Blue Babies
(or Baby Blues)

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Take-Away Ideas

Five “T”s

✧ Tetralogy…is its conus
✧ TGA: counter-intuitive, including PGE
✧ TAPVC: sickest possible children
✧ Tricuspid atresia: infinite variety
✧ Truncus: Pas & the truncal valve
General Information on Cyanotic Congenital Heart Disease

Definition: Perception of bluish discoloration of skin, sclerae and mucous membranes caused by > 4.5 grams of de-saturated Hb in arterial circulation.

Cong Hrt Disease - Diff Dx

Acyanotic

L→R shunts

Obstructions

Combo

Blue Babies (5 “T”s)

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Cyanotic Congenital Heart Disease
The Five “T”s

<table>
<thead>
<tr>
<th>% Freq</th>
<th>Lesion</th>
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</thead>
<tbody>
<tr>
<td>≈75%</td>
<td>Tetralogy of Fallot (ToF)</td>
</tr>
<tr>
<td>≈15%</td>
<td>Transposition of the Great Arteries (TGA)</td>
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<tr>
<td>≈10%</td>
<td>Truncus Arteriosus</td>
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<tr>
<td></td>
<td>Tricuspid Atresia (+ Ebstein’s)</td>
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ToF Pathology

If you truly understand this picture, you comprehend ToF.

1. Subpulmonary stenosis
2. Malalignment VSD
3. Over-riding aorta
4. RVH
Domed, thickened pulmonary valve

SubPS

Mal-alignment VSD

Over-riding Aorta (hard to see in this image)

Interventricular septum

RV
ToF Physiology

1. ALL ToF patients have an unrestrictive VSD.
2. ToF patients virtually never have LVOT obstruction.
3. Therefore, RV pressure always = LV pressure, no more, no less.
4. ToF pathophysiology is dominated by the degree and variability of the RVOT obstruction, which determines pulmonary blood flow ($Q_p$).
5. A PDA → a variable but unreliable source of $Q_p$. 
ToF Physiology - Total RVOT Obstruction

Pulmonary Atresia

What the neonatologist Usually sees.

Ductal-Dependent Pulmonary blood flow.

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ToF Physiology - Mod/Severe Obstruction

PVR is almost always low.

VSD is always unrestrictive.

Degree of RVOT obstruction $\rightarrow Q_p$, which $\rightarrow$ arterial $O_2$ saturation.
Mild muscular RVOT conal obstruction can constrict into severe stenosis under certain conditions. This can produce a *tet spell*. 

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A ToF Spell

Tet spell is *NOT* simply a large drop in arterial saturation. It is a specific clinical entity and a medical *emergency*. Occurs when RVOT obstruction ↑ and Qp ↓ < critical level.

**Clinical signs & symptoms**
- Unconsolable. Air hungry.
- Cyanotic, hyperpneic.
- Pale, diaphoretic.
- Essentially no murmur

**Treatment [Why each?]**
- Oxygen
- Volume
- NaHCO₃
- β-blockade

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Therapy in ToF

Ductal-dependent $Q_p$
- Usually a neonate. Give PGE
- GOB-T shunt (& ligate PDA).
- Usually pulm atresia: will need conduit for repair.

Survivable $Q_p$ through RVOT
- $\beta$-blockade.
- Open RVOT & close VSD.
- Trans-anular patch & close VSD.
- RV-PA conduit +/- close VSD.
Systemic-to-Pulmonary Shunt

Neonate with ToF/PA (1.9 kg)

Note finger.

Shunt sewn onto Innominate Art.

Shunt connects Ao with PA.
Repair of ToF

Pre-op

Preferred Repair

Repair across the anulus.

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Repair: ToF/Pulm Atr

Pre-op

Preferred Repair

Pulm atresia requires conduit.
Cyanotic Lesions

Tetralogy of Fallot
* Transposition of the Great Arteries
* Total anomalous pulmonary venous connection

Tricuspid atresia (*sometimes*)

Truncus arteriosus

* When the connections are not normal (RA-RV-PA etc), R-to-L and L-to-R are ambiguous and should not be used.
When the AO arises from the RV, AND, The Pulmonary Artery Arises from the LV,

You have Transposition of the Great Arteries (TGA).
TGA - Definition

Normal  TGA  ?TGA ?DORV

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TGA - Echo Diagnosis
Part 1 of 2

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TGA - Echo Diagnosis
Part 2 of 2

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Unsurvivable condition

IV lines are dangerous: Clots, bugs, bubbles go to brain.

Usual TGA physiology
TGA - Chest X-ray

What makes this *classic* for TGA?

1. Increased $Q_p$
2. Enlarged heart
3. Egg-on-a-string shape (narrow mediastinum)
4. No pulm process.

In a *blue* neonate without much distress.
TGA cannot survive on the PDA

Ductus can only flow one of two directions.

1. DAO-to-PA
2. PA-to-DAO

How does oxygenated blood get to the AAO?!
TGA and PGE

Dilates PDA (& $\rightarrow \downarrow$ PVR)

$\uparrow$ DAO-to -PA flow

$\uparrow Q_p$

$\uparrow$ LA volume

$\uparrow$ pO$_2$ in RA, RV, and thus AAO.

$\uparrow$ Shunt LA $\rightarrow$ RA
Clinical Algorithm in TGA

Simplest of all.

- Diagnosis by echo (in experienced hands)
- BAS occasionally needed (usually not)
- Repair within first month of life
- Concurrent repair of associated lesions
  (VSD; Coa; PDA; …PS)
TGA - Repair

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Presentation-TAPVC

- Variable degree of cyanosis
- +/- murmur. $S_2$: wide-to-single
- Often isolated (non-syndromic)
- CXR: $\uparrow$ to $\uparrow\uparrow Q_p$ & *variable* hrt size

Nine month (!) old

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Embryology of PVs/LA (1 of 3)
Embryology of PVs/LA (2 of 3)
Embryology of PVs/LA (3 of 3)
Supracardiac TAPVC

Old technology

Ancient Technology
Physiology: Blood flows to the path of least resistance.

1. **Blue** blood $\rightarrow$ RA $\leftarrow$ SVC

2. **Red** blood $\star$ LA $\rightarrow$ Inn. Vn

   Systemic vein (VV)

3. How do LA/LV/Ao get blood??

   **Ans:** Via the PFO.

4. What determines $Q_p$, therefore filling of RA and the **push** for blood to go from RA to LA???

   **Ans:** Net (!) PVR.
Infracardiac TAPVC

Physiology
1. PVR starts to ↓.
2. ↑ Blood into lungs
3. Obstruction!... The Liver.
4. Blood trapped in lungs
5. Large $Q_p$ NOT → RA.
6. RA→LA shunt NOT large
7. Results:

Low $Q_p$ → severe cyanosis.

↓ RA vol. → ↓ low LV filling → ↓ CO

ACIDOSIS!
Surgical Repair

Finish constructing the heart.

(Do not touch pulmonary veins.)

- Connect CPV to LA.
- Ligate unnecessary vein.
- Reposition atrial septum
“Tricuspid Atresia”

Generally applied to all hypoplastic right heart syndromes:

- Pulmonary atresia with intact ventricular septum (PAcIVS)
- Tricuspid Atresia
- Unbalanced A-V Canal
- Ebstein’s anomaly
- VSD ?
- Great Arteries ?
- Semi-lunar valve stenosis ?
“Tricuspid Atresia”

- Lesions with wide variety of pathology & physiology
- Only common element: obstruction to flow (usually $Q_p$)
- Must start long-term planning in NICU
- Almost all will ultimately have Fontan repair
- Three lesions discussed
  - PAcIVS
  - Tric Atr - sml VSD, NRGA
  - Tric Atr - small VSD & TGA
Pulm Stenosis v. Atresia

The Valve in critical PS

Pulmonary Atresia
Major features

- Mandatory atrial R→L shunt
- Ductal-dependent Q_p
- Variety of abnormalities of TV
- Hypoplasia & hypertrophy of RV
- RVp > or >> LVp
- Well-formed PA system
- May have:
  - RV-coronary artery connections
  - DAO-PA collaterals
Tricuspid atresia with

Small VSD & NRGA

Anatomy & Physiology

- Small RV gives rise PA
- Many obstructions to $Q_p$
- Ductal-dependent $Q_p$
- NEEDS:
  - Good ASD
  - Reliable source of Low pressure $Q_p$
Tricuspid atresia with Small VSD & TGA

Anatomy & Physiology
- Small RV gives rise to AO
- Small AAO & Coa (usually)
- Pulmonary Art Hypertension
- Similar to HLHS
- Ductal-dependent Qs
- NEEDS Good ASD
- Low pressure in PAs
- Unobstructed Qs
Tricuspid atresia - Clinical algorithms

- Approach driven by *precise* anatomy/pathophysiology
- Often do not need diagnostic cath, but may need BAS
- Assure adequate outflows (*plural*):
  1. From: RA &/or RV
  2. TO: Systemic and Pulmonary arteries
- Prepare for future events, i.e., cavo-pulmonary & Fontan
Cavo-pulmonary Anastomosis

Why not do in neonates? (Why do shunt first?)

We have learned NOT to do this anymore.

“Logic = a way to err with confidence.”
Late Follow-up

1985

Tricuspid atresia
S/p shunt

2002 Junior Olympics qualifier

Only has one ventricle

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Truncus Arteriosus

Defined as a congenital cardiac defect where, above a mal-alignment VSD, a single arterial trunk gives rise to all THREE arterial circulations: Systemic, Pulmonary, Coronary.

Truncus is subdivided into four types.
Types of Truncus
Repair of Truncus

Conus

RPA

LPA

MV

Truncus
Type I

VSD

Patch
Repair of Truncus

Incision In RVOT

Transect PAs

RV-PA valved conduit

Close AAO
“Take-Away”
From Five T’s

✧ Tetralogy…is all about the conus
✧ TGA: counter-intuitive, including PGE
✧ TAPVC: sickest, but curable children
✧ Tricuspid atresia: infinite variations
✧ Truncus: PA & truncal valve anatomy