Discordant Atrioventricular Connections

Morphology-Imaging-Surgical Correlation

Christopher Z Lam, MD

Department of Diagnostic Imaging

The Hospital for Sick Children and University of Toronto, Canada
No disclosures
Outline

➢ Introduction to Discordant AV Connections

➢ Congenitally Corrected Transposition of the Great Arteries (ccTGA)
  ➢ Typical Morphologic Features
  ➢ Conduction Axis
  ➢ Associated Anomalies
    ➢ VSD
    ➢ Tricuspid Valve Lesions
    ➢ mLVOTO/Pulmonary Stenosis
    ➢ Coronary Anomalies
  ➢ Atrial Situs
  ➢ Ventricular Relationship
  ➢ Surgical Strategy

➢ Other forms of Discordant AV Connections
Atrioventricular Connections

Concordant AV Connections

Discordant AV Connections
Atrioventricular Connections

Discordant AV Connections

Concordant AV Connections

Discordant AV connections = anatomic alignment, not a specific disease
ccTGA

- Most discordant AV connections will be ccTGA
ccTGA = Discordant AV and VA Connections
ccTGA

**Typical Features (Not Always)**

- Mitral and pulmonary valve fibrous continuity
- Subaortic infundibulum
  - No subpulmonary infundibulum
- Parallel outflows/arterial trunks, aorta anterior & left of PA (L-TGA)
- Larger membranous septum than normal
  - Especially when ventricular septum is intact
- Abnormal AV node position
ccTGA in Short Axis

Basal SAX

Mid SAX

Ao
PA
MV
TV
mRV
mLV
MV-PV Continuity / Subaortic Infundibulum
MV-PV Continuity / Subaortic Infundibulum
Great Arterial Relationship

Typical orientation in ccTGA (not always)
L-TGA / Parallel Arterial Trunks
ccTGA: Conduction Axis
What is the typical anatomy of the conduction axis in ccTGA?

- Wedged PA leads to atrioventricular septal malalignment gap
  - Filled with large membranous septum or pmVSD

- Functional AV node in anterolateral position
  - (regular node hypoplastic at apex of triangle of Koch)

- Conduction axis courses underneath PV annulus, anterosuperior to membranous septum

- Risk of AV block
  - Congenital, post-op, or progressive (fibrosis)
  - 1.3 – 2.0% annual risk of complete heart block

What is the typical anatomy of the conduction axis in ccTGA?

- Wedged PA leads to atrioventricular septal malalignment gap
  - Filled with large membranous septum or pmVSD

- Functional AV node in anterolateral position
  - (regular node hypoplastic at apex of triangle of Koch)

- Conduction axis courses underneath PV annulus, anterosuperior to membranous septum

- Risk of AV block
  - Congenital, post-op, or progressive (fibrosis)
  - 1.3 – 2.0% annual risk of complete heart block

Anderson RH et al. Pediatric Cardiology. 2010. 3rd Ed.
What is the typical anatomy of the conduction axis in ccTGA?
Are there other arrangements of the conduction axis in ccTGA?

Hypothesis: In PS/PA, AV node & conduction axis can be normal.

Deep PA = septal malalignment

PS/PA = no septal malalignment?

Are there other arrangements of the conduction axis in ccTGA?

➢ Separate AV nodes can connect individually to anterior & posterior conduction bundles of His

➢ Can form so-called Monckeburg sling of conduction tissue around VSD

Exceptions & rare variations exist, which can be mediated by underlying morphology
ccTGA: Associated Anomalies

- VSD, Tricuspid Valve Lesions, mLVOTO/PS, Coronary Anomalies
- Combination of associated anomalies dictate clinical presentation
How frequent are associated anomalies in ccTGA?

<table>
<thead>
<tr>
<th>Associated Anomaly</th>
<th>Frequency</th>
</tr>
</thead>
<tbody>
<tr>
<td>VSD</td>
<td>50-90%</td>
</tr>
<tr>
<td>mLVOTO / Pulmonary Stenosis</td>
<td>33-66%</td>
</tr>
<tr>
<td>Tricuspid Valve Lesions</td>
<td>90% at autopsy (33-50% significant)</td>
</tr>
<tr>
<td>Anomalous Coronaries</td>
<td>20-25%</td>
</tr>
</tbody>
</table>
What is the typical type of VSD in ccTGA?

- **Typically perimembranous VSD**
  - Subpulmonic
  - Often extends to mLV inlet
  - Conduction axis courses anterosuperior to VSD

- **Muscular VSD**
  - If subpulmonic, conduction axis also courses anterosuperior to VSD
  - If between outlets, conduction axis may course posteroinferiorly (as usual)

- **Doubly committed VSD**
  - Common in Asian populations

Anderson RH et al. Pediatric Cardiology. 2010. 3rd Ed.
Perimembranous VSD in ccTGA

- PA
- MV
- LV
- pmVSD
- anterior AV bundle
- anterosuperior LV recess
- LBB
- RV
- TV
- Ao
- inf
- RBB
- pmVSD

- LV recess
Conduction axis intimately associated with mLVOTO → at risk during resection
mLVOTO from aneurysmal fibrous interventricular membranous septum
Tricuspid Valve Lesions

- Tricuspid Valve Dysplasia
- Ebstein Malformation
- Straddling and Overriding

Risk of progressive TR that contributes to systemic RV dysfunction
Ebstein Malformation of TV in ccTGA

- Associated VSD in 75%
- Can be associated with aortic arch obstruction with severe TR
- Atrialized RV usually smaller & not as thinned compared with isolated Ebstein
- Anterior leaflet not sail-like in most cases

ccTGA – Overriding / Straddling TV – DILV Spectrum
ccTGA – Overriding / Straddling TV – DILV Spectrum
ccTGA – Overriding / Straddling TV – DILV Spectrum

Double Inlet Left Ventricle
DILV with transposition

Basal View – Inlets

Basal View – Septum
Leiden Classification for Coronary Arteries

Sinus 1 = To the right of a surgeon standing in non-coronary sinus
Sinus 2 = To the left

R = RCA
L = LAD
Cx = Circumflex
Coronaries in Discordant AV Connections

- n=46
- Typical coronary pattern: 1R2LCx
- ~50% with prominent conal branch
- 20-25% anomalous coronaries

ccTGA: Associated Lesions

- SVC
- RA
- FO
- IVC
- CS
- LV
- MV
- pm

- PA
- LV
- conduction axis
- valvar tissue tag (across pmVSD)
- MV-PV continuity
- fine trabeculations

* pmVSD
ccTGA: Associated Lesions

- RV
- LA
- S
- A
- Ao
- RV
- S
- P
- inf
- Ebstein TV
- coarse trabeculations
- mVSD
- pmVSD
- inf
- mVSD
- RBB
- S
- P
- Ebstein TV

(Labeled anatomical parts and lesions in heart diagrams)
ccTGA: Atrial Situs
How common are situs abnormalities in ccTGA?

- Atrial situs inversus in ~10%
- Dextrocardia / mesocardia with situs solitus also common
What is the significance of inversus in ccTGA?

- n=38
- 8 situs inversus
- 1/8 complete AV block
  - More often normal conduction axis
- 0/8 Ebstein TV

↓ atrial arrhythmia, TR, & systemic ventricular dysfunction in inversus
What is the significance of inversus in ccTGA?

Situs inversus ccTGA is not just the mirror image of situs solitus ccTGA
ccTGA: Ventricular Relationship
Abnormal ventricular relationships can be seen in ccTGA

Twisted AV Connection / Criss-Cross Heart
- rotation of ventricular mass

Superior-Inferior Ventrices
- tilting of ventricular mass
ccTGA with twisted AV connection
ccTGA: Surgical Strategy
What are the main determinants of the natural history of ccTGA?

1. Systemic mRV function
2. Systemic tricuspid valve function
3. Development of arrhythmias

Modified by impact of associated anomalies
# ccTGA: Surgical Strategy

<table>
<thead>
<tr>
<th>Surgical Strategy</th>
<th>Description</th>
<th>Issues</th>
</tr>
</thead>
</table>
| Physiologic (“Classical”) Repair | Repair all associated anomalies  
➢ Close VSD  
➢ Relieve LVOTO or LV-PA conduit  
➢ Repair/replace tricuspid valve | ➢ Systemic mRV failure  
➢ Progressive TR |
| Anatomic Repair           | Double-Switch  
➢ Atrial Switch (Mustard/Senning)  
➢ Arterial Switch  
➢ PS/PA: Rastelli, Nikaidoh, REV | ➢ Suitability for LV re-training  
➢ Poor success > 2 years-old  
➢ Systemic mLV failure  
➢ Atrial baffle / RV-PA obstruction  
➢ Neo-aortic regurgitation |
| Palliation                | ➢ PA Band: theoretically ↓ TR  
➢ Fontan: ↓ reintervention rates, ↓ complete heart block  
➢ 1½: anatomic repair + BCPC to unload mRV, ↓ baffle obstruction, theoretically ↓ strain on mLV | ➢ Specific to chosen palliation |
Other forms of Discordant AV Connections
Other forms of Discordant AV Connections

**Discordant AV Connections with Pulmonary Atresia**
- Relationships similar to ccTGA but with pulmonary atresia
- With VSD: can baffle mLV to Ao
- With IVS: hypoplastic mLV $\rightarrow$ single ventricle pathway

**Discordant AV Connections with Double Outlet Right Ventricle**
- Usually with malposed great arteries and PT close to VSD; PS common
- Therefore, very similar to ccTGA with VSD

**Discordant AV Connections with Concordant VA Connections**
- Variable infundibular morphology
- Physiology similar to transposition of the great arteries $\rightarrow$ atrial switch
Discordant AV Connections with Concordant AV Connections
Summary

➢ Most discordant AV connections will be ccTGA

➢ Morphology informs physiology & natural history of ccTGA:
  ➢ Systemic RV failure
  ➢ Systemic tricuspid valve regurgitation
  ➢ Development of arrhythmia and AV block

➢ Modified by associated anomalies:
  ➢ VSD: typically pmVSD, conduction axis runs anterosuperior
  ➢ mLVOTO/PS: conduction axis at risk during repair
  ➢ TV dysplasia / Ebstein malformation: TR is long-term risk factor
  ➢ Coronary anomalies: may impact suitability for arterial switch
  ➢ Inversus: more often normal conduction & less TV lesions = better prognosis

➢ Understanding morphology helps determine optimal surgical strategy
Thank you!
christopher.lam@sickkids.ca

Special Acknowledgements
Shi-Joon Yoo, MD PhD
Konstantin Krutikov, MD PhD
David Chiasson, MD