Adult Congenital Heart Disease

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ACHD Outline

- Anatomy
  - Common lesions, Eisenmenger's
  - Surgical shunts
- Physiology
  - PVR
  - SVR
- Perioperative risk factors
- Anesthesia
  - Techniques
  - Drugs
- Specific concerns
  - Hyperviscosity
  - Antibiotics

Case 1

Q1

42 y.o. female, uncorrected ASD, with ovarian cancer for elective laparoscopic oopherectomy.

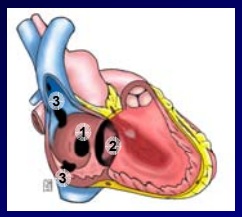
Which statement is true?

A. She is most likely to have an ASD primum
B. She has normal life expectancy
C. She requires referral to a tertiary care hospital
D. She may become cyanotic intraoperatively
E. She requires an open procedure

Atrial Septal Defects

- 1/1500 live births

1. Secundum - 75%
2. Primum - 15%
3. Sinus Venosus -10%
4. Coronary sinus - rare



Asymptomatic: murmur, abnormal ECG/CXR
Symptomatic: dyspnea/CHF, CVA/emboli, A Fib

Case 1

Q2

Which is not an independent risk factor for perioperative morbidity in this patient with an uncorrected ASD?

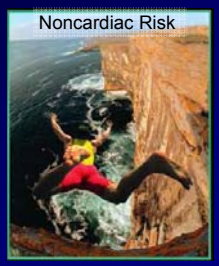
A. Pulmonary hypertension
B. Arrhythmias
C. Cyanosis
D. Ventricular function
E. ASD size

CHD Perioperative Morbidity

Cardiac Surgery
Independent Risk Factors

- Pulmonary hypertension
- Arrhythmias
- Cyanosis
- Ventricular function
- Reoperation

Noncardiac Risk



Warnes. J Thorac Cardiovasc Surg 2003; 126:1048-52

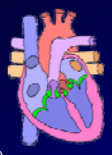
CHD Guidelines

- ACC/AHA
  - http://www.americanheart
- Canadian Society
  - http://www.cachnet.org
- European Society
  - http://www.escardio.org

- Useful anatomy and overall management
- No anesthesia input, nor specific helpful

Chassot PG. J Cardiothorac Vasc Anesth 2006;20:414-37
Cannesson M et al. Anesthesiology 2009;111:432-440

CHD Spectrum (AHA)



Simple
- AV, MV
- ASD, VSD
- Mild PS
- Small PDA

Moderate
- AS (sub/supra),
- SOVA, Aorta LV fistula
- PAPVD
- AVSD, 1° ASD, VSD
- Ebstein's
- TOF, RVOT infundibular
- PV regurge/stenosis
- PDA

Complex
- All cyanotic hearts
  - TGA
  - Tricuspid atresia
  - Truncus arteriosus
  - Pulmonary atresia
  - Single ventricle
- Double outlet ventricle
- Mitral atresia
- Conduits: valved or non
- Fontan
- Eisenmenger's

Warnes, et al. J Am Coll Cardiol 2008;52:143-263

Adult CHD

Untreated	Palliated	Corrected
+ + + +	+ + + Symptoms +	+ Normal

- Cyanotic CHD
- Fontan
- PAH
- Complex CHD residua
  - heart failure
  - valve disease
  - anticoagulation
- Malignant arrhythmias
- Childhood corrected
  - Normal ventricle
  - Life expectancy
  - No further intervention
- ASD
- VSD
- PDA

CHD Multi-system Effects

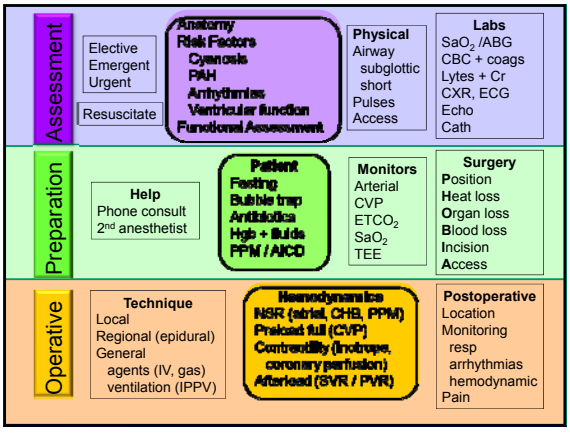
- Neuro: stroke, abscess, developmental
- Cardiac: CAD, PAH, Arrhythmias, vent function
- Respiratory: 1°, vasculature, V/Q
- Renal: renal dysfunction/failure (16%)
- Hematologic: Hgb, plts, coags
- MSK: position, lung

- 2-3% congenital anomaly
- 0.8% CHD
  - 85% isolated
  - 15% syndrome

Warnes. JACC 2005; 46:1-8

Laboratory

- BLOODS
  - Hgb (R→L shunt)
  - HCT (Fe def)
  - Coags (plt, INR)
  - Lytes
  - ABG/SaO_2
- ECG
  - Rhythm
  - Chamber size
- CXR
  - Heart shape + size
  - Pulmonary vascularity
- ECHO/MRI
  - Anatomic defect
  - Shunts
  - Ventricular function
  - Valve function
- CATH
  - Quantify stenosis
  - Measure SaO_2
  - Quantify shunt



Case 1

Q3

42 y.o. female, uncorrected ASD, has RA SaO_2 of 93%. She is asymptomatic.

During a GA, which statement is false ?

A. Induction with intravenous drugs is faster
B. She has pulmonary hypertension
C. $ETCO_2$ overestimates Parterial CO_2
D. Peripheral SaO_2 is an adequate shunt monitor
E. Bubble traps prevent paradoxical emboli

ASD R to L Shunt

- RV dilated + TR
- Cyanotic, ↓ SaO_2, ↑ Hct
- Shunt monitor: SaO_2
- $ETCO_2$ underestimates $PaCO_2$
  - Less PBF
- Induction: IV fast, gases slow

↑ PBF = ↑ SaO_2
- ↑ SVR
- ↓ PVR

Case 1

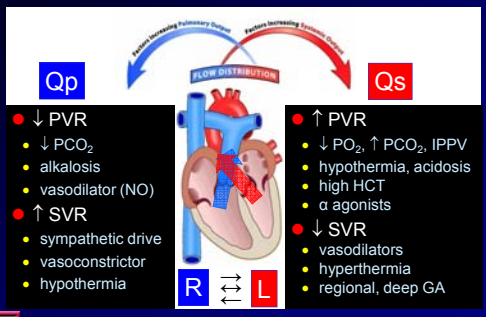
Q4

The patient has RA SaO_2 of 93%.
During her GA the SaO_2 drops to 85%.

Which strategy will worsen her SaO_2 ?

A. Increasing FIO_2
B. Increasing PEEP
C. Return to spontaneous ventilation
D. Reducing PVR
E. Increasing SVR

PVR and SVR



- ↓ PVR
  - ↓ PCO_2
  - alkalosis
  - vasodilator (NO)
- ↑ SVR
  - sympathetic drive
  - vasoconstrictor
  - hypothermia

- ↑ PVR
  - ↓ PO_2, ↑ PCO_2, IPPV
  - hypothermia, acidosis
  - high HCT
  - α agonists
- ↓ SVR
  - vasodilators
  - hyperthermia
  - regional, deep GA

Cannesson M et al. Anesthesiology 2009; 111:432-40

PVR Dependent Lesions

↑↑↑PVR→ ↓ SaO_2 or ↓ SBP

- TOF + PI + ↓↓RV
- Ebstein's
- Glenn
- Fontan
- Eisenmenger's

→

- PCO_2 25-30mmHg
- PH > 7.45
- FIO_2 100%
- Low intrathoracic P
- Inhaled: NO, Prostacyclin
- Mg SO_4
- RV function

Hemodynamics

	Goal	SVR/PVR	↓ SaO_2	↓ BP
L→R	↓ PBF	↓ / ↑	↑ FIO_2 ↓ PVR	↑ SVR
L>>R				↓ FIO_2 ↑ PVR
R →L	↑ PBF	↑ / ↓	↑ FIO_2 ↓ PVR	↑ PVR ↑ SVR
Eisen				
Fontan		↑ CVP/ ↓		

Case 2

Q5

19 y.o. male, uncorrected VSD urgent repair of a leaking femoral false aneurysm:

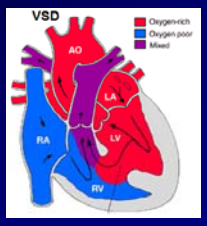
- High Qp:Qs ratio >3:1
- Room air SaO_2 97%
- Moderate ventricular dysfunction

Which statement is false?

A. This patient has pulmonary hypertension
B. Increasing FIO_2 will worsen acidosis
C. IV induction is slower in this patient
D. This patient will have a normal hemoglobin
E. High FIO_2 will improve peripheral perfusion

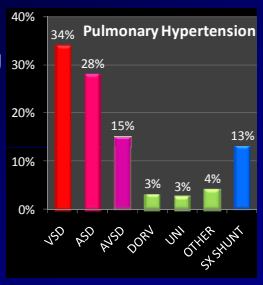
Unrestricted L→ R Shunts

- Qp : Qs > 3/1
- ↑ PBF → PAH
- High SaO_2 ≠ adequate O_2 delivery
- O_2 will ↑ PBF but ↓ systemic blood flow, worsening acidosis
- ↓ FIO_2 may ↑ peripheral perfusion



CHD Pulmonary Hypertension

- Mean PAP > 28mmHg
- PVR > 300dynes
- 4.2% adult CHD
- ↑ flow or pressure
- Fixed vs reactive
- Eisenmenger's 1-2%



Duffels et al. Int J Cardiol 2007;120:198-204

Case 2

Q6

If this patient does not undergo VSD repair he will develop Eisenmenger's syndrome.

What is not a feature of Eisenmenger's?

A. Pulmonary vaso-occlusive disease
B. Airway hemorrhage
C. Brain abscess
D. Polycythemia
E. Coagulopathy/platelet consumption

Eisenmenger's Syndrome

- Final pathway L→R shunt (Qp:Qs >2:1)
- Pulmonary vaso-occlusive disease (PVOD)
  - Severe PHT (PVR > 800dynes/s/cm^5)
  - Progressively equal ventricular pressures
  - Bidirectional shunt, RV dilated +TR
- Cyanosis → Erythrocytosis ≠ polcythemia
- Coagulopathy/platelet consumption
- Cerebral microemboli, brain abscess
- Airway hemorrhage

Case 2

Q7

After fasting, a patient with Eisenmenger's syndrome has Hgb 180 (Hct 60%) and a headache.

Perioperative management of this patient includes ?

A. Hemodilution to avoid intraoperative stroke
B. Phlebotomy
C. Transfusion to maintain Hgb 165
D. Minimize transfusion to avoid antibodies
E. Colloids to maintain intravascular volume

Hyperviscosity Syndrome

- Erythrocytosis ≠ polcythemia
  - High Hgb best indicator R→L shunt size + chronic
  - Stroke risk: a fib, HBP, microcytosis (Fe def)
  - Hyperviscosity syndrome: headache, "TIA"
    - Hct > 65%: 2° erythrocytosis
    - Hct < 65%: Fe def, dehydration
- Phlebotomy if (Hct > 65%):
  - Moderate - severe hyperviscosity syndrome from 2° erythrocytosis
  - Perioperative for autologous blood transfusion

Oechslin E, Int J Cardiol 2004;97:109-115

Case 2

Q8

Patient with uncorrected VSD and Eisenmenger's.

Anesthesia technique of choice for an elective femoral-femoral cross-over is ?

A. General anesthesia with spontaneous ventilation
B. General anesthesia with IPPV
C. Spinal anesthesia
D. Epidural anesthesia
E. Combined neuroaxial and general anesthesia

Regional Anesthesia

- ↓↓ SVR (dramatically)…Spinal>>>>>Epidural
- ↓ Preload (↓ venous return + ↑ capacitance)
- ↓ HR if Level T4
- Complications
  - Shunt worsen R→L, AP collaterals
  - Hypotension
  - Failed technique
  - Coagulation

Martin J et al. Reg Anesth Pain Med 2002;27:509-13

Case 3

Q9

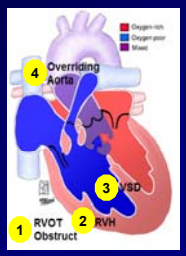
42 y.o. female, corrected Tetralogy of Fallot (TOF) for urgent treatment of a pulmonary hemorrhage.

This patient is unlikely to have?

A. Pulmonic insufficiency
B. Atrial arrhythmias
C. Residual VSD
D. RVOT obstruction
E. Symmetric radial pulses

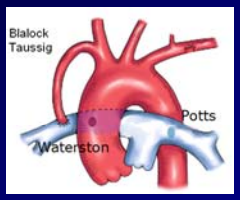
Uncorrected TOF

- 4 components
- Cyanosis: overriding aorta
  - ↑SVR → ↓R shunt → ↑SaO_2
- RVOTO dynamic (= HOCM)
- Aortopulmonary collaterals
  - Lower diastolic pressure
  - ↑ LV volume
- Unoperated < 3% at 40 years

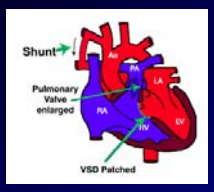


Shunts to Augment PBF

- Arterial shunts
- BP dependent
  - ↓ SVR → ↓ PBF (↓SaO_2)
- ↑↑ L → R shunt
  - ↓ coronary perfusion
  - ↓ peripheral perfusion
  - worsen LV function
- Arterial line/BP monitor



Corrected TOF



- PI (RV dysfunction)
- Arrhythmias (VT)
- Residual VSD
- RVOT obstruction
- Conduit stenosis (RVH)

Case 3

Q10

Corrected TOF with severe PI + RV dysfunction. She needs rigid bronchoscopy for pulmonary hemorrhage. She has no drug allergies.

What antibiotics should she receive?

A. None
B. Ampicillin + gentamycin
C. Clindamycin
D. Cefazolin
E. Vancomycin

Antibiotics CHD

- Unrepaired or palliated cyanotic CHD
- CHD repair < 6 months (no endothelialization)
- CHD repair but local residua prevents endothelial
- Prosthetic heart valves

Dental or upper airway
Ampicillin 2.0 gm IV
Cefazolin 1 gm IV
Ceftriaxone 1gm IV
Clindamycin 600mg IV

Circulation. Oct 9 2007;116(15):1736-54.

Case 3

Q11

She needs a rigid bronchoscopy for pulmonary hemorrhage. The infusion of choice for a total intravenous anesthetic is:

A. Fentanyl + versed
B. Propofol + remifentanil
C. Etomidate
D. Methohexital
E. Ketamine

Intravenous Anesthetic Agents

- Induction

	SVR	PVR	HR	MAP
Propofol	↓↓	↓	↓	↓↓
Ketamine	↑/↓	↑/↓	↑↑	↑
Etomidate	↓	↓↓	Ø	↓
Methohexital	↓	Ø	↑	↓

- Opioids
  - Fentanyl 5-20ug/kg
  - Sufentanyl 5-10ug/kg
- Amnestics
- Muscle relaxant

Ketamine
- Secretions
- Risk coronary ischemia
- IV induction 1-2mg/kg
- Sedation

Inhalational Agents

- Induction: Halothane or Sevo
- Maintenance: abolish PVR

	Contract	SVR	HR	MAP
Halothane	↓↓	↓↓	↓	↓↓
Iso	↓	↓	↑	↓
Sevo	↓	↓	--	↓
Des	↓	↓	↑	

- N_2O
  - Air emboli (shunts)
  - Mild ↓ CO, no change PVR
  - Reconsider: ↓PBF, PHT, ↓ myocardial function

Shared Airway + Jet Ventilation

- FIO_2 limitations
  - 100% O_2 for non laser cases
  - Room air or < 30% for laser (airway fire)
- No regulation or limitation of Paw
- ET CO_2 unmonitored
  - Apnea, ↑↑PCO_2
- Challenge for PVR dependent patients

Case 3

Q12

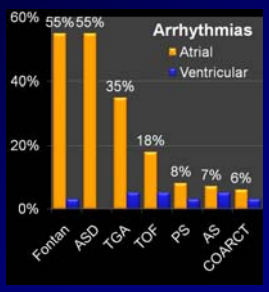
During the rigid bronchoscopy under TIVA the patient has short runs of ventricular tachycardia.

Optimal management of this patient is :

A. Deepen anesthetic
B. Administer amiodarone
C. No treatment, normal for patient
D. Hyperventilate
E. Wake the patient up

CHD Cardiac Arrhythmias

- Atrial: large size
- Ventricular: TOF
- Blocks: PPM
- Morbidity + mortality
- Management
  - Medical
  - Ablation
  - Surgery



Vander Veld ET et al. Eur J Epidemiol 2005;20:549-57

Case 4

Q13

20 y.o. male,univentricular heart, Fontan circulation in atrial flutter for elective cardioversion.

Systemic perfusion for this patient depends on:

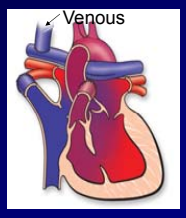
A. RA to LA pressure gradient
B. High PVR
C. High SVR
D. Subpulmonic ventricular function
E. Intracardiac shunt

Univentricular Heart

- 1 ventricle, 2 atria, 2 great vessels
  - Hypoplastic RV or LV
  - Atretic valve: MV, TV
- Volume overload = round ventricle
- Ratio PVR: SVR → Qp: Qs flow
- Palliative procedure
  - Shunt: ASD, Glenn, Blalock-Taussig
  - Fontan: no pulmonic ventricle, flow to PA

Glenn Shunt

- SVC → PA: upper body
  - IVC → RA: lower body
  - Pulmonary AVM
  - ↓ SaO_2 75-85%
- No access to heart from IJ
  - CVP insertion (RIJ careful)
  - IV (upper and lower)

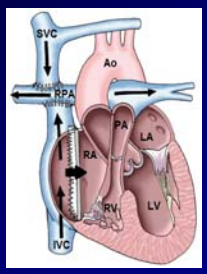


Fontan Procedure

- Indication: “single or common” ventricle
  - Tricuspid Atresia, Hypoplastic Right/Left Heart, Double Outlet RV, Double Inlet LV
  - Systemic venous return directly to PA
- Staged Procedure
  1. Bi-directional Glenn or hemi-Fontan
  2. Fontan procedure with IVC directed to PA
     - Older: includes RA in circuit
     - Newer: bypass RA

Fontan Circulation

- Passive PA filling
  - CVP – LAP: 8-10mmHg
  - Avoid ↑ intrathoracic P (IPPV, PEEP)
  - Avoid ↑ LAP: LV failure, MR
- Sinus rhythm
  - SSS or tachy-brady
  - Enlarged RA (thrombus)
- Good ventricular function
- Atriovent valve competent



Case 4 Q14

Anesthesia for elective cardioversion in this patient with a Fontan would include:

A. Faster IV induction
B. Positive pressure ventilation
C. Ketamine to maintain PVR
D. Propofol to reduce PVR
E. Fentanyl for pain

Fontan Circulation

- Avoid hypovolemia
  - CVP 15-18 mmHg, monitor CVP
- PVR low (SaO_2 95% RA, 90% = poor flow)
  - Spontaneous ventilation, ↓ $PaCO_2$, ↑ PaO_2
  - IPPV: low respiratory pressures, ↓ $PaCO_2$
- Avoid ↑ LAP: LV dysfunction, MR, loss NSR
  - Poor ventricular reserve (inotropes)
- Epidural more controlled then spinal
  - Bleeding (hepatic), thromboembolic (coumadin)

Case 4 Q15

Monitoring during elective cardioversion in this patient with a Fontan would not include:

A. Arterial line
B. ECG
C. Saturation monitor
D. End tidal CO_2
E. Central venous pressure

Case 4 Q16

Following cardioversion the patient is hemodynamically unstable with BP 70/40 and HR 50 NSR. You should immediately:

A. Administer ephederine
B. Administer volume
C. Pace the patient
D. Administer phenylepherine
E. Administer epinepherine

Cardioversions in CHD

- Emergent vs elective
- Fraught with complications
  - Mustard, TOF, Fontan
- Monitoring
- Location
- Clear “Plan B”
- TEE to r/o thrombus
- Pacing options if ↓ HR

Balling G, et al. J Thorac Cardiovasc Surg 2000;119:745-52

Summary

Assessment
Anatomy
Risk Factors
Functional Assess
Airway, pulses, SaO_2
CBC, Echo, Cath

Preparation
Fasting (Hgb + fluids)
Antibiotics
Bubble trap
PPM/AICD
Monitors

Operative
Technique (simple)
Ventilation: (SaO_2, $ETCO_2$)
Hemodynamics
Afterload (SVR / PVR)
Postop monitored care

Thank You