Viscero-atrial Situs

- Situs refers to the position of the atria and viscera relative to the midline
- Three types: solitus, inversus, ambiguous
### Visceral Situs

<table>
<thead>
<tr>
<th>Situs solitus</th>
<th>Situs inversus</th>
<th>Situs ambiguous</th>
</tr>
</thead>
<tbody>
<tr>
<td><img src="image1" alt="Diagram" /></td>
<td><img src="image2" alt="Diagram" /></td>
<td><img src="image3" alt="Diagram" /></td>
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<tr>
<td><img src="image4" alt="Diagram" /></td>
<td><img src="image5" alt="Diagram" /></td>
<td><img src="image6" alt="Diagram" /></td>
</tr>
</tbody>
</table>

### Atrial Situs

<table>
<thead>
<tr>
<th>Atrial situs solitus</th>
<th>Atrial situs inversus</th>
<th>Atrial situs ambiguous</th>
</tr>
</thead>
<tbody>
<tr>
<td><img src="image7" alt="Diagram" /></td>
<td><img src="image8" alt="Diagram" /></td>
<td><img src="image9" alt="Diagram" /></td>
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<td><img src="image11" alt="Diagram" /></td>
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Heterotaxy or Situs Ambiguous

Heteros (Greek): Other
Taxis: Arrangement
Other than the usual order or arrangement
• Situs other than solitus or inversus
• Present in 3% of neonates with CHD
• Not a specific disease, but a constellation of cardiac, vascular, and visceral abnormalities
• No single finding is pathognomonic

Heterotaxy – Defining features

• Situs ambiguous of the viscera
• Lung symmetry
• Atrial appendage symmetry
• Anomalous systemic venous return
• Anomalous pulmonary venous return
• (Associated intra-cardiac defects: functional common atrium, common AV canal, functional single ventricle, outflow tract obstruction )
Heterotaxy

• Tendency towards syndromic clustering of defects
  – Asplenia or right isomerism
  – Polysplenia or left isomerism
  – Single right-sided spleen

Anything that can go wrong will go wrong!!
Heterotaxy
Possibilities at each segmental level

- IVC
- Hepatic Veins
- Pulmonary veins
- SVC
- Pulmonary artery and aorta
- Intracardiac defects

Inferior Vena Cava

- Continuous IVC with tortuous/curved hepatic segment of IVC
- IVC on same side of abdominal aorta (asplenia)
- Bilateral suprarenal IVC (asplenia)
IVC and aorta on same side

Bilateral Suprarenal IVC

IVC Anatomy

<table>
<thead>
<tr>
<th></th>
<th>Asplenia</th>
<th>Polysplenia</th>
</tr>
</thead>
<tbody>
<tr>
<td>Continuous IVC</td>
<td>Almost 100%</td>
<td>20%</td>
</tr>
<tr>
<td>Interrupted IVC</td>
<td>Very rare, but reported</td>
<td>80%</td>
</tr>
</tbody>
</table>
Hepatic Veins

- To unilateral IVC
- To ipsilateral IVC
- To IVC and to atrium
- To unilateral atrium
- To ipsilateral atria
Hepatic veins

Pulmonary Venous Connections

<table>
<thead>
<tr>
<th></th>
<th>Asplenia</th>
<th>Polysplenia</th>
</tr>
</thead>
<tbody>
<tr>
<td>To LA or Common Atrium</td>
<td>34%</td>
<td>60%</td>
</tr>
<tr>
<td>To RA</td>
<td>2%</td>
<td>20%</td>
</tr>
<tr>
<td>To ipsilateral atrium</td>
<td></td>
<td>15%</td>
</tr>
<tr>
<td>To systemic vein</td>
<td>64%</td>
<td>5%</td>
</tr>
</tbody>
</table>
Pulmonary venous anomalies in heterotaxy

Superior Vena Cava

- Asplenia
  - Bilateral SVC 36% (none to coronary sinus)
  - Absent coronary sinus septum (95%)
- Polysplenia
  - Bilateral SVC 41% (61% of those have one SVC to coronary sinus)
  - Absent coronary sinus septum (28%)
  - 26% of single SVCs connect to coronary sinus
Left hepatic vein to coronary sinus, malposition of septum primum, occult polysplenia

Typical Cardiac Abnormalities

- Common AV Canal
- Single Ventricle (R>L)
- Conotruncal anomalies
- Outflow tract obstruction
- Right Aortic Arch
- Cardiac Malposition (e.g., dextrocardia)
Common AV Canal

Subpulmonary obstruction and subaortic conus in a patient with asplenia and DORV
Severe aortic and subaortic obstruction in polysplenia, s/p Norwood procedure

Heterotaxy syndrome: Imaging

- Echocardiography: primary diagnostic modality
- Excellent tool in expert hands
- Limitations
  - Narrow field of view, depends on acoustic windows
  - Limited ability to delineate extra-cardiac anatomy, especially pulmonary veins, systemic veins, arterial collaterals
  - Limited delineation of complex spatial relationships
- Pulmonary venous anatomy not delineated by echo in 42% of patients with heterotaxy (Geva T et al, Circulation 1995)
MRI approach to heterotaxy

- Two schools of thought 😊
  - ‘Throw the kitchen sink’ approach
  - ‘MRA is all you need’ approach

? RATIONAL APPROACH

Heterotaxy - Clinical Scenarios

- Neonatal period (prior to immediate palliation)
- Screening for immediate post-operative complications
- Screening prior to Glenn or Fontan procedures (prior to permanent palliation)
- Late post-operative monitoring for morphology and function (after permanent palliation)
Heterotaxy Indications for MRI (n = 50)

Prior to initial palliation:
Indications for MRI (n=18/50)
Prior to initial palliation: MRI protocol

- Temporarily stable or unstable patient
- IV access: arm or leg. UVC not preferred
- Coil: Smallest coil that covers relevant anatomy
- Sequences:
  - Cine TFE (cine GRE): axial (3-4 mm with overlap)
  - Black blood: coronal and axial (2-3 mm)
  - Gd 3d MRA (approx 1 mm isotropic)

Prior to initial palliation: Indications for MRI

Pulmonary veins

- All pulmonary veins not identified
- TAPVR insertion not visualized
- Atypical or mixed TAPVR
- Unusual morphology
Prior to initial palliation: Indications for MRI Pulmonary veins

TAPVR to left SVC with varix
Prior to initial palliation: Indications for MRI
Pulmonary arteries

- Pulmonary atresia: source of PBF
- Confluent branch pulmonary arteries in pulmonary atresia
- Systemic collaterals
- Branch Pulmonary artery stenosis

Source of pulmonary blood flow and pulmonary artery confluence
Ductal dependent pulmonary blood flow in pulmonary atresia

Unsedated newborn, Scan time: 0.35 seconds

Prior to initial palliation: Indications for MRI

- Aorta
  - Aortic stenosis or atresia
  - Screen for coarctation
  - Coronary anatomy
  - Define complex spatial relationships
Polysplenia, {A,L,I} common atrium, ventricular inversion, inversus normal great arteries, CAVC, coarctation
Asplenia, DORV, l-malposed aorta, s/p PA banding, double aortic arch

Complications following initial palliation:
Indications for MRI (n=5/50)

• Recurrent pulmonary vein obstruction after TAPVR repair
s/p TAPVR repair, PV stenosis

Conclusion

- Plan for MRI adapted to different clinical scenarios in heterotaxy
- MRI has potential to become one-stop shop in heterotaxy
- Anything that can go wrong will go wrong (well, almost!). And the eye sees only what the mind knows! So, planning and supervision essential for success.