Making Sense of Complex Repairs 20 years Later

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Disclosures

- No financial disclosures
- Some drugs discussed are an off-label application

Objectives

1. Identify the anatomy of complex congenital heart repairs
2. Differentiate uncomplicated from complicated repairs with respect to physiology and anesthetic management
3. Discuss Single ventricle, TGA and Eisenmenger syndrome physiology and anesthetic management

ACHD

- CHD 5 - 9 of 1,000 live births
  - 1.5 of 1,000 live births have complex CHD
- More adults than children live with CHD in USA
  - ~1 - 3 million adults with CHD in USA & Canada
  - ~1.8 million adults with CHD in Europe
- Improvement in survival over the past 20 years
  - 90% of children survive to adulthood
- USA estimates 500,000 adults with complex CHD
  - Only 10% receive follow-up care in a ACHD center

[26] Rouine-Rapp 2012

What is so unique about ACHD population?

- Society has already invested a large amount of resources to achieve survival to adulthood
- Young adults with CHD have the potential to contribute to the GDP for 30-40 years
- The period of early adulthood is relatively uneventful in terms of complications and resource utilization compared with early childhood and later adult life

[35] Williams 2011

However……..

- Many young adults with ACHD do not receive cardiology follow-up
  - Re-location with school and work
  - Health insurance
  - Perception that they are doing well
  - Lack of transition from pediatric to adult programs
- This lack of preventive care may increase the overall costs of care

Perioperative Outcomes

- Major non-cardiac surgery in ACHD
  - Greater morbidity and mortality
  - ACHD independent predictor of increased mortality
- Vulnerable population
  - 50% of adults with CHD can not correctly name or describe their diagnosis
  - Majority of anesthesia providers do not have the knowledge and are not comfortable looking after patients with ACHD, especially as complexity increases

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Anesthesia providers and ACHD

- ACHD patients presenting for non-cardiac surgery
- Highest knowledge and comfort scores for:
  - Fellowships in cardiac anesthesia and pediatric anesthesia
  - Increased frequency of CPB cases
  - Increased frequency of providing care for patients under 2yrs of age
- Implications for training

[19] Maxwell 2014

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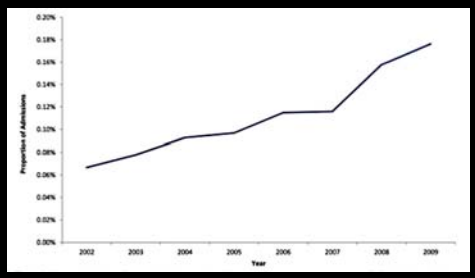
Increasing non-cardiac surgery in ACHD patients



[20] Maxwell 2013

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Outcomes

	ACHD Cohort n = 10,004	Comparison Cohort n = 37,581		
Outcome	n (%)	n (%)	*P* Value	OR (95% CI)
Death	407 (4.1)	1,355 (3.6)	0.031	1.13 (1.01–1.27)
LOS (median [IQR])	4.8 (2.4–10.4)	2.9 (1.5–5.6)	<0.001	
Total charges (median [IQR])	$42,171 ($22,918–$93,847)	$26,982 ($15,814–$46,784)	<0.001	
ARF	620 (6.2)	1,826 (4.9)	<0.001	1.29 (1.18–1.42)
Pneumonia	942 (9.4)	2,998 (8.0)	<0.001	1.20 (1.11–1.29)
Respiratory failure	916 (9.2)	2,933 (7.8)	<0.001	1.19 (1.10–1.29)
DVT/PE	405 (4.1)	773 (2.1)	<0.001	2.01 (1.78–2.27)
Stroke	607 (6.1)	1,168 (3.1)	<0.001	2.01 (1.82–2.23)
MI/cardiac arrest	431 (4.3)	1,307 (3.5)	<0.001	1.25 (1.12–1.40)
Composite	2,145 (21.4)	6,003 (16.0)	<0.001	1.44 (1.36–1.52)

Values are reported as number (percentage) unless otherwise denoted as median (IQR). Composite = ARF, pneumonia, respiratory failure, DVT/PE, stroke, MI, and cardiac arrest.
ACHD = adult congenital heart disease; ARF = acute renal failure; DVT = deep venous thrombosis; IQR = interquartile range; LOS = length of stay; MI = myocardial infarction; OR = odds ratio; PE = pulmonary embolus.

[20] Maxwell 2013

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Mortality by lesion type

		Died
	n	n (%)
Atrial septal defect	4,068	155 (3.8)
Congenital aortic stenosis/aortic insufficiency	1,789	53 (3.0)
Congenital mitral stenosis/regurgitation	85	3 (3.5)
Congenital conduction defect*	469	10 (2.1)
Congenital coronary anomaly	248	9 (3.6)
Pulmonic stenosis	239	13 (5.4)
Tetralogy of Fallot	121	7 (5.8)
Ventricular septal defect	831	52 (6.3)
Ebstein anomaly	65	4 (6.2)
Others	1,745	76 (4.4)
Combined complex†	344	25 (7.3)

[20] Maxwell 2013

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Single Institution Data

Mayo Clinic, 2013

- All patients who had undergone Fontan palliation (n = 1,133)
- Patients > 16yrs, Fontan, for non-cardiac surgery
- 39 GAs given to 31 patients
  - 31% had perioperative complications
  - One death

[24] Rabbitts 2013

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ACHD: Anesthetic Considerations

- Detailed knowledge of anatomy & physiology
- Multidisciplinary team
- Increased perioperative risk
  - CHF
  - Pulmonary Hypertension
  - Cyanosis
  - Bleeding & thrombosis
  - Dysrhythmias

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ACHD and CHF

CHF defined as:

- VO_2<25ml/kg/min & NT-pro-BNP >100pg/ml
  - 26% of ACHD, mostly young 30-40yrs
  - Increases as lesion complexity increases
  - Greatest risk of CHF:
    - Single ventricle (R > L)
    - Tetralogy of Fallot s/p repair with PI
    - TGA

[7] DiNardo 2013

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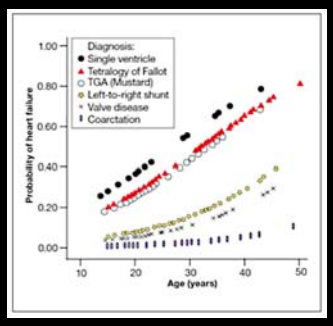
Probability of Heart Failure by CHD



[7] DiNardo 2013

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Relative Risk for Specific Arrhythmias in Common Congenital Heart Defects

	IART	AF	WPW	VT/SCD	SA Node Dysfunction	Spontaneous AV Block	Traumatic AV Block
VSD	+			+			+
ASD	+	+					
TOF	++			++			+
AS		+		++			+
D-TGA (M&S)	+++			++	+++		
CAVC	+					+	++
SING V (F)	+++	+		+	+++		
L-TGA	+		++	+		++	+++
Ebstein's anomaly	++		+++	+			

AF indicates atrial fibrillation; WPW, Wolff–Parkinson–White syndrome; SCD, sudden cardiac death; SA, sinoatrial; VSD, ventricular septal defect; ASD, atrial septal defect; TOF, tetralogy of Fallot; AS, aortic stenosis; M&S, after the Mustard or Senning operation; CAVC, common AV canal defect; SING V (F), single ventricle after the Fontan operation; +++, high risk; ++, moderate risk; and +, slight risk.

[33] Walsh 2007

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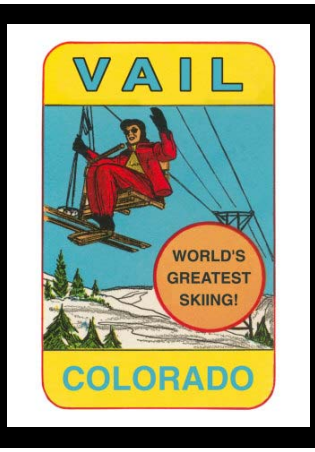

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Complex Lesions and Physiology

- Single ventricle palliated to Fontan physiology
- Transposition of the Great Arteries (TGA)
- Eisenmenger syndrome

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Single Ventricle Physiology

"So in whatsoever creature there is lungs, there is likewise in them two ventricles of the heart, the right and the left"

William Harvey 1638

However…..

1940s recognition that across species, PAP was 25/10 mmHg and venous pressure alone may be sufficient to move blood through the lungs

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ACHD: Single Ventricle Physiology

- Palliative path for a univentricular heart
  - Stage 1: BT shunt, Norwood procedure, Sano shunt, PA Banding
  - Stage 2: Glenn shunt
  - Stage 3: Fontan
- The single ventricle may be morphologic left or right

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CHD leading to single ventricle pathway

Pathology	Pulmonary Blood Flow	Why Biventricular Repair Is Not Possible
SV		
LV (DILV)	↓ more often than ↑ depends on pulmonic or aortic obstruction	RV hypoplastic
RV (DORV)	↓ more often than ↑ depends on pulmonic or aortic obstruction	LV hypoplastic or Inlet VSD, straddling TV
Unbalanced AVC	↑ more often than ↓	Inlet VSD, straddling TV
Heterotaxy syndrome*	↓ more often than ↑ depends on pulmonic or aortic obstruction	Common AV valve, common atrium
Tricuspid Atresia		TV absent, RV hypoplastic
Type I - GA-NR	↓ most common	
Type II - D-TGA	↑ more common	
Type III - L-TGA	↓ or ↑	
HLHS	↑	LV and ascending aorta hypoplastic

[18] Leyvi 2010

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Stage 1 Too much PBF

Tighten to PA Band to ½ systemic BP (Trusler's rule) and SaO_2 of 80%

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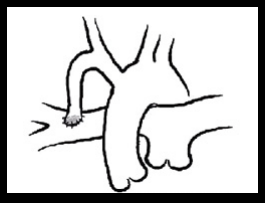
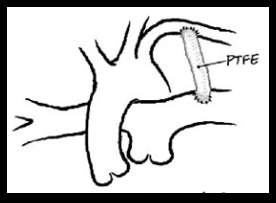
Stage 1 Too little PBF



Classic BT shunt



Modified BT shunt

Establish a reliable source of PBF - balance Qp:Qs with Sats 80%

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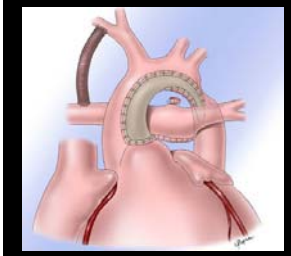
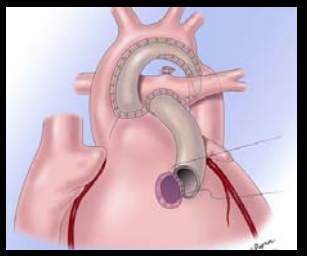
Stage 1 Norwood



BT-Shunt



Sano Modification

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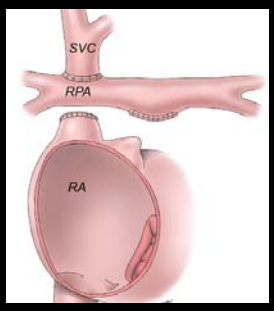
Qp/Qs

$Qp / Qs = (SaO_2 – SvO_2) / (SpvO_2 – SpaO_2)$

– Assume $SpvO_2$ 100%
– Measure $SaO_2 = SpaO_2 = 80\%$
– Measure $SvO_2 = 60\%$

$Qp / Qs = (SaO_2 – SvO_2) / (SpvO_2 – SpaO_2)$
$= (80 – 60) / (100 – 80)$
$= 20 / 20$
$= 1 / 1$

Stage 2 Glenn Shunt



- 2-6 months of age (PVR has decreased)
- Decrease volume load on the heart
- Sats 80%
- Physiology of PBF
- Pulmonary arterial venous AVMs develop due to lack of hepatic factor to lungs
- Monitoring issues

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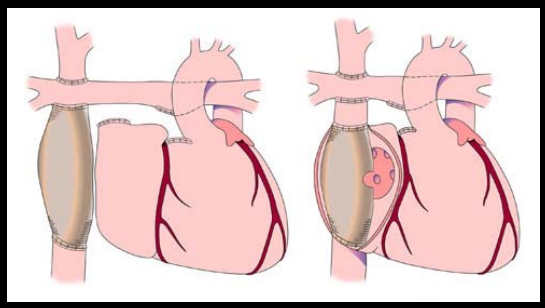
Stage 3: Fontan



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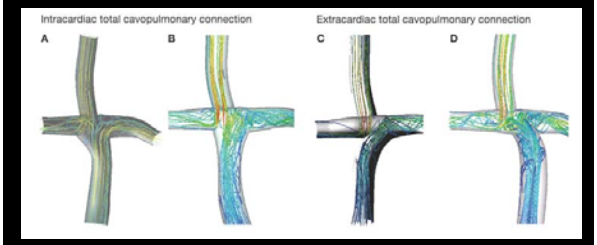
Fontan Fluid Dynamics



[4] de Leval 2005

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Stage 3: Fontan

Extracardiac Conduit

- Preserve fluid energy
- Preserve normal atrial pressures
- Eliminates extensive atrial suture lines (decrease arrhythmias)
- Fenestration allows systemic preload to be maintained (at the expense of saturation) in the event of increased PVR. May be closed in cath lab at a later time.

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Fontan Physiology and PBF

1. Spontaneous Ventilation

- Increased venous return and PBF
- BUT avoid hypercarbia, hypoxia, atelectasis and acidosis
  – Increase PVR
  – Decrease PBF and CO

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Fontan Physiology and PBF

2. Positive Pressure Ventilation
- PBF during the expiratory phase
  - Limit PIP < 20 cmH_2O
  - Low RR (<20 bpm)
  - Short inspiratory times
  - Avoid high PEEP (but avoid atelectasis)
  - Tidal volumes 10ml/kg
  - Early extubation
- Adequate intravascular volume

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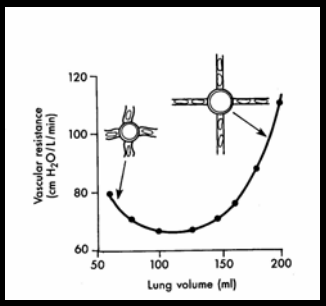
Optimizing Ventilation



[34] West

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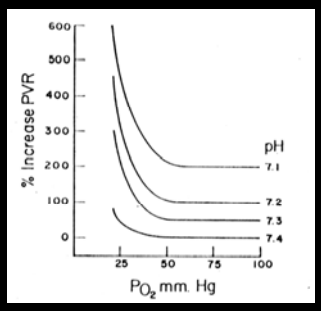
Prevent an increase in PVR



[27] Rudolph 1966

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Fontan Failure

1. Cardiac
- Arrhythmias
- Congestive heart failure (Increased work of single ventricle)
  - Problems with ECHO estimations
- AV valve regurgitation

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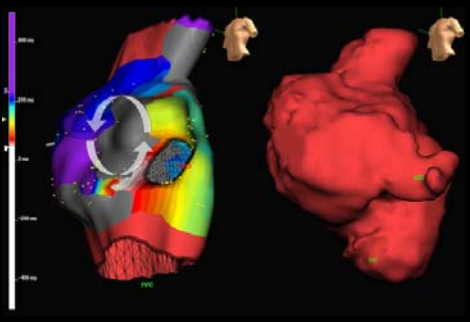
Fontan Arrythmias



[22] Mondesert 2013

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Fontan Failure

2. Pulmonary
- Increasing PVR
- Cyanosis
  - Pulmonary AVMs, fenestration
- Pleural effusions
- Plastic bronchitis

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Plastic Bronchitis

[30] Singhal 2013

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Fontan Failure

3. Hepatic
- Dysfunction
  - Synthetic function decreased
- Protein-losing enteropathy
  - Loss of proteins, immunoglobulins
  - Ascites
- Esophageal varices

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Fontan Failure

4. Hematolgic
- Thromboembolic
  - Hypercoagulability, atrial arrhythmias
  - Passive venous flow
- Often on aspirin and/or Coumadin

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Fontan: Perioperative decreased CO

- Hypovolemia (NPO status)
- Positive pressure ventilation
- Hypercarbia
- Hypoxemia
- Increase venous capacitance (anesthetics)
- Ventricular dysfunction
- Arrhythmias
- Increase PVR (effusion, ascites, hypothermia, pain)

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Fontan: Periop strategies to increase CO

- Optimize ventilation
  - FiO_2 1.0, pCO_2 30 mmHg, pH 7.45
  - Consider iNO 20-40ppm
- Adequate anesthesia & analgesia
- Normothermia
- Inotrope support
  - Milrinone
  - Dobutamine

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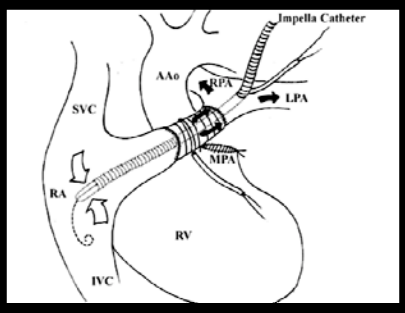
Future Fontan



VanArsdale AHA Abstract 2013

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Complex Lesions and Physiology

- Single ventricle palliated to Fontan physiology
- Transposition of the Great Arteries (TGA)
- Eisenmenger syndrome

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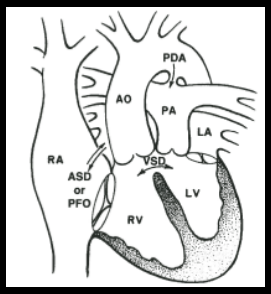
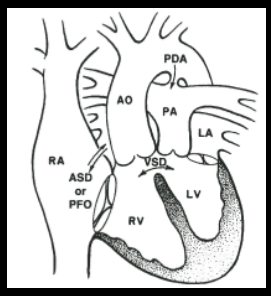
Anatomy of TGA

- Atrio-ventricular *concordance*, ventriculo-arterial *discordance*
- d-TGA
  - Aorta is anterior and to the right of the pulmonary trunk (as opposed to posterior)
  - Refers to embyrological looping



TGA: diagnosis in fetal life

- Chronic cerebral hypoxia – delayed brain maturation
- Antenatal diagnosis (75% in modern centers) has a beneficial effect on pre-op status and post-op outcome
- High pre-op lactate predicts poor neurological outcome



Types of TGA

1. TGA with intact ventricular septum (TGA/IVS)
   - 85% of cases
2. TGA with ventricular septal defect (TGA/VSD)
   - 10% of cases
   - Associated with other abnormalities: right side aortic arch, IAA, coarctation of aorta
3. TGA/VSD with left ventricular outflow tract obstruction (TGA/VSD/LVOTO)
   - Abnormal pulmonary valve not suitable for arterial switch operation (ASO)
4. Congenitally corrected TGA (ccTGA)

Coronary artery anatomy

- The coronary arteries will need to be moved during the ASO
- Anomalies of the coronary arterial course which are most challenging:
  - Intramural
  - Coronary artery stretches over RVOT
  - Lying close to a commissure

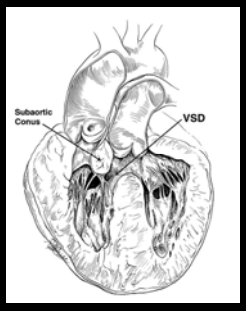
TGA: early management

- Balloon atrial septostomy (BAS) (Miller & Rashkind 1964) may predispose to cerebral embolism but is needed in the majority of infants
- Failure of cyanosis to resolve after PGE and BAS may indicate pulmonary hypertension (12%)

Taussig-Bing malformation

1949 Helen Taussig and Richard Bing

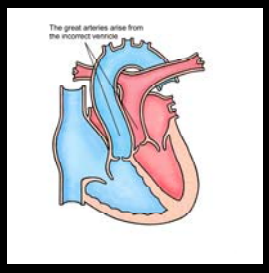
- DORV with both great vessels from the RV
- VSD streams blood from LV to PA and RV to Aorta
- Repair by tunnelling VSD to PA (create TGA) and then perform ASO (undo the TGA)

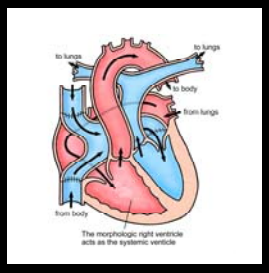


TGA: options for surgical correction

- 1959 Senning procedure
  - Complex re-routing of blood in the atria
- 1964 Mustard procedure
  - Simpler atrial baffling
    - Right blood into the right great artery but from the wrong ventricle
    - Problems with:
      - Conduction system
      - Baffle leaks and obstruction
      - RV failure
      - Systemic (tricuspid) AV valve failure

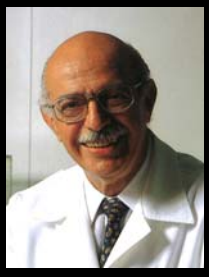
TGA: Senning-Mustard Repair



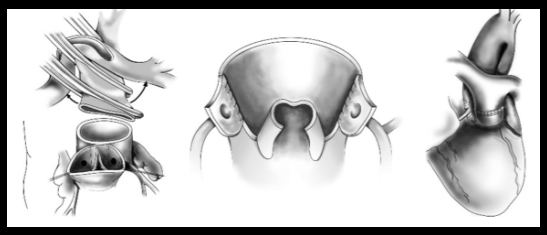


Arterial Switch Operation (ASO)

- 1975 Adib Jatene performed the first ASO in Brazil
- True anatomical correction of TGA



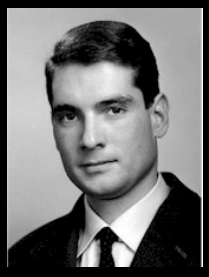
TGA: ASO



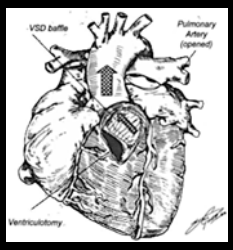
Lecompte Maneuver: the PAs are brought anterior to the aorta

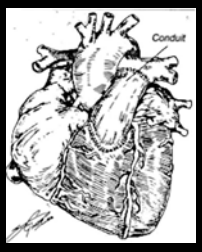
TGA/VSD/LVOTO

- 1969 Giancario Rastelli Mayo Clinic
- Pulmonary valve is no good so precludes the ASO
- Rather than closing the VSD it is baffled to the aorta and an RV-PA homograft is placed (requires replacement)



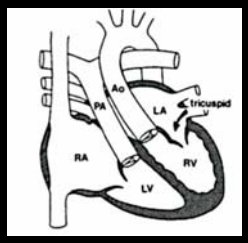
Rastelli Procedure





ccTGA

- Double discordance
  - Atrioventricular
  - Ventriculoarterial
- Physiologically corrected transposition
- L-TGA
- High incidence of Ebstein like dysplasia of the systemic (tricuspid) AV valve

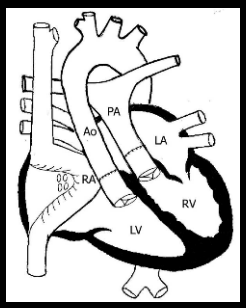


ccTGA

- Often survive to age 50yrs uncorrected BUT
  - Develop AV valve regurgitation
  - Impaired RV function
  - Rhythm disturbance
- Classic repair
  - Fix any intracardiac defects such as VSD, repair tricuspid (systemic) AV valve
  - Pacemaker placement
  - Leave the RV systemic

The double switch for ccTGA

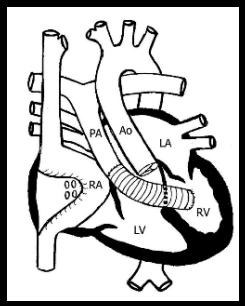
1. ASO
2. Senning-Mustard



The double switch for ccTGA

- What if there is LVOTO?

1. Senning – Mustard
2. Rastelli



Outcomes of ASO

- Survival approaching 100%
  - Mild developmental delay
- Pulmonary artery stenosis
  - Re-operation or cardiac cath in 30% of long term survivors
  - Be careful with pulmonary artery stents: compression of aorta and/or coronary artery

Late complications: adults after ASO

1. Arrhythmias
   - More common in baffle procedure
   - 10% including AVN block, SVT and VT (increased with VSD)
2. Coronary artery dysfunction
   - Clinically silent ischemia due to denervated heart
   - Usually present with pallor, VT or sudden death
   - Significant problems in 10% patients so far
   - Screening
3. Dilation of the neo-aortic root and AI

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Complex Lesions and Physiology

- Single ventricle palliated to Fontan physiology
- Transposition of the Great Arteries (TGA)
- Eisenmenger syndrome

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Eisenmenger Sydrome

- Victor Eisenmenger 1897
  - 32yr old man with cyanosis and hemoptysis
  - Large VSD on post-mortem
- Paul Wood 1958
  - Coined the term Eisenmenger syndrome
  - Presence of a congenital heart defect permitting increased pulmonary blood flow/pressure resulting in increased PVR and reversed shunt

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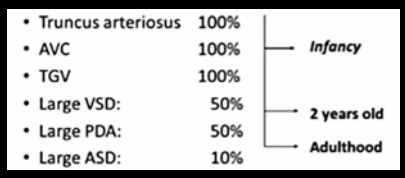
CHD and the likelihood of developing PH if not repaired within the designated time frame



Pre-tricuspid shunt (increase in flow) and post-tricuspid shunt (increase in flow and pressure)

[25] Rosenzweig 2012

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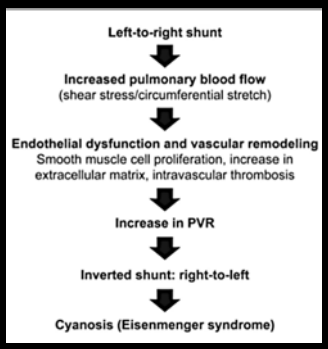
Development of Eisenmenger Syndrome



[2] Beghetti 2009

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Clinical manifestations of ES

Abnormalities	Clinical manifestations
Elevated and fixed pulmonary vascular resistance	Exercise intolerance, dyspnoea, syncope, sudden death
Secondary erythrocytosis	Hyperviscosity, relative iron, folic acid and vitamin B_{12} deficiency
Bleeding diathesis	Hemoptysis, cerebral hemorrhage, menorrhagia, epistaxis
Right ventricular failure	Liver enlargement, oedema
Arrhythmias	Syncope, sudden cardiac death
Altered red cell rheology and thrombotic diathesis	Cerebrovascular events, such as stroke or transient ischaemic attacks, intrapulmonary thrombosis
Renal dysfunction	Increased blood urea nitrogen, hyperuricaemia and gout
Hepatobiliary dysfunction	Calcium bilirubinate gall stones, cholecystitis
Infections	Endocarditis, cerebral abscess
Skeletal disease	Scoliosis and hypertrophic osteoarthropathy

[15] Kumar 2009

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Treatment of ES

1. Pharmacological
   A. Digoxin, diuretics, anticoagulants
   B. PAH pathways
      i. Treat and repair?
2. Phlebotomy
3. Transplantation
   A. Lung Tx with correction of cardiac defect
   B. Heart-Lung Tx

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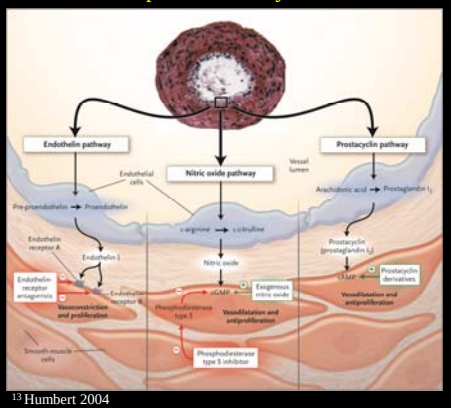
Therapeutic Pathways in PH



[13] Humbert 2004

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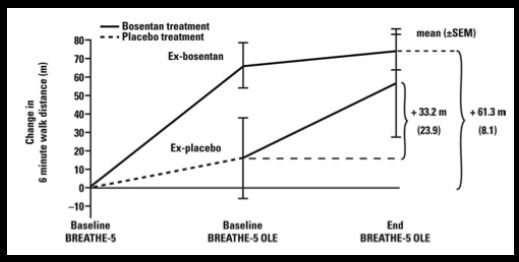
BREATHE-5
Bosentan Randomized Trial of ERA Therapy-5



[12] Gatzoulis 2008

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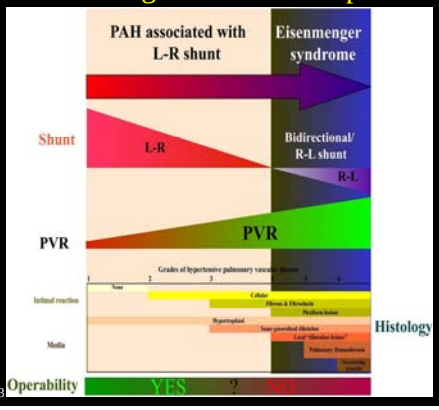
Eisenmenger: Treat and Repair?



[17] Lanigan 2013

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Conclusions

- Increasing number of adult survivors with complex CHD
- Perioperative care may be in smaller centers with little pre-op information
- Understand the plumbing!
- Consider CHF, arrhythmias, pulmonary hypertension, bleeding/thrombosis, and cardiopulmonary interactions

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Thankyou!

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