JACC Vol. 6, No. 1
July 1985:234–6

Ductus Arteriosus Aneurysm Presenting as Pulmonary Artery Obstruction: Diagnosis and Management

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The occurrence of pulmonary artery obstruction in an 8 day old infant as a complication of an aneurysm of a nonpatent ductus arteriosus is reported, together with the echocardiographic and angiographic findings. To relieve the obstruction, the aneurysm and an intrapulmonary thrombus were successfully removed with the use of cardiopulmonary bypass when the infant was 3 months old.

(J Am Coll Cardiol 1985;6:234–6)

An aneurysm of a nonpatent ductus arteriosus is an uncommon cardiovascular anomaly. Sequelae that have been reported (1,2) include rupture, embolism, infection and thrombosis of the aortic arch. Obstruction of the pulmonary artery tree has not been previously described in association with this abnormality. This report describes the physical findings, diagnosis and management of a 3 month old infant with obstruction to the main and left pulmonary arteries secondary to an aneurysm of the nonpatent ductus arteriosus and a thrombus extending into the pulmonary artery.

Case Report

A male infant weighing 3.3 kg was born at term with 1 and 5 minute Apgar scores of 9 and 10, respectively. Vomiting associated with feeding occurred from the first day of life, but the infant was otherwise well and was discharged at 3 days. At age 8 days, he was readmitted to the hospital with lethargy, poor feeding and vomiting. Shortly after admission, "cardiovascular collapse" occurred. The infant was resuscitated and transported to the medical center. On arrival, peripheral perfusion was good and a grade 3/6 ejection systolic murmur was audible in the pulmonary area with radiation into the left lung field. Marked hepatomegaly was present in addition to hyperbilirubinemia and prolonged prothrombin and partial thromboplastin times. Renal failure was indicated by the presence of elevated blood urea nitrogen and serum creatinine levels and decreased urinary output. An electrocardiogram showed right ventricular hypertrophy and right axis deviation. A chest X-ray film revealed a normal-sized heart with a round density in the left upper mediastinum and decreased pulmonary flow to the left lung. Pulmonary flow to the right lung was normal (Fig. 1).

A two-dimensional echocardiogram demonstrated a solid mobile mass within the main pulmonary artery (Fig. 2). This mass appeared to partially occlude the left pulmonary artery but the right pulmonary artery was patent. The aortic arch was normal, but the ductus arteriosus could not be visualized from the suprasternal position.

Catheterization findings. After an exchange transfusion, cardiac catheterization was performed. The right ventricular pressure was suprasystemic at 95/4 mm Hg and the left ventricular pressure was 75/4 mm Hg. The left pulmonary artery pressure distal to the mass was 18/8 mm Hg. A right ventriculogram (Fig. 3A) showed a nonopacified smooth mass within the main pulmonary artery that partially occluded the left pulmonary artery. A left ventriculogram (Fig. 3B) demonstrated patency of the aortic end of the ductus arteriosus and a large aneurysm of the ductus. No left to right shunting into the pulmonary artery was seen. An aneurysm of a nonpatent ductus arteriosus with obstruction to the main and left pulmonary arteries due to thrombosis was thought to be the most likely diagnosis, and surgical relief of the obstruction was recommended. Because of persistent hepatic and renal failure and, subsequently, the development of non-A, non-B hepatitis, surgery was

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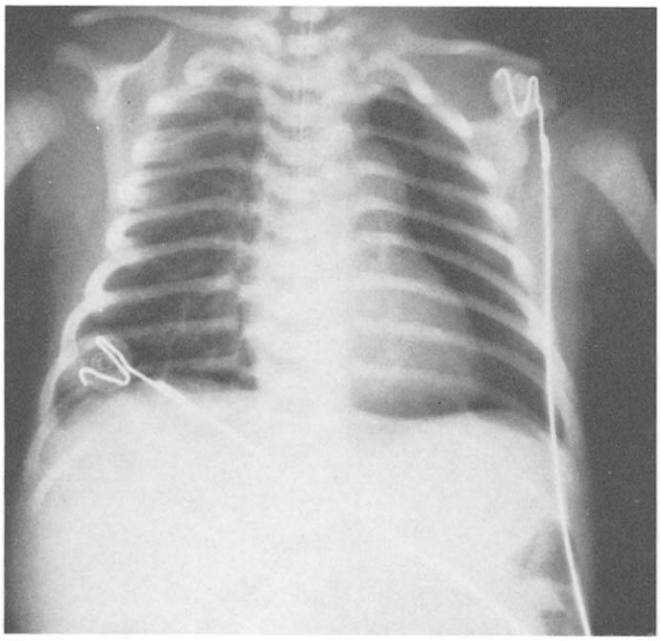
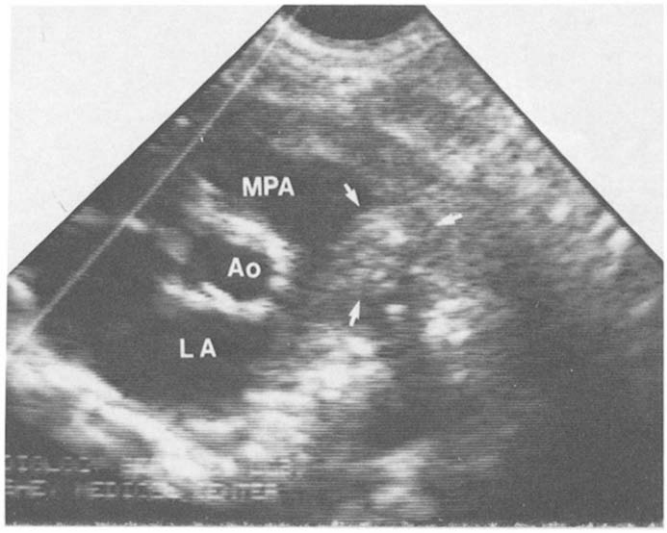


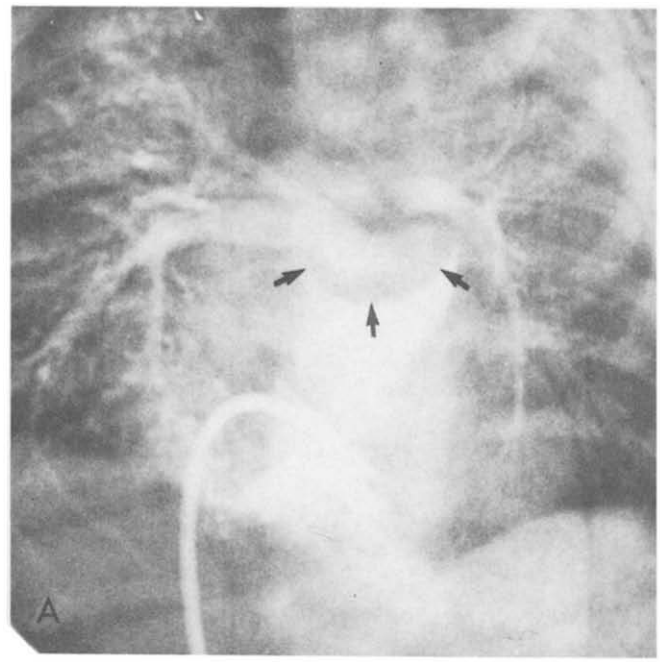
Figure 1. Anteroposterior chest X-ray film showing the soft tissue density in the left upper mediastinum.

delayed until the patient was 3 months of age. During this time, the infant ate and gained weight in a normal manner.

Surgical procedure and findings. The operation was performed through a left lateral thoracotomy and revealed an aneurysm of the ductus arteriosus (Fig. 4). The pulmonary end of the ductus contained a mass that could be palpated within the main pulmonary artery. The aortic end of the ductus arteriosus was divided and oversewn. Cardio-

Figure 2. Two-dimensional echocardiographic short-axis view at the level of the great arteries showing the mass (**arrows**) within the main pulmonary artery (MPA) with extension toward the left pulmonary artery. Ao = aorta; LA = left atrium.





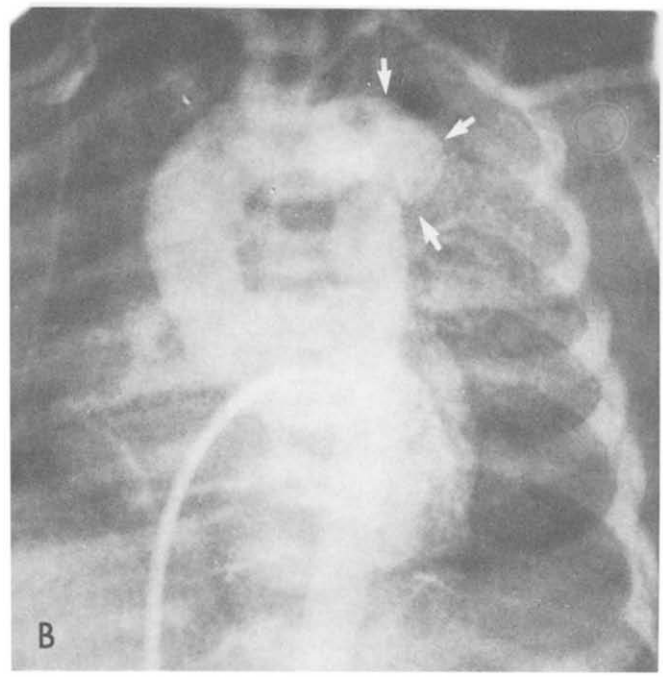


Figure 3. Angiocardiograms. **A**, Right ventriculogram showing a smooth nonopacified mass (**arrows**) within the main pulmonary artery. **B**, Left ventriculogram showing the aneurysm of the ductus arteriosus (**arrows**) with absence of opacification of the pulmonary arteries.

pulmonary bypass was established by inserting a large cannula into the right ventricle through the proximal pulmonary artery while the aorta was perfused in the mid descending portion. The main pulmonary artery was opened after it was occluded just distal to the cannula by a vascular clamp. The mass was dissected free of the main and proximal left pul-

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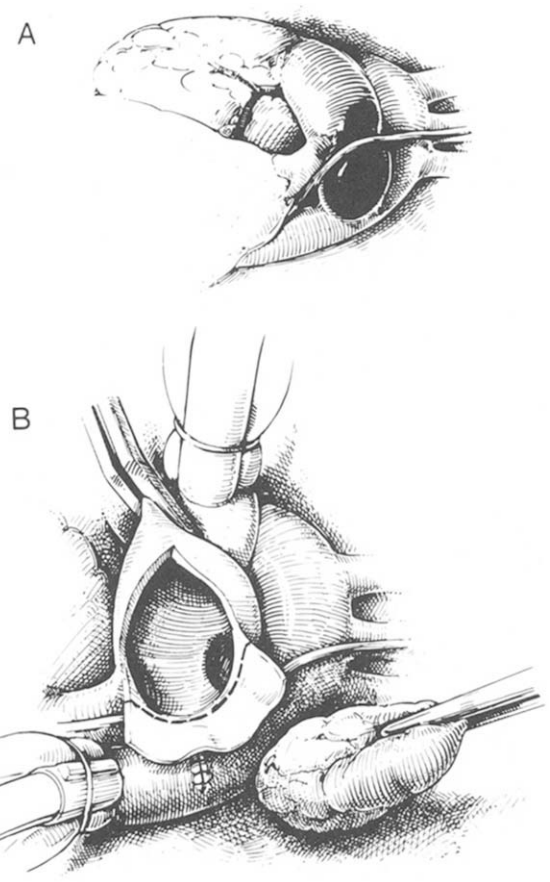

Figure 4. **A**, Illustration of the ductal aneurysm between the aorta and main pulmonary artery as observed at operation. **B**, Illustration of the technique used to remove the thrombus within the aneurysm. The proximal main pulmonary artery and descending aorta are cannulated for cardiopulmonary bypass. The aortic end of the ductus has been divided and oversewn.

monary arteries and what appeared to be an organized thrombus was removed (Fig. 4). After closure of the pulmonary artery, the patient was weaned from cardiopulmonary bypass. His postoperative course was uneventful and he was discharged 7 days later with a normal cardiac examination. Follow-up study at 5 months of age revealed the presence of a left diaphragmatic palsy, but the patient was otherwise well.

Discussion

Incidence. The incidence of aneurysm of the nonpatent ductus arteriosus has been reported to be low. However, Heikkinen et al. (3) retrospectively reviewed 1,138 chest X-ray films of newborn infants and made the diagnosis in approximately 1% of the infants based on the presence of a characteristic rounded mass in the left upper mediastinum. In two of these infants, the diagnosis was confirmed by surgical intervention.

Diagnosis. Pulmonary artery obstruction as a result of an aneurysm of a nonpatent ductus arteriosus had not been described. Despite this, the most likely diagnosis in our patient on the basis of the chest roentgenogram, echocardiogram and angiocardiogram was an aneurysm of the ductus arteriosus. The differential diagnosis consisted of a pulmonary embolus, thrombus or a tumor within the pulmonary artery. The mobility of the mass as seen on the echocardiogram indicated it was unlikely to be an embolus, and the ductal aneurysm noted on angiocardiography suggested that the mass within the pulmonary artery was an extension of a thrombus within the aneurysm rather than a tumor or an isolated thrombus.

Echocardiography revealed the cause of the pulmonary stenosis murmur and of the differential flow to the right and left lungs but did not reveal the ductal aneurysm. This may have been due to the more anterolateral and cranial position of the aneurysm when compared with the normal ductus arteriosus. Angiography was required to make an exact anatomic diagnosis of the ductal aneurysm.

Complications. A review of the world literature by Falcone et al. (1) revealed a complication rate of 43% in 61 patients with an aneurysm of the ductus arteriosus. Death occurred in 31% of these patients and rupture of the aneurysm, the most frequent complication, occurred in 18%. Embolism and infection were the other major complications described. McFaul et al. (2) reported on an infant with aortic arch thrombosis secondary to the extension of a thrombus from an aneurysm of the ductus arteriosus. Hoarseness and left phrenic nerve palsy may also result from a ductal aneurysm.

Pulmonary artery obstruction should be considered a complication of a ductal aneurysm due to thrombosis with the thrombus extending into the pulmonary artery. It can probably be diagnosed noninvasively by chest roentgenogram and an echocardiogram. Angiocardiography is indicated, however, if uncertainty remains. Surgical removal of the aneurysm and relief of the obstruction can be performed with the use of cardiopulmonary bypass.

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