Introduction: Anomalous origin of the left coronary artery from the pulmonary artery is a rare congenital anomaly occurring in 0.26% of patients with congenital heart disease. Patent ductus arteriosus is more common with an incidence of 0.02%-0.006% of live births. The presence of these two anomalies occurring together is obviously extremely rare.

We present the case of a patient with unrecognized ALCAPA with pulmonary hypertension who underwent PDA ligation for failure to thrive. Hyperventilation of the patient and closure of the duct resulted in a decreased flow to the left coronary artery with subsequent myocardial ischemia and ventricular fibrillation.

Case Report: A 1 month old boy, full term, 2.7 kg was scheduled for PDA ligation. The baby was born at 42 weeks gestation but was small for gestational age (2.1 kg). He was noted to have dysmorphic features (abnormal digits, hypertelorism, low set ears, webbed neck, widely-spaced nipples) and genetic work-up revealed a 49 XXXXY anomaly. The baby had significant failure to thrive and subsequent echocardiogram showed: large PDA with predominantly left to right flow, multiple small apical muscular VSDs, a large secundum ASD and PFO, severe pulmonary hypertension with systemic RV pressure, and mild mitral regurgitation. The left main coronary artery was noted to arise from ectopically from the rightward portion of the left sinus of Valsalva. The patient was scheduled for elective surgical PDA ligation.

After placement of a 24 gauge intravenous catheter, anesthesia was induced with etomidate, remifentanil and vecuronium. A 3.0 endotracheal tube and a right radial arterial line were placed without difficulty. Anesthesia was maintained with sevoflurane and a remifentanil infusion 0.1mcg/kg/min. Ventilation was initially FiO2 0.5 with air. Initial blood gas analysis revealed significant respiratory acidosis with a pH of 7.12, PaCO2 of 89mmHg and PaO2 126mmHg. Ventilation was adjusted in order to correct the acidosis and the patient at this point remained hemodynamically stable. Subsequent blood gas analysis after one hour revealed an improved pH of 7.32, PaCO2 of 56mmHg and PaO2 of 199mmHg. An initial attempt at video assisted thoracoscopic approach was abandoned due to technical difficulties. The PDA closure was achieved via left thoracotomy. The patient remained hemodynamically stable throughout the procedure, however as the team was preparing to transfer him to the Intensive Care Unit, the patient suddenly developed ventricular fibrillation. The rhythm was rapidly identified, he was defibrillated and received 3 rounds of epinephrine and lidocaine. He converted to sinus rhythm but required dopamine 5-7.5 mcg/kg/min for blood pressure control and was started on a prophylactic lidocaine infusion 40 mcg/kg/min. Emergency TTE showed severe dysfunction of LV and global ST elevations were present on ECG. The patient was transported to the cardiac ICU uneventfully. Over the next 8 hours the ECG changes slowly improved although inotropic support remained necessary. Repeat echocardiogram revealed only slightly improved ventricular function. No further delineation of coronary anatomy was possible. ALCAPA was suspected and the decision was made to perform a cardiac catheterization to further delineate the coronary anatomy. The catheterization revealed a large ASD and several small apical muscular VSDs, a Qp/Qs of 4:1 and an anomalous LMCA arising from the main PA. Transposition of the LCA to the aorta and closure of the ASD were scheduled and performed successfully the next day. The patient was extubated 2 days later and discharged home on captopril and furosemide after 5 days.

Discussion: Mortality for surgical repair of PDA is low (<1%) and the common complications include inadvertent ligation of PA or descending aorta, recurrent laryngeal nerve injury, excessive bleeding due to PDA disruption, and chylothorax. Myocardial ischemia prior to PDA can be anticipated in the presence of tachycardia and low aortic diastolic blood pressure. Myocardial ischemia following PDA ligation is an unexpected finding.

Dramatic changes occur in the coronary circulation in all children with ALCAPA within the first weeks of life. While the fetus remains in utero, the heart may develop quite normally. Initially after birth, the still relatively high pulmonary vascular resistance and pulmonary artery pressure allow the myocardium...
supplied by the anomalous artery to remain well perfused. In the few days after birth, pressure in the pulmonary trunk falls. Subsequent compromise of LV subendocardial perfusion develops due to both reduced antegrade flow and to frank retrograde flow in the LMCA with resultant coronary steal. In addition, all antegrade coronary flow is de-oxygenated mixed venous blood. Severe left ventricular dysfunction results with a 90% mortality if untreated. In this patient, the presence of a large PDA with resultant left to right shunting provided maintenance of high pulmonary artery pressure and antegrade delivery of relatively oxygenated blood to the LMCA. Surgical ligation of the PDA acutely reduced coronary perfusion pressure and oxygen delivery to the left ventricle with resultant ventricular fibrillation.

**Conclusion:** Suspicion of ALCAPA arose in this patient based on the clinical course following PDA ligation and the unusual origin of LMCA as delineated by echocardiography. Cardiac catheterization remains the gold standard for delineation of coronary artery anomalies. The objective of angiography is to establish the integrity and source of the coronary arteries. Anomalous origin of a coronary artery from the PA should be considered when coronary ischemia follows an acute reduction in PA pressure. The diagnosis of ALCAPA, once made, is an indication for urgent surgery with excellent surgical results being reported following re-establishment of a normal antegrade dual coronary artery system.