Pulmonary Embolism CT in Various Stages of Single Ventricle Palliation: Challenges and Technique Optimization

Maryam Ghadimi Mahani M.D.
Clinical Assistant Professor
Department Of Radiology
C.S. Mott Children’s Hospital
University Of Michigan
No disclosure

University of Michigan
Department of Radiology
Acknowledgement

• Images and illustrations are used with permission from:

  - Ghadimi Mahani M. Agarwal PP. Rigsby CK  Lu JC. Fazeli Dehkordy S. Wright RA. Dorfman A and Krishnamurthy R.

  - CT for Assessment of Thrombosis and Pulmonary Embolism in Multiple Stages of Single-Ventricle Palliation: Challenges and Suggested Protocols.

Teaching Points

1. Describe Fontan pathway and inherent challenges in optimal pulmonary arterial opacification on Computed Tomography (CT)

2. Illustrate common pitfalls in the diagnosis of pulmonary embolism and Fontan pathway thrombosis

3. Describe CT protocols aimed at providing optimal scan quality
Table of Contents

• Introduction
• Staged Surgical Approach to Fontan procedure
• Etiologies of Pulmonary embolism in Fontan procedure
• Challenges in optimizing pulmonary arterial enhancement in patients undergoing staged Fontan procedure
• Case examples of common pitfalls in imaging and diagnosis of thrombi and pulmonary embolism in these patients
• Suggested techniques for CT
• Conclusions
Fontan Procedure

• Multistage surgical palliative repair procedure

• Modified since its introduction by Fontan and Baudet, in 1971

• Systemic venous flow bypasses the right ventricle and enters the pulmonary circulation directly

• Performed in single ventricle physiology or any congenital heart disease that double ventricle physiology cannot be maintained such as: Hypoplastic ventricle (right or left), tricuspid atresia, double inlet ventricle,...
Staged Surgical Approach to Fontan Procedure
Hypoplastic Left Heart Syndrome

LV: Left ventricle
Ao: Native aorta
RV: Right ventricle
RA: Right atrium
LA: Left atrium
PA: Pulmonary artery
SVC: Superior Vena Cava
IVC: Inferior Vena Cava
RPA: Right Pulmonary Artery
LPA: Left Pulmonary Artery
MPA: Main Pulmonary Artery
Stage 1. Norwood Procedure

1. Divide Pulmonary Artery
2. Create modified Blalock-Taussig (BT) shunt
3. Create neoaorta (neoA) by using MPA root as homograft
4. Create Atrial Septal Defect
Stage 2: Cavopulmonary Shunt

1. Take down modified BT shunt
2. Create Bidirectional Glen

OR
Stage 2: Cavopulmonary Shunt

1. Take down modified BT shunt

2. Create HemiFontan (Baffle between the right atrium and RPA)
Stage 3: Modified Fontan Procedure

Lateral tunnel Fontan

- Tunnel created in the right atrium by using prosthetic material to direct blood flow from systemic vein to the lung
Extracardiac conduit

- IVC separated from right atrium and a polytetrafluoroethylene tube graft created from IVC to PA beside the right atrium (rather than inside it)

- Fenestration or small opening (not shown) may be created between the venous pathway and right atrium (in either extracardiac or lateral tunnel Fontan)

Stage 3: Modified Fontan Procedure
Etiologies of Pulmonary Embolism in Patients With Fontan Palliation

- Exact etiology not known
- Possibly multifactorial due to:
  - Atrial arrhythmia
  - Protein losing enteropathy
  - Sluggish, non laminar blood flow in Fontan pathway
  - Abnormal liver function test
  - Coagulation abnormalities
Challenges of PE CT

1. Optimizing homogenous pulmonary arterial enhancement
   - **Unopacified blood:** injecting through one upper/or lower extremity will result in mixture of unopacified blood from IVC or left SVC (if present) and cause filling defects resembling thrombosis
   - **Optimal delay scan time**
Challenges of PE CT

2. Errors in recognizing the anatomy and surgical procedure in these patients
   - Using neo-aorta (native MPA used as the homograft for constituting the neo-aorta) as the region of interest for bolus tracking

3. Not being aware of additional lesions
   - Such as left SVC
Case Examples

Pitfalls
Injection through Access from One Extremity

- Different timing for the opacification of the left and right pulmonary arteries (Fig. 1)

- Mixture of the unopacified blood from lower extremity veins through IVC resulting flow artifact resembling filling defect in the central pulmonary arteries (Fig. 1)
Case example

Fig. 1. (A) CT pulmonary angiography in a 20 year old male with hypoplastic left heart syndrome status post lateral tunnel Fontan operation with shortness of breath. Note unopacified left lower lobe pulmonary arteries (arrows) which was interpreted as pulmonary embolism by the emergency department radiologist.
Fig. 1. (B and C) The injection was performed only from the left upper extremity (not shown) and at the time of obtaining the images the left lower lobe pulmonary arteries were not opacified yet (as opposed to right lower lobe branch pulmonary arteries) since the left PA is mainly getting its blood flow from IVC side of Fontan pathway. For the flow artifact seen in the central PA(*) it was considered pulmonary embolism, since they misinterpreted the unopacified left lower lobe pulmonary arteries as pulmonary embolisms. (AO, native aorta) neoA (neo-aorta)
Fig. 3. (A) Time resolved MRA in a 11 year old male shows delayed and incomplete filling of the inferior Fontan baffle with pseudo-filling defects simulating thrombus (arrow).

(B) Axial CT image following a lower extremity injection for the CT angiogram (CTA), there is a pseudo-filling defect (arrow) in the proximal RPA form unopacified blood from the SVC baffle.
Suggested technique 1

- **Dual injection technique** from both upper extremity and lower extremity

- Upper extremity with slower injection rate compared to the lower, (e.g. upper 2-2.5 ml/s; lower 3-4 ml/s)

- Using bolus track technique for determination of delay scanning at the level of left pulmonary artery branch for timing (if evaluating the pulmonary arteries) (Fig 4)
Fig. 4. Use of bolus track for CT pulmonary angiography patient with Fontan palliation. Timing bolus technique is not preferred due to variability in flow velocities in these patients. Note the placement of the region of interest in the left pulmonary artery (A, arrow). It is important to understand the anatomy and physiology of Fontan circulation and not to use the neo-aorta (neoA) for bolus tracking technique (B).
**Suggested technique 1**

- Prepare a delay scan in advance
- Preferably monitor the study as it is obtained by the technician
- If the opacification is not optimal, delay scan can be performed (usually with 60-65 second delay) (Fig 5)
- For evaluating the Fontan pathway for thrombosis more delay in scanning is needed (about 2-3 minutes) (Fig 6)
Fig. 5. Axial CT angiography images of the chest using intravenous contrast agent in early (A) and delayed phases (B). Note apparent filling defect (arrow) on the early phase due to sluggish flow in SVC-Fontan pathway mimicking thrombosis. This appearance resolves on subsequent delayed image 1 minute post injection.
Fig. 6. Axial CT angiography images through Fontan pathway using intravenous contrast agent in a 10 year old female with hypoplastic left heart syndrome status post Fontan operation; in early (A) and delayed phases (B and C). CT was urgently requested for evaluation of thrombus at IVC Fontan pathway junction suspected on echocardiogram. Note apparent filling defect (arrow) on the early phase due to sluggish flow and unopacified blood in Fontan pathway mimicking thrombosis. This appearance resolved on subsequent delay images obtained 1 minute (B) and three minutes (C) post injection.
Fig. 7. (A) Diagnostic CT Pulmonary angiography in a 5 year old male with multiple congenital heart defects including hypoplastic left ventricle, double outlet right ventricle, status-post multiple cardiac surgeries including lateral Fontan operation, presented with frequent desaturation and recurrent croup.

Note optimal enhancement of the branch pulmonary arteries and SVC. Double Injection Technique (suggested technique 1) was used. Native (AO) to neo-aorta (neo A) anastomosis (Damus-Kaye-Stansel, DKS, anastomosis, arrow)
Fig. 7. (B, C) Diagnostic CT Pulmonary angiography in a 5 year old male with multiple congenital heart defects. Lateral tunnel Fontan (arrow in B)
Suggested technique 2

- Single upper extremity injection

- Using 130 ml of contrast (adult size patient; 3 ml/kg in children up to 40 kg), inject at a rate of 2.5 ml/sec (Fig 8)

- Scan at about 70 second or a little longer (75 second) if there is evidence of significant atrioventricular valve regurgitation by echocardiogram
Fig. 8. Axial CT angiography images in a 3 year old male with crescentic peripheral thrombus in Fontan pathway (arrow in A) that got worse over time in follow up CT a month later (arrow in B).

Eventually the baffle thrombosed completely seen on catheter angiogram (not shown) despite anticoagulation.
Steady State Imaging with blood pool contrast agents

Fig. 9. Current gold standard for MRA in the setting of Fontan is the use of blood pool contrast agent, which ensures optimal filling of all the vessels without any streaming artifact.
Conclusion

- Pulmonary embolism and thrombosis are common in patients with Fontan operation.
- If CT is performed, precise technique is needed to avoid flow artifact misread as pulmonary embolism or thrombosis in Fontan Pathway.
Conclusion

- Either use both upper or lower extremity injections and/or use delayed scan technique with 70 seconds delay for evaluation of pulmonary arteries.

- Familiarity with anatomy and flow pattern of post Fontan operation patient is crucial for correct diagnosis.

- Direct monitoring of the study at the time of obtaining the examination is suggested to modify the exam as needed.


Acknowledgement

- Special thanks to Ms. Danielle Dobbs for drawing the illustrations.
- Corresponding Author: Maryam Ghadimi Mahani M.D.

maryamg@med.umich.edu