HISTORY: , The patient is a 4-month-old who presented with respiratory distress and absent femoral pulses with subsequent evaluation including echocardiogram that demonstrated severe coarctation of the aorta with a peak gradient of 29 mmHg and associated dilated cardiomyopathy with fractional shortening of 16%. A bicuspid aortic valve was also seen without insufficiency or stenosis. The patient underwent cardiac catheterization for balloon angioplasty for coarctation of the aorta.,PROCEDURE: ,After sedation and 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 percutaneous technique a 4-French 8 cm long double lumen central venous catheter was inserted in the left femoral vein and sutured into place. There was good blood return from both the ports.,Using a 4-French sheath a 4-French wedge catheter was inserted into the right femoral vein and advanced through the right heart structures out to the branch of pulmonary arteries. The atrial septum was not probe patent.,Using a 4-French sheath a 4-French marker pigtail catheter was inserted into the left femoral artery and advanced retrograde to the descending aorta ,ascending aorta and left ventricle. A descending aortogram demonstrated discrete coarctation of the aorta approximately

8 mm distal to the origin of the left subclavian artery. The transverse arch measured 5 mm. Isthmus measured 4.7 mm and coarctation measured 2.9 x 1.8 mm at the descending aorta level. The diaphragm measured 5.6 mm. The pigtail catheter was exchanged for a wedge catheter, which was then directed into the right innominate artery. This catheter was exchanged over a wire for a Tyshak mini 6 x 2 cm balloon catheter which was advanced across the coarctation and inflated with complete disappearance of discrete waist. Pressure pull-back following angioplasty, however, demonstrated a residual of 15-20 mmHg gradient. Repeat angiogram showed mild improvement in degree of aortic narrowing. The angioplasty was then performed using a Tyshak mini 7 x 2 cm balloon catheter with complete disappearance of mild waist. The pigtail catheter was then reintroduced for a pressure pull-back measurement and final angiogram.,Flows were calculated by the Fick technique using an assumed oxygen consumption.,Cineangiograms were obtained with injection in the descending aorta.,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 pediatric intensive care unit in satisfactory condition. There were no complications.,DISCUSSION: , Oxygen consumption was assumed to be normal. Mixed venous saturation was low due to mild systemic arterial desaturation and anemia. There is no evidence of significant intracardiac shunt. Further the heart was desaturated due to

VQ mismatch.,Phasic right-sided pressures were normal as was the right pulmonary artery capillary wedge pressure with the A-wave similar to the normal left ventricular end-diastolic pressure of 12 mmHg. Left ventricular systolic pressure was mildly increased with a 60 mmHg systolic gradient into the ascending aorta and a 29 mmHg systolic gradient on pressure pull-back to the descending aorta. The calculated flows were mildly increased. Vascular resistances were normal. A cineangiogram with contrast injection in the descending aorta showed a normal left aortic arch with normal origins of the brachiocephalic vessels. There is discrete juxtaductal coarctation of the aorta. Flow within the intercostal arteries was retrograde. Following balloon angioplasty of coarctation of the aorta, there was slight fall in the mixed venous saturation and an increase in systemic arterial saturation as the fall in left ventricular systolic pressure from 99 mmHg to 92 mmHg. There remained a 4 mmHg systolic gradient into the ascending aorta and 9 mmHg systolic gradient pressure pull-back to the descending aorta. The calculated systemic flow fell to normal values. Final angiogram with injection in the descending aorta demonstrated improved caliber of coarctation of the aorta with mild intimal irregularity and a small left lateral filling defect consistent with a small intimal tear in the region of the ductus arteriosus. There is brisk flow in the descending aorta and appropriate flow in the intercostal arteries. The narrowest diameter of the aorta measured 4.9 x 4.2 mm.,DIAGNOSES: ,1. Juxtaductal coarctation of the aorta.,2. Dilated cardiomyopathy.,3. Bicuspid aortic valve.,4.

Patent foramen ovale.,INTERVENTION: , Balloon dilation of coarctation of the aorta.,MANAGEMENT: , The case will be discussed at combined Cardiology and Cardiothoracic Surgery Case Conference. The patient will be allowed to recover from the current intervention with the hopes of complete left ventricular function recovery. The patient will undoubtedly require formal coarctation of the aorta repair surgically in 4-6 months. The further cardiologic care will be directed by Dr. X.