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INTRODUCTION
Ductus arteriosus is an essential conduit in fetal life. After birth the duct will close, which results in connecting the pulmonary and systemic circulation in the series. In healthy term newborn babies, the functional closure of the duct commences within a few hours of birth and is followed by an anatomical closure over the next few weeks. A similar, but delayed pattern is seen in late preterm babies. Postnatal closure of the duct is important in an anatomically normal heart, however, its patency is essential in duct‑dependent congenital heart lesions. This is achieved by prostaglandin infusion.

Thrombus in Patent Ductus Arteriosus (PDA) or pulmonary arteries is extremely rare and can lead to pulmonary hypertension, shock, and refractory cardiac arrest in duct‑dependent congenital heart disease. We report here, a case of a preterm infant, with antenatal diagnosis of Tetralogy of Fallot (TOF) and pulmonary atresia, who developed a rapidly progressive thrombus in the left pulmonary artery and ductus arteriosus. Timely intervention with anticoagulation resulted in the quick resolution of the thrombus and averted life‑threatening complications.

CASE REPORT
Our patient was a female infant, born to a 28‑year‑old primigravida, with a past medical history of hypothyroidism and atrial septal defect, repaired at five years of age. Her antenatal serology was protective. Her blood sugars were normal. She was treated for pregnancy-induced hypertension with labetalol from 30 weeks of pregnancy. There was no family history of thrombophilia. The fetal echocardiogram at 19 weeks of gestation confirmed TOF with double outlet right ventricle (DORV)-type of ventriculoarterial connection and pulmonary atresia.

The infant was delivered at 34 + 3/7 weeks gestation by an emergency Cesarean section on account of fetal distress. She was vigorous at birth and had APGAR scores of 6 and 8, at one and five minutes, respectively. She did not need any resuscitation. Her birth weight was 1770 grams. The cord arterial blood gas was normal. She was cyanotic with pre‑ and postductal saturations of 73 and 76%, respectively.

We report a case of a premature baby, who had an antenatal diagnosis of pulmonary atresia, with a double outlet right ventricle (DORV). This baby was managed with prostaglandin infusion to keep the ductus patent. During the initial echo, the cardiologist noted a thrombus in the thin, long, and tortuous duct, which rapidly progressed to partially occlude the left pulmonary artery. This was treated with unfractionated heparin and the thrombus resolved within two hours of commencing the heparin infusion. This was followed by low molecular heparin for six weeks. Had this been missed, the pulmonary circulation would have been severely compromised and would have led to refractory cardiac failure. This article highlights one of the possible causes of failure of prostaglandin infusion in maintaining ductal patency in duct‑dependent cardiac lesions.

Key words: Congenital heart disease, heparin, patent ductus arteriosus, prostaglandin, thrombus
Her cardiovascular examination revealed normal S1, single S2, and a grade three out of six continuous murmur at the left upper sternal border. The rest of the physical examination was unremarkable.

At one hour of age she received low flow oxygen at 200 ml/minute due to decreasing oxygen saturation (SpO2). Infusion of prostaglandin E1 (PGE1) was started at a dose of 0.05 mcg/kg/minute and later increased to 0.1 mcg/kg/minute, to attain an SpO2 between 80 and 85%. Umbilical arterial and venous catheters were placed and the tip position was confirmed on X-ray. She had to be intubated due to apnea following prostaglandin infusion. An echocardiogram (ECHO) was performed by the attending cardiologist at two hours of age, which confirmed the antenatal diagnosis. Ductus arteriosus was tortuous, thin, and long. During the course of the ECHO, a thrombus was noted in the left pulmonary artery partially occluding its lumen, which rapidly enlarged and extended into the ductus arteriosus [Figures 1 and 2].

After confirming the absence of an intracranial bleed and a normal coagulation profile, a bolus of unfractionated Heparin 75 units/kg was given followed by 28 units/kg/hour as an infusion. Hematocrit was 55%. Thrombophilia workup was not done as she was already receiving heparin through the umbilical arterial line. A repeat ECHO done at four hours of life and, one hour after initiation of heparin infusion, demonstrated complete resolution of the thrombus from both the duct and left pulmonary artery [Figure 3]. Heparin infusion was discontinued and low molecular weight heparin (enoxaparin) was commenced to maintain the anti-Xa levels between 0.2 and 0.4 (prophylactic regimen). Enoxaparin was continued for six weeks, without any complication. No recurrence of thrombus was noted in the ECHO after two weeks of discontinuation of enoxaparin. The infant underwent a surgical repair at eight weeks and was discharged home at 10 weeks of life.

Discussion: The exact incidence of neonatal pulmonary artery thrombosis is not known and to the best of our knowledge, this is the first case report of a rapidly progressive pulmonary artery and ductus arteriosus thrombosis in a neonate with duct-dependent congenital heart lesion. Occlusion of the ductus arteriosus by the thrombus would have been catastrophic in this setting. There is a high index of suspicion that thrombus in the duct or pulmonary artery can easily be missed on clinical examination or by ECHO unless there is a high index of suspicion. No other cases of thrombus in ductus arteriosus and main pulmonary artery or its branches by echocardiography in term newborns presented with hypoxemia, respiratory distress, cyanosis, and systolic murmur. Three of these babies needed surgical removal of the thrombus, as anticoagulant management failed. One of them was successfully managed with low molecular weight heparin and one on long-term aspirin. None of these babies had congenital heart disease, thrombotic diseases, or central lines. Thrombus in ductus and pulmonary arteries in term infants, presenting as congenital heart disease needing surgical thrombectomy has been reported. None of them had any identifiable risk factors for thrombosis. Gen Niwayana has reported an autopsy series of six babies who had thrombus in the ductus and pulmonary artery, who failed to respond to both medical and surgical treatments. Monica et al., have argued that premature closure of the duct in the face of an immature coagulation system in newborns might tilt the balance toward formation of the thrombus in the closing duct and its extension into the pulmonary artery. The ductus arteriosus in babies with pulmonary atresia is thin and tortuous. This causes a turbulent blood flow. Experimental evidences suggest that turbulence in blood flow can predispose to thrombosis. The histological tissue characteristics are also altered in infants receiving PGE1. Premature closure of the duct, immature coagulation system in newborns, tortuous ductus arteriosus with turbulent flow in the setting of pulmonary atresia, and the presence of vascular catheters, might have all been involved in the mechanism of thrombus formation and rapid progression in our case.

Therapeutic options: Anticoagulation, thrombolytics, and surgical thrombectomy are the therapeutic options available in term and preterm neonates. Surgical thrombectomy

![Figure 1: Echocardiogram showing growing thrombus in the duct and LPA](image-url)
is indicated if medical therapy fails\cite{13,14} and carries a higher risk of complications.

Unfractionated Heparin and Low molecular weight Heparin (Enoxaparin) alter the prothrombotic status in the infant and prevent progression of the thrombus. Unfractionated heparin molecules have higher binding sites and initiate anticoagulation more rapidly than low molecular weight heparin. In acute life-threatening situations, unfractionated heparin may have an advantage.\cite{15} In our case we have used unfractionated Heparin in the initial phase of treatment, due to its rapidity of action and good safety profile. With Enoxaparin therapeutic levels may be difficult to achieve in a limited time frame.\cite{16} However, it has a predictable and sustained action in the long term and it needs less frequent monitoring of the coagulation status.\cite{17} We initiated it as soon as resolution of the thrombus was demonstrated. We did not the use the recombinant tissue plasminogen activator (r-TPA) as first-line intervention, because of the lower safety profile in the premature infant.

Left pulmonary artery (LPA) coarctation due to closing ductus arteriosus is a dangerous complication and a cause of LPA occlusion in babies with TOF with pulmonary atresia.\cite{18,19} This case suggests that thrombosis of the LPA may be an additional mechanism for LPA occlusion that needs to be considered in patients deteriorating on prostaglandin infusion.

In our case, if an ECHO had not been conducted in time, the clinical condition would have only deteriorated and ended up in an increased dose of prostaglandins and respiratory support, without much benefit.

**CONCLUSIONS**

- Thrombus formation in the branch pulmonary arteries or ductus arteriosus may be a cause of persistent or acute desaturation in babies with duct-dependent cardiac lesions on prostaglandin infusion. Rapid access to echocardiography is of immense benefit in this setting

  - Initial therapy with unfractionated Heparin may be beneficial while getting organized for other treatments like recombinant tissue plasminogen activator and surgery.

**REFERENCES**


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