Pulmonary Venous Anomaly - Embryology and Anatomy

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Pulmonary Vein Anomaly

- Total Anomalous Pulmonary Venous Connection
- Partial Anomalous Pulmonary Venous Connection
- Common Pulmonary Vein Stenosis or Atresia
- Unilateral Pulmonary Vein Stenosis or Atresia
The development of pulmonary vein begins at 27~29 days of gestation. The vein arise from the lung buds that are part of the vascular plexus of the foregut, the splanchnic plexus. There are multiple connections to the umbilicovitelline and cardinal venous system. Coalesce to form four vessels that join with common pulmonary vein that emerges from the back wall of atrium.
A. Splanchnic plexus drains the lung buds. It shares the root of drainage of splanchnic plexus, cardinal venous system, and the umbilicovitelline systems.

B. The common pulmonic vein originates as an invagination from the left atrial side of the common atrium and establishes communication with the splanchnic plexus.
C. No longer necessary, the primitive pulmonary venous connections disappear.

D. Finally, the individual pulmonary veins are incorporated into the left atrium, and the common pulmonary vein no longer exists.
Embryology

The Systemic venous tributaries identified in molecular terms expression of the transcription factor Tbx18.

The Pulmonary veins → do not contain this protein.
Embryology - 4 wks

- Right venous confluence
- Gut
- Left venous confluence
- Dorsal mesocardium
Embryology – 5 wks

- Pulmonary ridges
- Valves of systemic venous sinus
- Left venous confluence
Embryology – 6 wks

- Pulmonary vein
- Left atrium
- Superior caval vein
- Left sinus horn
- Inferior caval vein

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Embryology – 51 wks
Embryology

- Vestibular spine
- Primary atrial septum
- Right atrial appendage
- Left atrium
- Systemic venous orifice
- Pulmonary vein
Embryology

- Right pulmonary vein
- Left pulmonary vein
- Left atrium
- Right atrium
- Right venous valve
- Left venous valve
- Tendon of Todaro
- Muscularising vestibular spine
Embryology

- Pulmonary venous component
- Left superior caval vein
- Systemic venous sinus
- Left atrium
- Primary atrial septum
- Muscularised buttress
- Right appendage
- Atrioventricular node
Embryologic Classification

I. Atresia of common pulmonary vein while pulmonary systemic venous connections are still present
   A. Partial anomalous pulmonary venous connection
   B. Total anomalous pulmonary venous connection

II. Atresia of the common pulmonary vein after pulmonary systemic venous connections are obliterated
   - Atresia of the common pulmonary vein

III. Stenosis of the common pulmonary vein
    - Cor triatriatum

IV. Abnormal absorption of the common pulmonary vein into the left atrium
   A. Stenosis of the individual pulmonary vein
   B. Abnormal number of pulmonary vein
Anomalous Connection

- One vein drains anomalously
- One lung drains anomalously
- All veins drain anomalously
Why Anomalous Connection?
Why Anomalous Connection?
Pulmonary Vein Anomaly

- Total Anomalous Pulmonary Venous Connection
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Total Anomalous Pulmonary Venous Connection

A pulmonary vein is connected anomalously only when it is attached to a site other than the morphologically left atrium.

- 1 % of all congenital heart disease
- Boy : Girl = 4 : 1
Total Anomalous Pulmonary Venous Connection

**Normal Heart**
- Pulmonary Veins from Lungs
- Superior Vena Cava
- Atrial Septum
- Tricuspid Valve
- Inferior Vena Cava
- Mitral Valve
- Aortic Valve
- Ventricular Septum

**Total Anomalous Pulmonary Venous Return**
- Pulmonary Veins Connect to Superior Vena Cava
- Opening Between Atria
- Tricuspid Valve
- Inferior Vena Cava

Legend:
- **Red**: Oxygen-rich Blood
- **Blue**: Oxygen-poor Blood
- **Purple**: Mixed Blood

AO = Aorta
PA = Pulmonary Artery
LA = Left Atrium
RA = Right Atrium
LV = Left Ventricle
RV = Right Ventricle
Different Sites of Anomalous Connection
Different Sites of Anomalous Connection

A. TAPVC to the LinV by way of the LVV
B. TAPVC to the CS
C. TAPVC to the portal system
Anatomy of TAPVC

- Anomalous connections
- Stenotic connections
- Abnormal numbers of pulmonary veins
Anatomy of TAPVC

KEY to the diagnosis of TAPVC
- the anatomic connections of all four pulmonary veins
- the size and location of each vein
- how and where the four veins enter the heart, to seek stenosis
Nature of the Structure

- Pulmonary artery
- Bronchus
- Pulmonary venous channel in vice
Anatomy of TAPVC

- to establish whether the anomalous pulmonary venous connection is:
  - an isolated malformation?
  - part of a more complex anomaly?
  - whether there are associated structural malformations of the pulmonary vasculature?
2/3 of patients:
TAPVC is an isolated anomaly, associated only with the required interatrial communication.

1/3 of patients:
significant other cardiac defects occur, truncus arteriosus, pulmonary atresia, AVSD, TGA, single ventricle physiology, or heterotaxy syndrome (asplenia or polysplenia).
Without severe pulmonary venous obstruction:
- Present in heart failure at 2~3 months of age.
- History of difficulty of feeding, pneumonia.
- Cyanosis is rare.

With severe pulmonary venous obstruction:
- Obvious severe Cyanosis
- Skin mottling reflecting poor peripheral perfusion and metabolic acidosis
- Tachypnea
Inheritance of TAPVC

- Cat eye syndrome – trisomy of the centromeric portion of chromosome 22q
- Association with a deletion of chromosome 2q31-q33
- Holt-Oram syndrome
- Asplenia syndrome
TAPVC - Supracardiac

- SCV
- Ascending vein
- Ventricular mass

Diagram:
1
2
3
TAPVC CASE
TAPVC- Supracardiac and Cardiac
TAPVC- Cardiac

- Pulmonary veins
- Left ventricle
- Left atrium
- Right atrial mouth of sinus
- Coronary sinus

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TAPVC - Cardiac
TAPVC- Right Isomerism

- Pulmonary veins to midline confluence
- Isomeric right appendages
- Morphologically right appendage
- Absence of coronary sinus
TAPVC- Infracardiac

Body of left atrium
Confluence
Descending vein
TAPVC - Infracardiac
TAPVC or not?

Differential Diagnosis

- Complete TGA with large VSD
- AVSD with common atrium
- Levoatrial cardinal vein associated with mitral atresia and intact atrial septum
Normal, but Abnormal Connection

- SCV
- Closed atrial septum
- Normally connected pulm. veins
- Imperforate mitral valve
Partial Anomalous Pulmonary Venous Connection
Different Sites of Anomalous Connection

- Right pulmonary veins to right SVC or azygos vein
- Right pulmonary veins to right atrium
- Right pulmonary veins to IVC (Scimitar syndrome)
- Left pulmonary veins to the left innominate vein.
CASE

- M/ 19 yr, Palpitation, DOE
- RLL, RML, part of RUL drained by SCMV connected to the intrahepatic IVC
- Scimitar variant with non-restrictive connection of meandering vein to LA
Common Pulmonary Vein Stenosis
- Cor Triatriatum

I. Accessory atrial chamber receives all pulmonary veins and communicates with left atrium
   A. no other connection – classic cor triatriatum
   B. other anomalous connection – RA or TAPVR

II. Accessory atrial chamber receives all pulmonary veins and does not communicate with left atrium
    A. Anomalous connection to right atrium directly
    B. With total anomalous pulmonary connection

III. Subtotal Cor triatriatum
    A. Accessory atrial chamber receives part of pulmonary veins and connect to left atrium
    B. Accessory atrial chamber receives part of the pulmonary veins and connects to right atrium
Common Pulmonary Vein Stenosis - Cor Triatriatum
Common Pulmonary Vein Atresia
Anomalous Pulmonary to Systemic Collateral Vein & Levoatrial Cardinal Vein
Stenosis or Atresia of Individual Pulmonary Vein

- Dilated left pulm. veins
- Narrowed veno-atrial junction
- Hypertrophied left atrium
Stenosis or Atresia of Individual Pulmonary Vein
CASE

- Baby at 2 hours after birth, tachypnea, cyanosis

Baby 1

Baby 2
CASE

- Baby at 2 hours after birth, tachypnea, cyanosis

Baby 1

Baby 2
Thank You!
감사합니다.