Coexisting Cardiac Disease in Pregnancy

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DISCLOSURE
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Scope of the problem

- Confidential Enquiries into Maternal Deaths in the UK, 2006-2008
- 2.29 million births, 261 deaths, 154 indirect deaths
  - 53 deaths due to cardiac disease (2.31/100,000 births, a new high)
    - 6 myocardial infarction
    - 7 aortic dissection
    - 9 peripartum cardiomyopathy
    - 10 “sudden arrhythmic death syndrome”-obesity and cardiomyopathy without hypertension?
- An additional 6 late deaths due to peripartum cardiomyopathy
Figure 10.1 Maternal mortality rates from cardiac causes of death; England and Wales 1952–84, United Kingdom 1985–2002
UK and Ireland Confidential Enquiries into Maternal Deaths and Morbidity

• 2009-2011 triennium
  • 2.4 million maternities
  • 51 deaths due to cardiac disease
  • 2.14 per 100,000 births

https://www.npeu.ox.ac.uk/mbrrace-uk
Outline

• Epidemiology of cardiac disease in pregnancy
• Signs and symptoms of cardiac disease in normal pregnancy
• Physiologic changes of pregnancy
• Congenital cardiac disease
  • VSD
    • Eisenmenger’s complex
  • Marfan Syndrome
• Acquired cardiac disease
  • Rheumatic valvular: MS
  • Idiopathic
    • Peripartum cardiomyopathy
    • Pulmonary hypertension
  • Surgically corrected cardiac disease
• Logistics of caring for the pregnant patient with cardiac disease
Epidemiology

• Incidence: Decreased from 3.6% to 1.6% since 1970’s
  • Prevention of rheumatic valvular disease
  • Increased survival in patients with congenital disease
  • Rheumatic:congenital 25:1 ⇒ 3:1

• Valvular: MS 90%, MR 6%, aortic <5%

Mangano DT. In Shnider and Levinson’s Anesthesia for Obstetrics (4th ed.), 2002
Epidemiology

- Increasing incidence of CAD
  - Delayed childbearing
  - Smoking
  - Increased prevalence in the general population
  - Cocaine abuse
Signs and symptoms of cardiac disease in the healthy parturient

- Decreased exercise tolerance, DOE, PND
- Near-syncope (aortocaval compression)
- Jugular venous distention
- Peripheral edema
- Bibasilar rales
- Systolic murmurs

Signs and symptoms of cardiac disease in the healthy parturient

- EKG abnormalities:
  - Arrhythmias: SVT, PACs, PVCs
  - ST-T depression
  - Q waves in III
  - Left and right axis deviation

Maternal physiologic changes

• Cardiovascular
  • Blood volume
    • ↑15% by 12 weeks
    • ↑50% by end of 2nd trimester
    • Minimal increase during third trimester

Bernstein IM. Obstet Gynecol 2001; 97:669
Maternal physiologic changes

• Cardiovascular
  • Systemic vascular resistance ↓20% at term
    • Prostacyclin (PGI₂)
    • Progesterone
    • Dilutional anemia ⇒ improved rheology
    • Low resistance circuit in parallel
Maternal physiologic changes

- Sum of parallel resistances:
  \[
  \frac{1}{R_1} + \frac{1}{R_2} = \frac{1}{R_T}
  \]
  - \(R_1=100, \ R_2=100, \ R_T=50\)
  - \(R_1=100, \ R_2=50, \ R_T=33.3\)
Maternal physiologic changes

• Cardiovascular
  • Cardiac output
    • ↑35-40% at end of 1st trimester
    • Initial change due to increased HR, followed by increased stroke volume
    • Intrinsic contractility (LVSWI) unchanged
    • ↑50% at term

Capeless EL. Am J Obstet Gynecol 1989; 161:1449
Maternal physiologic changes

• Intrapartum changes in cardiac output
  • ↑60%-sympathetic stimulation-modified by analgesia
  • Additional increase-autotransfusion
  • Postpartum-uterine involution
    • Implication for elective cesarean in cardiac patients
Maternal physiologic changes

• Respiratory
  • Oxygen consumption
    • ↑60%-fetus, placenta, uterus
    

  • Oxygen supply
    • FRC ↓20%

  • Implication: apnea ⇒ rapid desaturation
Maternal physiologic changes

- Gastrointestinal
  - LES incompetence, ↑intragastric pressure (20 weeks)
  - ↓gastric emptying in labor
    - Pregnancy per se: no effect
  - Acidity and volume of gastric secretions appear to be unchanged despite elaboration of ectopic gastrin by the placenta

These changes imply an increased risk of aspiration pneumonitis and thus the need for rapid sequence induction
Physiologic consequences of anesthesia

• General anesthesia
  • Propofol: myocardial depression, vasodilatation
  • Laryngoscopy and intubation:
    • Tachycardia
    • Systemic and pulmonary hypertension
    • Myocardial ischemia, LV failure
• Narcotic technique?
  • Maternal aspiration
  • Newborn depression
Physiologic consequences of anesthesia

- Regional anesthesia
  - Venous dilatation $\Rightarrow$ ↓ venous return $\Rightarrow$ ↓CO
  - Dilatation of arteriolar bed $\Rightarrow$ ↓SVR $\Rightarrow$ shunt reversal
  - SAB > LEA
  - Dilute epidural LA: minimize, do not completely eliminate sympathetic block
  - IT opioids and sympathectomy
Elective C/S vs. “stress of labor”

- Postpartum hemodynamic changes (uterine involution) equivalent in both groups
- Fluid shifts greater in major abdominal surgery
- Regional block necessary for C/S ⇒ profound sympathetic blockade
Ventricular septal defect

• Most common congenital cardiac defect
• Often little hemodynamic disturbance
• In most severe form, causes hemodynamic alterations that contraindicate pregnancy
VSD-pathophysiology

- Small defect
  - RV, PA pressures unchanged
  - Minimal increase in pulmonary blood flow
  - Jet effect-endocarditis risk
    - Prophylaxis not indicated for NSVD

Van Mook WNKA. Curr Opin Crit Care 2005; 11:435
VSD-pathophysiology

• Moderate defect:
  • Pulmonary blood flow increased
  • RV, PA pressures below systemic levels
  • LV volume overload $\Rightarrow$ CHF
  • Vascular disease unlikely
VSD-pathophysiology

• Large defect
  • No resistance to flow across defect
  • RV and LV pressures equalize
  • Pulmonary vascular disease
VSD-pregnancy outcome

• Well tolerated in asymptomatic women
• CHF and arrhythmias with moderately sized defects
• >10% incidence of VSD in offspring of affected mothers (and >4% in offspring of affected fathers)

Uebing A. *BMJ* **2006**; 332:401
VSD-anesthetic management

• Small defects
  • No specific alterations
  • Invasive monitoring not required
  • SBE prophylaxis: operative vaginal delivery, C/S

• Moderate defects
  • Untreated pain ⇒ ↑SVR ⇒ ↑L to R shunt, CHF
  • CVP (not PA) monitoring
  • Epidural anesthesia recommended for labor, C/S
    • Potential for shunt reversal?
VSD-anesthetic management

- Moderate defects
  - GA: prevent increases in SVR and HR that might increase shunt
  - Avoid myocardial depressants in setting of CHF
Eisenmenger’s complex

• “Pulmonary hypertension at systemic level, due to high pulmonary vascular resistance, with reversed or bi-directional shunt through a large VSD”
  

• Exposure of pulmonary vasculature to high pressures
  • Initial pulmonary vasoconstriction (reversible)
  • Eventual irreversible obliterative changes and fixed increases in PVR
Eisenmenger’s complex

- As PVR increases, RV pressure increases $\Rightarrow$ shunt reversal
- Direction, magnitude of shunt depends on SVR:PVR ratio
Eisenmenger’s complex

• Widely considered an absolute contraindication to pregnancy, with reported mortality 25-50%
  
  
  
  Van Mook WNKA. *Curr Opin Crit Care* **2005**; **11**:430

• 30 pregnancies in 10 women (total 35)
  • 13 TOP, 4 SAB, 1 IUFD at 23 weeks
  • 12 live births
    • 10 preterm, 2 term
  • 9/10 patients alive at long term follow up despite significant functional deterioration

Eisenmenger’s-anesthetic management

- Maintain right sided filling pressures
- Maintain adequate SVR
- Avoid factors increasing PVR

Hypercarbia
Hypoxemia
Hypothermia

Acidosis
High airway pressures
Eisenmenger’s-anesthetic management

• Invasive monitoring
  • CVP adequate measure of RV preload
  • Pulmonary artery catheterization
    • PA rupture
    • Initiation of poorly tolerated arrhythmias
    • Little additional information vs. CVP
Eisenmenger’s-anesthetic management

- Labor
  - Epidural analgesia with dilute LA
  - Increase SVR if $O_2$ saturation decreases
    - Pulse oximetry indirectly monitors shunt
  - Intrathecal opioids

- Cesarean section
  - Use of LEA has been described
  - Inhalation agent can adversely effect shunt
  - High dose narcotics can maintain hemodynamics *but*
    - Neonatal depression
    - Aspiration
Eisenmenger’s—anesthetic management

- Intensive postoperative surveillance
  - Most common time of death is immediate postpartum period (small PE)
Marfan syndrome

- Autosomal dominant disorder of fibrillin formation (chromosome 15)
- Family history present in 65-75%
- Prevalence 4-6/10,000, unrelated to sex, race, or ethnicity
- Mean life expectancy 32 years
  - Aortic dissection, aortic rupture, cardiac failure cause of death in >90% of patients

Marfan syndrome

- Clinical manifestations
  - Ocular
    - Subluxation of the lens
  - Musculoskeletal
    - Dolichostenomelia and arachnodactyly
    - Pigeon chest
    - Scoliosis
  - Cardiovascular
    - Weakness of the aortic media-root dilatation, dissection
    - Aortic insufficiency
    - Mitral valve prolapse
Marfan syndrome and pregnancy

- 50% of children will be affected
- 1% risk of fatal complication, rising with increasing aortic root diameter
  

- Pregnancy does not worsen long-term outcome in the absence of risks for dissection
  

- Most dissections occur during the third trimester
  
Marfan syndrome and pregnancy

- In patients with pre-pregnancy aortic root diameter < 42 mm, outcome almost universally favorable
  

- Obstetric complications: increased risk of spontaneous abortion, PTL, PPH, cervical incompetence, placenta previa
Medical management

• Beta blockade
• Serial echocardiography
  • Will determine mode of delivery
• Early initiation of epidural analgesia
  • Be aware of increased incidence of scoliosis
• Shortened second stage
• Elective cesarean section:
  • Aortic root diameter > 40 mm
  • Progressive dilatation during pregnancy

Lipscomb KJ. *Br J Obstet Gynaecol* 1997; 104:201
Pathophysiology of mitral stenosis

- Proximal to valve
  - Compliant left atrium progressively dilates due to incomplete emptying
    - Onset of atrial arrhythmias
    - Formation of atrial clot
  - Increased pulmonary venous pressure (↓LA compliance)
    - Fluid transudation and pulmonary symptoms
    - Fixed increases in PVR and RV failure
Pathophysiology of mitral stenosis

• Distal to valve
  • Chronic LV underloading
  • Decreased wall thickness
  • By LaPlace’s law, wall tension=$P\times R/2h$, wall tension is increased.
  • Afterload sensitive LVEF impaired
Interaction of pregnancy with mitral stenosis

- Increased blood volume predisposes to pulmonary edema
  - Common presentation at end of second trimester
- Limited ability to increase cardiac output due to impaired LV filling
  - Cannot increase CO to compensate for decreased BP secondary to vasodilatation
- Normal ↑ in HR poorly tolerated
  - Shortened diastolic time further impairs LA emptying causing pulmonary edema
• Sudden increase in preload after delivery can produce pulmonary edema
• Hypovolemia (e.g., hemorrhage) poorly tolerated
Maternal outcome in mitral stenosis

- Maternal cardiac complications seen in 35% of pregnancies
  - 67% in severe
  - 38% in moderate
  - 26% in mild

  Silversides CK. *Am J Cardiol* 2003; 91;1382

- Maternal death has become quite uncommon
  - 0/195 cases

  Harnett M, Tsen LC. Cardiovascular Disease. In Chestnut’s Obstetric Anesthesia, 4th Ed.
Anesthetic goals

• Avoid tachycardia
• Maintain SVR
• Avoid ↑PVR
• Invasive monitoring
Epidural analgesia for labor

- Avoid epinephrine test dose
- Minimize sympathetic blockade
- Phenylephrine for hypotension
- Analgesia during second stage (prevent Valsalva)
Anesthetic management C/S

- Cesarean section: obstetric indications only
- Epidural anesthesia
  - Slow titration with 0.5% bupivacaine, 0.75% ropivacaine
- Spinal anesthesia
  - Rapid onset of sympathetic block compared to LEA
  - Continuous spinal?
General anesthesia

- Induction: avoid sympathetic stimulation, myocardial depression
  - Opioid, etomidate, non-depolarizing relaxant
- Treat atrial fibrillation aggressively
  - Cardioversion, verapamil, digoxin
- Avoid drugs producing tachycardia
  - Atropine, ketamine, meperidine
- Hypotension due to tachycardia: β-blockade
Treatment of decompensated MS

- Valve replacement under CPB
- Open commisurotomy
- Closed commisurotomy
- Balloon valvuloplasty
Balloon valvuloplasty

• 19 patients NYHA III-IV
• Mean EGA 30 weeks
• Mean valve area $0.83 \text{ cm}^2 \Rightarrow 2.4 \text{ cm}^2$
• All completed pregnancy in an uncomplicated fashion
• All NYHA class I at one year

Patel JJ. Am Heart J 1993; 125:1106
Balloon valvuloplasty

- 21 valvuloplasty
- 24 open commissurotomy
- Similar improvement in symptoms
- Fetal mortality
  - Commisurotomy 8
  - Valvuloplasty 1

De Souza JA. J Am Coll Cardiol 2001; 37:900
Balloon valvuloplasty

- 36 women, mean age 25.8
- 25 NYHA II, 11 NYHA III
- Successful in 35/36 patients (improved by >1 functional class, 0% mortality)
- Mean valve area $0.74 \rightarrow 1.59 \text{ cm}^2$
- All had uncomplicated deliveries
- All remained NYHA I-II at 33 month followup

Sivadasanpillai H. *Am J Cardiol* 2005; 95:1504
Peripartum cardiomyopathy (PPCM)

• Diagnostic criteria
  • Cardiac failure in the last month or within six months of delivery
  • Absence of a demonstrable cause for cardiac failure
  • Impaired LV systolic function by echocardiogram
Peripartum cardiomyopathy

• Exclusion criteria
  • Infectious, toxic, or metabolic disorders
  • Ischemic or valvular disease
  • Vascular disease (aortic dissection)
  • Complications of pregnancy
    • Amniotic fluid embolism
    • Pulmonary embolism
    • Preeclampsia
Peripartum cardiomyopathy

- Incidence 1:15,000 (US), 1:1000 (South Africa), 1:400 (Haiti)
  
  Fett JD. *Am J Obstet Gynecol* 2002; 186:1005

- Reported risk factors:
  
  Maternal age > 30  
  African-American descent  
  Poor nutrition  
  Multiple gestation  
  History of PIH  
  Enterovirus infection  
  Selenium deficiency  
  Guillain-Barré syndrome
Peripartum cardiomyopathy

- Evidence for a unique disorder:
  - Age distribution
    - Idiopathic: middle age
    - PPCM: disease of the young
  - Relation to EGA
    - Pre-existing cardiomyopathy: second trimester (peak C.O.)
    - PPCM: Late gestation or postpartum
Peripartum cardiomyopathy

- Evidence for a unique disorder
  - Postpartum resolution
    - Pre-existing cardiomyopathy: extremely uncommon
    - PPCM: common
  - Recurrence with subsequent pregnancy
    - Unique to PPCM

Lang RM. Peripartal cardiomyopathy.
In Cardiac Problems in Pregnancy (3rd ed), 1998
Peripartum cardiomyopathy

- Distinct disorder, distinct etiology?
  - Nutritional
    - Gross malnutrition, specific deficiency never identified
  - Pre-eclampsia
    - Unlikely--transient LV dysfunction, resolves within days
  - Immunologic
    - More common in multiparas and twin gestations
Peripartum cardiomyopathy

- Clinical presentation
  - Orthopnea, DOE, PND
  - Peripheral edema, JVD, gallop rhythm
  - Pulmonary, systemic thromboembolism
  - Echo: dilated, hypokinetic ventricle
Peripartum cardiomyopathy

- **Treatment**
  - Sodium restriction
  - Digoxin
  - Anticoagulation (mural thrombus)
  - Normal activity level as tolerated
Peripartum cardiomyopathy

• Treatment
  • Afterload reduction
    • Hydralazine ante-partum
    • ACE inhibitors post-partum
  • Cardiac transplantation
    • Patients that undergo transplantation tolerate subsequent pregnancies well

Miniero R. J Heart Lung Transplant 2004; 23:898
Peripartum cardiomyopathy—natural history

• It has been suggested that future pregnancy is contraindicated only if LV function fails to return to normal after the index pregnancy.
• Even with apparent return of normal contractility, outcome may be worse than was once thought.
Peripartum cardiomyopathy-natural history

• 44 women with subsequent pregnancies
  • 28 normal function (Group 1)
  • 16 persistent LV dysfunction (Group 2)

• Heart failure
  • Group 1: 21%
  • Group 2: 44%

• Mortality
  • Group 1: 0%
  • Group 2: 19%

Peripartum cardiomyopathy-natural history

- Subclinical diminished cardiac reserve
  - Dobutamine stress echocardiography as screening tool?

Anesthetic management

- Invasive monitoring
- Suspend anticoagulation
- Epidural analgesia for labor
- Epidural anesthesia for cesarean section
- General anesthesia:
  - Avoid myocardial depressants
  - Blunt sympathetic response to noxious stimuli
Idiopathic pulmonary arterial hypertension

- Mean pulmonary artery pressure of >25 mm Hg at rest, or more than 30 mm Hg during exercise
- Diagnosis of exclusion:
  - Left sided valvular disease
  - Myocardial disease
  - Congenital heart disease
  - Respiratory disease
  - Connective tissue disorder
  - Thromboembolic disease
Idiopathic pulmonary arterial hypertension

• Pathophysiology
  • Vasoconstriction
  • Smooth-muscle cell and endothelial-cell proliferation
  • Thrombosis
• Imbalance of vascular effectors
  • Prostacyclin/thromboxane imbalance
  • ↑endothelin-1
  • ↓NO synthase
  • Elevated serotonin

Farber HW. *New Engl J Med* 2004; 351:1655
Idiopathic pulmonary arterial hypertension

- Demographics
  - 63% of patients in NIH registry were female
  - Mean age 36 years
- Usual presentation late in disease, with death occurring within a few months to several years
Idiopathic pulmonary arterial hypertension

• Presentation:
  • Early disease: exertional dyspnea, fatigue, chest pain, palpitations, syncope
  • Late disease: dyspnea at rest, hemoptysis, hoarseness (compression of recurrent nerve by dilated PA)
Idiopathic pulmonary arterial hypertension

- Laboratory findings
  - Polycythemia, hypercoagulability
  - ↓total lung capacity, FVC
  - Right axis deviation
  - CXR: Cardiomegaly, dilated pulmonary trunk, decreased pulmonary vascular markings
  - Echocardiogram: enlarged, hypertrophied RV, small LV
Idiopathic pulmonary arterial hypertension

• Relation to pregnancy
  • Maternal mortality 40%, usually within a few hours to several days after delivery
  • Outcome cannot be predicted on the basis of pre-pregnancy maternal condition
  • IUGR common secondary to low fixed cardiac output
Idiopathic pulmonary arterial hypertension

• Patients responding to a trial of nitric oxide, IV adenosine, or IV PGE$_2$ may have a sustained response to oral calcium channel blockers

• Continuous infusions of prostacyclin and PGE$_2$
Idiopathic pulmonary arterial hypertension

- Intrapartum management
  - Central monitoring (PA catheter vs. CVP)
  - Frequent blood gases
  - Mode of delivery determined by obstetric indications, clinical status of mother during labor
  - “It is important to provide effective analgesia for labor and delivery”

Idiopathic pulmonary arterial hypertension

• Principles of anesthetic management
  • Avoid ↑PVR
  • Avoid ↓RV filling pressure
  • Avoid ↓SVR
    • Unable to increase cardiac output to compensate for hypotension
    • RV very sensitive to perfusion pressure (ischemia)
  • Avoid negative inotropes
Idiopathic pulmonary arterial hypertension

- Epidural analgesia for labor
  - Meticulous attention to maintaining filling pressures and C.O.
- Intrathecal opioids (single shot or continuous catheter techniques)
- Cesarean section:
  - GA usually recommended
  - Use of CSEA has been described with good results

Bonnin M. *Anesthesiology* 2005; 102:1133
Surgically corrected cardiac disease

- Is the patient anticoagulated?
- Is antibiotic prophylaxis indicated?
  - Bacteremia unlikely with NSVD, but….
- Is contractility impaired?
  - Residual effect of corrected lesion (AI)
  - Consequence of surgical approach (TOF repair)
- Is repair associated with high incidence of arrhythmias?
  - Senning interatrial baffle for TGV
Surgically corrected cardiac disease

- Is a residual structural defect present? (mitral stenosis after annuloplasty, infundibular obstruction after TOF repair)
- Are there persistent structural changes in the pulmonary circulation?
- What is the anatomy of the repair?
  - Fontan procedure-passive filling of PA and need to maintain preload
Logistics of care

• Identification of the patient with significant cardiac disease by obstetrician
• Consultation with primary cardiologist, evaluation of baseline function
• Anesthesiology consultation at 20-24 weeks
• Consensus of caregivers
• Dissemination of anesthetic/obstetric plan
Logistics of care

- The myth of the “prophylactic” cesarean
- Planned vaginal delivery
  - Should induction be scheduled?
  - Where will labor, delivery, and recovery occur?
    - Are ICU beds available?
    - Can L&D care for critically ill patients with invasive monitors?