Obstructive Total Anomalous Pulmonary Venous Return Masqueraded as Severe Meconium Aspiration Syndrome in a Newborn

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Abstract
Certain congenital heart diseases (CHD) may be difficult to diagnose when associated with poor precordial window or concurrent critical condition. We report a case of supracardiac type TAPVR with obstruction, of which the initial diagnosis was meconium aspiration syndrome (MAS) complicated with persistent pulmonary hypertension of the newborn (PPHN). The patient’s critical condition required extracorporeal membrane oxygenation (ECMO) support, and the diagnosis of TAPVR was made incidentally when an open heart surgery was required to remove a thrombi in the aortic arch. The patient recovered well after a rerouting surgery of TAPVR. The possibility of making an earlier diagnosis from clinical signs, reversed differential cyanosis and high oxygenated blood in the superior vena cava (SVC) is further explained. (J Pediatr Resp Dis 2013;9:65-68)

Key words: meconium aspiration syndrome, extracorporeal membrane oxygenation, total anomalous pulmonary venous return.

INTRODUCTION
Total anomalous pulmonary venous return (TAPVR) is a cyanotic congenital heart disease (CHD) in which pulmonary veins do not enter the left atrium directly. Instead, all pulmonary veins drain to systemic veins, an atrial septal defect (ASD) or patent foramen ovale (PFO) must be present to shunt adequate blood flow from the right atrium (RA) to the left atrium (LA). The incidence of TAPVR is 10.6 in 100,000 Taiwanese live births, ranking 10th in all CHDs.1,2 Prenatal diagnosis is difficult. Newborn infants with obstructed TAPVR may present with marked cyanosis, tachypnea, dyspnea, hypoxemia, and metabolic acidosis must receive immediate corrective surgery. In addition to echocardiographical study for the cardiovascular anomaly, some clinical features can help to make an earlier diagnosis of TAPVR.

We herein report a case of supracardiac type TAPVR with obstruction, that was initially considered as a case of meconium aspiration syndrome (MAS) complicated with persistent pulmonary hypertension of the newborn (PPHN).

CASE REPORT
A 40-week-and-6-day old male infant presented with meconium stained amniotic fluid after a vaginal delivery. His birth body weight was 2,610 g (below the 5th percentile of Taiwanese birth weight curve), body length was 48 cm (at the 10th percentile), and head girth was 30 cm (below the 10th percentile). The Apgar score at one and five minutes after birth were 6 and 8, respectively. His breathing showed suprasternal and subcostal retractions. The pulse oximeter (SpO2) showed 50-60% of oxygen saturation. The oral and nasal suctioning evacuated a lot of sticky meconium and his respiratory distress improved. His heart rate

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was 100-110 bpm, respiratory rate 60/min, and the limbs were cold with poor perfusion. The patient was immediately intubated and transferred to neonatal intensive care unit (NICU).

A chest radiograph showed bilateral pneumothorax with ground-glass appearance of the lung fields, (Figure 1) which was managed with thoracentesis and a right side chest tube was established. (Figure 2) Arterial blood gas (ABG) examination revealed hypoxemia, metabolic acidosis with elevated lactate (pH 7.241, PaCO$_2$ 28.9 mmHg, PaO$_2$ 35.8 mmHg, HCO$_3$- 12.5 mmol/L, SaO$_2$ 59.9%, lactate 11.3 mmol/L). Under the impression of MAS with PPHN, the patient received sodium bicarbonate infusion to correct severe metabolic acidosis, high frequency oscillatory ventilator (HFOV), inhaled nitric oxide (iNO) and inotropic support (dopamine and milrinone). However, his SaO$_2$ still remained around 50-60%. To compare the preductal and posductal O$_2$ saturation, two SpO$_2$ monitors were placed at right upper limb and one lower limb, which showed 40% and 60%, respectively, consistent with a pattern of reversed differential cyanosis.

Echocardiography was arranged to exclude structural heart disease. Because of pneumothorax and poor lung condition, the precordial echo window was very poor. He had mild right atrial and right ventricular enlargement, slightly subnormal left ventricular contractility with flat septal motion, secundum type ASD with bidirectional shunt, large perimembranous type ventricular septal defect (VSD), and a large patent ductus arteriosus (PDA), both with right to left shunt. (Figure 3) The spatial relationship and the connection of the great arteries was normal. Because of the persistent hypoxemia, a venoarterial extracorporeal membrane oxygenation (VA-ECMO) was urgently set up, with V cannula into the right internal jugular vein and A cannula into the right internal carotid artery, the centrifugal pump could provide a steady blood flow of 0.4 L/min by a pump speed 1950 rpm.

At 3 days of age, an echocardiography follow-up showed an thrombi in the aortic arch. Thereupon, emergent surgery to remove the thrombi and to total repair of his VSD and PDA was performed. (Figure 4) Unexpectedly, high oxygenated blood was found in the superior vena cava (SVC) while weaning off the cardiopulmonary bypass. Further examination of the pulmonary venous anatomy showed no connection between PVs and the LA. Instead, the confluened PV drained to the SVC and all the PVs looked small. A diagnosis of obstructive type TAPVR was made and a total correction by rerouting the PV confluence to the posterior wall of left atrium, VA-ECMO was then removed successfully. The post-operative course was uncomplicated. He was extubated 1 week after the operation and was discharged from the hospital 3 weeks later.

**DISCUSSION**

Because of poor precordial echo window, TAPVR was not found by the first echocardiography and urgent ECMO was set up to rescue his hypoxemia. Besides, the typical findings of TAPVR, including
enlarged right side chambers, and compressed left side chambers, were not shown in four chamber view. Because the VSD provided blood flow to the left ventricle, so the typical small LV was not detected. Under ECMO support, a couple of image studies could have been arranged to reveal the cardiovascular anomalies, which include TEE, CHD-CT and cardiac catheterization. Poor precordial window can be overcome through transesophageal approach, but transesophageal echocardiography (TEE) is limited by the size of the infant. If the initial echocardiography is indeterminable, CT-CHD should be considered. However, the risk of transferring a newborn with unstable vital sign is high.

Newborns with PPHN have right-to-left shunting across a PFO/ASD, PDA, or both, causing significant hypoxemia. If the shunting is exclusively at PDA, a gradient in the arterial oxygen pressure of greater than 20 mmHg or oxygen saturation of greater than 5% preductally (upper limbs) and postductally (lower limbs) could be observed. This phenomenon is called differential cyanosis. This phenomenon could be explained by the following mechanism. The oxygenated blood returning to the right atrium from anomalous pulmonary veins to the SVC is preferentially directed to the right ventricle and further flows through PDA to the lower limbs. Deoxygenated blood returning from the inferior vena cava (IVC) is preferentially directed to the left atrium across PFO/ASD and further supplying the upper limbs. However, due to the association of VSD, this mechanism may not be reliably to explain the reverse differential cyanosis in this case.
In conclusion, we reported a newborn with obstructive TAPVR and VSD, initially treated as MAS with PPHN, and once rescued by ECMO. The diagnosis was made during a surgery to repair VSD and to remove intra-aortic thrombi. Concomitant VSD made the diagnosis of TAPVR difficult. When congenital cardiovascular anomaly is suspected on a patient under ECMO support, further image studies, including TEE, MDCT or cardiac cath, should be performed.

REFERENCES