Pulmonary Artery Thrombus Presenting as Cyanotic Cardiac Defect in a Neonate

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Abstract:
Background: Pulmonary artery thrombosis presenting as pulmonary arterial hypertension (PAH) is a known entity in adult patients but presentation in neonate is very rare.
Case characteristics: Two days old neonate, presented with PAH with cyanosis, diagnosed with thrombus involving main and left pulmonary artery causing complete occlusion of left.
Intervention: Pulmonary artery thrombolysis was done with tissue plasminogen activator (tPA) followed by heparin for 3 months.
Outcome: There was complete dissolution of thrombus after thrombolysis.
Message: When assessing a neonate with cyanosis and PAH, pulmonary artery thrombi should be ruled out before labeling it as PPHN.
Abbreviations: MPA: main pulmonary artery, LPA: left pulmonary artery, RPA: right pulmonary artery, RV: right ventricle

1. INTRODUCTION
The prevalence of symptomatic neonatal arterial thrombosis is approximately 1 in 40,000 births, with 90% of cases linked to indwelling intra-arterial catheters (1-3). Other risk factors are sepsis, polycythemia, maternal diabetes, asphyxia, and inherited thrombophilias (1-3). Neonate presenting with isolated pulmonary artery thrombosis with severe pulmonary arterial hypertension is a rare event (4). We are reporting such a term neonate presenting with congestive heart failure (CHF) and cyanosis and diagnosed as thrombus involving main and left pulmonary artery.

2. CASE DESCRIPTION
A 4-day-old neonate, 1st in birth order, product of 35 weeks caesarian section delivery (indication fetal distress with previous caesarian and maternal short stature) with birth weight of 2 kg. Baby was apparently well for 1st 48 hrs of life when parents noticed fast breathing, feeding difficulty and bluishness. Baby was admitted for evaluation and stabilization with local pediatrician. With suspicion of cardiac defect; he was referred to our hospital for further evaluation on day 4th of life. On examination, there was tachycardia, tachypnea, and cyanosis with severe respiratory distress. Chest X-ray showed cardiomegaly, no parenchymal lung lesion. Echocardiography done with Phillips IE33, broadband transducers (S12, S8), showed dilated right atrium and right ventricle, patent foramen ovale with right to left shunting, confluent pulmonary arteries, and echogenic shadow in main pulmonary artery extending to origin of left pulmonary artery (Figure 1a). Doppler interrogation showed moderate tricuspid regurgitation with peak gradient of 70mmHg. There was small flow in right pulmonary artery while no flow seen in left pulmonary artery on color flow mapping suggestive of almost complete occlusion of left pulmonary artery. A CT pulmonary angiography confirmed the presence of a thrombus partially occluding main pulmonary artery and completely occluding the left pulmonary artery.

Baby was kept In intensive care unit. After sending workup for prothrombotic state (protein C, protein S activity, antithrombin III level, factor V Leiden mutation, anticardiolipin antibodies, homocystein and serum ANA), he was started on thrombolytic therapy with tissue plasminogen...
activator (tPA) at 0.3 mg/kg/hour infusion. Blood investigations showed low level functional antithrombin III (55%, normal 80-120%), protein C (37%, normal 70-140%) and protein S (54%, normal 50-140%).

After 4 hours of infusion, echocardiography showed small flow in left pulmonary artery. At the same time, child had bradycardia with hypotension and was put on ventilatory support. Metabolic profile (serum electrolytes, blood sugar, and serum calcium) was normal. Ultrasound head did not show any abnormality. CT head was also done to rule out any intracranial bleed.

tPA infusion was stopped and child was started on heparin infusion (20 unit/kg/hr). Review echo after 24 hrs showed good flow in left pulmonary artery (figure 1b). Heparin infusion was given for 48 hrs and later shifted to subcutaneous heparin. The neonate was discharged home on daily subcutaneous low-molecular-weight heparin. It was given for 3 months. Pro-coagulant work-up done at 3 months showed improvement in level of functional antithrombin III (75%, normal 80-120%), protein C (59%, normal 70-140%) and protein S (69%, normal 50-140%).

At 3-month follow-up he was doing well and there was no thrombus in the pulmonary artery.

![Fig 1a](image1a.png) ![Fig 1b](image1b.png)

**Figure 1a.** Small flow seen in right pulmonary artery with complete occlusion of left pulmonary artery

**Figure 1b.** Post thrombolytic therapy; Color flow mapping showing good flow in both pulmonary arteries

3. DISCUSSION

There are few case reports about neonatal thrombosis including pulmonary artery(4) and also of aorta(5). With large pulmonary artery thrombus, presentation is with respiratory failure and cyanosis while with thrombus in aorta is with coarctation of aorta (4,5). The current data on modes of presentation in the neonates with pulmonary artery thrombosis are very limited.

Neonates are more to development of thrombosis as the naturally occurring inhibitors of coagulation, antithrombin, heparin cofactor II, protein C, and protein S, are also reduced at birth, although á2 macroglobulin is significantly increased(6). The fibrinolytic system also appears different, with plasminogen concentrations around 50% of adult values (7). These features all tend to be gestationally dependent and are therefore more pronounced in the preterm infant (8). Our child was borderline preterm (35 weeks gestational age) and born by cesarean section for fetal distress. Blood investigations showed low level of functional antithrombin II, protein C and protein S. All these factors made him high risk candidate for development of spontaneous thrombosis.

The diagnosis of pulmonary artery thrombus is suspected on echocardiography only and it requires a high index of suspicion. In all newborn with echocardiography features of pulmonary arterial hypertension with no structural heart defect, proximal or distal pulmonary artery thrombus should be ruled out before labeling it as persistent pulmonary arterial hypertension of neonate (PPHN). Thrombus in proximal pulmonary artery can be seen by echocardiography while for detailed definition and in case of thrombus beyond hilum, CT pulmonary angiography is needed to diagnose and quantify the thrombus (3).
Treatment of neonatal spontaneous arterial thrombosis is controversial. An expert panel on the management of arterial thromboembolic events in neonates recommended that therapy should be individualized based on the extent of thrombosis and the urgency of the clinical situation (9).

Treatment options are thrombolytic therapy with tissue plasminogen activator, anticoagulation with heparin, low-molecular-weight heparin surgery, and catheter-based embolectomy. In case of large thrombus load causing hemodynamic compromise, thrombolytic therapy seems to be the most preferred treatment option (10). Drugs used are tPA, urokinase and streptokinase. tPA is preferred agent in neonate because of improved clot lysis in vitro (10). Most important complication is bleeding.

In our case, with tPA and heparin infusion, there was complete canalization of left pulmonary artery. We had to discontinue tPA after 4 hrs of treatment as baby had neurological event. In 4 hrs only, there was partial recanalization of left pulmonary artery flow. After ruling out any intracranial bleed, heparin infusion was started. With heparin infusion there was complete dissolution of pulmonary artery thrombus. Baby was discharged on maintenance therapy with subcutaneous heparin for 3 months.

4. CONCLUSION

When assessing a neonate with cyanosis and pulmonary arterial hypertension, in a structurally normal heart, it is important to look for pulmonary artery thrombi before labeling it as PPHN. Pulmonary artery thrombus, if diagnosed and treated on time, has good outcome.

REFERENCES


