Multidisciplinary Management and High-Risk Cesarean Delivery in Mosaic Turner Syndrome with Complex Aortic Pathology: A Case Report

Payton Gore, MD; Zikora Akah, BS; Daniel Conti, MD
UC Davis Anesthesiology and Pain Medicine

Introduction

Turner syndrome

- chromosomal abnormality (45,X)
- occurs in approximately 3.2 per 10,000 female live births.

It is commonly associated with:

- premature ovarian insufficiency with “streak” ovaries, or ovaries with no or few atresic follicles
- Short stature
- congenital cardiac abnormalities including bicuspid aortic valve and aortic disease
- Autoimmune conditions

Turner syndrome mosaicism

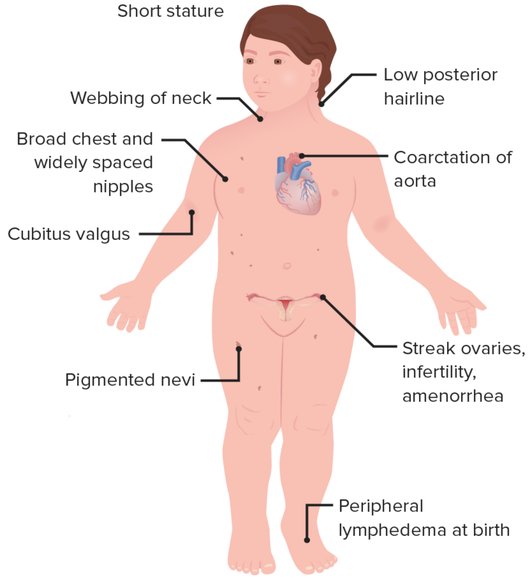
- Mosaic 45,X/46,XX genotype
- generally present with a milder phenotype with
  - fewer congenital defects, and
  - Preserved fertility
  - typically diagnosed later in life if and when complications arise.

Turner Syndrome and Fertility

- ~10% of individuals with Turner syndrome are capable of achieving spontaneous pregnancy and this is generally associated with the presence of mosaicism.

Turner Syndrome and Pregnancy

- The structural heart defects associated with Turner syndrome can lead to complications such as aortic dilation and dissection which can be further exacerbated during pregnancy due to the increase in cardiac output, blood volume, and vascular wall stress.



Case Presentation

A 28-year-old G4P2112 at 32w1d with a history of mosaic Turner syndrome and bicuspid aortic valve status post repaired type A and B aortic dissections with residual, chronic abdominal aneurysm presents with worsening shortness of breath. Vitals: HR 64-105, BP 80s/50s - 130s/70s. Physical examination notable for jugular venous distention and worsening peripheral edema.

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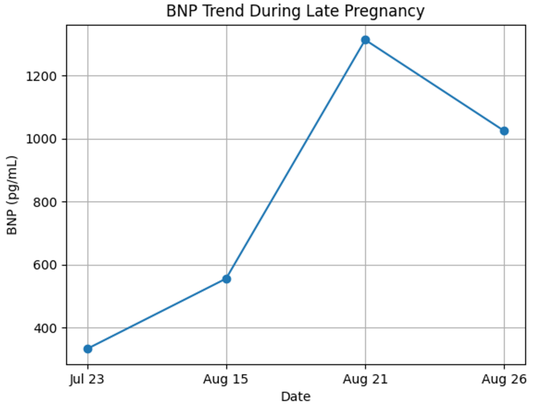
HPI

- 28-year-old G4P2112 at 31w2d with progressive dyspnea and chest pain beginning at 28w gestation
- Hx of bicuspid aortic valve that was discovered after an acute type A aortic dissection-- 3 years after delivery of second child (2x prior NSVD)
  - S/p emergent surgical repair with bioprosthetic aortic valve replacement and ascending aortic graft
  - C/b residual thoracic aorta dissection requiring TEVAR during admission
  - C/b dilated cardiomyopathy EF 25% which recovered after surgery to 65%
  - 7 months prior to the current admission, surveillance CT showed aneurysmal degeneration of the descending thoracic aorta and she had endovascular repair
  - CT prior to admission showed a chronic aortic dissection distal to the graft, extending to the level of the renal arteries

BNP	Gestation
333	27+2
556	29+4
1313	30+3
1024	31+1



Objective

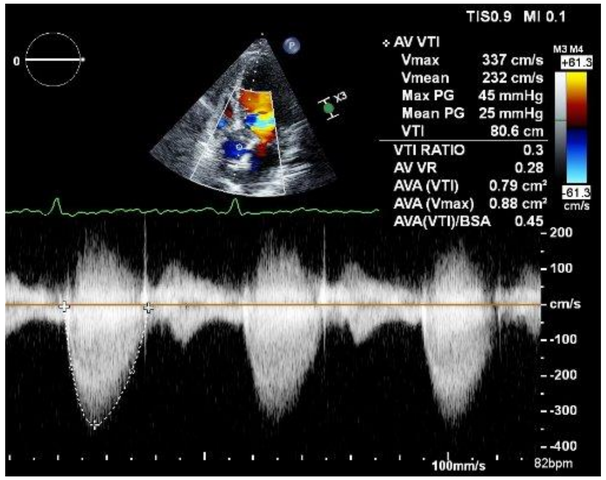
- Troponin wnl, EKG normal sinus rhythm, CXR with no pulmonary edema
- repeat TTE @ 31w2d showed preserved EF 70%, well seated bioprosthetic valve, no regurg or stenosis, normal RV
- MRA chest/abdomen on August 28 showed stable 2.2 cm dissection from T11-L1

Predelivery Management

Multidisciplinary care team engaged early:
- cardiology, MFM, vascular surgery, CV surgery, OB and cardiac anesthesia

Diuresis
- furosemide
- no improvement in dyspnea and chest pain
- only small decrease in BNP

Delivery:
- Overall clinical picture suggested increased physiologic strain
- proceeded with scheduled CS and bl salpingectomy at 32w1d for risk reduction



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Intraoperative and Postpartum Course

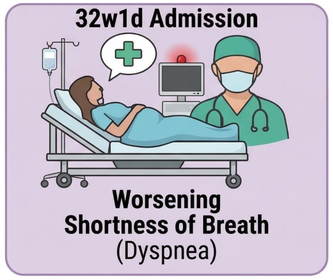
Anesthetic goals:	• Minimize aortic shear stress • Support cardiac output • Minimize volume overload
Preoperative arterial line placed	• Strict BP and HR control, SBP<=120 • Phenylephrine and clevidipine in line
Modified combined spinal epidural	• reduced intrathecal dose • 1.2 ml hyperbaric bupivacaine + IT morphine and fentanyl
Uterotonics:	• Avoid methylergonovine and carboprost • no additional uterotonics were required

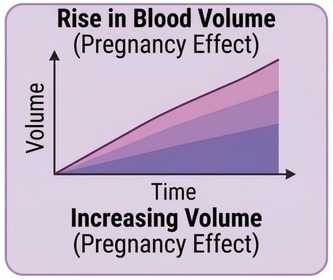
Uncomplicated low transverse cesarean delivery,

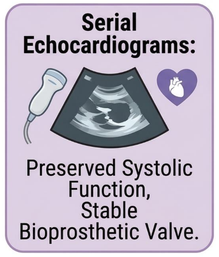
- transferred to ICU for close monitoring x24 hours
- discharged on POD 2

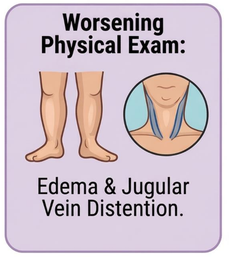
At cardiology follow up:

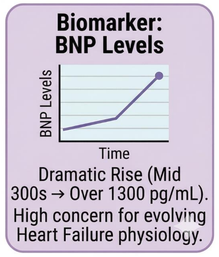
- sustained symptomatic improvement
- had not required diuretic therapy since discharge











Discussion

Pregnancy in patients with mosaic Turner syndrome confers substantial cardiovascular risk, particularly in the third trimester

Cardiac risk scoring models (mWHO, ZHARA, CARPREG) have focused on anatomic lesions, in this case clinical decision making was guided by functional capacity

Recent literature supports a possible role for NYHA class in cardiac risk modeling.

Importance of early anesthesia consultation, multidisciplinary coordination, and an individualized anesthetic

Consider sending young and otherwise healthy female patients who present with aortic dissection for genetic testing