Double outlet right ventricle (DORV)

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Double outlet right ventricle

• Complex lesion, represents 3% of CHD seen in fetus

• Both Great Arteries arise completely or predominantly from the morphological right ventricle

• Associates with increased NT, genetic and extracardiac anomalies
Double outlet right ventricle

- Double outlet right ventricle is a spectrum of abnormalities
  - almost always has a VSD
  - heterogeneous with respect to size and position of VSD and relationship of GA
  - individual anatomy dictates surgical approach and outcome

- It poses a number of challenges
  - Definition
  - Anatomic Variability (*incl. normal or abnormal AV connections*)
  - Surgical options
What is in the name?
Double outlet right ventricle

DORV

TOF type DORV

“Simple” DORV
- Always VSD:
  - subaortic, subpulmonary or uncommitted;
  - +/- PS

Complex DORV
- Unbalanced Ventricles
  - AVSD (often RAI or LAI)
  - MV atresia
- Coarctation, IAA
- Discordant AV connections
- Criss-cross heart
Double outlet right ventricle
What is in the name?

Normal atrial situs
Patent, concordant AV connections

DORV

Both GA (parallel) from RV
Subpulmonary, subaortic or uncommitted VSD

Overriding aorta
“50% rule”
TOF type DORV

Repair depends from GA relationship with VSD

Anatomical repair-
VSD closure & PS release

LV to Ao baffle
ASO with baffling VSD to neo-Ao
Rarely BV repair is unfeasible
“Tetralogy of Fallot type” DORV and Tetralogy of Fallot have the same surgical approach

- True override is independent of septal axis
  - postnatally is assessed relative to chord of circle in short axis*

*BR Wilcox, AC Cook, RH Anderson. Surgical anatomy of the heart, 2004
Double Outlet Right ventricle v. Tetralogy of Fallot

- Perimembranous VSD, **overriding aorta**
- Pulmonary artery < aorta (PA st)
Double outlet right ventricle (DORV)

This is not a single diagnosis but a description of many different ones!
DORV- both Great Arteries arise completely from the morphological right ventricle

- VSD closer to the aorta
Double outlet right ventricle

- Subpulmonary VSD
Double outlet right ventricle

• ?Noncommitted VSD
Cross-sectional sweep of thorax

Traditional approach

- 2D echocardiography

- Echocardiographic “sweeps” used to determine
  - Position of VSD
  - Position of great arteries
  - Straddling valves
  - Outflow tract obstruction
### Echocardiographic features of DORV with normal atrioventricular connections

<table>
<thead>
<tr>
<th>Feature</th>
<th>Normal heart</th>
<th>DORV</th>
</tr>
</thead>
<tbody>
<tr>
<td>4-chamber view</td>
<td>Normal</td>
<td>Normal/ VSD/straddling AV valves</td>
</tr>
<tr>
<td>LVOT</td>
<td>Aorta directed towards right shoulder</td>
<td>Highly variable</td>
</tr>
<tr>
<td>Relationship of GA</td>
<td>Aorta and PA cross each other with PA anterior</td>
<td>Highly variable: normally related, transposed</td>
</tr>
<tr>
<td>Size of GA</td>
<td>Aorta similar to PA</td>
<td>Highly variable: equal sized, Aorta larger than PA or vice versa</td>
</tr>
<tr>
<td>3-VTV</td>
<td>Aortic arch and ductal arch meet in a “V” shape</td>
<td>Aortic arch left or right sided</td>
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Schematic of AV valves and great arteries in DORV

Courtesy of JM Simpson
Favourable routing

Courtesy of JM Simpson
Unfavourable Inlet VSD

Courtesy of JM Simpson
Favourable VSD for “routing” but unfavourable GA position – more favourable towards switch

Courtesy of JM Simpson
Published work

Three-dimensional fetal echocardiography for prediction of postnatal surgical approach in double outlet right ventricle: a pilot study

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A Systematic Three-Dimensional Echocardiographic Approach to Assist Surgical Planning in Double Outlet Right Ventricle

More equivocal
DORV- traps
DORV - aorta anterior, parallel GAs, MV straddle

Courtesy of JM Simpson
Mitral atresia with DORV
Mitral atresia with DORV, ?IAA
## Double outlet right ventricle (DORV)

<table>
<thead>
<tr>
<th>Associations</th>
<th>Postnatal management according to anatomy</th>
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<tbody>
<tr>
<td>Extracardiac and genetic anomalies</td>
<td>Baffling of LV to aorta using a patch which also effects closure of VSD</td>
</tr>
<tr>
<td>22q11.2 deletion</td>
<td>• enlargement of VSD may be needed</td>
</tr>
<tr>
<td>aneuploidies</td>
<td>Arterial switch operation and VSD closure</td>
</tr>
<tr>
<td>• e.g T13, T18</td>
<td>Not possible to fully repair</td>
</tr>
<tr>
<td>• VACTREL and CHARGE</td>
<td>• Anatomy of VSD</td>
</tr>
<tr>
<td></td>
<td>• Size of ventricles and function of AV valves</td>
</tr>
<tr>
<td></td>
<td>• Position of great arteries</td>
</tr>
<tr>
<td></td>
<td>Prognostication difficult</td>
</tr>
</tbody>
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Conclusions

- Systematic, thorough approach to all segments essential
- Understand the surgical considerations
- Some of the newer modalities may assist in difficult cases
THANK YOU!

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