HISTORY: ,The patient is a 5-1/2-year-old with Down syndrome, complex heart disease consisting of atrioventricular septal defect and tetralogy of Fallot with pulmonary atresia, discontinuous pulmonary arteries and bilateral superior vena cava with a left cava draining to the coronary sinus and a right aortic arch. As an infant, he was initially palliated with the right and modified Blalock-Taussig shunt in October of 2002 and underwent atrioventricular septal defect and repair of pulmonary artery unifocalization and homograft placement between the right ventricle and unifocalized pulmonary arteries. He developed a significant branch of pulmonary artery stenosis for which on 07/20/2004, he underwent a bilateral balloon pulmonary arterioplasty and stent implantation at the San Diego at Children's Hospital. This was followed on 09/13/2007 with replacement of pulmonary valve utilizing a 16-mm Contegra valve. A recent echocardiogram demonstrated a significant branch of pulmonary artery stenosis with the predicted gradient of 41 to 55 mmHg and a well-functioning Contegra valve. The lung perfusion scan from 11/14/2007 demonstrated 47% flow to the left lung and 53% flow to the right lung. The patient underwent a repeat catheterization in consideration for further balloon angioplasty of the branch pulmonary arteries.,PROCEDURE: , After sedation, the patient was placed under general endotracheal anesthesia breathing 50% oxygen throughout the case. 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, 6-French wedge catheter was inserted into the right femoral vein and advanced through the right heart structures out to the branch pulmonary arteries. This catheter was exchanged over wire. A 5-French marker pigtail catheter was directed into the main pulmonary artery. A second site of venous access was achieved in and the left femoral vein with the placement of 5-French sheath.,Using a 4-French sheath, a 4-French pigtail catheter was inserted in the right femoral artery and advanced retrograde to the descending aorta, ascending aorta and left ventricle. Angiogram with injection in the main pulmonary artery demonstrated stable stent configuration of the proximal branch pulmonary arteries with intimal ingrowth in the region of the proximal stents. The distal right pulmonary measured approximately 10 mm in diameter with a mid stent section measuring 9.4 mm and the proximal stent near the origin of the right pulmonary artery of 5.80 mm. The distal left pulmonary measured approximately 10 mm in diameter with a mid stent measuring 10.3 mm and the proximal stent near the origin of the left pulmonary artery is 6.8 mm diameter. The left femoral venous sheath was exchanged over wire for a 7-French sheath. Guidewires were then advanced through the respective venous sheath into the branch pulmonary arteries and simultaneous balloon pulmonary arterioplasty was performed using the two Z-Med

12 x 4 cm balloon catheter was advanced into the branch of pulmonary arteries and inflated maximally to 9 hemispheres of pressure on 5 occasions near complete disappearance of proximal waist. The balloon catheter was then exchanged for a 5-French Mistique catheter for pressure pull-back and measurement in the angiogram. The catheter's wires were then removed and final hemodynamic assessment was made with the wedge catheter.,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 angiograph injection in the main pulmonary 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 in normal. Mixed venous saturation that was not normal with no evidence of intracardiac shunt. Left side of the heart was mildly desaturated following a part to parenchymal lung disease with the partial pressure of oxygen of only 82 mmHg. Aphasic right atrial pressures were normal with an A-wave similar to the normal right ventricular end-diastolic pressure. Left ventricular systolic pressure was moderately elevated at 70% of systemic level and there was no obstruction into the proximal main pulmonary artery. There was a 20 mmHg of peak systolic gradient across the branch

pulmonary artery stents to the distal artery. Right and left pulmonary artery capillary wedge pressures were normal with an A-wave similar to the mildly elevated left ventricular end-diastolic pressure of 13 mmHg. Left ventricular systolic pressure was systemic. No outflow constriction 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 injection in the main pulmonary artery showed catheter induced pulmonary insufficiency, well functioning Contegra valve with no appreciable calcification. The proximal narrowing of the distal main pulmonary artery was appreciated. Neointimal ingrowth within the proximal stents were appreciated. There is good distal growth of the pulmonary arteries. Arborization appeared normal. Levophase contrast returned to the heart appeared normal with a well-functioning left ventricle and the right aortic arch. Following the branch pulmonary artery angioplasty that was increased in the mixed venous saturation, as well as an increase in the systemic arterial saturation. Right ventricular systolic pressure felt slightly to 40 mmHg with an increase in systemic arterial pressure with a systolic pressure ratio of 54%. The main pulmonary pressures remained similar. There was 10 mmHg systolic gradient into the branch of pulmonary arteries. There is an increase in distal branch of pulmonary arteries with the mean pressure increased from 16 mmHg to 21 mmHg. Final angiogram with injection in the main pulmonary artery showed a competent Contegra valve. A

brisk flow through the proximal branch stents with the improved caliber of the branch pulmonary artery lumens. There was no evidence of intimal disruption.,DIAGNOSES: ,1. Atrioventricular septal defect.,2. Tetralogy of Fallot with the pulmonary atresia.,3. Bilateral superior vena cava. The left cava draining to the coronary sinus.,4. The right aortic arch.,5. Discontinuous pulmonary arteries.,6. Down syndrome.,PRIOR SURGERIES AND INTERVENTIONS: ,1. Right modified Blalock-Taussig shunt.,2. Repair of tetralogy of Fallot with external conduit.,3. The atrioventricular septal defect repair.,4. Unifocalization of branch pulmonary arteries.,5. Bilateral balloon pulmonary angioplasty and stent implantation.,6. Pulmonary valve replacement with 16-mm Contegra valve.,CURRENT DIAGNOSES: ,1. Mild-to-moderate proximal branch pulmonary stenosis.,2. Well-functioning Contegra valve and current intervention. A balloon dilation of the right pulmonary artery.,3. Balloon dilation of left pulmonary artery.,MANAGEMENT: , The case will be discussed at Combined Cardiology and Cardiothoracic Surgery Case Conference and conservative outpatient management will be pursued. Further cardiologic care be directed by Dr. X.