HISTORY: , The patient is a 19-year-old boy with a membranous pulmonary atresia, underwent initial repair 12/04/1987 consisting of pulmonary valvotomy and placement of 4 mm Gore-Tex shunt between the ascending aorta and pulmonary artery with a snare. This was complicated by shunt thrombosis _____ utilizing a 10-mm balloon. Resulting in significant hypoxic brain injury where he has been left with static encephalopathy and cerebral palsy. On 04/07/1988, he underwent heart catheterization and balloon pulmonary valvuloplasty utilizing a 10-mm balloon. He has been followed conservatively since that time. A recent echocardiogram demonstrated possibly a significant right ventricle outflow tract obstruction with tricuspid valve regurgitation velocity predicting a right ventricular systolic pressure in excess of 180 mmHg. Right coronary artery to pulmonary artery fistula was also appreciated. The patient underwent cardiac catheterization to assess hemodynamics associated with his current state of repair.,PROCEDURE:, The patient was placed under general endotracheal anesthesia breathing on 30% oxygen throughout the case. 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. The right femoral vein advanced through the right heart structures out to the branch pulmonary

arteries. This catheter was then exchanged over wire for a 5-French marker pigtail catheter, which was directed into the main pulmonary artery.,Using a 5-French sheath, a 5-French pigtail catheter was inserted in the right femoral artery and advanced retrograde to the descending aorta, ascending aorta, and left ventricle. This catheter was then exchanged for a Judkins right coronary catheter for selective cannulation of the right coronary artery.,Flows were calculated by the Fick technique using a measured assumed oxygen consumption and contents derived from Radiometer Hemoximeter saturations and hemoglobin capacity.,Cineangiograms were obtained with injection of the main pulmonary artery and right coronary artery.,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 normal with no evidence of intracardiac shunt. Left-sided heart was fully saturated. Phasic right atrial pressures were normal with an A-wave similar to the normal right ventricular end-diastolic pressure. Right ventricular systolic pressure was mildly elevated at 45% systemic level. There was a 25 mmHg peak systolic gradient across the outflow tract to the main branch pulmonary arteries. Phasic branch pulmonary artery pressures were normal. Right-to-left pulmonary artery capillary wedge pressures were normal with an A-wave

similar to the normal left ventricular end-diastolic pressure of 12 mmHg. Left ventricular systolic pressure was systemic with no outflow obstruction to the ascending aorta. Phasic ascending and descending pressures were similar and normal. The calculated systemic and pulmonary flows were equal and normal. Vascular resistances were normal. Angiogram with contrast injection in the main pulmonary artery showed catheter induced pulmonary insufficiency. The right ventricle appeared mildly hypoplastic with a good contractility and mild tricuspid valve regurgitation. There is dynamic narrowing of the infundibulum with hypoplastic pulmonary annulus. The pulmonary valve appeared to be thin and moved well. The median branch pulmonary arteries were of good size with normal distal arborization. Angiogram with contrast injection in the right coronary artery showed a non-dominant coronary with a small fistula arising from the proximal right coronary artery coursing over the infundibulum and entering the left facing sinus of the main pulmonary artery.,INITIAL DIAGNOSES:,1. Membranous pulmonary atresia.,2. Atrial septal defect.,3. Right coronary artery to pulmonary artery fistula.,SURGERIES (INTERVENTIONS): ,1. Pulmonary valvotomy surgical.,2. Aortopulmonary artery central shunt.,3. Balloon pulmonary valvuloplasty.,CURRENT DIAGNOSES: ,1. Pulmonary valve stenosis supplemented to hypoplastic pulmonary annulus.,2. Mild right ventricle outflow tract obstruction due to supple pulmonic narrowing.,3. Small right coronary artery to main pulmonary fistula.,4. Static encephalopathy.,5. Cerebral palsy.,MANAGEMENT: , The

case to be discussed with combined Cardiology/Cardiothoracic Surgery case conference. Given the mild degree of outflow tract obstruction in this sedentary patient, aggressive intervention is not indicated. Conservative outpatient management is to be recommended. Further patient care will be directed by Dr. X.