CONOTRUNCAL ANOMALIES
A Case Based Review

Beverley Newman M.D.
Professor of Radiology
Lucile Packard Children’s Hospital at Stanford University

Thanks to Frandics Chan for supplying several cases
The outflow tract (conotruncus) of the tubular embryonic univentricular heart begins as a common outlet. Spiral flow through the arteriovenous connection and differential conal growth result in the creation of separate left and right ventricular outflow tracts which normally connect to the aorta and main pulmonary artery respectively.
Conotruncal Anomalies

- Transposition of the Great Arteries
- Double Outlet Right Ventricle
- Truncus Arteriosus
- Tetralogy of Fallot
Aberrant coronary arteries associated with conotruncal anomalies

- Surgically relevant
  - Aberrant LAD anterior to RVOT (4% of TOF)
  - Aberrant coronary in TGA

Courtesy Frandics Chan
Q1. The most common plain film radiographic appearance of complete transposition of the great arteries in the newborn is:

1. Normal
2. Cardiomegaly and shunt vascularity
3. Egg on a string
4. Bulge of left superior heart border
5. Coeur en Sabot
Complete Transposition of the great Arteries (D-transposition)
Congenitally Corrected Transposition (L-Transposition) (dextrocardia 25%)
Truncus Arteriosus
Tetralogy of Fallot
Q2 This configuration of the aortic arch is characteristic of which of the following congenital cardiac anomalies

1. Truncus Arteriosus
2. Transposition of the Great Arteries
3. Congenitally Corrected Transposition
4. Double Outlet Right Ventricle
5. Tetralogy of Fallot
Transposition of the Great Arteries

- Discordant connections of the ventricles and great arteries
- CCTGA- also atrioventricular discordance: associated with VSD, ASD, tricuspid abnormality (Ebstein’s), pulmonic stenosis, conduction abnormalities
- TGA – associated with VSD, ASD, PDA, pulmonary stenosis/atresia, Ao coarctation/interruption, AV valve abnormalities and RV hypoplasia or single ventricle
Transposition of the Great Arteries (TGA)
TGA with VSD and PDA
Q3 Which of the following surgical procedures are used to repair Double Outlet Right Ventricle

1. Arterial switch
2. Rastelli
3. VSD closure and PA outflow tract patch
4. RV to PA conduit
5. Fontan
6. All of the above
Double Outlet Right Ventricle

- Subaortic VSD (50%)
- Subpulmonic VSD (30%) (Taussig-Bing)

Doubly Committed VSD
- Remote/uncommitted VSD

Frank et al. Radiographics: 30;2010
Double Outlet Right Ventricle

- Both great arteries arise predominantly from the right ventricle. Both great arteries have subvalvular conal muscle.
- Associated with VSD, pulmonary stenosis or atresia, TGA, mitral stenosis or atresia, hypoplastic LV, straddling AV valves, subaortic/aortic valve obstruction/coarctation.
- May physiologically resemble TOF (subaortic VSD and PS); Transposition (Subpulmonic VSD); VSD (subaortic VSD); Hypoplastic or univentricular heart.
Double Outlet Right Ventricle

- **Surgery:**
  - Subaortic VSD – Rastelli tunnel through VSD connecting LV with Ao
  - RVOT obstruction may necessitate patch similar to TOF repair or RV – PA conduit
  - Subpulmonary VSD – Switch and VSD patch
  - Doubly committed VSD- Rastelli or switch depending on LV /Ao relationship
  - Remote VSD – may need single ventricle palliation and eventual Fontan
DORV with subaortic VSD and PS – like TOF
DORV with Transposition and subpulmonic VSD

- Ao: Aortic Valve
- PA: Pulmonary Artery
- VSD: Ventricular Septal Defect
- Outlet Septum
- Muscular Septum
- Mus. VSDs
- Conal Muscle
- Aortic Valve
DORV WITH SUBAORTIC VSD AND PS – RASTELLI REPAIR

DECR RV FX, TR

Gaca, A Radiology 2008. 248:1;44
13yo Asplenia, abnormal situs, dextrocardia, unbalanced AVC with hypoplastic LV, DORV, Lt sided Glenn/Fontan

FONTAN- COMMON PATHWAY FOR VERY COMPLEX HEART DISEASE
Q5 Which of the following syndromes are most often associated with Truncus Arteriosus

1. Di George Syndrome
2. Heterotaxy with Asplenia
3. Alagille Syndrome
4. Williams Syndrome
5. Down syndrome
TRUNCUS ARTERIOSUS

- Single arterial vessel arises from the heart which gives rise to aortic, pulmonary and coronary arteries
- Association with VSD, truncal valve abnormality, RAA, interrupted aortic arch (11-14%), abnormalities of the mitral valve and anomalous pulmonary venous connections
Truncus Arteriosus Type 1 - 22q11 deletion
(~ 35% of cases)
Alagille Syndrome
WILLIAMS SYNDROME
3d old
Heterotaxy with right Isomerism - Asplenia
Q4 Which of the four features of Tetralogy of Fallot is the most consistently present but also the most variable in severity

1. Overriding Aorta
2. Ventricular Septal Defect
3. Right Ventricular Hypertrophy
4. Pulmonary Outflow Obstruction
Tetralogy of Fallot

- Most common cyanotic congenital heart defect
- Anterior malposition of the conal septum results in: overriding aorta; pulmonary outflow obstruction; ventricular septal defect and right ventricular hypertrophy
- Associated with VSD, Right aortic arch (25%), pulmonary atresia, PDA, major aorticopulmonary collaterals, ASD, CAVC
TOF post repair

RPA 2.1cm
LPA 1.3*1 – 2.5cm
TOF PA MAPCA’S
Tetralogy of Fallot, absent pulmonary valve
Conotruncal Anomalies

Summary

- **Transposition of the Great Arteries**
  The aorta moves upward and to the right instead of pulmonary artery

- **Truncus Arteriosus**
  Truncal Septum fails to develop

- **Double Outlet Right Ventricle**
  Arterial trunk divides but stays over on the right side

- **Tetralogy of Fallot**
  Truncal septum deviates anteriorly
References


5. Partridge J. Cardiac Embryology for Imagers. www.cardiacradiology.co.uk/presentations/cardiac_embryology.ppt


References


