Case report

Successful multimodality management of severe pulmonary arterial hypertension during pregnancy with VA-ECMO and atrial septostomy using stent

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SUMMARY

A 30-year-old Thai woman (gravida 1, para 0) at 33 weeks gestation was referred to our hospital due to acute right ventricular failure. Pulmonary vasodilators were gradually administered before delivery. On the verge of sudden postpartum cardiac circulation collapse, she was resuscitated with veno-arterial extracorporeal membrane oxygenation (VA-ECMO). Abdominal compartment syndrome was developed in the early period of the mechanical support. Knowledge of pathophysiology about pulmonary arterial hypertension during pregnancy was applied. Atrial septostomy was the effective procedure for discontinuing mechanical support (VA-ECMO) corresponding to the suitable timing for maximal effect of pulmonary vasodilators. The patient and her child were safe and discharged in 2 months after the admission.

BACKGROUND

Pulmonary arterial hypertension (PAH) in pregnancy is rare due to contraceptive education and even uncommon for mothers of identical twins who were coincidentally pregnant at almost the same period. PAH is known to be associated with significantly high morbidity and may lead to maternal death during pregnancy, labour and postpartum period. According to consensus guidelines, termination of early pregnancy is strongly recommended. However, our patient was in the third trimester of her pregnancy and in need of multidisciplinary management after the postpartum death of her identical twin. The challenging case required careful preparation in every stage of treatments.[1 2]

CASE PRESENTATION

A 30-year-old Thai woman (gravida 1, para 0) at 33 weeks gestation with a good history of antenatal care programme was referred to our Critical Care Medicine Department after gradually developing dyspnoea on exertion, orthopnoea and legs pitting oedema for 1 month. The previous health status of the patient was normal. She was able to do hard work in a grocery store before admission. The patient had an identical twin whose gestational age was 2 weeks older. Her identical twin also developed acute hypoxic respiratory failure and fetal distress. She underwent emergent caesarean section and died of acute right ventricular failure the next day after giving birth. Fortunately, our patient was recognised and asked for her referral by the primary doctor who concerned that she might find herself in the same situation as her twin. The patient was admitted into medical intensive care unit followed by her transthoracic echocardiogram (TTE) (table 1; video 1). Other lab results including arterial blood gas and serum lactate values were normal. The multidisciplinary team including pulmonologist, cardiologist, cardiothoracic surgeon, anesthesiologist, obstetrician and paediatrician agreed to wait for maternal medical preparation and antenatal glucocorticoid therapy for premature infant. Over 1 week of admission, high flow oxygen, inhaled iloprost and intravenous milrinone were initiated with gradual dose titration under close blood pressure monitor with arterial line placement. Subcutaneous low-molecular-weight heparin was prescribed to prevent hypercoagulability, commonly used for pregnancies in the presence of PAH. She was doing well with our treatment. We did not monitor pulmonary capillary wedge pressure but TTE outcome was of insignificant change. Despite the patient's uterine contraction which unavoidably worsen maternal circulation, the team took a decision on caesarean delivery with spinal anaesthesia blockage in cardiothoracic operative room as veno-arterial extracorporeal membrane oxygenation (VA-ECMO)[3 4] cannulation was put on standby. After 40 min at the recovery room, the patient suddenly lost consciousness without sign of hypotension, and asystole cardiac arrest was detected. Cardiopulmonary resuscitation was performed in 5 min with a return of spontaneous circulation. Meanwhile, right femoral artery and vein cannulas were immediately inserted with initiated 3.5 L/min ECMO flow rate at 3400 rpm, fractional delivered oxygen 60% and sweep gas flow 2 L/min. The patient remained haemodynamically stable on ECMO and fully gained of neurocognitive functions without hypothermia activation.

TREATMENT

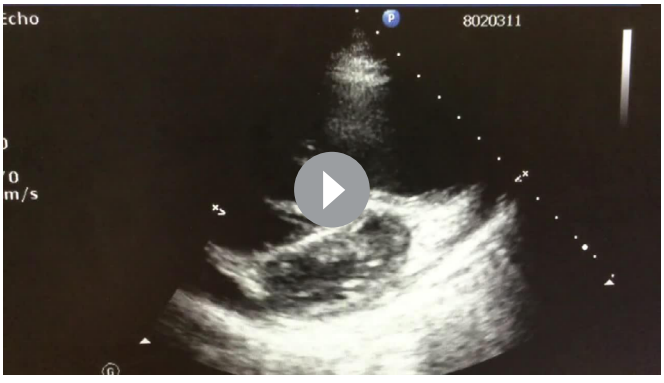
Targeted pulmonary vasodilators included inhaled nitric oxide gradually titrated up to 60 ppm, inhaled iloprost at 22.5 μg/day, intravenous continuous drip of milrinone 0.7 μg/kg/min, intravenous continuous drip of epinephrine 0.1–0.2 μg/kg/min, weekly intravenous of levosimendan 0.2 μg/kg/min, oral

Table 1 TTE at the first day of admission and after VA-ECMO decannulation (1 month after admission)

TTE findings	The first day of admission	After VA-ECMO decannulation (1 month after admission)
Cardiac chamber	Normal size of left atrium Enlarged size of right atrium Enlarged size of right ventricle Small left ventricular cavity with D-shape	Normal size of left atrium Mild enlarged right atrium Mild enlarged right ventricle Improved D-shape of left ventricle Well-seated atrial septostomy stent with bidirectional flow
Left ventricular ejection fraction	84%	75%
Diastolic function	Dysfunction grade I	Normal
Cardiac valves	Moderate to severe tricuspid regurgitation	Mild tricuspid regurgitation
PAP (mm Hg)	Systolic PAP 72 Diastolic PAP 29 Mean PAP 44	No report
Right atrial pressure (mm Hg)	15	8
Right ventricular systolic pressure (mm Hg)	71.8	52.8
Tricuspid annular plane systolic excursion (cm)	1.7	19
Tricuspid regurgitation peak velocity (m/s)	4.2	3.3
E/E′	7	7.6

PAP, pulmonary artery pressure; TTE, transthoracic echocardiogram; VA-ECMO, veno-arterial extracorporeal membrane oxygenation .

bosentan 250 mg/day and oral sildenafil titrated up to 300 mg/day. All drugs were simultaneously given.[5] On postoperative day 5, the patient complained lower abdominal pain, caused by an increase of intra-abdominal pressure to 20 mm Hg that was indirectly measured from the bladder. The patient's haemoglobin dropped from 10 to 7 mg/dL in 6 hours. Her platelet dropped to 70×10^9/L while her activated clotting time (ACT) and partial thromboplastin time (PTT) were stable in controlled range. Bedside abdominal ultrasound detected an amount of fluid in lower half of abdominal cavity. The blood pressure was still stable with tachycardia. Meanwhile, the VA-ECMO showed signs of volume depletion; ECMO flow decreased from 3.5 to 2.8–3 L/min swinging back and forth of cannulas (chattering of lines), and central venous pressure decreased to 4 mm Hg.



Video 1 Parasternal short axis and four-chamber view of TTE in the first day of admission. TTE, transthoracicechocardiogram.

Active intra-abdominal bleeding with abdominal compartment syndrome was diagnosed. Intravenous heparin was temporarily held and corrected all coagulopathy followed by thromboelastography. Small-sized pigtail catheter was performed at the bedside due to the high-risk surgical procedures. After 4 hours, urine output, intra-abdominal pressure and ECMO flow were recovered. The team discussed the issue of VA-ECMO decannulation due to the unpredictable intra-abdominal bleeding; however, right ventricular function from TTE monitor did not significantly improve. Accordingly, right to left shunt was initiated as bridging procedure to release pressure from right-sided heart chamber. At the second week of admission, the use of atrial septostomy at a stent diameter of 3 mm to discontinue mechanical haemodynamic support was failed.

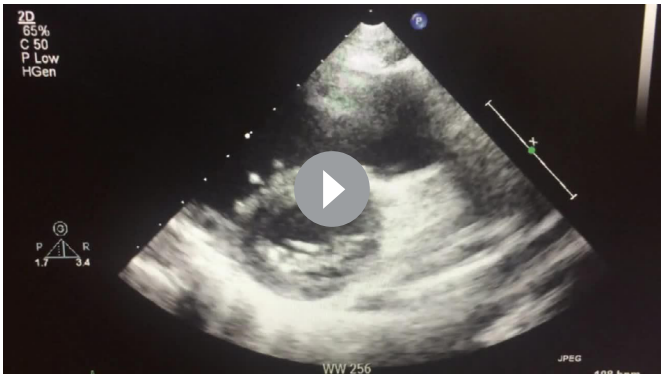
OUTCOME AND FOLLOW-UP

VA-ECMO was totally placed for 1 month. Atrial septostomy at a stent diameter of 8 mm was used and it eventually made a successful attempt to discontinue VA-ECMO support. Accordingly, right to left shunt was initiated as bridging procedure for releasing pressure from right-sided heart chamber. She was finally discharged without major complications from our hospital after 2 months of admission with home oxygen cannula and oral medications (sildenafil 300 mg/day, bosentan 250 mg/day, selexipaq 1600 mg/day, ivabradine 5 mg/day and dabigatran 220 mg/day).

DISCUSSION

Our patient was suspected familial PAH with previous normal status. Physiological changes of pregnancy included an increase of circulatory volume, oxygen demand and coagulability as well as a decrease of systemic vascular resistance[6] . All changes slowly occurred over the course of gestation, easily detected in the antenatal care programme.[5] The loss of our patient's identical twin taught her a bitter lesson, yet gave her a survival chance in careful preparation for delivery. According to the mortality, postnatal maternal deaths usually occur within 1 hour to 7 days due to volume shifts secondary to relief of inferior vena cava compression acutely altered cardiac preload, which cause acute right ventricular decompensation in patient with vulnerable right-sided cardiac function.

Another sudden effect of cardiac failure after delivery was a drop of serum prostaglandin level increasing pulmonary vascular resistance (PVR). Nowadays, a caesarean section is a recommended surgical procedure to deliver baby because it eliminates the possibility of uncontrolled vaginal bleeding and haemodynamic effects under spinal anaesthesia. Although we prepared some pulmonary vasodilators 5 days before delivery, the sudden cardiac arrest from acute right ventricular failure occurred 1 hour after delivery. After the femoral VA-ECMO was immediately inserted, the circulation and neurocognitive function achieved recovery. As for the prenatal and perinatal VA-ECMO support, we agreed that it would rather increase risks of bleeding and site complications than provide benefits. Timing for weaning from ECMO for the patient with pulmonary hypertension was slower than patients with other aetiologies, increasing the risk of possibility of dynamic shunt and right heart failure. Since we were faced with unpredictable massive intra-abdominal bleeding, the period of decannulation of VA-ECMO was shorter than our expectation. Atrial septostomy at a stent diameter of 3 mm was performed to maintain haemodynamic by releasing some right-sided heart pressure but it resulted in circulation failure without mechanical support. Despite gradual cardiac improvement from



Video 2 Parasternal short axis and four-chamber view of TTE after VA-ECMO decannulation (1 month after admission). TTE, transthoracicechocardiogram; VA-ECMO, veno-arterialextracorporeal membrane oxygenation.

TTE monitor (table 1; video 2), we did not have enough experience to decide when the suitable time for decannulation would be, particularly in the second chance. Right-to-left shunt was later performed with a larger stent diameter of 8 mm to avoid unanticipated problems. The patient was stable after weaning and safely decannulated from ECMO. However, she became one of the emergent candidates in the heart–lung transplantation list. Permanent contraception was seriously discussed in the family, and familial PAH was suspected in these identical twins. The genetic counselling of PAH will be discussed again among the outpatient clinic personnel so that PAH in twin children will be definite after classification.

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Learning points

- Bleeding is the most common complication of extracorporeal membrane oxygenation that requires a careful balance between risks and benefits.
- The pathophysiology of pulmonary arterial hypertension during pregnancy is required for multimodality management.
- The optimal and suitable of timing, maximal effect of pulmonary vasodilators and mechanical support were challenging.[7]
- Multidisciplinary team approach to complex and critically ill patients is a key to successful therapeutic management.
- The atrial septostomy may be the additional successful method rescuing in severe pulmonary hypertension patient with maximum medications.[8]

8 Baruteau A-E, Barnetche T, Morin L, *et al*. Percutaneous balloon atrial septostomy on top of venoarterial extracorporeal membrane oxygenation results in safe and effective left heart decompression. *Eur Heart J Acute Cardiovasc Care* 2018;7:70–9.