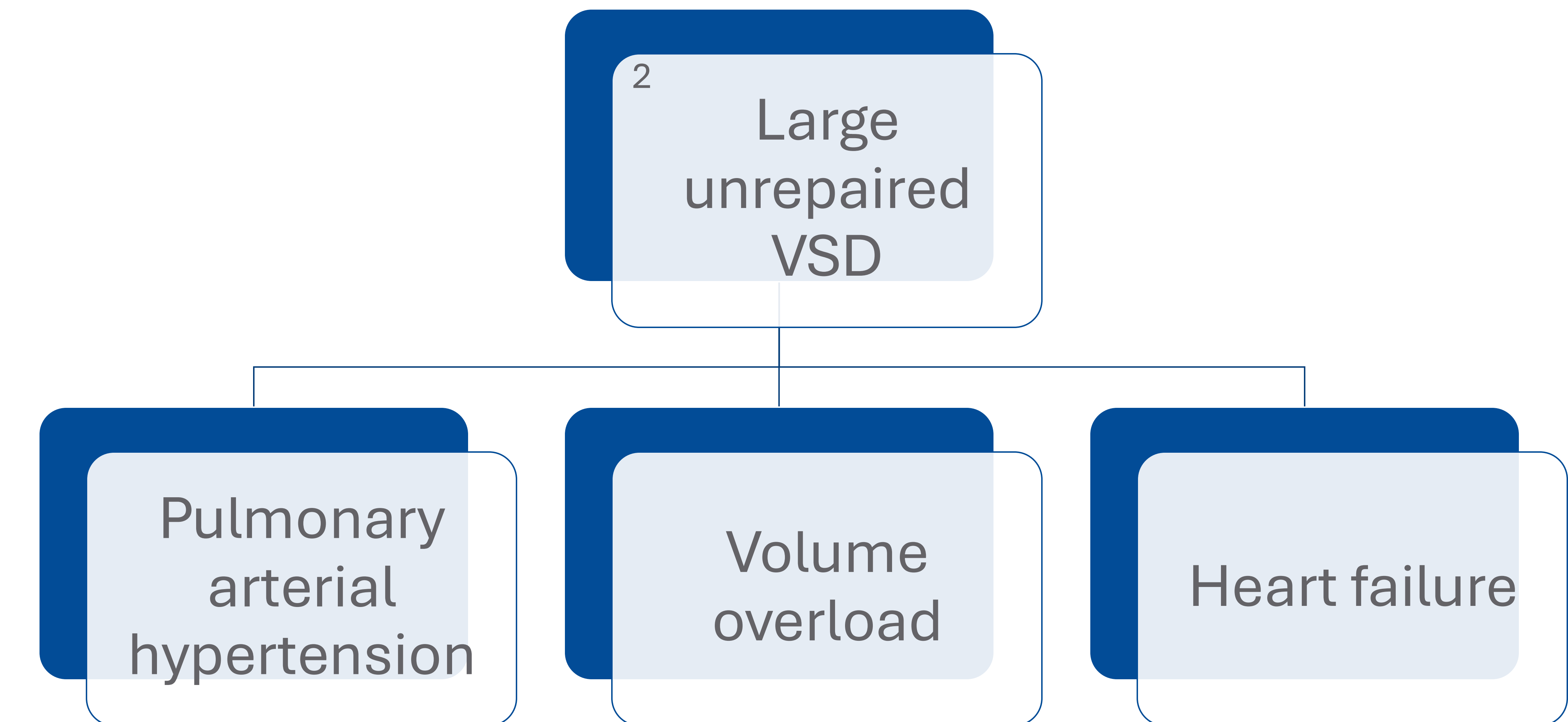


Managing cesarean delivery complicated by severe pulmonary arterial hypertension secondary to newly diagnosed conotruncal ventricular septal defect

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Introduction

- Ventricular septal defect (VSD) is the most common congenital heart disease
- ~30% isolated defects and occurs up to 50% with other defects¹
- Conotruncal VSDs occur in the outlet portion of the ventricular septum
 - Associated with defects such as tetralogy of Fallot¹
- Pregnancy unmasks morbidities and increases risk of maternal & fetal complications^{2,3}



Case Report

Pre-operative

- 19-year-old primigravida diagnosed at 33w3d with heart murmur
- Later developed dyspnea and was admitted for optimization
- Echocardiogram (Fig. 1) and cardiac catheterization
 - **Biventricular dysfunction** (RVEF 29%, LVEF 50%), **severe PAH** (RVSP 76 mmHg)
- Initiated pulmonary vasodilators: sildenafil and epoprostenol

Intra-operative

- Scheduled cesarean delivery at 36w3d

Epidural

Arterial Line

Vasopressors

- Epinephrine
- Vasopressin

Inhaled nitric
oxide (iNO)

Femoral ECMO
microcannulation

- Complicated by postpartum hemorrhage (PPH) secondary to uterine atony and coagulopathy
 - Treated with oxytocin, tranexamic acid, 2U pRBC, norepinephrine to prevent hypotension



Figure 1. Patient's transthoracic echo with large perimembranous VSD and overriding aorta. The RV is hypertrophic and dilated, suggesting pressure and volume overload

Post-operative

- Admitted to the intensive care unit (ICU)
- Weaned off epoprostenol and discharged on postoperative day 9

Discussion

- Unrepaired VSD carries high risk of maternal HF, arrhythmia, and hemorrhage³
- PAH and RVEF can be optimized with **pulmonary vasodilators** and **inotropic support**⁴
- Delivery planning requires **multidisciplinary discussion** with maternal-fetal medicine, anesthesiology, and cardiology
 - Cesarean delivery: associated with more hemodynamic shifts, but provides controlled environment
 - Vaginal delivery: prolonged labor and Valsalva maneuvers can precipitate cardiovascular collapse
- Elevated risk of PPH due to **coagulopathy** caused by iNO and prostacyclins⁴
 - Recognition and treatment important to prevent hypovolemia-mediated worsening of cardiac output

Intraoperative Goals

- Avoid sudden hemodynamic changes
- Maintain euvolemia
- Preserve biventricular function

References

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3. BJOG 2010; 117(16):683-69
4. BJA Education 2023; 23(1): 24-31