

BACKGROUND

Double Outlet Right Ventricle (DORV):

- 1/10,000 live births¹
- Features:² (Figure 1)
 - Pulmonary artery and aorta arise from the right ventricle
 - Concomitant VSD
 - Hypoplastic left ventricle
- Location of the VSD, presence of pulmonary stenosis, and other concomitant cardiac abnormalities determine hemodynamics and palliation strategy of the DORV

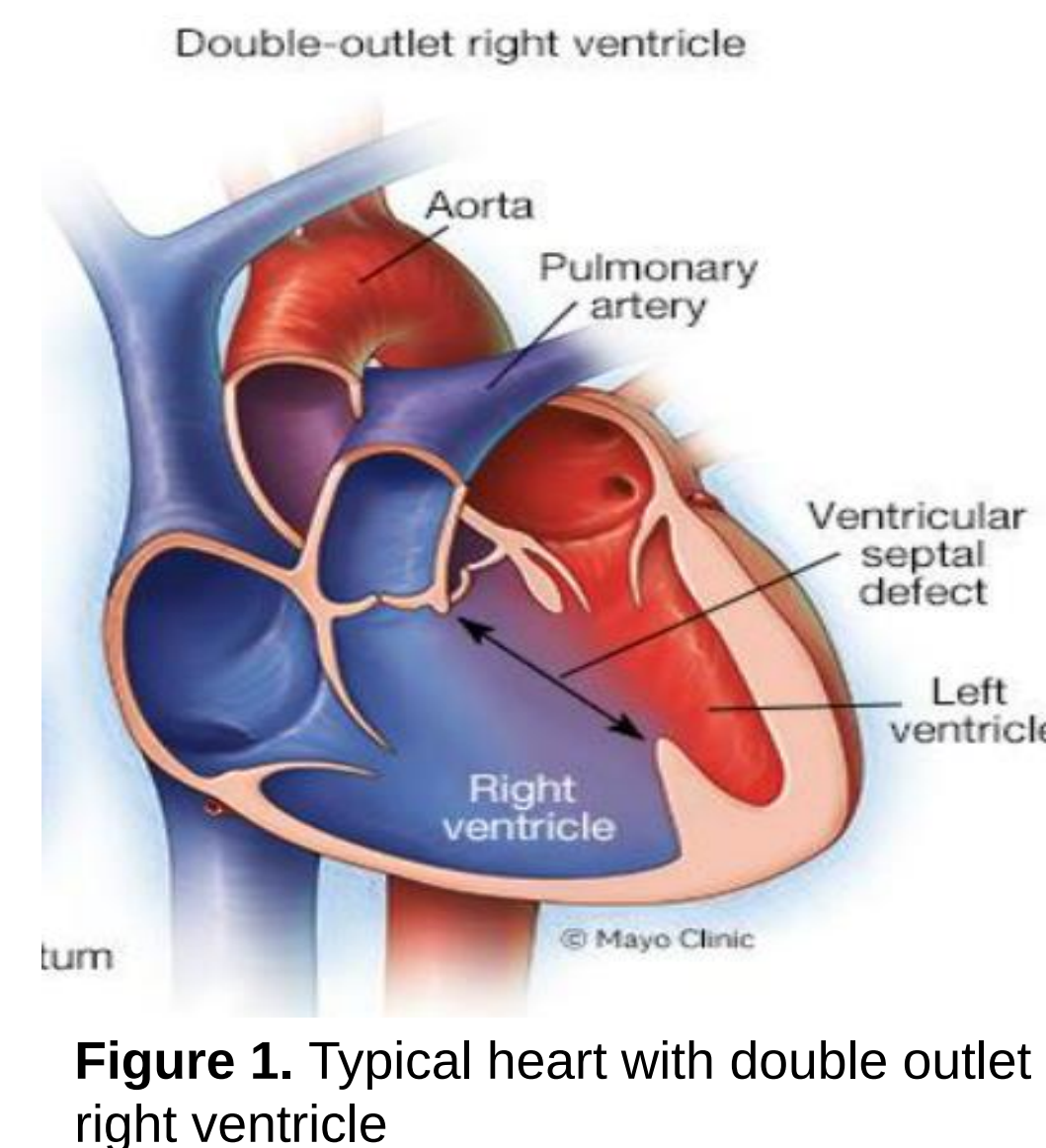


Figure 1. Typical heart with double outlet right ventricle

Procedural interventions:³

- Biventricular repair
- Single ventricle palliation
 - Glenn, Fontan

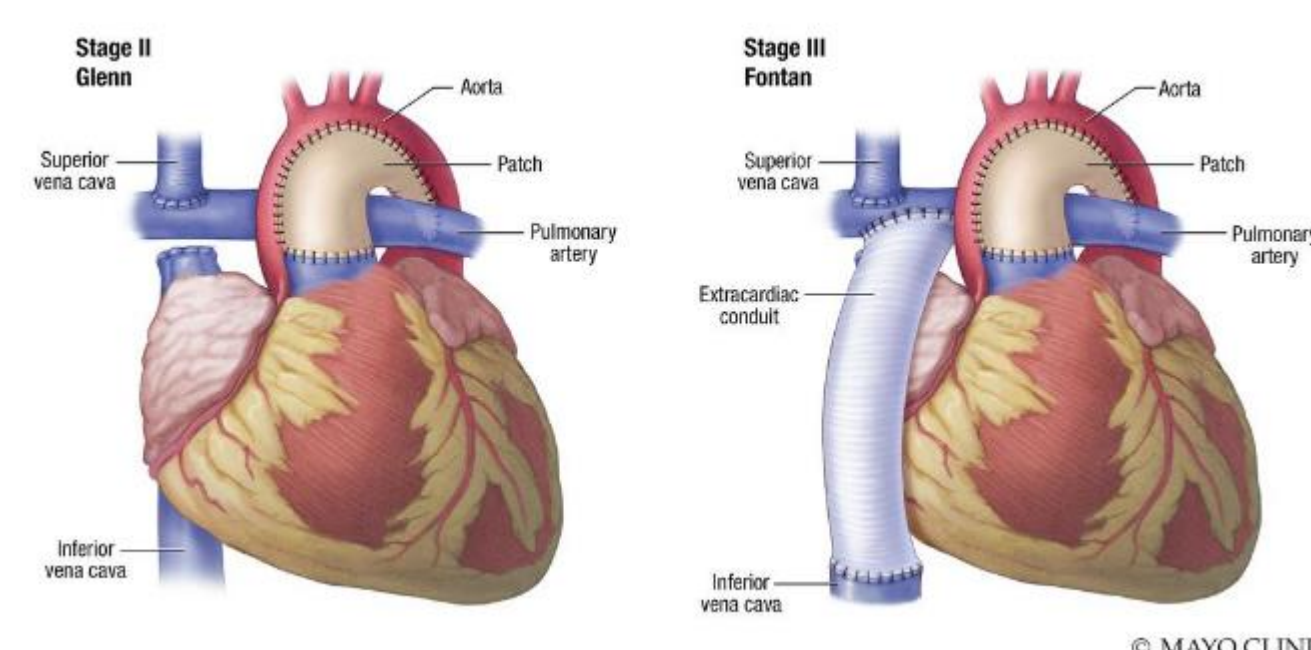


Figure 2. Staged palliation for DORV

Long term complications of single ventricle palliation:⁴

- Arrhythmia
- Protein-losing enteropathy
- Atrioventricular valve insufficiency
- Plastic bronchitis
- Conduit stenosis

CASE DESCRIPTION

37-year-old female underwent Fontan angiography and conduit stenting

Past Medical History:

- DORV, unbalanced AV canal defect
 - s/p bilateral bidirectional Glenn (1993)
 - s/p extracardiac Fontan procedure (1996) (Figure 3)
 - s/p mechanical atrioventricular valve (2005) on warfarin
- Multi-system manifestations: liver cirrhosis, right MCA stroke

Intraoperative course:

- Deep sedation (propofol and dexmedetomidine infusions), spontaneously breathing (SpO₂ 97%)
- Spontaneous pulmonary hemorrhage after endovascular stenting and balloon dilation of Fontan conduit

Challenges and management strategies:

Emergent intubation in airway hemorrhage and lung isolation in Fontan

Hypovolemia in single ventricle physiology

Treatment of bleeding in the presence of a mechanical AV valve

- Immediate single lumen endotracheal tube placement
- Bronchial blocker placed in right lower lobe, left deflated
- Bronchoscopy revealed right lower lobe hemorrhage
- Protamine administered (ACT 318 --> 151)

- 2 units pRBC, epinephrine 0.01 mcg/kg/min
- INR 1.2, Hb 15.9 (baseline 15.3), lactate 1.8

- Repeat bronchoscopy: evidence of clot formation
- Restarted heparin infusion
- Angiography demonstrated no contrast extravasation

Postoperative complications:

- ARDS secondary to pneumonia - initiation of VV ECMO (POD 4)
- Subarachnoid hemorrhage, no recurrent pulmonary hemorrhage



Figure 3. Extracardiac Fontan conduit seen on CT

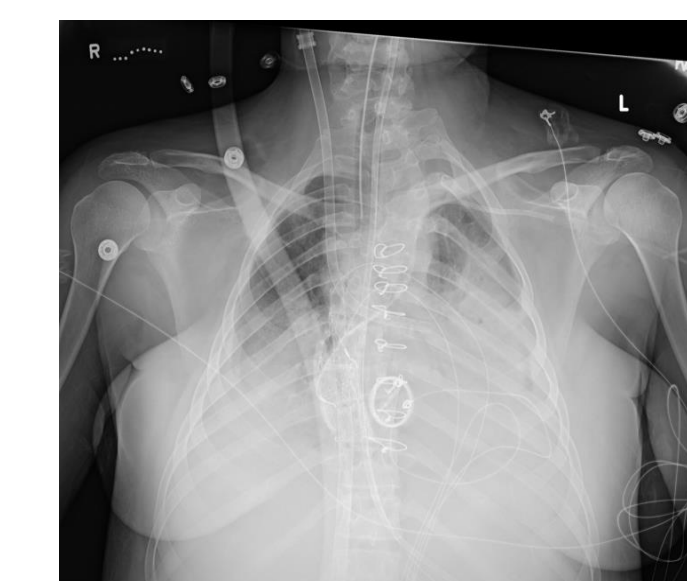


Figure 4. CXR demonstrating near complete opacification of both lungs

DISCUSSION

- Spontaneous respirations is most physiologically favorable due to maintaining negative intrathoracic pressure.
- Patients with Fontan circulation are more prone to pulmonary hemorrhage given increased pulmonary collaterals⁵ and the use of anticoagulants.
- Lung isolation techniques may be needed for lung protection but the resulting shunt can increase pulmonary vascular resistance.
- Acute hypovolemia and increased pulmonary vascular resistance can rapidly precipitate hypoxia, hypercarbia, decreased cardiac output, ischemia of the single ventricle and ultimately lead to cardiac arrest.⁶
- Resuscitation efforts should focus on inotropic support of single ventricle, preload maintenance, and PVR reduction.
- Early consideration of ECMO; limited vascular access, and prior repeated sternotomies make initiation of ECMO challenging.

REFERENCES

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