*Cardiology 2018*

# Direct Measurement of Thoracic Duct Pressure in Patients with Fontan Physiology

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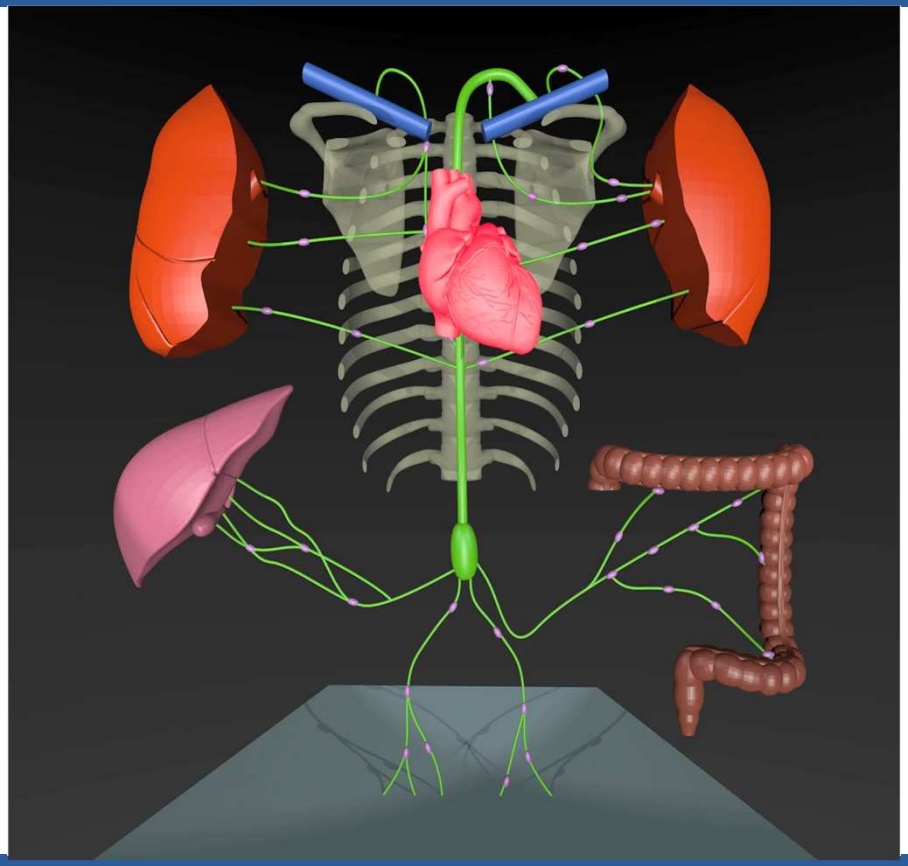
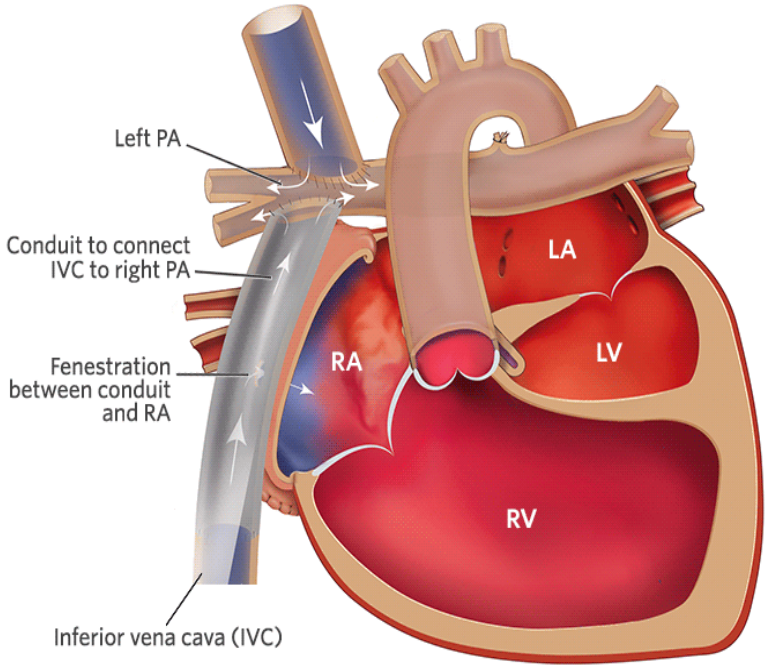
# WHY measure thoracic duct pressure?

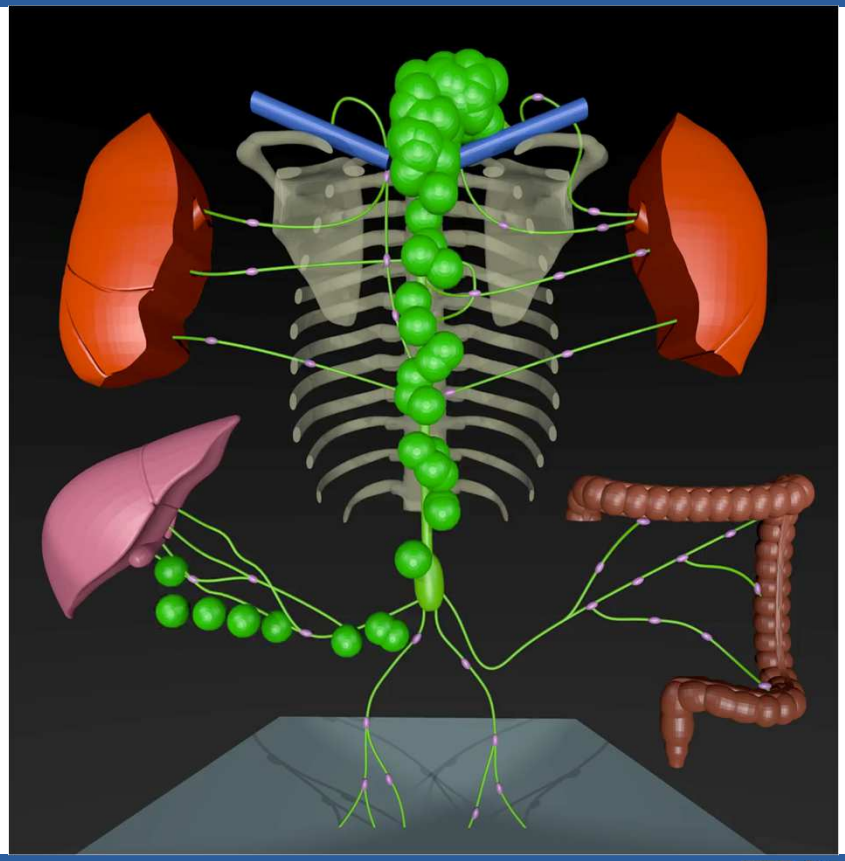
- Fontan complications due to lymphatic abnormalities:
  - Chylothorax
  - Chylopericardium
  - Plastic bronchitis
  - Protein losing enteropathy
- Limited understanding of pressure & flow in the lymphatic system (“lympho-dynamics”)

# Normal lymphatic anatomy

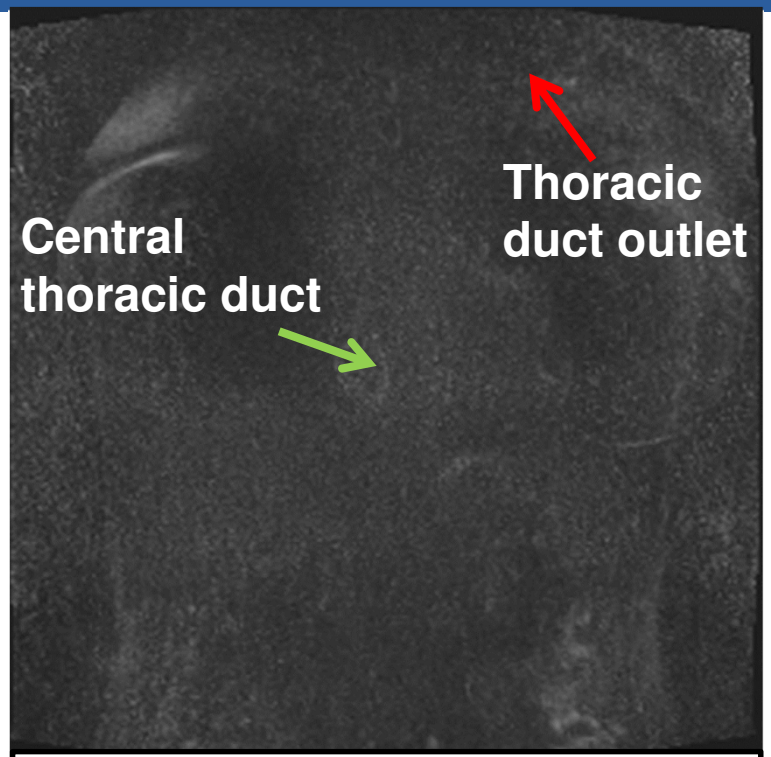


# Fontans = ↓ lymph drainage & ↑ production

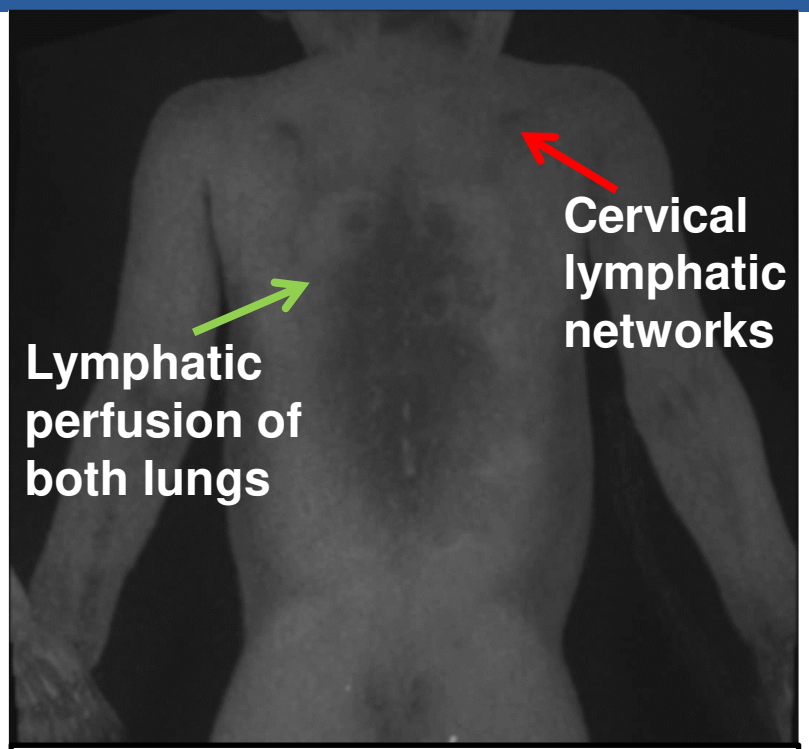


# Dynamic MR lymphangiogram




Normal lymphatic flow




Pulmonary Lymphatic Perfusion Syndrome (PLPS)

# Objectives

1) Measure thoracic duct (TD) pressure directly
   - Implications for interstitial pressure → related to edema
2) Compare TD pressure to Fontan pressure
   - Elevated TD pressure → lymphatic complications
3) Assess change in TD pressure with acute occlusion
   - Simulate changes that occur after TD embolization

# Methods

- Retrospective review
- 29 Fontan patients from April 2016 – Sept 2017
- TD pressures have been routinely measured during lymphatic interventions (prior to TD embolization) at our center since 2016

# Inclusion Criteria

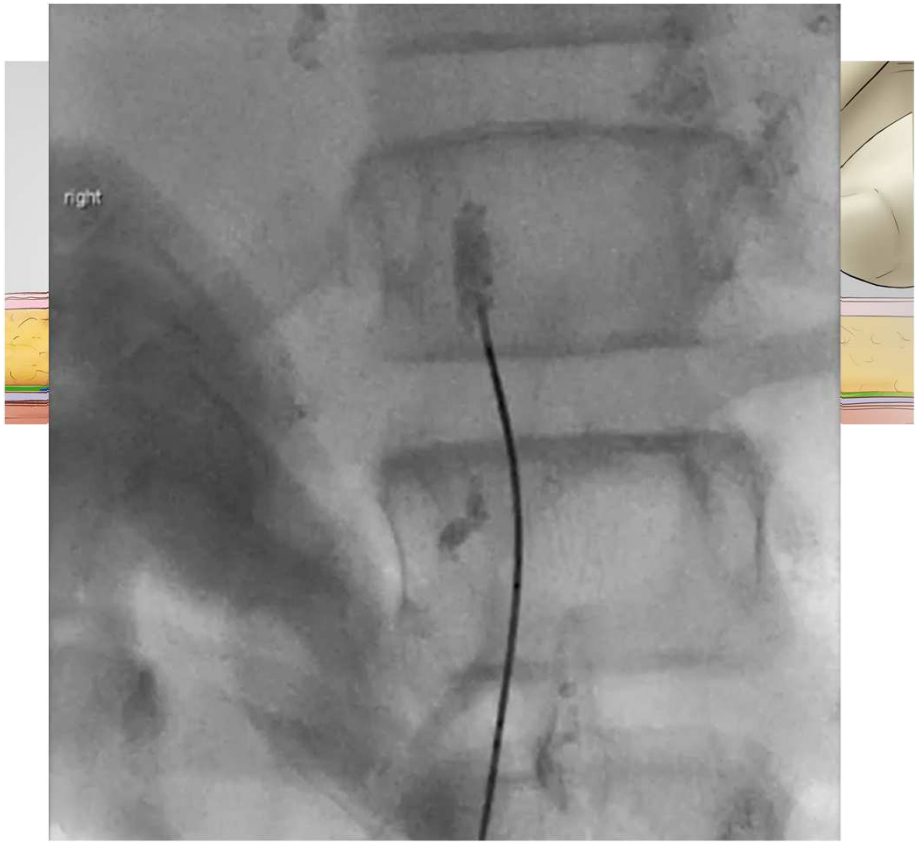
- Fontan physiology
- “Pulmonary lymphatic perfusion syndrome” (PLPS) as the indication for lymphatic intervention
- Simultaneous recording of TD and Fontan pressure

# Measurements

- All patients had positive pressure ventilation
  - Nadir used to estimate mean TD pressure when respiratory variation was present
- Acute TD outlet occlusion testing performed with external compression maneuver
  - Evaluate for previously-undetected lymphatic channels

# How to access the thoracic duct?

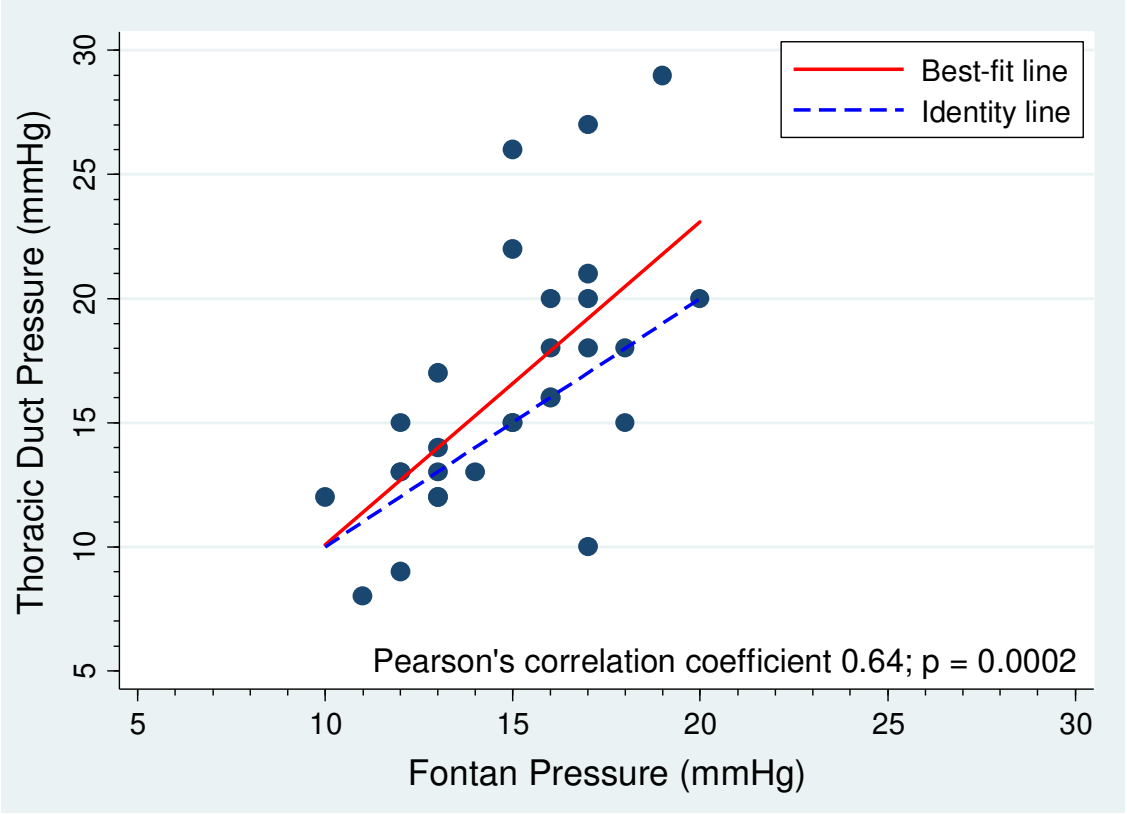
1) Ultrasound-guided inguinal lymph node access
2) Intranodal lymphangiography
3) Percutaneous transabdominal TD cannulation



# Demographics

| Patient characteristics (N=29) | Median (IQR) or count |
|---|---|
| Age at intervention (years) | 6.8 (5.1 – 10.4) |
| Male | 19 (66%) |
| Indication for intervention:<br>Pulmonary lymphatic perfusion syndrome (PLPS) | 29 (100%) |
| Clinical diagnoses: | |
| Chylothorax alone | 6 (21%) |
| Plastic bronchitis ± history of chylothorax | 17 (58%) |
| Plastic bronchitis + concurrent PLE | 6 (21%) |

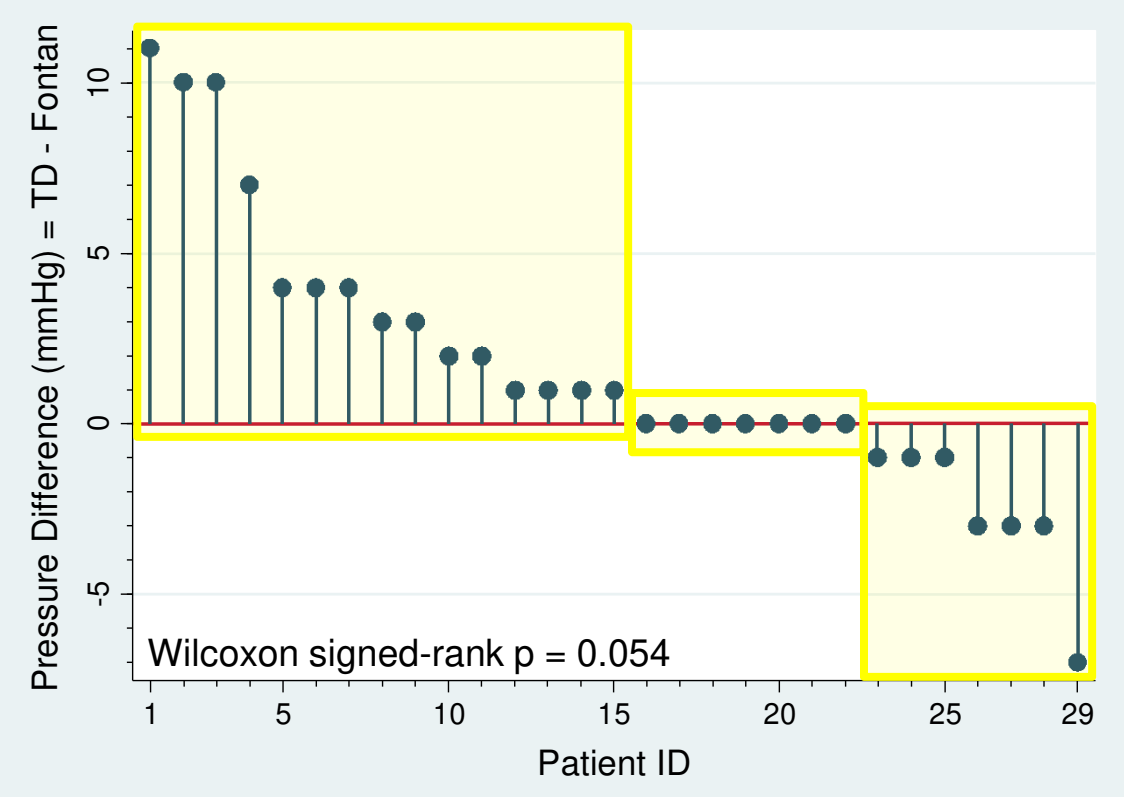
# Results (at baseline)




## Overall cohort:

- Significant correlation ↑ Fontan and ↑ TD pressures, p = 0.0002
- *Non-pulsatile* TD pressure waveform at baseline

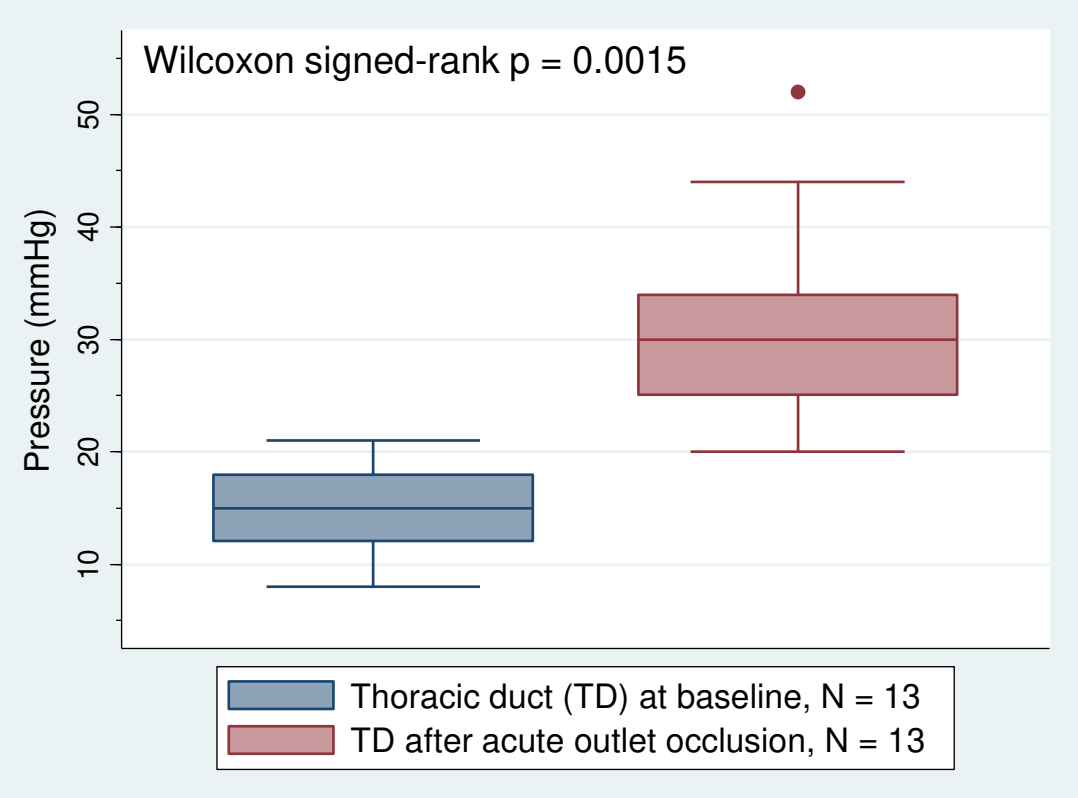
# Thoracic Duct – Fontan Pressure = ?




Individually:

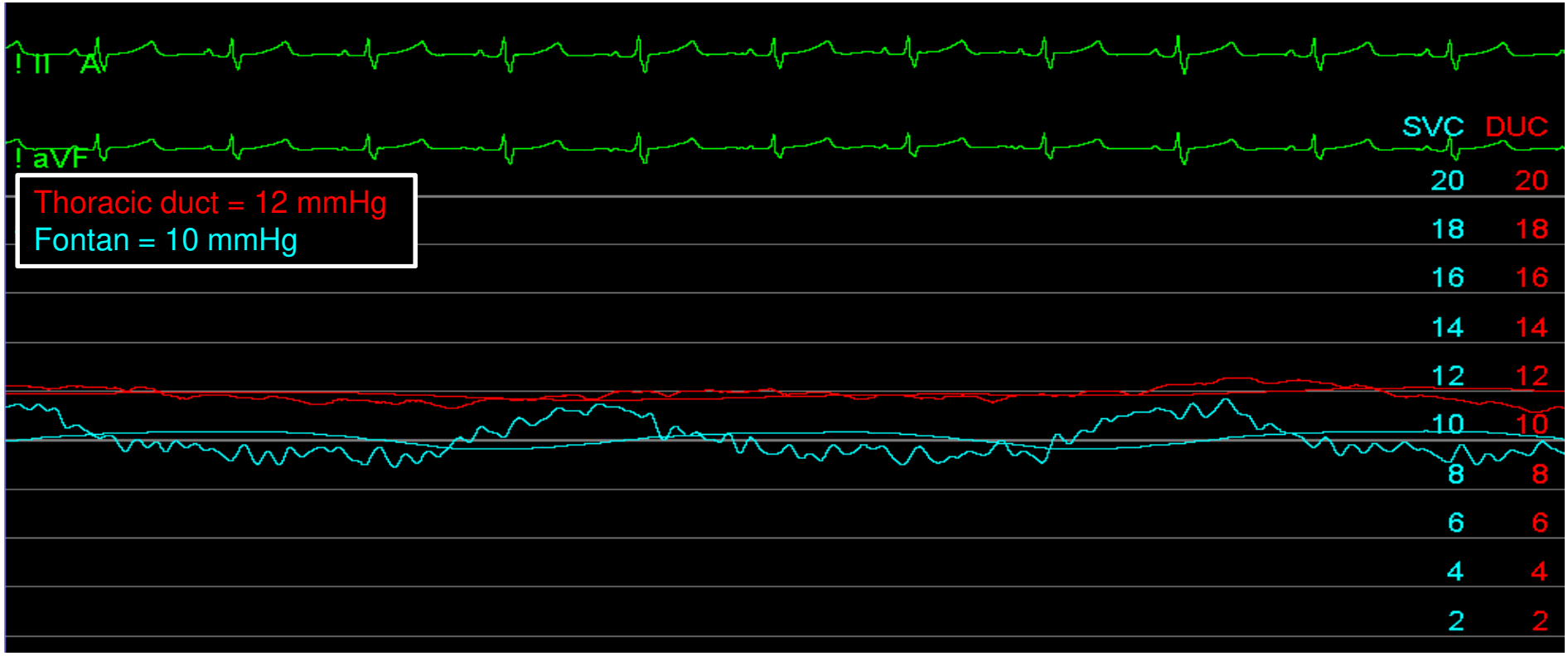
- TD **>** Fontan in 15
- TD **=** Fontan in 7
- TD **<** Fontan in 7

# Acute TD outlet occlusion testing

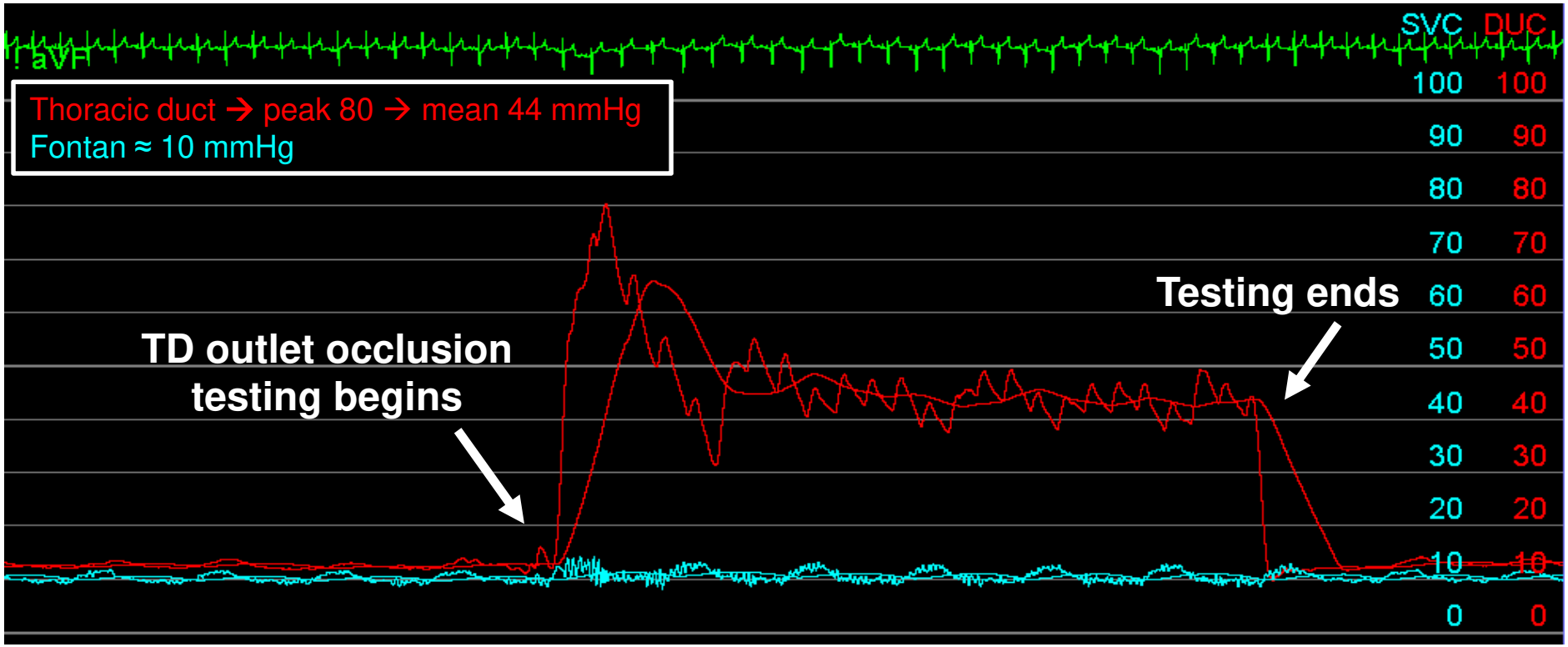



- Significant ↑ TD pressure from median 15 → 30 (IQR 25-34), p = 0.0015
- *Pulsatile* TD pressure waveform

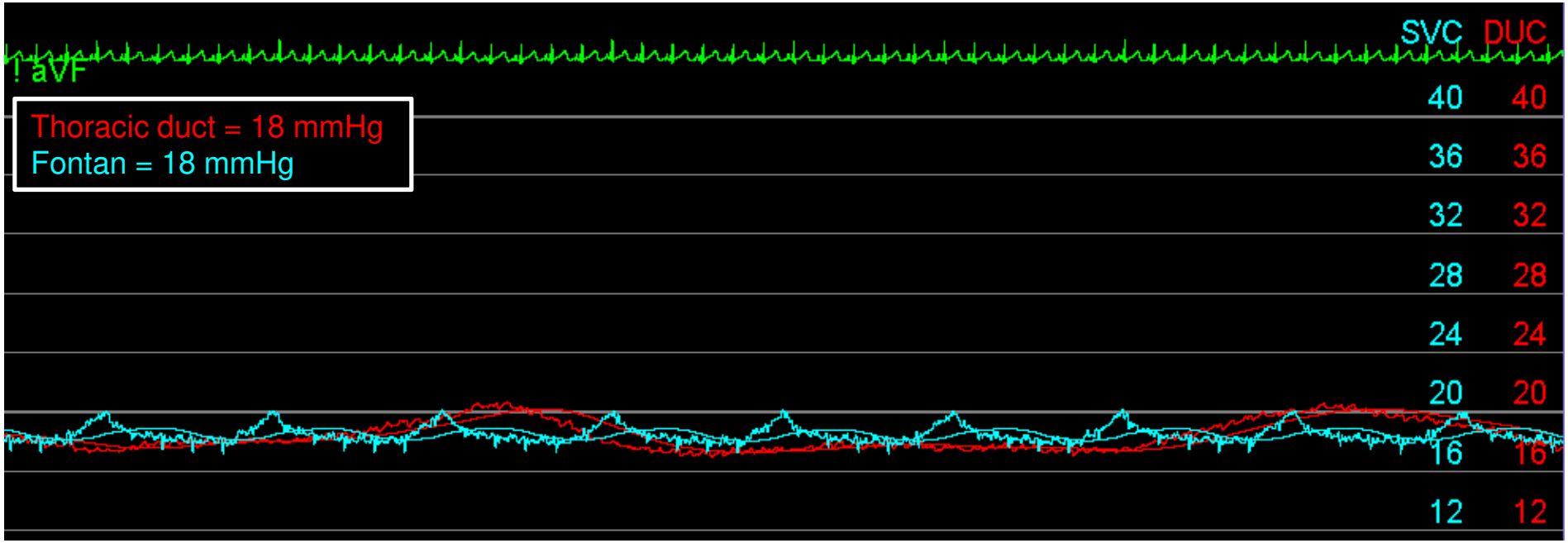
# Patient 1 (at baseline)



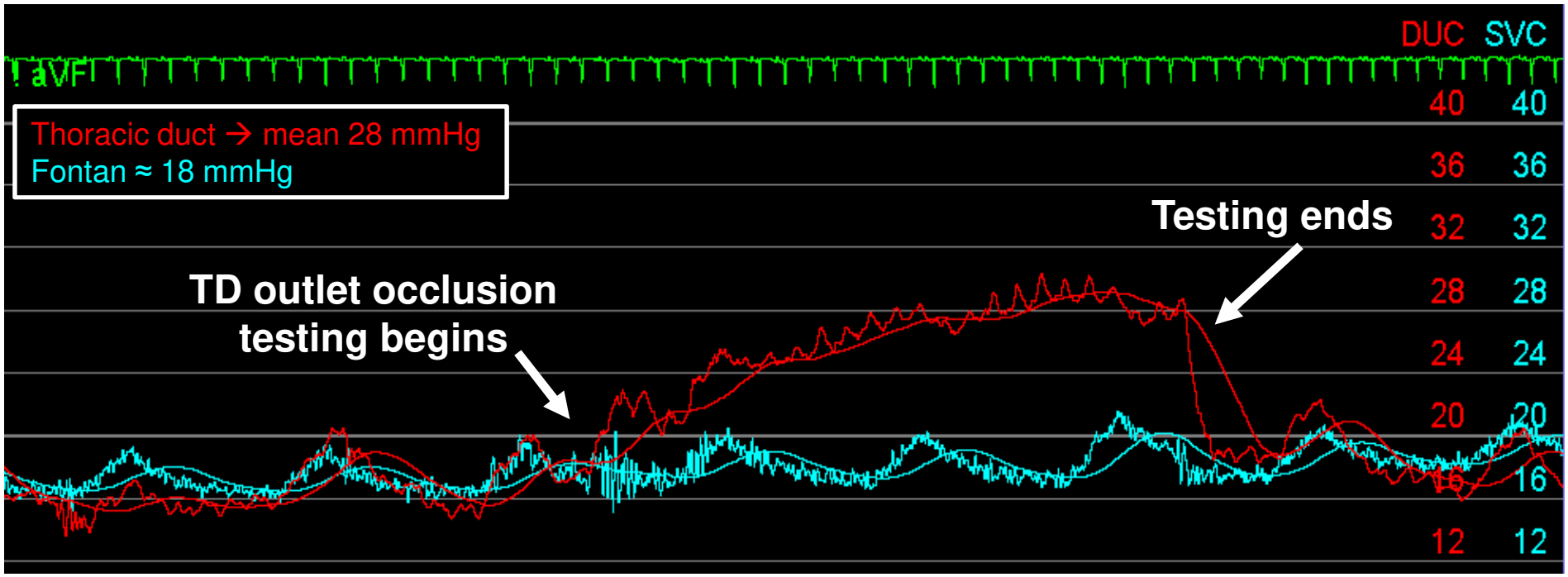
# Patient 1 (with outlet occlusion testing)



# Patient 2 (at baseline)



# Patient 2 (with outlet occlusion testing)



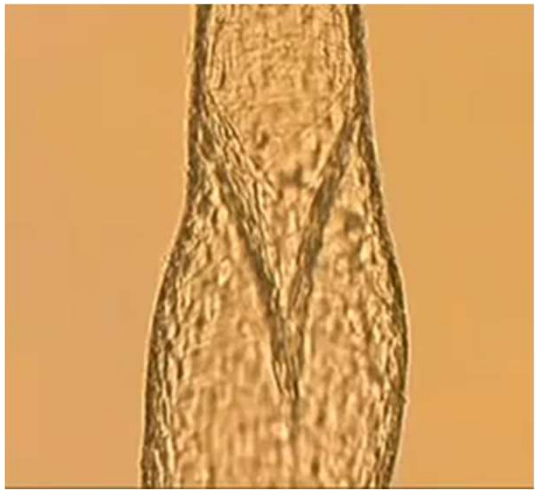
# Conclusions

1) TD pressure correlates with Fontan pressure overall
   - In half of our patients, TD > Fontan pressure
   - When TD < Fontan, is it due to # decompressing channels?
   - Does TD pressure relate to the *severity* or *chronicity* of clinical lymphatic complications?

# Conclusions

2) With acute TD outlet occlusion, TD pressure increases significantly and becomes pulsatile

- If the TD can generate high pressures, why does it get overwhelmed and manifest clinically?
- Is the evoked pulsatility we observed due to intrinsic lymphatic contractility?



# Thank you!

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# Additional information

| Patient characteristics (N=29) | Median (IQR) or count |
|---|---|
| Anatomic cardiac diagnosis: | |
| Hypoplastic left heart syndrome (HLHS) | 12 (41%) |
| Double inlet left ventricle (DILV) | 7 (24%) |
| Unbalanced common AV canal | 4 (14%) |
| Pulmonary atresia with intact ventricular septum (PA-IVS) | 3 (10%) |
| Tricuspid atresia | 2 (7%) |
| Other | 1 (3%) |