Anomalies of Pulmonary Arteries and Veins

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Pediatric/Congenital Heart Disease Boot Camp

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- No financial disclosures
- Off label use of gadolinium in children
Learning Objectives

- Recognize common anomalies of:
  - Pulmonary arteries
  - Pulmonary veins
- Describe utility of cardiac MRI in evaluation
Pulmonary Artery Anomalies
Pulmonary Artery Anomalies

PA anomalies

- Anomalies of number
- Anomalies of location
- Anomalies of size
Anomalies of Number
Unilaterally Absent PA

- Associated with congenital heart disease:
  - Absent LPA with tetralogy of Fallot
- Distal pulmonary artery often normally distributed
Ductal Origin of Branch PA

Ductal origin of LPA

CTA

RPA from MPA

PDA

LPA

Ductal origin of LPA
Anomalies of Location

Origin of LPA from RPA (PA sling)

Origin of LPA from RPA (PA sling)

- Airway anomalies common
  - Complete membranous rings
  - Tracheomalacia
  - Abnormal lung lobulation
  - Bronchus suis
Anomalies of Location
Aortic Origin of Branch PA

- Branch pulmonary artery origin from the dorsal ascending aorta
- Right >> left
- Associations:
  - Tetralogy of Fallot
  - Patent ductus arteriosus
- “Hemitruncus”

![Image with labeled structures: Asc. Ao, MPA, RPA, LPA]
Aortic Origin of RPA
Anomalies of Size
Branch PA Stenosis/Hypoplasia

- Isolated
- Associated congenital heart disease
  - Tetralogy of Fallot
- Congenital rubella
- Williams syndrome
  - Supravalvar
  - Branch PA

Bilateral branch PA stenosis – Williams syndrome
Pulmonary Atresia with Ventricular Septal Defect (PA-VSD)

- Intra-cardiac anatomy similar to tetralogy of Fallot
- Pulmonary blood supply variable
  - PDA
  - Major aortopulmonary collaterals
PA-VSD: Goals of MRI

- Sources of pulmonary blood flow
- PA size and confluence
- Collateral communication with PAs
Pulmonary Vein Anomalies
Pulmonary Vein Embryology

- Lung buds from foregut – splanchnic drainage
- Left atrium connects to PV
- Regression of primitive splanchnic connections
Classification of PV Anomalies

- Totally anomalous pulmonary venous connection (TAPVC)
- Partially anomalous pulmonary venous connection (PAPVC)
- Sinus venosus defect
- Cor triatriatum
- Pulmonary vein stenosis/atresia
TAPVC

Types:
- Supracardiac
- Cardiac
- Infracardiac
- Mixed

Confluence of PV draining into posterior right atrial wall
PAPVC

Common types:

- Left PV to innominate vein
- Right PV to SVC
  - With sinus venosus defect
  - Without sinus venosus defect
- Right PV to IVC (scimitar syndrome)
Left PV to Innominate Vein

- Ascending vertical vein
- LPA
- Normal left lower PV
- LA
- Innominate vein
- RPA
Scimitar syndrome

- Shadow on X-ray resembles Turkish sword (scimitar)
- Hypoplasia of right lung
- Pseudo-sequestration of right lower lobe of lung
- Aortopulmonary collaterals

Mesoposed heart, hypoplastic right lung and scimitar sign (arrows)
Scimitar Syndrome

Dextroposed heart, large LPA, small RPA

Descending anomalous vein (arrow)
Sinus Venosus Defect

- **Unroofing of right upper PV to SVC**
- **Defect is dilated orifice of right upper pulmonary vein**
- **PAPVC of right PV to SVC (blue arrow)**
Cor Triatriatum

- Membrane in LA (black arrow)
- Proximal PV “chamber”
- Stenotic connection to LA
Unilateral PV Atresia

No perfusion to left lung

Time resolved Gd-MRA

Atretic left PV

No
Post-Operative Problems
Limitations of Echocardiography

Branch pulmonary arteries by 2D echo

Post-operative deterioration of acoustic windows!
Post-Operative PA Stenosis

Severe stenosis of RPA after repair of truncus arteriosus
Post-Operative PV Stenosis/Atresia

- May follow pulmonary vein surgery
- Progressive
- Focal or diffuse

Bilateral PV stenosis
Warden Procedure for Repair of Sinus Venosus Defect

SVC to RA appendage

RPV to LA baffle
Conclusion

- Reviewed important anomalies of Pulmonary arteries and Pulmonary veins.
- MRI particularly well suited because:
  - Accurate anatomical details
    - Gadolinium MRA
    - Black/bright blood imaging
  - Functional information
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