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CASE REPORT
Cardiovascular surgery

A 1-month-old infant with pulmonary hypertensive crisis after cardiac surgery was successfully rescued with connection of left atrial and right ventricular pressure measurement tubes: a case report

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Background Pulmonary hypertensive crisis is a complication with extremely high mortality after surgery of congenital heart disease. However, there are still no treatment guidelines or expert consensus on the standard treatment of pulmonary hypertensive crisis, and the effect of conventional treatment is still unsatisfactory. We present a case of a patient who developed pulmonary hypertensive crisis after cardiac surgery, and was successfully rescued with a pioneering method, which has never been reported so far.

Case summary An infant with congenital heart disease had undergone cardiac surgery successfully. Due to obvious myocardial oedema, sternal closure was delayed. The left atrial and right ventricular pressure monitoring tubes, both of which were connected through a triplet, were inserted into right pulmonary vein and pulmonary artery, respectively, and the triplet was in closed condition. On the night of the surgery, pulmonary hypertensive crisis occurred. Emergency bedside thoracotomy was given, and the triplet was turned on urgently to make the left atrial and right ventricular pressure monitoring tubes connected. Meantime, conventional treatment was performed. Eventually, the pulmonary hypertensive crisis was quickly relieved, and the infant was discharged 9 days later.

Discussion The left atrial and right ventricular pressure monitoring tubes are placed intraoperatively in patients who both need delayed sternal closure and have high risk factors for pulmonary hypertensive crisis, by which could not only monitor the pressure of left atrium and right ventricle in real time but also effectively relieve the right ventricular pressure instantaneously when pulmonary hypertensive crisis occurs, as well as remedy ischaemia of systemic and coronary circulation.

Keywords Pulmonary hypertensive crisis • Pulmonary hypertension • Congenital heart disease • Cardiac surgery • Case report

ESC curriculum 6.4 Acute heart failure • 6.7 Right heart dysfunction • 7.5 Cardiac surgery • 9.1 Aortic disease • 9.6 Pulmonary hypertension

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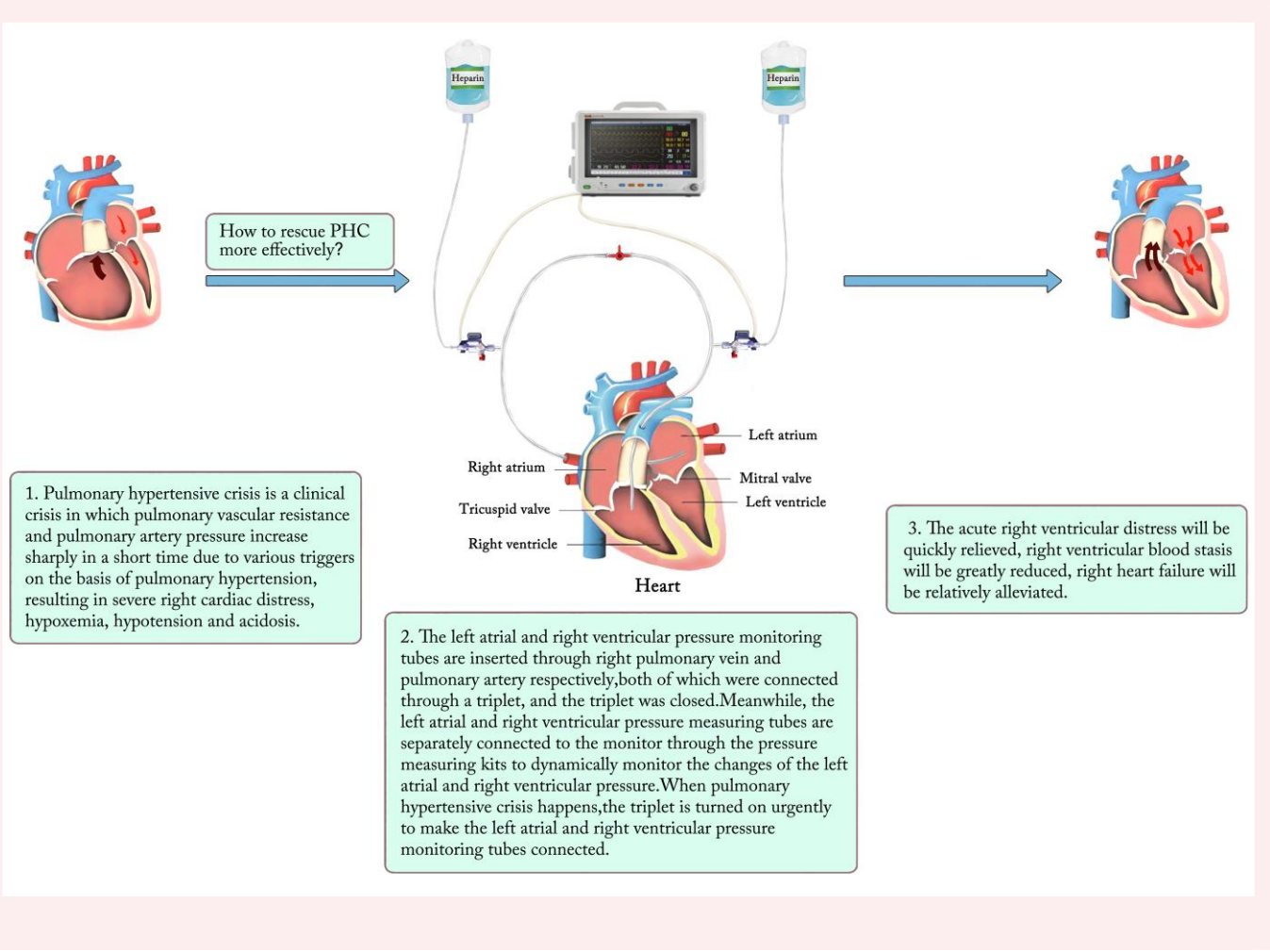
Learning points

- Pulmonary hypertensive crisis is a complication with extremely high mortality after surgery of congenital heart disease, but the effect of conventional treatment is still unsatisfactory.
- The left atrial and right ventricular pressure monitoring tubes are placed intraoperatively in patients who both need delayed sternal closure and have high risk factors for pulmonary hypertensive crisis, by which could effectively relieve pulmonary hypertensive crisis.

Introduction

Pulmonary hypertensive crisis is a clinical crisis in which pulmonary vascular resistance and pulmonary artery pressure increase sharply in a short time due to various triggers on the basis of pulmonary hypertension, resulting in severe hypocardiac discharge, hypoxaemia, hypotension, and acidosis, which is also a serious complication of children after surgery of congenital heart disease.[1–5] It progresses so fast that rapid deterioration or even death is likely to occur if it is not treated timely and effectively. However, there are still no treatment guidelines or expert consensus on the standard treatment of pulmonary hypertensive crisis so far. Most cardiac centres manage such emergencies by full sedation, inhalation of pure oxygen and nitric oxide,[6] or drugs to reduce pulmonary vascular resistance and pulmonary artery pressure, but the therapeutic effect is still unsatisfactory. Here, we present a case of a 1-month-old infant with interruption of aortic arch, ventricular septal defect, patent ductus arteriosus, and pulmonary artery hypertension (IAA/VSD/PDA/PH) who developed a pulmonary hypertensive crisis after cardiac surgery. Using this case report as a foundation, we discuss the mechanism and effect of the innovative therapy that has never been reported so far in order to provide an exploratory scheme for more effective treatment of pulmonary hypertensive crisis.

Summary figure



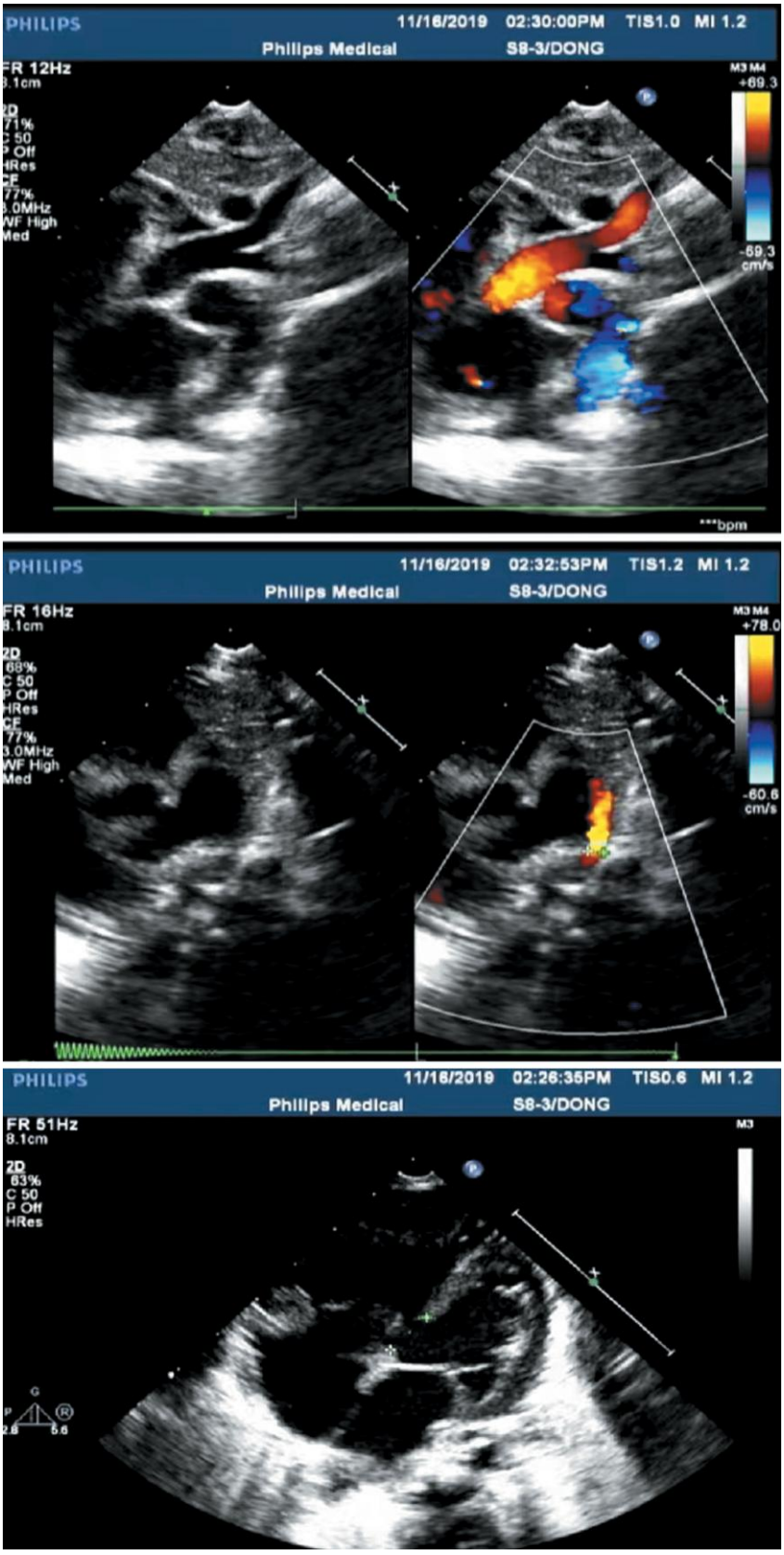


Figure 1 Transthoracic echocardiography figure for the pre-operative status of the infant.

Case presentation

A 1-month-old infant (sex, female; height, 51.2 cm; body weight, 3.1 kg; body surface area, 0.21 m^2) was hospitalized due to IAA/VSD/PDA/PH (*Figure 1*).

Cardiovascular CT revealed a ventricular septal defect of 13 mm and the continuity of the aortic arch at the distal end of the left subclavian artery was interrupted, where the descending aorta was connected to the left pulmonary artery via an arterial catheter with an internal diameter of about 5 mm. Meantime, the ratio of pulmonary to systemic blood pressure measured by cardiac ultrasound was 0.95, which met the diagnostic criteria of congenital heart disease combined with severe pulmonary hypertension.

After completing relevant examinations, aortic arch interruption correction, ventricular septal defect repair, ductus arteriosus cut-off, and suture were performed and the operation was successful. Due to obvious myocardial oedema in the infant, sternal closure was delayed. The left atrial and right ventricular pressure monitoring tubes, both of which were connected through a triplet, were inserted into right pulmonary vein and pulmonary artery, respectively, and the triplet was in closed condition (*Figures 2* and *3*). When the infant returned to intensive care unit, ventilator-assisted breathing was received, and the non-invasive cardiac monitoring system (Most-Care system) was used to closely monitor the vital signs. Meanwhile, the left atrial and right ventricular pressure measuring tubes were separately connected to the monitor through the pressure measuring kits to dynamically monitor the changes of the left atrial and right ventricular pressure (*Figure 3*).

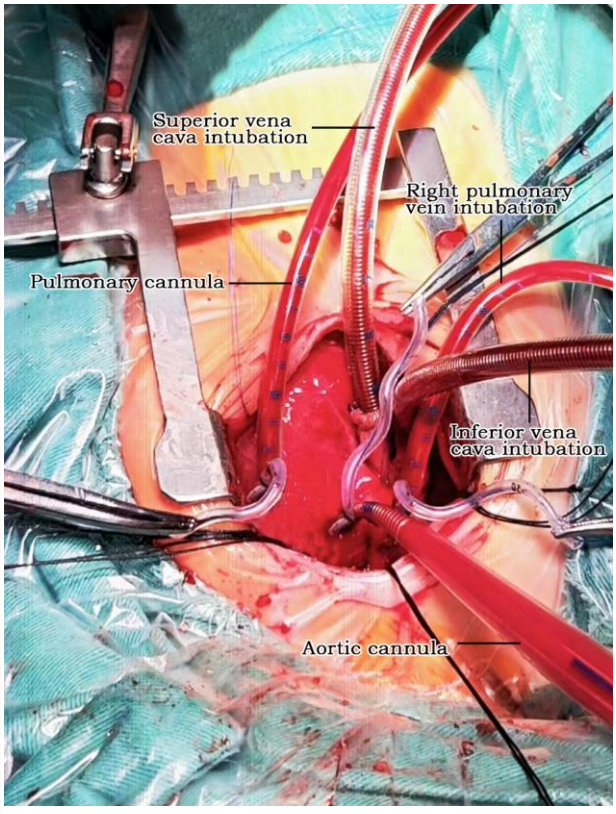


Figure 2 The left atrial and right ventricular pressure monitoring tubes were inserted through right pulmonary vein and pulmonary artery, respectively.

According to the infant's condition, selective pumping of vasoactive drugs such as dopamine, dobutamine, milrinone, epinephrine, and other vasoactive drugs were given to improve coronary perfusion and cardiac function.[7,8]

On the night of the surgery, pulmonary hypertensive crisis was triggered by endotracheal intubation with sputum aspiration. Pulmonary artery pressure soared from 18–22/13–15 mmHg to 80/53 mmHg, systemic circulation pressure plunged from 78–83/44–49 mmHg to 40/22 mmHg, blood oxygen saturation (SpO_2) decreased from 98% to 84%, and heart rate dropped from 150–158 b.p.m. to 51 b.p.m. simultaneously (*Table 1*). Emergency bedside thoracotomy was given, by which it is found that the right atrium and right ventricle were significantly enlarged with distress, and the triplet was turned on urgently to make the left atrial and right ventricular pressure monitoring tubes

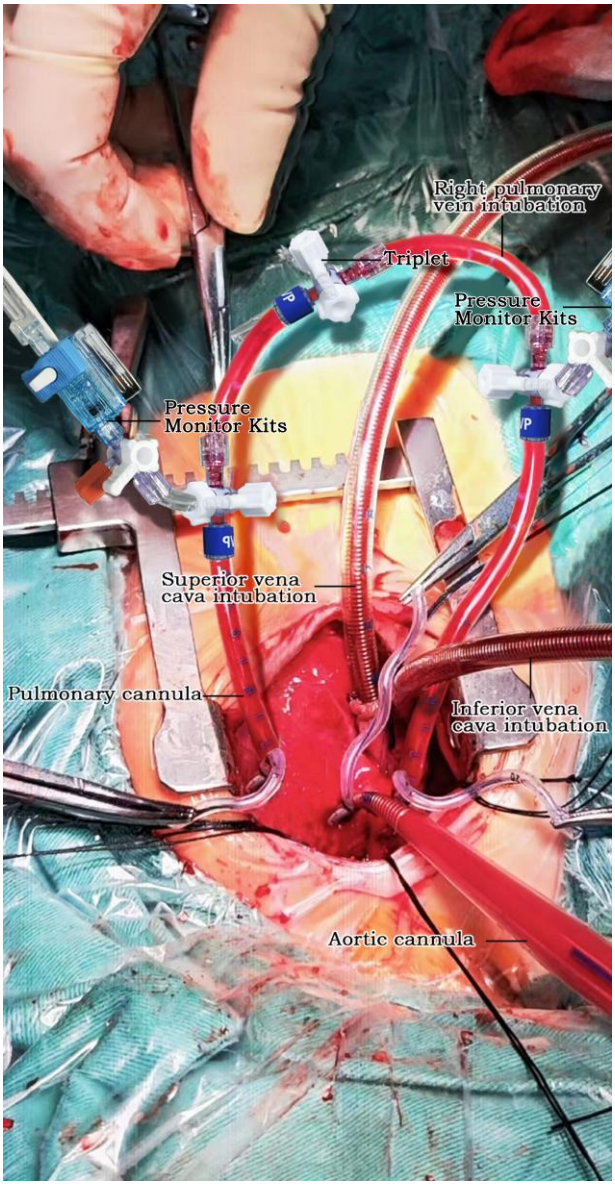


Figure 3 The left atrial and right ventricular pressure monitoring tubes were connected with a triplet, both of which were connected to the pressure measuring kits, respectively.

Table 1 Data before and after rescue of pulmonary hypertensive crisis

	Before PHC	Beginning of PHC	In rescue	After rescue
BP (mmHg)	78–83/44–49	43/22	68–72/44–50	83–88/50–53
PAP (mmHg)	18–22/13–15	80/53	65–70/40–52	19–22/15–17
LAP (mmHg)	5–7	1	10–13	4–7
SpO_2 (%)	96–98	84	83–89	95–99
CI (L/min/m^2)	3.18–3.25	1.15	2.58–2.70	3.13–3.28
HR (b.p.m.)	150–158	51	83–110	155–163

PHC, pulmonary hypertensive crisis; PAP, pulmonary artery pressure; LAP, left atrial pressure; SpO_2, blood oxygen saturation; CI, cardiac index; HR, heart rate.

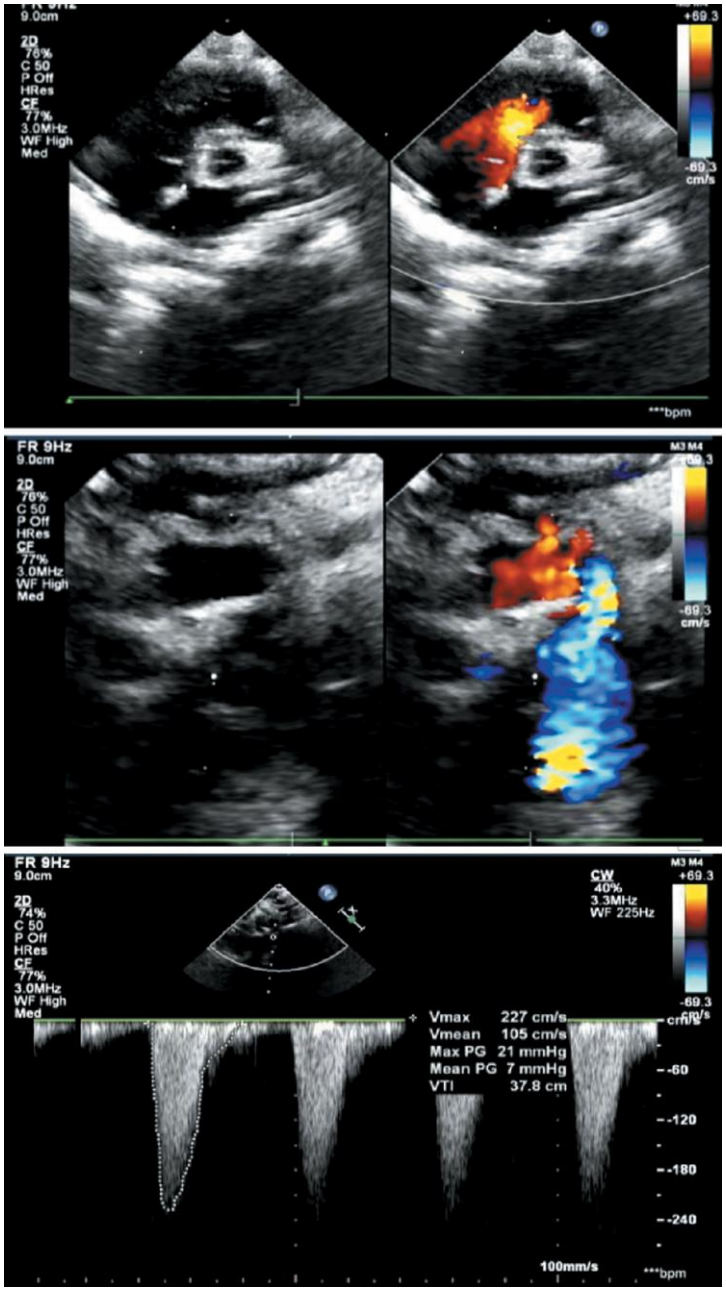


Figure 4 Transthoracic echocardiography figure of the infant one year after surgery.

connected. Meantime, pure oxygen and nitric oxide (20 ppm) were inhaled by breathing bag, and fentanyl, midazolam, rocuronium bromide, and other drugs were given to enhance sedation and muscle relaxation therapy. The systemic blood pressure increased instantly to 68–72/44–50 mmHg, right cardiac distress relieved immediately, and the heart rate soon increased to 83–110 b.p.m. Three minutes later, the SpO_2 increased to 89%, and the pulmonary artery pressure decreased to 19–22/15–17 mmHg. The triplet was turned off, and ventilator was given to assist breathing subsequently, and then the SpO_2 increased to 95–99%, blood pressure to 83–88/50–53 mmHg, and heart rate to 155–161 b.p.m. (*Table 1*). After 20 min of observation, the infant's vital signs were stable and there was no recurrence of pulmonary hypertensive crisis, so the infant was given delayed sternal closure again.

Thirty-six hours after surgery, bedside sternal closure was performed, the left atrial and right ventricular pressure monitoring tubes were removed smoothly, and the infant was discharged 9 days later. One year after surgery, transthoracic echocardiography showed the infant recovered well (*Figure 4*).

Discussion

Pulmonary hypertensive crisis is a complication with extremely high mortality after surgery of congenital heart disease. The central link of its onset is the sharp increase of pulmonary artery pressure caused by various causes, eventually resulting in right heart failure and low systemic hypotension.[9,10] There are many causes of pulmonary hypertensive crisis after surgery of congenital heart disease, including infection, hypoxia, sputum stimulation, inadequate sedation, surgery, and etc. Some children with congenital heart diseases are complicated with severe pulmonary hypertension before operation, and a large number of inflammatory factors are produced during cardiopulmonary bypass with pulmonary interstitial exudation. After surgery, they are more prone to pulmonary hypertensive crisis, resulting in extreme contraction of pulmonary vessels, obvious increasing of right ventricular afterload, and acute right heart failure. Meanwhile, due to left ventricular ischaemia, coronary perfusion is significantly reduced, and total heart failure will also occur subsequently.

At present, the treatment of pulmonary hypertensive crisis in most heart centres includes: (i) remove the incentive; (ii) keep the airway unobstructed and provide adequate oxygen; (iii) calm down to reduce emergency stimuli; (iv) correct right heart failure and hypotension; and (v) use of drugs to reduce pulmonary hypertension: including nitric oxide for inhalation; prostacycline analogues, such as iloprostaglandin; endothelin receptor antagonists, such as bosentan; vasopressin, which could produce better haemodynamic changes in the treatment of pulmonary hypertensive crisis[11]; and phosphodiesterase-5 inhibitors, such as sildenafil.

Nevertheless, acute right ventricular distress caused by pulmonary hypertensive crisis cannot be immediately alleviated by the conventional treatment methods above. However, the creative therapy, namely left atrial and right ventricular pressure tube bridging, has an immediate effect in the treatment of acute right cardiac distress. When pulmonary hypertensive crisis occurred, the left atrium and right ventricular pressure tubes were connected instantly, by which acute right ventricular distress was quickly relieved, right ventricular blood stasis was greatly reduced, right heart failure was relatively alleviated, systemic blood volume through right-to-left shunt were supplied timely, and thereby alleviating systemic circulation and coronary circulation ischaemia. Although the right-to-left blood shunt is not conducive to the maintenance of SpO_2, we found that the SpO_2 of the infant did not decrease significantly during the whole rescue process.

When pulmonary hypertensive crisis is relieved, the left atrial and right ventricular pressure tubes need to be gradually disconnected, so there is no permanent right-to-left shunt, and is still a normal physiological circulation state eventually.

Conclusion

In conclusion, the left atrial and right ventricular pressure monitoring tubes are inserted intraoperatively in patients who both need delayed sternal closure and have high risk factors for pulmonary hypertensive crisis, by which could not only monitor the pressure of left atrium and right ventricle in real time but also effectively relieve the right ventricular pressure instantaneously when pulmonary hypertensive crisis occurs, as well as remedy ischaemia of systemic and coronary circulation. The effect is significant, which is worthy of further large clinical sample study.

Lead author biography

Dr Hailong Song is a senior associate chief physician of cardiac surgery in the First Hospital of Hebei Medical University. He is good at surgical treatment of congenital heart diseases in children.

Consent: The legal guardian of the infant has given a full consent to the publication of this manuscript in accordance with COPE guidelines.

Conflict of interest: None declared.

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Data availability

The data underlying this article will be shared on reasonable request to the corresponding author.

References

1. Bernier ML, Romer LH, Bembea MM. Spectrum of current management of pediatric pulmonary hypertensive crisis. *Crit Care Explor* 2019;**1**:e0037.
2. Friesen RH, Alswang M. Changes in carbon dioxide tension and oxygen saturation during deep sedation for paediatric cardiac catheterization. *Paediatr Anaesth* 1996;**6**:15–20.
3. de Caen AR, Berg MD, Chameides L, Gooden CK, Hickey RW, Scott HF, *et al.* Part 12: Pediatric Advanced Life Support: 2015 American Heart Association Guidelines Update for Cardiopulmonary Resuscitation and Emergency Cardiovascular Care. *Circulation* 2015;**132**:S526–S542.
4. Bronicki RA, Baden HP. Pathophysiology of right ventricular failure in pulmonary hypertension. *Pediatr Crit Care Med* 2010;**11**:S15–S22.
5. Wheller J, George BL, Mulder DG, Jarmakani JM. Diagnosis and management of postoperative pulmonary hypertensive crisis. *Circulation* 1979;**60**:1640–1644.
6. Germann P, Braschi A, Della Rocca G, Tuan Dinh-Xuan A, Falke K, Frostell C, *et al.* Inhaled nitric oxide therapy in adults: European expert recommendations. *Intensive Care Med* 2005;**31**:1029–1041.
7. Blaise G, Langleben D, Hubert B. Pulmonary arterial hypertension pathophysiology and anesthetic approach. *Anesthesiology* 2003;**99**:1415–1432.
8. Salehi A. Pulmonary hypertension: a review of pathophysiology and anesthetic management. *Am J Ther* 2012;**19**:377–383.
9. Haddad F, Doyle R, Murphy DJ, Hunt SA. Right ventricular function in cardiovascular disease, part II: pathophysiology, clinical importance, and management of right ventricular failure. *Circulation* 2008;**117**:1717–1731.
10. Brunner N, Jesus Perez VA, Richter A, Haddad F, Denault A, Rojas V, *et al.* Perioperative pharmacological management of pulmonary hypertensive crisis during congenital heart surgery. *Pulm Circ* 2014;**4**:10–24.
11. Sugawara Y, Mizuno Y, Oku S, Goto T. Effects of vasopressin during a pulmonary hypertensive crisis induced by acute hypoxia in a rat model of pulmonary hypertension. *Br J Anaesth* 2019;**122**:437–447.