# Management of a Parturient with Unrepaired Tetralogy of Fallot

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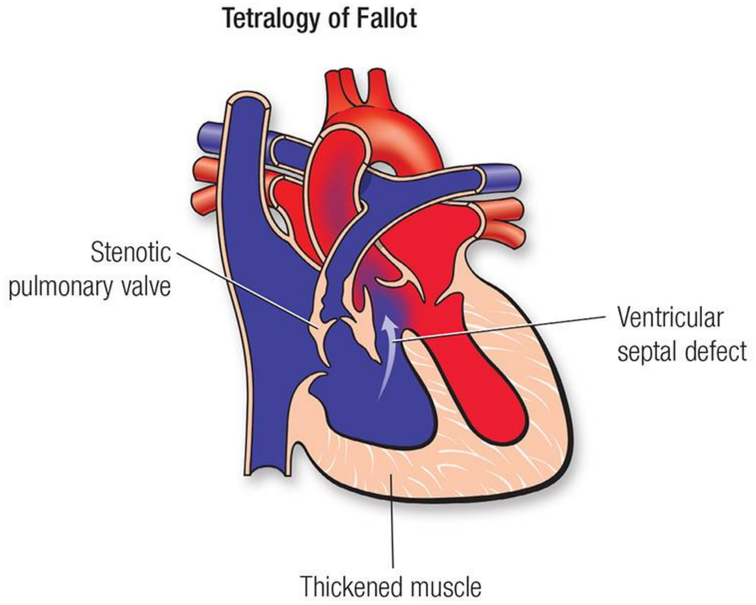
## Introduction

- Unrepaired Tetralogy of Fallot (ToF) is a rare congenital cardiac anomaly consisting of 4 heart problems
- While TOF is often diagnosed in utero or shortly after birth, depending on the severity of the disease, it may not be diagnosed until adulthood.
- Often patients with TOF will have “tet spells” which presents with cyanosis.
- The strain of pregnancy on the heart can lead to significant morbidity and mortality due to right ventricular dilation and failure, thromboembolism, and tachyarrythmias.
- While patients with repaired TOF generally can tolerate pregnancy, patients with unrepaired TOF, especially those who remain cyanotic, are considered high risk and pregnancy is generally not recommended.

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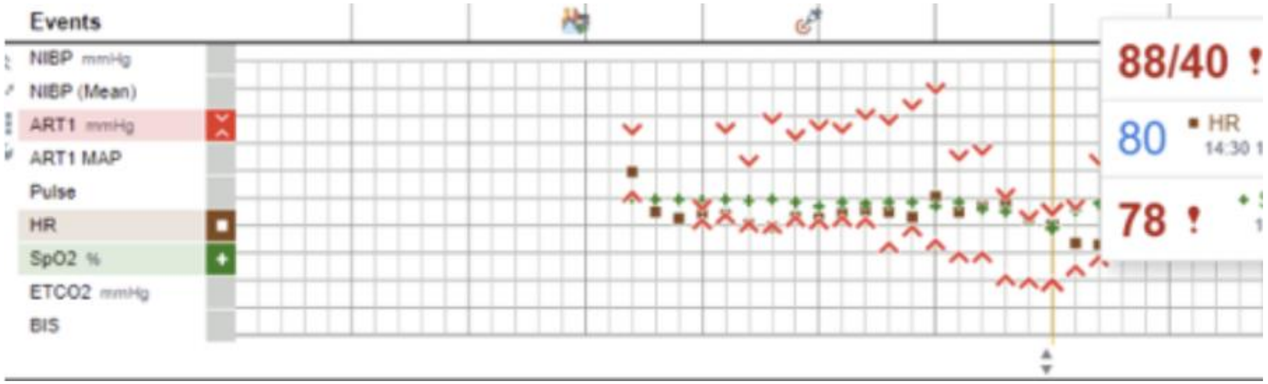
# The Case

- 26 y.o. G2P1 who presented at 20 weeks with a history of unrepaired ToF
- **Medical History:** Unrepaired TOF (never offered valve repair or replacement), 1 prior c/s in South America (37 weeks uncomplicated)
- **Consults:** Maternal Fetal Medicine (MFM), Cardiology, ICU, Anesthesiology

– TTE at 29 weeks: normal biventricular size and function, unrepaired ToF with secundum ASD with left to right shunt, large anterior malalignment VSD (predominantly left to right shunting), moderate-severe PV stenosis, and dynamic LV outflow tract obstruction.




- **Labor Course:** Admitted for scheduled cesarean delivery at 37w0d. Risks and benefits of cesarean section with neuraxial anesthesia with a backup for general anesthesia were explained

–Standard ASA monitors, arterial line, 2x 18g IV

–Epidural at L4/L5. Following a test dose with 3 mL lidocaine 2%, an additional 5 mL was given 5 minutes later, and a phenylephrine infusion was started at 0.2mcg/kg/min. Within 1 minute of injection, the parturient became hypotensive (MAP 50 nadir) and simultaneously became hypoxic (SpO2 78%). Patient was asymptomatic.

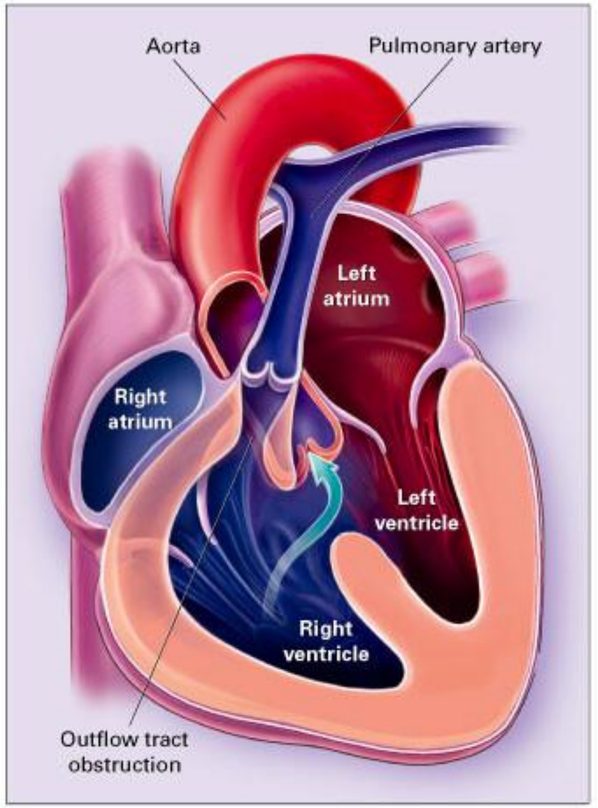
– 200mcg of phenylephrine was bolused, infusion increased to 1.0mcg/kg/min, supplemental oxygen provided. Within 2 minutes, MAPs and oxygen normalized.

– Epidural was slowly loaded with an additional 11mL of lidocaine 2%, and the case proceeded uneventfully

- **Postpartum Course:** Admitted to the ICU and discharged to home on post-operative day 3

# Learning Points

- Patients with unrepaired ToF are tenuous. Rapid changes in SVR can temporarily reverse right-to-left shunt leading to hypoxia.
- Epidural anesthesia is often considered a preferred option in parturients with congenital heart defects like TOF, as it allows for controlled pain relief while minimizing the risk of the hemodynamic instability associated with general anesthesia.
- Administration of phenylephrine restored the pressure differential by increasing SVR and slowing the heart rate, allowing for greater filling time and improved left ventricular preload, which restored the dominant left-to-right shunt.
- Interdisciplinary collaboration is critical in managing high-risk pregnancies.
- While the patient in this case was well-compensated and had a relatively favorable outcome, careful management remains crucial, as a deterioration in cardiac function or sudden obstetric complications can pose significant risks.
- With careful planning, early consultation with specialists, and prompt intervention in response to hemodynamic changes, it is possible to safely navigate labor and delivery.




Brickner ME, Hillis LD, Lange RA. Congenital heart disease in adults. First of two parts. *N Engl J Med*. 2000;342(4):256-263.

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