The Sequential Segmental Analysis of the Malformed Heart

A Pathological Categorization

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• As early as the 1970’s. (Shinebourne et al in 1976)


• Some aspects remain controversial.

• 2 major schools.
In all Anderson’s publications, his main rival, Richard Van Praagh from Boston, appears to drive Anderson to nearly obsession. These two leviathans have been battling for years over the language & methods used to describe CHD, & between them, they have made enormous contributions to our understanding of cardiac pathology & subsequently to the clinical practice of pediatric cardiology. As a result, the pediatric cardiology community has been divided into self-proclaimed Andersonians & Van Praaghians, although most of us probably fall somewhere in the middle.

Brent J. Barber, M.D.
David J. Sahn, M.D.
Oregon Health & Science University
Portland, Oregon USA
... describes CHD through his system of "sequential segmental analysis," which categorizes recognizable anatomical facts, avoiding speculative embryological assumptions.
1. Atrial Component.
2. Ventricular Component.
3. Arterial Component.

1. A-V Junction.
2. V-A Junction.

3 Segments

2 Junctions
Lev (1954) proposed that the chambers of the heart should be called by *what* they were, rather than by *where* they were; that each chamber had characteristic that identified it, no matter where it was in relation to the rest of the heart.
8 Steps are adopted:

1. Determine Atrial Arrangement.
3. Analysis of A-V Junction.
5. Analysis of V-A Junction.
6. Analysis of Cardiac Position.
8. Associated Malformations.

So, Where are we exactly?
Step 1:

1. Determine **Atrial Arrangement.**

2. Determine **Ventricular Mass Structure.**
3. Analysis of **A-V Junction.**
4. Analysis of **Arterial Segment.**
5. Analysis of **V-A Junction.**
6. Analysis of **Cardiac Position.**
7. Analysis of **Other Body Organs.**
8. **Associated Malformations.**
The morphology of a given chamber should be determined on the basis of its most constant component.
<table>
<thead>
<tr>
<th><strong>MRAA</strong></th>
<th><strong>MLAA</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>Broad + Traingular + Wide-based</td>
<td>Tubular + Narrow + Small junction</td>
</tr>
<tr>
<td>Pectinate ms extend all around the vestibule of A-V Junction.</td>
<td>Pectinate ms are confined to the ant quadrant of the vestibule.</td>
</tr>
<tr>
<td></td>
<td>Post part of vestibule &amp; P venous compartment are smooth</td>
</tr>
</tbody>
</table>
4 possible Topographic Patterns

LATERALIZATION

SITUS SOLITUS (Usual)
- IVC & AO on opposite sides of spine
- MRA on same side as IVC

SITUS INVERSUS (Mirror Image)

ISOMERISM

RA-ISOMERISM
- IVC & AO on same side of spine (Lt)
- IVC slightly anterior

LA-ISOMERISM
- AO is midline
- IVC: interrupt & contin via Azygos system poster to AO
Step 2:

1. Determine *Atrial Arrangement*.

2. Determine *Ventricular Mass Structure*.

3. Analysis of *A-V Junction*.
4. Analysis of *Arterial Segment*.
5. Analysis of *V-A Junction*.
6. Analysis of *Cardiac Position*.
7. Analysis of *Other Body Organs*.
8. *Associated Malformations*. 

*So, Where are we exactly?*
Definition:

Ventric mass extends betn A-V → V-A Junctions

Components:

1. INLET:
A-V junction → distal attachment of the tension apparatus

2. APICAL TRABECULAR

3. OUTLET:
Which supports the arterial valve
The Ventr is diagnosed on basis of **Apical Trabecular Component ONLY**

What about the AV-valve?
The valve follows the ventricle

Not

The ventricle follows the valve
The **M valve** – by definition is a bi-leaflet valve.

The **T valve** has chordal attachments to the IVS.

The **T valve** has a slightly lower insertion on the IVS compared to the M valve.
Step 3:

1. Determine *Atrial Arrangement*.
2. Determine *Ventricular Mass Structure*.

3. Analysis of *A-V Junction*.

4. Analysis of *Arterial Segment*.
5. Analysis of *V-A Junction*.
6. Analysis of *Cardiac Position*.
7. Analysis of *Other Body Organs*.
8. *Associated Malformations*. 

*So, Where are we exactly?*
Analysis of A-V Connexion

Type of connexion (Myocardial Junction) (Union or No-Union)

Bi-Ventricular AV connexion

Mode of connexion (Valve Morphology)

Univentricular AV Connexion

Uni-Atrial Univentricular Connexion

Separate AV valves

Common AV valve
Uni-ventricular A-V connection

Uni-Atrial
Uni-ventricular A-V connection

Bi-ventricular A-V connection

Uni-ventricular A-V connection

Uni-ventricular A-V connection
One Atrium is **NOT** connected directly with the Ventricular mass

as in:
**Situs Solitus**
- LA connected to Domin LV
- Absent Rt A-V connex

= **Tr Atresia**
Analysis of A-V Connexion

Type of connexion (Myocardial Junction) (Union or No-Union)

Mode of connexion (Valve Morphology)

Separate AV valves
Common AV valve
Valve Morphology

2 separate valves

Common valve

Stenotic

Regurgitant

Imperforate (R or L)

Overriding

Straddling

partial

complete
Absent A-V connex & Imperforate Valve

Hemodynamically: same
Morphologically: different

Absent Connection

Imperforate valve
Absent connexion

Imperforate valve
2 perforate valves

1 common valve

2 valves:
Imperforate Rt

All have Concordant A-V connexion
Valve Morphology

2 separate valves

- Stenotic
- Regurgitant
- Imperforate (R or L)

Common valve

- Overriding
- Straddling

partial

complete
OVERRIDING / STRADDLING
OVERRIDING / STRADDLING

Overriding right AV valve without straddling

Straddling right AV valve in DILV

Ventricular septum
Step 4:

1. Determine *Atrial Arrangement*.
2. Determine *Ventricular Mass Structure*.
3. Analysis of *A-V Junction*.
4. Analysis of *Arterial Segment*.
5. Analysis of *V-A Junction*.
6. Analysis of *Cardiac Position*.
7. Analysis of *Other Body Organs*.
8. *Associated Malformations*.
- no intrinsic feature
to differentiate AO from PT

These are distinguished on basis of **branching**

<table>
<thead>
<tr>
<th>1. AO:</th>
<th>3. <strong>Common Art Trunk:</strong></th>
</tr>
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<tbody>
<tr>
<td>asc. Portion → arch + &gt; 1 cor art + bulk of systemic vessels</td>
<td>Single great art that - gives rise in its asc portion to cor &amp; PA’s, - then continues as desc AO to give systemic arts.</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>2. P.T. :</th>
<th>4. <strong>Solitary Art Trunk:</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>&gt; PA. If 2 → a bifurcating vessel.</td>
<td>with P Atresia &amp; - complete absence of PT &amp; - complete absence of intra-peric PA’s, The lung supply is via: Duct(s) Collateral arts.</td>
</tr>
</tbody>
</table>
Step 5:

1. Determine *Atrial Arrangement*.
2. Determine *Ventricular Mass Structure*.
3. Analysis of *A-V Junction*.
4. Analysis of *Arterial Segment*.
5. Analysis of *V-A Junction*.
6. Analysis of *Cardiac Position*.
7. Analysis of *Other Body Organs*.
8. *Associated Malformations*. 
Analysis of the V-A Junction

1. Type of V-A Conexión:
   - 2 semilunar valves
   - Common valve

2. Infundibular Morphology

3. Valve Morphology

4. Arterial Trunk relation
1. Type of V-A Connexion

2 Arterial Valves

- Conc V-A Connexion.
- Discord V-A Connexion.
- D.O. V-A Connexion
  (>50% of an Art Trunk is committed to a ventr)

Common Arterial Valve + Comm Arter Trunk
Single Outlet from heart

Can exist in:
2. AO + atretic PA
3. PT + atretic AO
4. Solitary Art Trunk: absent PT & can’t differ existing trunk: AO or Comm Trunk
2. Infundibular Morphology

- Sub-P Infundibulum
- Sub-AO Infundibulum
- Bilater Infundibulum
- Bilat deficient Infundibulum
3. Valve Morphology

- **Stenotic**
- **Incompetent**
- **Imperforate:** but with:
  - recognizable connexion bet ventr mass & Art Trunk
  - Balloons into Art during systole
- **Overriding:** valve is assigned to the ventr which supports its greater circumf
Straddling **NEVER** occurs in semilunar valves
4. Arterial Trunk relationship

Both trunks leave the heart in either:

- Spiral fashion
- Parallel fashion

Describe the origin of AO in relation to PT:

- Rt/Lt coordinates
- Ant/Post coordinates
Step 6:

1. Determine Atrial Arrangement.
3. Analysis of A-V Junction.
5. Analysis of V-A Junction.
6. Analysis of Cardiac Position.
8. Associated Malformations.

So, Where are we exactly?
Position of the Heart:

- Rt Chest.
- Midline.
- Lt. Chest

Direction of the Apex:

- To the Rt.
- To Midline.
- To the Lt.
Confusing Terms

**Levocardia:**
Position of the heart in the left hemithorax + apex directed to the left.

**Dextrocardia:**
Position of the heart in the right hemithorax + apex directed to the right.
(usually + Situs Solitus)

**Dextroversion:**
Position of the heart in the right hemithorax

**Mesocardia:**
The heart is in the midline.
Step 7:

1. Determine Atrial Arrangement.
3. Analysis of A-V Junction.
5. Analysis of V-A Junction.
6. Analysis of Cardiac Position.
8. Associated Malformations.
• **Lungs & Bronchi:**
  * Usual arrangement.
  * Mirror image.
  * Rt Isomerism.
  * Lt Isomerism.
Liver:
- Predominently Rt.
- Predominently Lt.
- Midline.

Spleen:
- Lt-sided
- Rt-sided
- Asplenia.
- Polysplenia.
Step 8:

1. Determine *Atrial Arrangement*.
2. Determine *Ventricular Mass Structure*.
3. Analysis of *A-V Junction*.
4. Analysis of *Arterial Segment*.
5. Analysis of *V-A Junction*.
6. Analysis of *Cardiac Position*.
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Major reference:
Anderson RH and Ho SY.
Sequential Segmental analysis – description and categorization for the millenium.
Cardiol Young 1997; 7(1):98-116