearance of the waist. Distal right upper pulmonary artery was then dilated and stent implanted utilizing a PG 1870 BPX Genesis stent premounted on 7-mm OptiProbe balloon catheter. Repeat angiograms were then performed. Attention was then directed to the large venous collateral vessel arising from the left superior vena cava with a contrast filling of a left-sided azygos vein. A selective angiogram demonstrated a large azygos vein of the midsection measuring approximately 9.4 mm in diameter. An Amplatzer 12 mm vascular plug was loaded on the delivery catheter and advanced through the flexor sheath into the azygos vein. Once stable device was confirmed, the device was released from the delivery catheter. The 4-French Bernstein catheter was then reintroduced and 5 inch empirical 0.038 inch, 10 cm x 8 mm detachable coils were then implanted above the vascular plug filling the proximal azygos vein. A pigtail catheter was then introduced into the left superior vena cava for final angiogram.,Flows were calculated

by the Fick technique using an assumed oxygen consumption and contents derived from Radiometer Hemoximeter saturations and hemoglobin capacity. Cineangiograms were obtained with injection of the coronary sinus of pulmonary veins, the innominate vein, superior vena cava, the main pulmonary artery, and azygos vein. 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, but was low due to systemic arterial desaturation of 79%. The pulmonary veins were fully saturated with partial pressure of oxygen ranging between 120 and 169 mmHg in 30% oxygen. Remaining saturations reflected complete admixture. There was increased saturation in the left pulmonary artery due to aortopulmonary collateral flow. Phasic right atrial pressures were normal with an A-wave somewhat to the normal right ventricular end-diastolic pressure of 9 mmHg. Left ventricular systolic pressure was systemic with no outflow obstruction to the ascending aorta. Phasic ascending, descending pressures were similar and normal. Mean Glenn pressures at initiation of the case were slightly elevated at 14 mmHg with a transpulmonary gradient of 9 mmHg. The calculated systemic flow was a normal pulmonary flows reduced with a QP:QS ratio of 0.6:1. The pulmonary vascular resistance was elevated at 4.4 Woods units. Following stent implantation in

the right caval pulmonary anastomosis and right pulmonary artery, there was a slight increase in the Glenn venous pressures to 16 mmHg. Following embolization of the azygos vein, there was increase in systemic arterial saturation to 84% and increase in mixed venous saturation. There was similar increase in Glenn pressures to 28 mmHg with a transpulmonary gradient of 14 mmHg. There was an increase in arterial pressure. The calculated systemic flow increased from 3.1 liters /minute/meter squared to 4.3 liters/minute/meter squared. Angiogram within the innominate vein following stent implantation demonstrated appropriate stent position without significant distortion of the innominate vein or proximal cava. There appeared unobstructed contrast flow to the right lower pulmonary artery of a 1-mmHg mean pressure gradient. There was absence of contrast filling of the right middle and right upper pulmonary artery. Final angiogram with a contrast injection in the left superior vena cava showed a forward flow through the right Glenn, a good contrast filling of the right lower pulmonary artery, and a widely patent left Glenn negative contrast washout of the proximal right pulmonary artery and left pulmonary artery presumably due to aortopulmonary collateral flow. Contrast injection within the right upper pulmonary artery following the stent implantation demonstrated widely patent proximal right pulmonary artery along the length of the implanted stents though with retrograde contrast flow.,INITIAL DIAGNOSES:  
,1. Asplenia syndrome.,2. Dextrocardia bilateral superior vena cava.,3. Atrioventricular septal defect.,4. Total anomalous

pulmonary venous return to the right-sided atrium.,5. Double outlet right ventricle with malposed great vessels.,6. Severe pulmonary stenosis.,7. Separate hepatic venous drainage into the atria.,PRIOR SURGERIES AND INTERVENTIONS: ,1.

Right modified Blalock-Taussig shunt.,2. Bilateral bidirectional Glenn shunt.,3. Patch augmentation of the main pulmonary to left pulmonary artery.,CURRENT DIAGNOSES: ,1.

Obstructed right caval pulmonary anastomosis.,2. Obstructed right proximal pulmonary artery.,3. Venovenous collateral vessel.,CURRENT INTERVENTION: ,1. Balloon dilation of the right superior vena cava and stent implantation.,2. Balloon dilation of the proximal right pulmonary artery, stent implantation.,3. Embolization of venovenous collateral vessel.,MANAGEMENT: , The case will be discussed in Combined Cardiology Cardiothoracic Surgery case conference. A repeat catheterization is recommended in 3 months to assess for right pulmonary artery growth and to assess candidacy for Fontan completion. The patient will be maintained on anticoagulant medications of aspirin and Plavix. Further cardiology care will be directed by Dr. X.