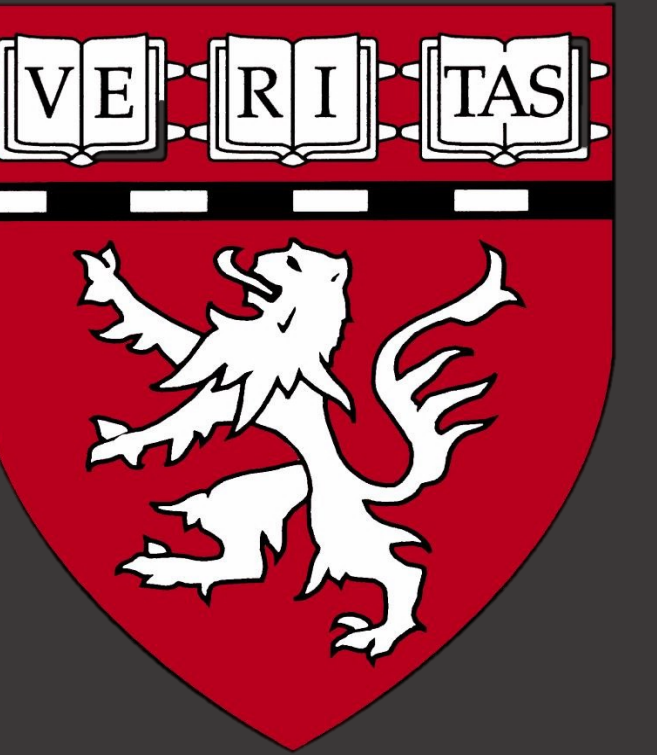




ANESTHESIA FOR CESAREAN DELIVERY IN A PATIENT WITH CONGENITALLY CORRECTED TRANSPOSITION OF THE GREAT ARTERIES

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INTRODUCTION

Congenitally corrected transposition of the great arteries (CCTGA) is a rare congenital heart disease characterized by atrioventricular and ventriculoarterial discordance. This double discordance results in an anatomic left ventricle supporting the pulmonary circulation, and anatomic right ventricle supporting the systemic circulation.

The pregnancy associated increase in circulating blood volume and cardiac output, as well as the stress of labor and delivery, may result in complications and requires specialized cardiology, obstetric, anesthesia and postoperative care. We report the perioperative planning for cesarian delivery of a patient with CCTGA.

CASE BACKGROUND

Our patient was a 31 year old G4P0030 with situs inversus totalis, ventricular inversion [I, D, D-MGV] double outlet right ventricle (RV) with associated subvalvular, valvular and right pulmonary artery (PA) stenosis, large inlet ventricular septal defect (VSD) and secundum atrial septal defect (ASD), chronic cyanosis and had two spontaneous abortions in that setting.

- At age 26 she underwent an ASD and VSD patch repair and pulmonary outflow tract reconstruction with left ventricle (LV) to pulmonary artery conduit (homograft) placement. Her course was complicated by postoperative complete heart block requiring pacemaker placement in her subpulmonary LV.

CASE BACKGROUND (continued)

- Over the next year, she had multiple hospital admissions for fluid overload and dysrhythmias in the setting of severe systemic tricuspid regurgitation and systemic RV dysfunction.
- She was medically optimized and counseled on the high maternal cardiovascular and fetal risks of pregnancy.
- After transitioning to a non-teratogenic heart failure regimen of digoxin, metoprolol and torsemide, she became pregnant.

ANATOMY

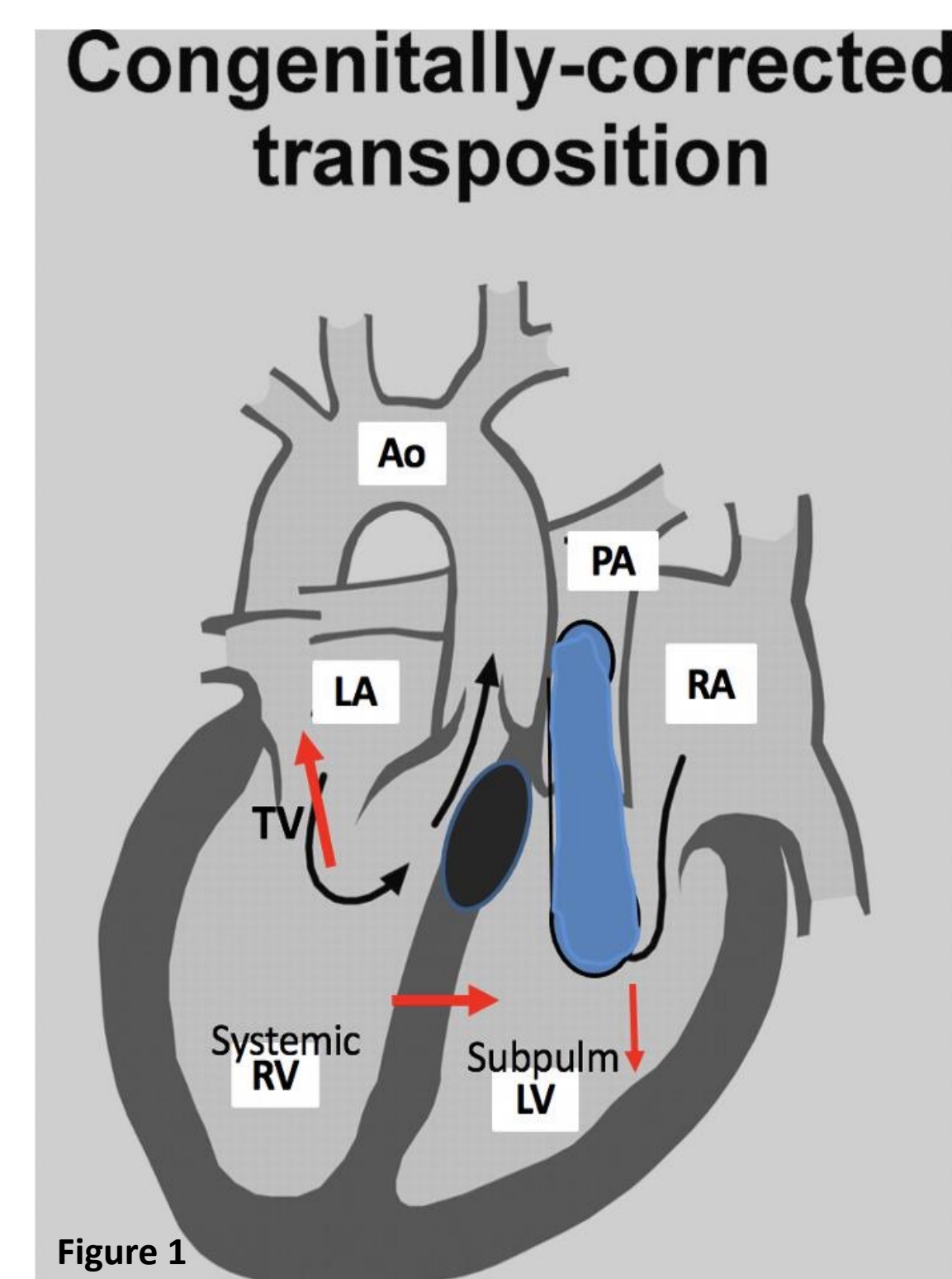


Figure 1. Our patient's anatomy including situs inversus totalis, congenitally corrected transposition (ventricular inversion), severe valvular pulmonary artery stenosis, ASD, VSD, s/p ASD and VSD patch repair and pulmonary outflow tract reconstruction with LV to pulmonary artery conduit. With relief of pulmonary stenosis post-operatively the septum shifted to the LV, tricuspid valve leaflets splayed, and tricuspid regurgitation increased.

PREOPERATIVE COURSE

At 27 weeks, she presented with volume overload and supraventricular tachycardia requiring diuresis and medication titration. Multidisciplinary planning with adult congenital cardiology, maternal fetal medicine, and obstetric and cardiac anesthesia teams occurred.

At 31 weeks, she was readmitted with worsening congestive heart failure.

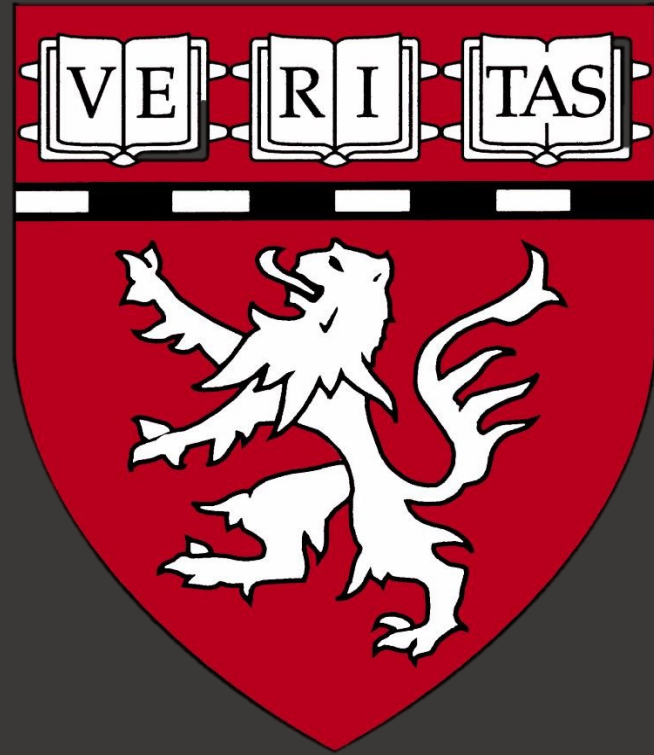
- Physical exam showed significant jugular venous distention, P2, S3 and S4 heart sounds, and orthopnea.
- TTE showed:
 - Dilated, hypertrophic RV with an ejection fraction of 47%
 - LV with moderate systolic dysfunction and pressures of 59mmHg
 - Severe tricuspid regurgitation
 - Mild mitral regurgitation
- IV bumetanide and metolazone was given for aggressive diuresis. The patient experienced frequent ectopy despite significant electrolyte repletion.
- Fetal position was transverse. Daily fetal non-stress tests were reassuring, and betamethasone was given for lung maturation.
- Out of concern for worsened fluid overload and thus cardiac function with ongoing pregnancy, a multi-disciplinary decision was made to proceed with delivery via c-section in a cardiac OR.



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TTE Imaging

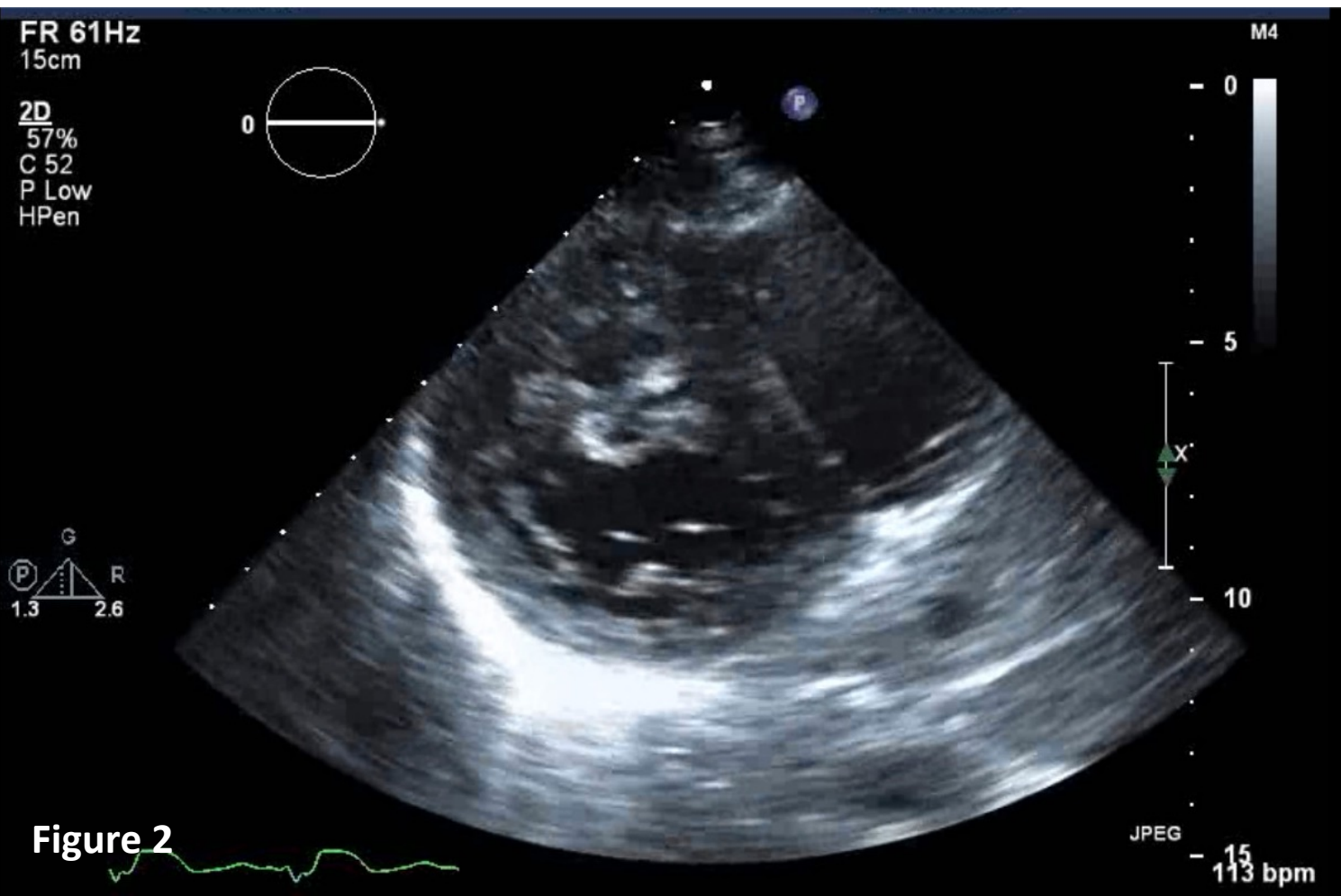


Figure 2. Short axis view of hypertrophied systemic RV and sub-pulmonic LV

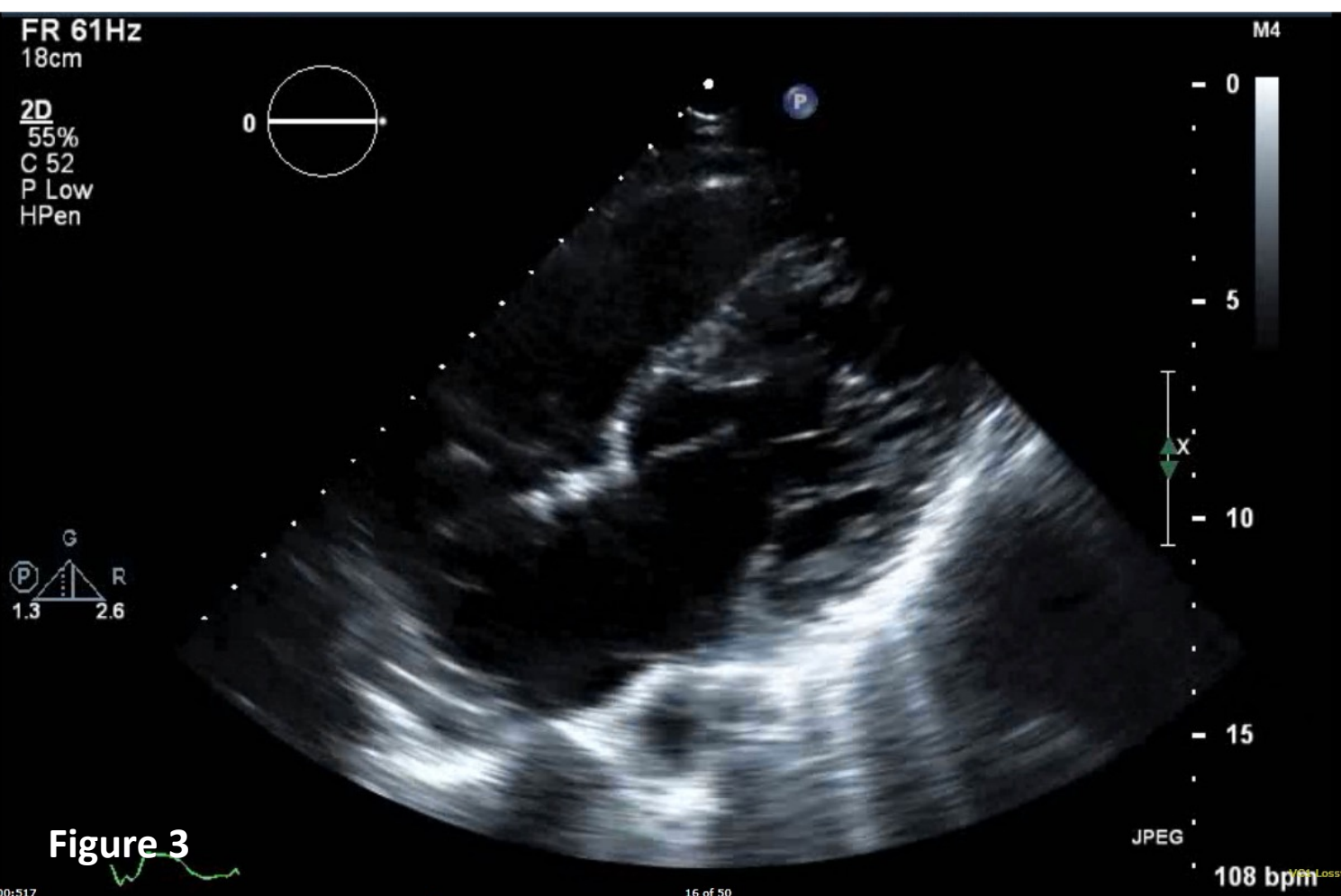


Figure 3. Subcostal view notable for hypertrophied systemic RV

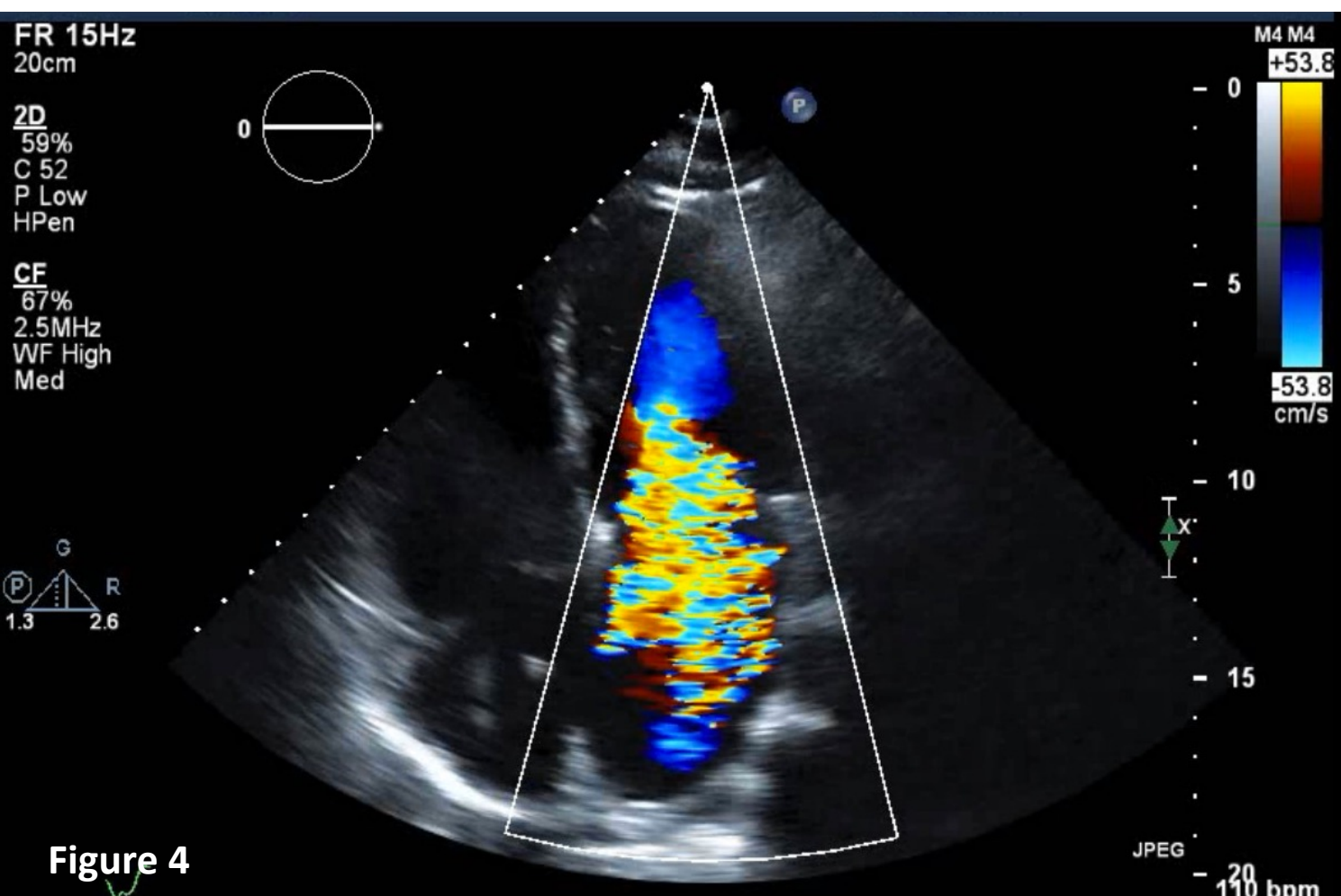


Figure 4. Apical four chamber view demonstrating severe systemic tricuspid regurgitation

INTRAOPERATIVE MANAGEMENT

- Her pacemaker was DDD-R, 100% V-pacing and left unchanged.
- Central and arterial lines were placed. TTE was available for hemodynamic management.
- Cardiac surgery and ECMO teams were on standby.
- Bumetanide infusion and electrolyte repletion was continued for ongoing ectopy, and milrinone was started on arrival to the OR.
- A combined spinal epidural was placed with 0.5mL intrathecal 0.75% hyperbaric bupivacaine, 150µg morphine and 15µg fentanyl.
- Once the patient was positioned with defibrillator pads in right uterine displacement with right precordial ECG leads, the epidural was dosed to a T4 sensory level with 2% lidocaine with epinephrine.
- At uterine incision, epinephrine was started for inotropic support.
- The neonate was delivered with Apgar scores of 8 and 9 and moved to the NICU for CPAP.
- A bumetanide bolus, IM oxytocin and nitroglycerin were given in anticipation of autotransfusion and increased vascular resistance.
- Uterine tone was strong and the epidural was removed at the end of the case.

POSTOPERATIVE COURSE

She was managed in the cardiac intensive care unit with continued IV diuresis and inotropic support that was weaned off over two days. She was medically optimized on lisinopril, metoprolol, digoxin, and torsemide as well as fluid and salt restriction.

She was transferred to the floor on postpartum day 4 and discharged on postpartum day 8.

CONCLUSION

Patients with CCTGA have complex congenital lesions that increase the risk of cardiac complications during pregnancy, labor, and delivery. As isolated lesions are rare, an individualized, interdisciplinary approach is crucial for safe delivery.

REFERENCES

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