CONGENITAL HEART DISEASE: NOMENCLATURE

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CONGENITAL HEART DISEASE: NOMENCLATURE
"FEAR NOT – MAKE IT SIMPLE"
CONGENITAL HEART DISEASE: NOMENCLATURE
"FEAR NOT – MAKE IT SIMPLE"
DISCLOSURES

• NOTHING TO DISCLOSE
OBJECTIVES

• To recognize the importance and understanding of basic terminology in CHD.
• To formulate the philosophy of the segmental analysis to describe cardiac anatomy.
• To identify the three segments of the heart and to describe their connections.
OUTLINE

• Introduction

• Segmental approach
  – Atrial arrangement / cardiac position
  – The three segments
  – Connections

• Looping

• Summary
CHALLENGES

• Congenital heart defects – very wide spectrum:
  – Various forms and combinations
  – Modification of the underlying anatomy / pathophysiology by the congenital heart surgeons!
  – Complex anatomy and morphology

• Terminology / language in CHD!
CHALLENGES

• Surgical / interventional procedures
  – Named after surgeons / physicians
  – Different types / modification of procedures for the same CHD

• Combination of different CHD

• Syndromes
Blalock Hanlon procedure
Waterston shunt
Sterling Edwards procedure
Blalock Taussig Thomas shunt
Potts shunt
Konno procedure
Baffes procedure
Senning / Mustard procedure
Kawashima procedure
Scimitar syndrome
Arterial switch procedure
Yasui procedure
Fontan procedure and its modifications
Shone complex
Eisenmenger syndrome
Holt-Oram syndrome
Williams syndrome
LEOPARD syndrome
Isomerism
Microdeletion 22q11
LEOPARD syndrome
Scimitar syndrome
Microdeletion 22q11
BEFORE YOU START......

• Surgical history
  – Surgical notes!
• Morphology / anatomy
• Pathophysiology
BEFORE YOU START......

- Surgical history
  - Surgical notes!
- Morphology / anatomy
- Pathophysiology

To understand the anatomy / connections and surgical procedures
To understand the specific long-term complications of each CHD and procedure
What Are You Looking For?
Sequential Segmental Analysis

Clear language
misunderstanding
There is a Congenital in the ER ....
?????
Everybody is confused.....
those who work with them every day
Philosophy of Segmental Analysis is Founded on MORPHOLOGY

• **Chambers** are recognized according to their **morphology**
  – Each chamber has **intrinsic features**
• The chambers are not in their anticipated location!
Segmental Approach

Atrial Arrangement / Position

Identify The Three Segments:
- (Atria) – Ventricles – Great Arteries

Define the Connections
- Atrio-ventricular / ventriculo-arterial
Position of the heart within the chest

Cardiac Position
- Levoposition
- Mesoposition
- Dextroposition

Cardiac Orientation
- Levocardia
- Mesocardia
- Dextrocardia

Dependent from many factors
- Cardiac malformation
- Mediastinal/thoracic structures
The **UNKNOWN** Patient: Sequential, Segmental Approach

S.Y. Ho, Cardiac Morphology and Nomenclature, p. 7-18
In: Gatzoulis, Webb, Daubeny (eds.)
Diagnosis and Management of Adult Congenital Heart Disease 2003
S I T U S - SIDEDNESS

- **Cardiac** Situs
- **Pulmonary** Situs
- **Abdominal** Situs
Atrial Arrangement
(Cardiac Situs)

• *Position* of the morphologic *RIGHT ATRIUM*, independent from:
  – Cardiac position
  – Cardiac orientation
  – Position of the ventricles / great arteries
Atrial Arrangement

Harmony between arrangement of the atrial appendages and thoraco-abdominal organs

Anderson RH, Paediatric Cardiology, 3rd edition, 2010; pages 3-16
Situs Inversus, Dextrocardia
Atrial Arrangement

- **Situs solitus**: Morphological RA is on the right of the morphological LA

- **Situs inversus**: Morphological RA is on the left of the morphological LA

- **Situs ambiguus**: Indeterminate sidedness in the setting of isomerism
Atrial Arrangement

- **Situs solitus**: Morphological RA is on the right of the morphological LA
- **Situs inversus**: Morphological RA is on the left of the morphological LA
- **Situs ambiguus**: Indeterminate sidedness in the setting of isomerism

Appendages: the landmark for morphologic **RIGHTNESS** and **LEFTNESS**
RIGHT ATRIAL APPENDAGE

- Triangular
- Broad base
- Pectinate muscles within the appendage extend all round the vestibule of the tricuspid valve!
Right Atrial Appendage

Right Atrium

- Appendage
  - Triangular
  - Broad base
- Terminal crest
**LEFT ATRIUM**

- **Left Atrium – Appendage**
  - Hook-shaped
  - Narrow entrance
  - Pectinate muscles are confined within the appendage

- **No terminal crest**
Left Atrial Appendage

Left Atrium
- Appendage
  - Hook-shaped
  - Narrow entrance
- No terminal crest
ISOMERISM

• Paired, *mirror-image* sets of normally single, *non-identical* organ systems
  – Atria
  – Lungs
  – Viscera
Isomerism

Paired morphologically right structures:
- Bilateral right bronchi
- Bilateral morphologic right atria
- Asplenia / transverse liver
- Other malformations

Isomeric lungs and atrial appendages abdominal organs are jumbled up

Anderson RH, Paediatric Cardiology, 3rd edition, 2010; pages 3-16
Left Isomerism
<table>
<thead>
<tr>
<th>Anatomic Feature</th>
<th>Right Atrium</th>
<th>Left Atrium</th>
</tr>
</thead>
<tbody>
<tr>
<td>Veins</td>
<td>IVC: constant</td>
<td>Pulmonary veins: variable</td>
</tr>
<tr>
<td></td>
<td>SVC/CS: variable</td>
<td></td>
</tr>
<tr>
<td>Appendage</td>
<td>Broad, triangular</td>
<td>Narrow, finger-like</td>
</tr>
<tr>
<td>Musculi pectinati</td>
<td>Many</td>
<td>Few</td>
</tr>
<tr>
<td>Terminal Crest</td>
<td>Present</td>
<td>Absent</td>
</tr>
<tr>
<td>Septal Surface</td>
<td>Septum secundum</td>
<td>Septum primum</td>
</tr>
<tr>
<td>Conduction system</td>
<td>Sinoatrial node</td>
<td></td>
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</table>
The **UNKNOWN** Patient:
Sequential, Segmental Approach

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Left Ventricle

Fibrous Continuity

Right Ventricle

Ventricular Crest
Ventricular Crest
Landmark for RV Morphology

- Muscular crest in the RVOT intervening between tricuspid and semilunar valve
- Demarcation the junction between the outlet septum and the pulmonary infundibulum
Normal heart

**Apical attachment**

of the septal tricuspid valve leaflet

Situs inversus ccTGA
VENTRICULAR SEGMENT

Tricuspid Valve / RV
- Apical attachment of the septal leaflet
- Ventricular crest
- Extensive septal leaflet tethering to the septum
- Moderator band
- Course trabeculations

Mitral Valve / LV
- No tendinous chords tethering to the septum
- Fibrous continuity to the semilunar valve
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The ARTERIAL Segment

Aorta

Common Arterial Trunk

Pulmonary Artery
The ARTERIAL Segment

- Aorta
  - Coronary arteries
  - Branches to the head

- Pulmonary arteries
  - Bifurcation to the *left* and *right* lung
The ARTERIAL Segment

• Common arterial valve
• Blood supply to:
  – Coronary arteries
  – Pulmonary arteries
  – Systemic arteries

Anderson RH, Paediatric Cardiology, 3rd edition, 2010; pages 3-16
Segmental Approach

Atrial Arrangement / Position

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CONNECTION vs DRAINAGE

CONNECTION
• *Anatomic* term
• Link between two structures
  – Veno-atrial
  – Atrio-ventricular
  – Ventriculo-arterial

DRAINAGE
• *Hemodynamic* term
• Blood flow direction
Left Isomerism
CONNECTIONS

Isomeric Connections

Concordant Connections

Discordant Connections

Univentricular Connections
AV CONNECTIONS

Discordant Connections

Concordant Connections
ISOMERIC CONNECTION

Isomeric right appendages

Right hand ventricular topology

Isomeric right appendages

Left hand ventricular topology
UNIVENTRICULAR Connections
THREE TYPES

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Double Inlet Left Ventricle

Malalignment of the atrial and ventricular septum

Concordance without overriding
Concordance with overriding
Double inlet LV with overriding
Double inlet LV without overriding

50% Rule

Double Inlet Left Ventricle
OVERRIDING

VS

STRADDLING
STRADDLING AV-valve

- Anomalous insertion of tendinous cords of papillary muscles into the contralateral ventricle (VSD!!!)
• *Malalignment* of the annulus of one AV-valve or semilunar valve relative to the ventricular septum
Freedom, RM, Mawson JB, Yoo SJ, Benson LN (eds.)
In: Textbook of Angiocardiography, 1997, page 110
DOUBLE OUTLET RIGHT VENTRICLE

• Both great arteries arise predominantly from the one ventricle
  – DORV
DOUBLE OUTLET RIGHT VENTRICLE

Malalignment of the outlet septum relative to the remainder of the interventricular septum

50% Rule

LOOPING
SV, Left horn sinus venosus
A, Future atrium
LV, Future left ventricle
RV, Future right ventricle
CT, Future conotruncus
A1, First aortic arch arteries
DAo, Dorsal aorta

Foregut
Dorsal
Cranial → Caudal
Ventral

Options
PACCSAP
AMERICAN COLLEGE OF CARDIOLOGY
Go To
D – Loop of the Heart Tube

(Audio Ref 9)

SV, Sinus venosus
A, Primitive atrium
LV, Presumptive left ventricle
RV, Presumptive right ventricle
CT, Conotruncus
S, Aortic sac
A1, First aortic arch arteries
A2, Second aortic arch arteries forming
fg, Foregut
DAo, Dorsal aorta

D-Loop (180°)
D-Loop

- *Inflow portion of the morphologic RV* lies to the *right* of the morphologic *LV*

L-Loop

- *Inflow portion of the morphologic RV* lies to the *left* of the morphologic *LV*
Situs solitus
ccTGA
L – LOOP
(95%)

Situs inversus
ccTGA
D – LOOP
(5%)
ccTGA

Situs solitus – 95%

Situs inversus – 5%

Wilcox B, et al. Surgical Anatomy of the Heart; 2005; p. 251
PUT IT TOGETER!
Atrial Arrangement?
SITUS SOLITUS

LAA

LA
SITUS SOLITUS
Ventricular Segment?
DISCORDANT AV CONNECTION
ARTERIAL SEGMENT?
ARTERIAL SEGMENT?
SITUS SOLITUS - CCTGA
SUMMARY

Knowledge and expertise
• Terminology / Anatomy / Morphology
• Surgical Procedures

Long-term complications
Segmental analysis makes CHD simple
WYSIWYG

What You See Is

What You Describe
“You need to know the journey of the red blood cells”
ACKNOWLEDGEMENT
CARDIAC ANESTHESIA TEAM