

Evaluation of the Failing Fontan

Shakeel A Qureshi

Evelina London Children's Hospital

London, UK

Disclosures

Consultancy:

NuMED Inc

Lifetech Inc

Venus Medtech

Proctor:

Medtronic Inc

St Jude Medical

Occlutech

Problems after Fontan operations

- Arrhythmias up to 40% over 10 yrs
- Protein-losing enteropathy
- Late ventricular dysfunction
- Late mortality
- Hypoxaemia from residual new R → L shunts
- Liver dysfunction
- Pathway obstruction
- Baffle leak

Early Failure of Fontan circulation

Presentation:

- low cardiac output
- pleural effusions
- chylothorax
- ascites

Evaluation of failing Fontan patients

- Echocardiography (TTE and TOE) - ventricular function, AV valve regurgitation
- CT or MRI - assess Fontan pathways, AV valve regurgitation and ventricular function
- Cardiac catheterisation - assess pressures, plan and proceed to interventions

Early Failure of Fontan circulation

Early assessment

- Identify any pathway obstruction and correct it
- Take-down of Fontan and place systemic-to-PA shunt or Glenn shunt
- Cardiac transplantation

Failing Fontan early postoperatively



- 4 year old with HLH, post Norwood I & II
- Had TCPC. 4 days later, severe renal failure, severe hepatic dysfunction, haemodynamic instability
- In catheter lab, no SVC obstruction, LA pressure mean 15, SVC and PA pressure mean 21 mmHg
- Fenestration completely occluded

Failing Fontan early postoperatively



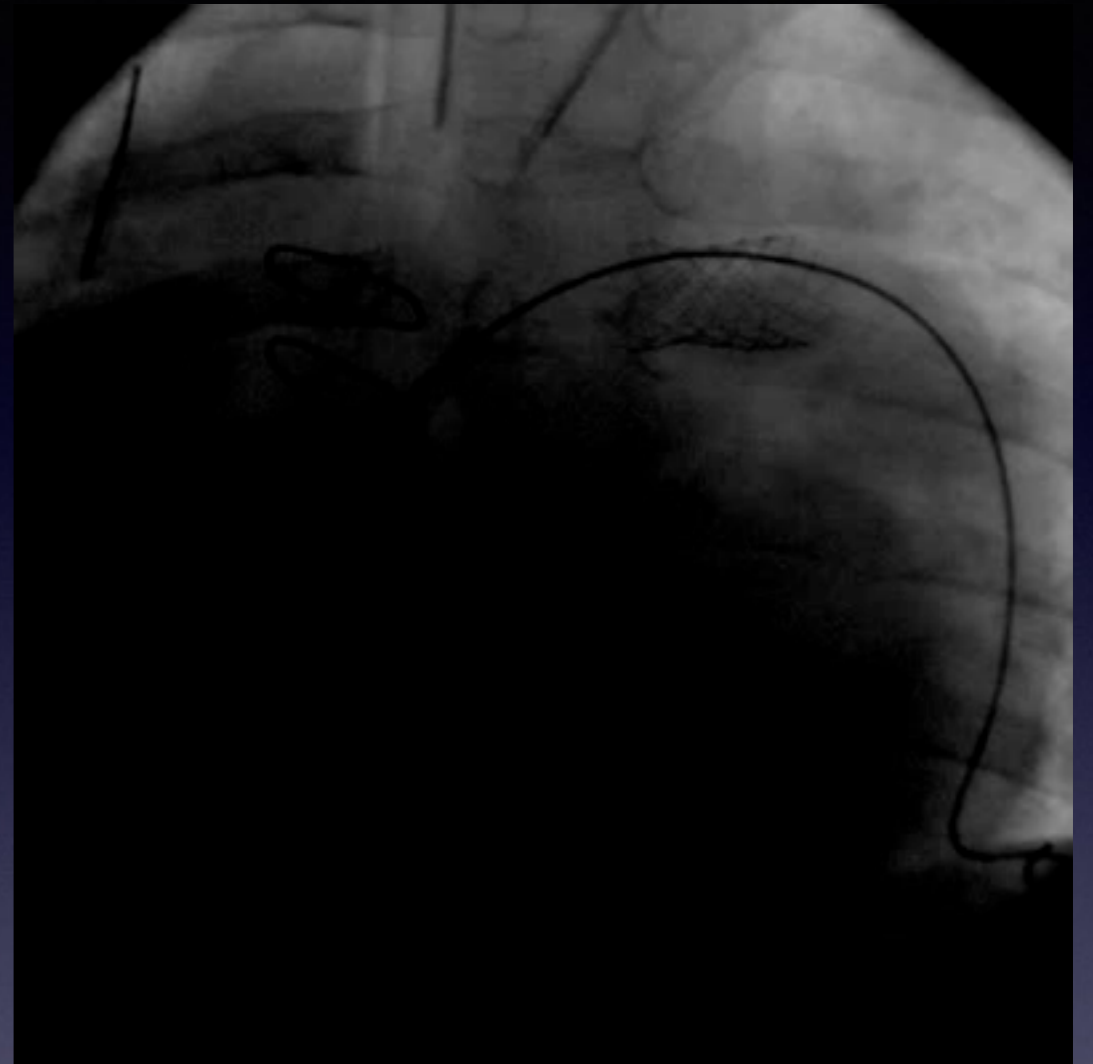
- 5mm Genesis stent implanted in fenestration
- PD for 2 days, renal and liver function improved
- Discharged home 12 days later
- 8 years later, well, O₂ saturation 90%, stent still patent, mild ankle oedema

Fenestration with Fontan operation

- Residual right-to-left shunt
- Reduces caval pressure, increased preload and CO
- At expense of cyanosis
- Reduces mortality and morbidity
- If circulatory failure noted after Fontan operation, be prepared to take down Fontan and convert to bidirectional Glenn shunt or B-T shunt

Early failing Fontan

Combination of LPA stenosis and IVC baffle obstruction

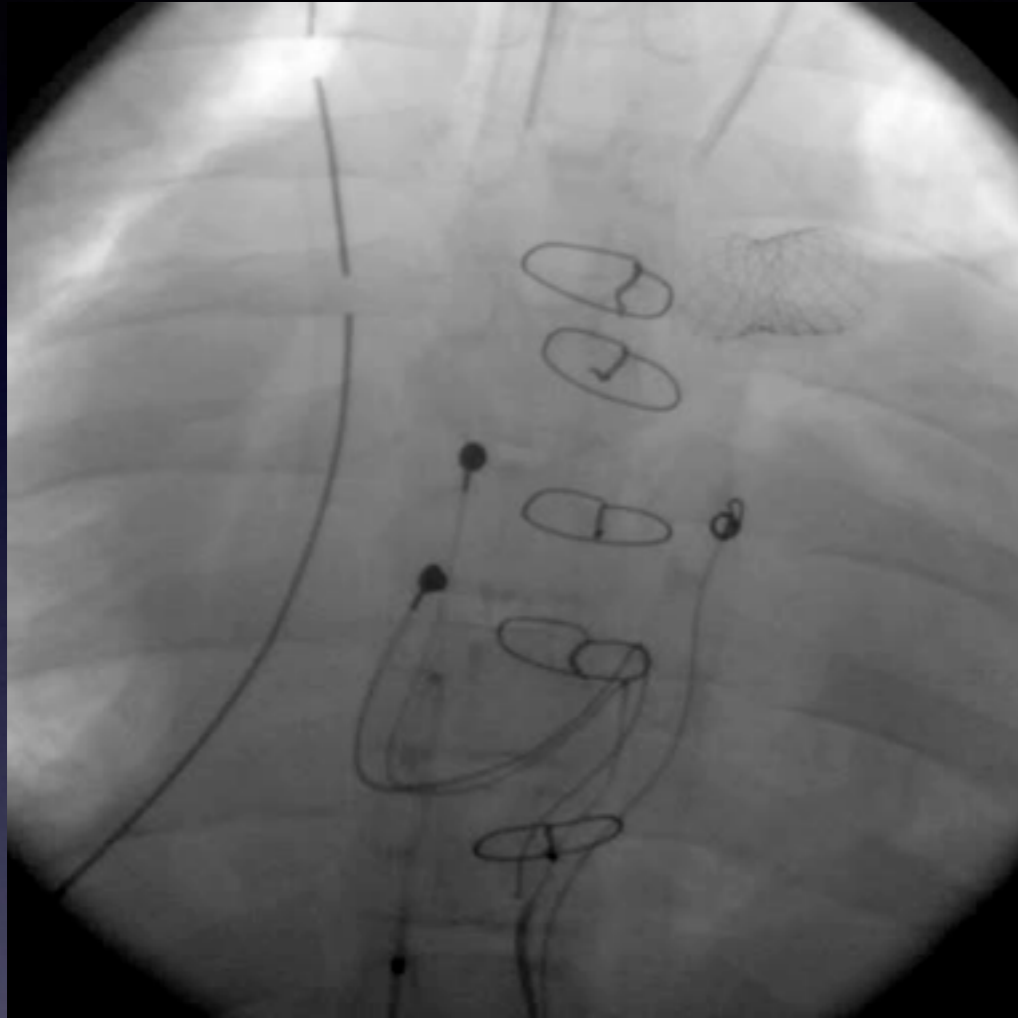


•5 days post Fontan

•Haemodynamic instability, pleural effusions, renal failure

Early failing Fontan

Treated by LPA stent and IVC pathway stent

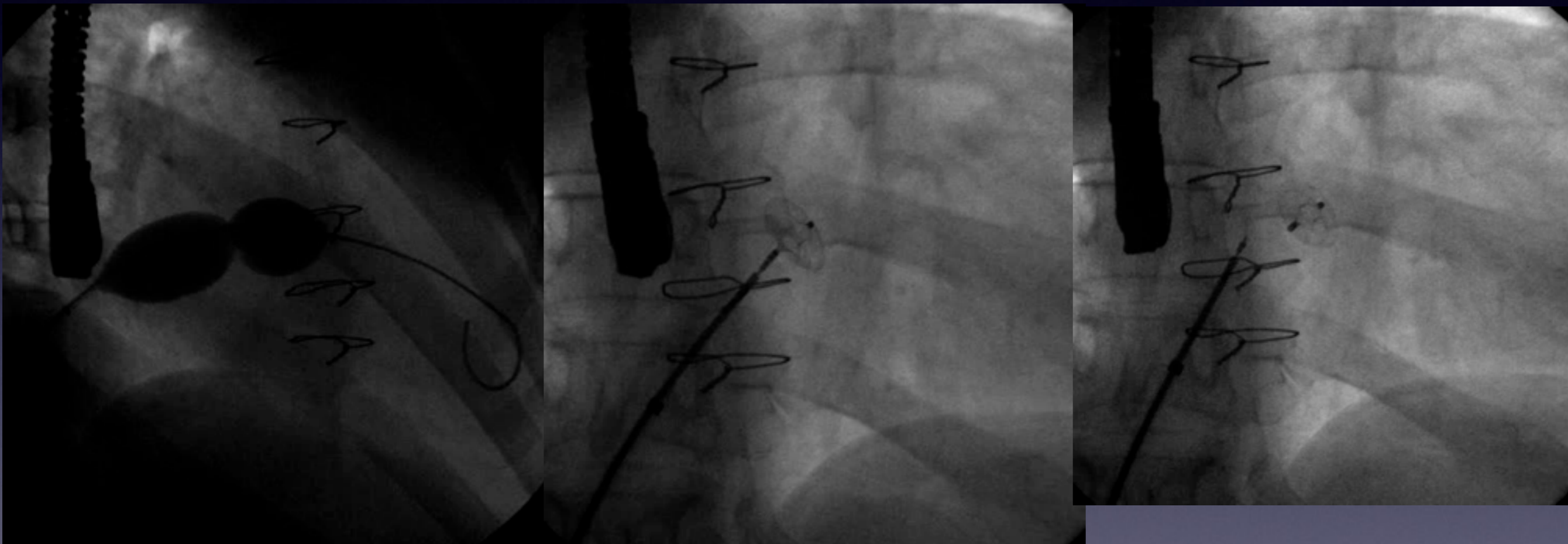


- After LPA stenting and lateral tunnel stenting, stabilised, pleural effusions resolved
- Discharged home 2 weeks later

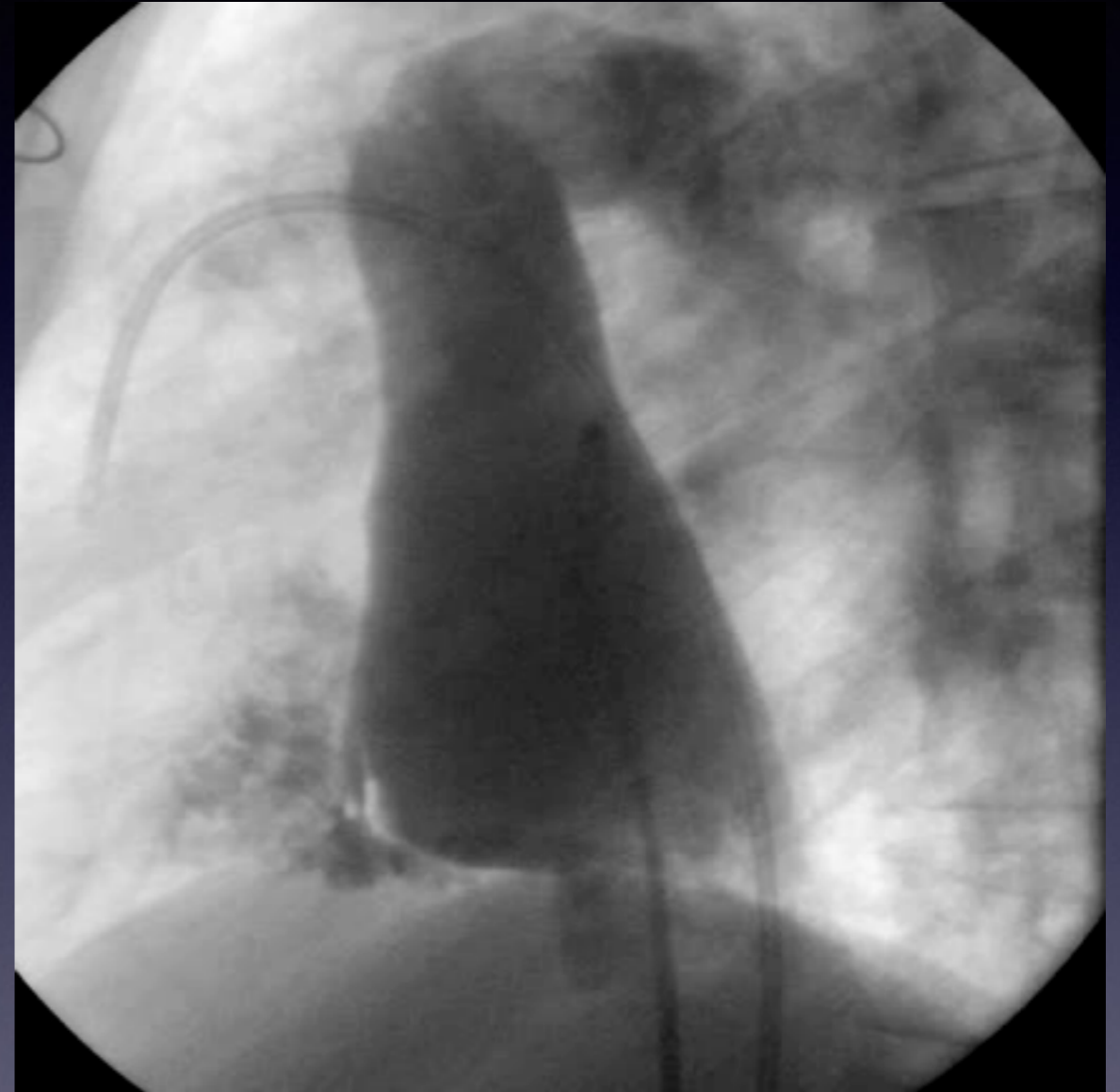
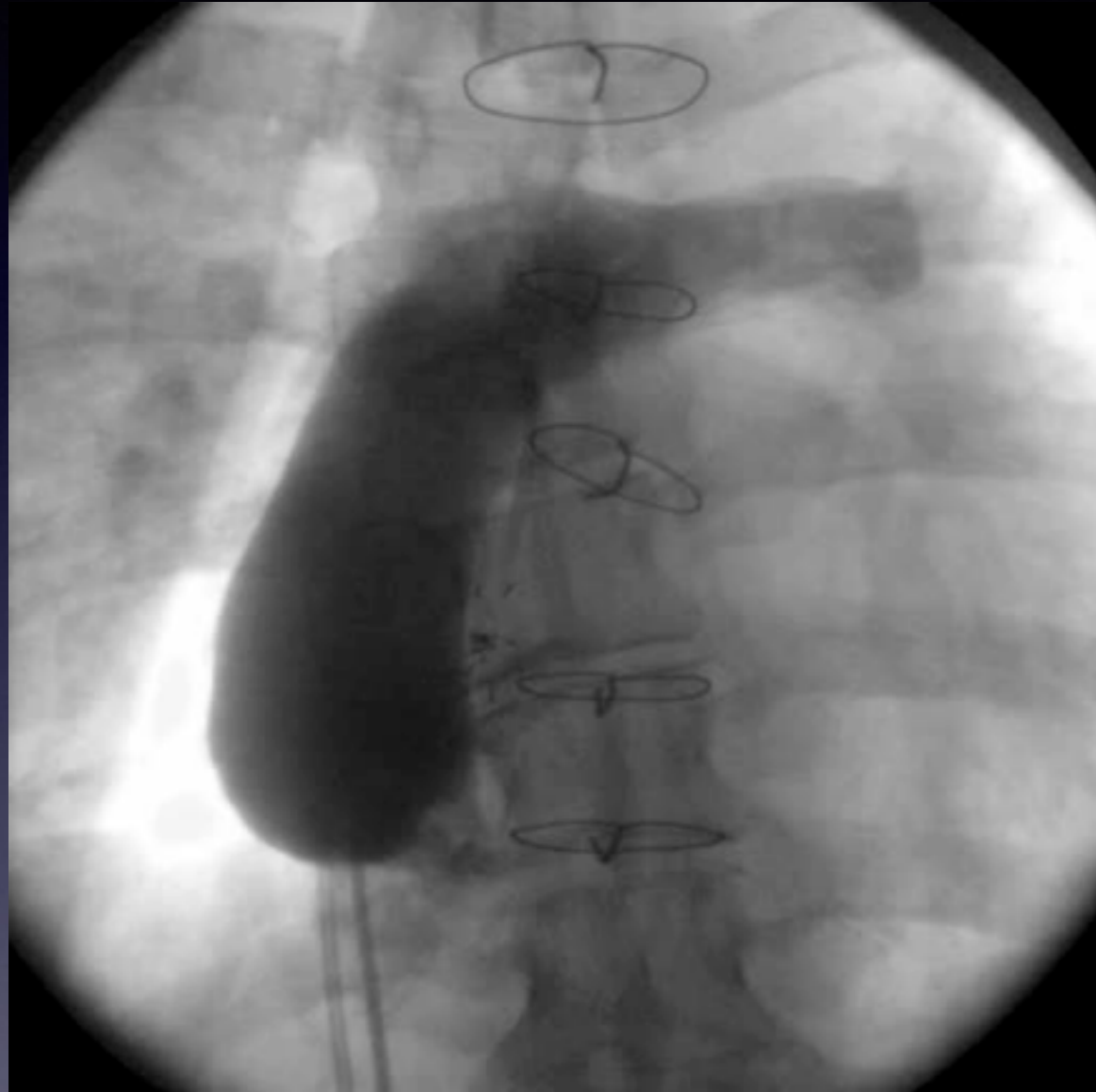
Cyanosis after Fontan

- Mild degree of desaturation (Fenestration/coronary sinus)
- More severe desaturation $< 90\%$ suggest AVMs, veno-venous collaterals or intracardiac R-L shunt (eg leak in lateral tunnel)
- May need intervention (catheter - coils/device occlusion)

Fontan operation Closure of fenestration



Early postoperative cyanosis: Pathway leak



Early postoperative cyanosis: Pathway leak



Late Fontan failure

- 12 year old with DILV, TGA, VSD
- Fontan operation age 3 years
- Previous cerebellar infarcts, and right hemiplegia
- Developed effort intolerance and increasing cyanosis on exercise

Failing Fontan

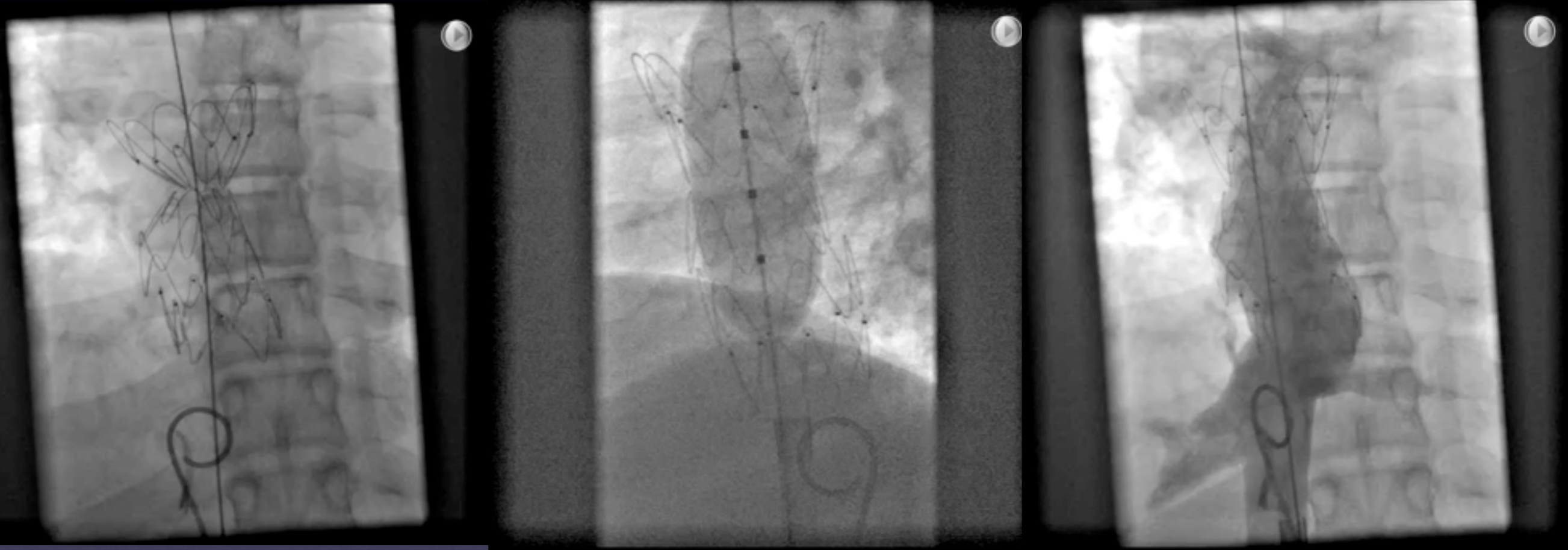
Cyanosis and IVC pathway obstruction



- IVC pressure mean 13 mm Hg
- Cook Z-stent implanted and dilated to 20 mm

Failing Fontan

IVC pathway obstruction - stent graft insertion



- Good result maintained 5 years later
- O₂ saturation 93%

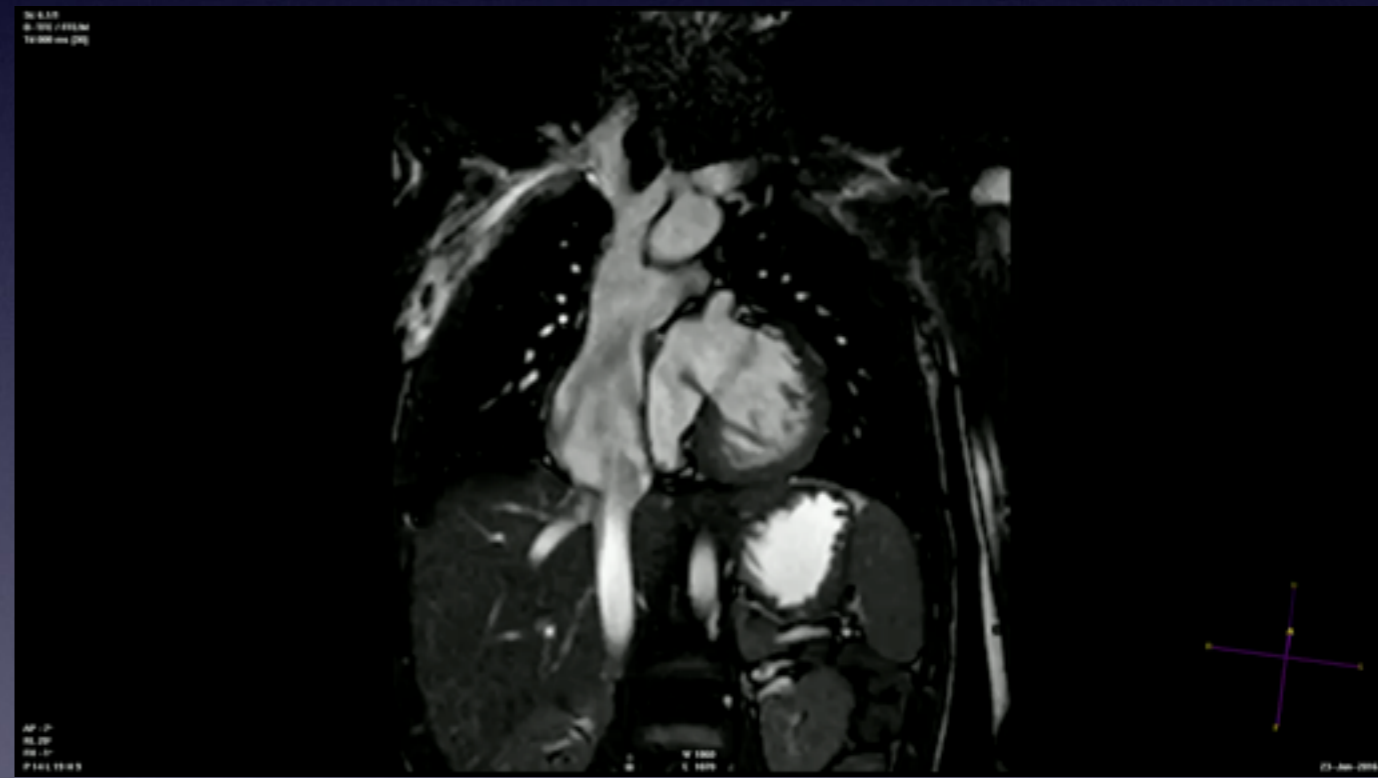
Late Failure of Fontan circulation

- Presentation (lymphatic dysfunction, PLE): pleural effusions, ascites, peripheral oedema, fat malabsorption, low albumin
- Early assessment and treatment: MCT diet, optimise cardiac rhythm, cardiac output, pulmonary vasodilator therapy, after load reduction, steroids, diuretics, anticoagulation, spironolactone, albumin infusions, immunosuppression, cardiac transplantation, creation of fenestration

Evaluation of patients with PLE

- Low cardiac output with Fontan circulation pressure > 15 mm Hg:
- Consider fenestration creation in addition to medical treatment (diuretics, PDE5 inhibitors)

Assessment of Fontan circulation



Failing Fontan

- HLHS - Norwood I, Glenn and lateral tunnel

Fontan (age 3 years)

- Severe protein-losing enteropathy 1 year after

Fontan

- MRI - LPA smaller than RPA, no other

obstruction, RVEF 73%

- SVC pressure 21 mm Hg, IVC 20 mm Hg, LA 15

mm Hg

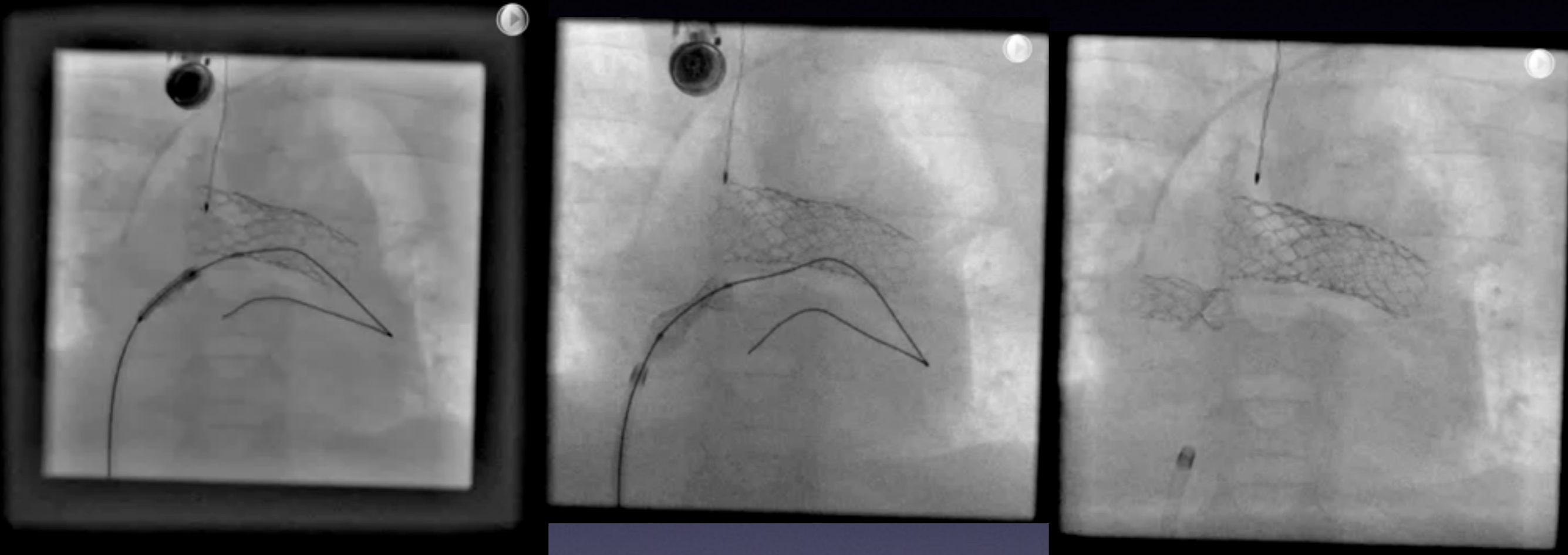
- LPA smaller than RPA



Failing Fontan - PLE 1 year after operation



Failing Fontan - LPA stenting combined with atrial septal stenting



- Pressure in tunnel 22 mmHg, LPA 21mm Hg, LA 14 mmHg
- Treated with prednisolone, diuretics, sildenafil, carvedilol
- Was put on transplant waiting list

LPA stent and fenestration in Fontan circulation

- PLE improved over 3 months
- 3 years later, mild ankle oedema but otherwise well
- Routine MRI and cardiac catheterisation:
 - SVC mean 12 mmHg, lateral tunnel mean 11 mm Hg, LPA mean 12 mm Hg, RPA mean 11 mm Hg, RVEDP 5 mm Hg
- Cardiac output 3.2 l/min/m², PVR 2.2 WU.m²

LPA stent and fenestration in Fontan circulation

- After Dobutamine 10 mcg/kg/min

- Cardiac output 4.3 l/min/m²

- After Dobutamine 20 mcg/kg/min

- RVEF 66%

- Cardiac output 4 l/min/m², PVR 2.35 WU.m²

Failing Fontan - ascites

- 7 year old with HLHS
- Norwood stage I at age 1 day
- Stage II age 6 months
- Stage III - fenestrated Fontan age 3 years (2005)

Failing Fontan

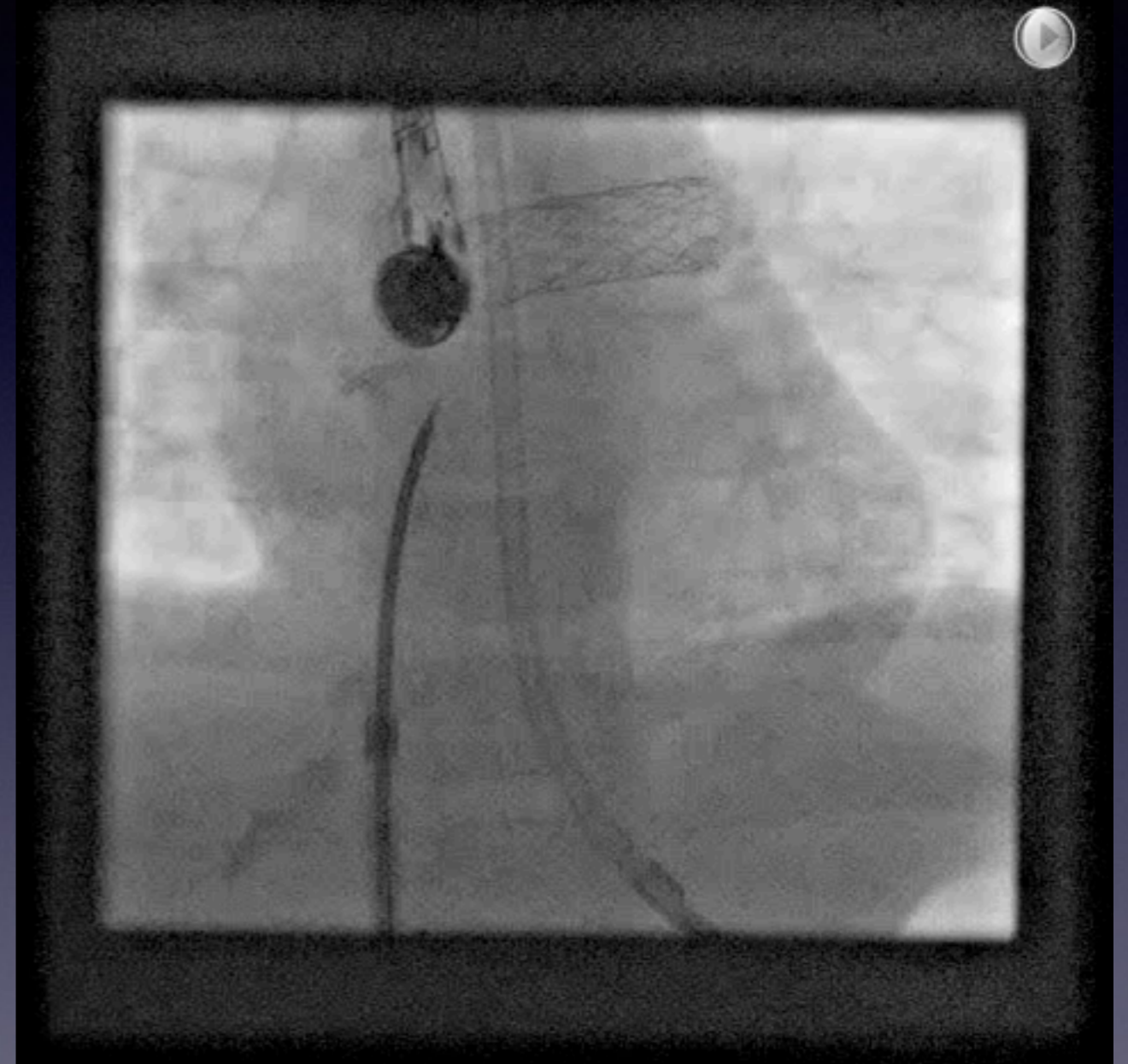
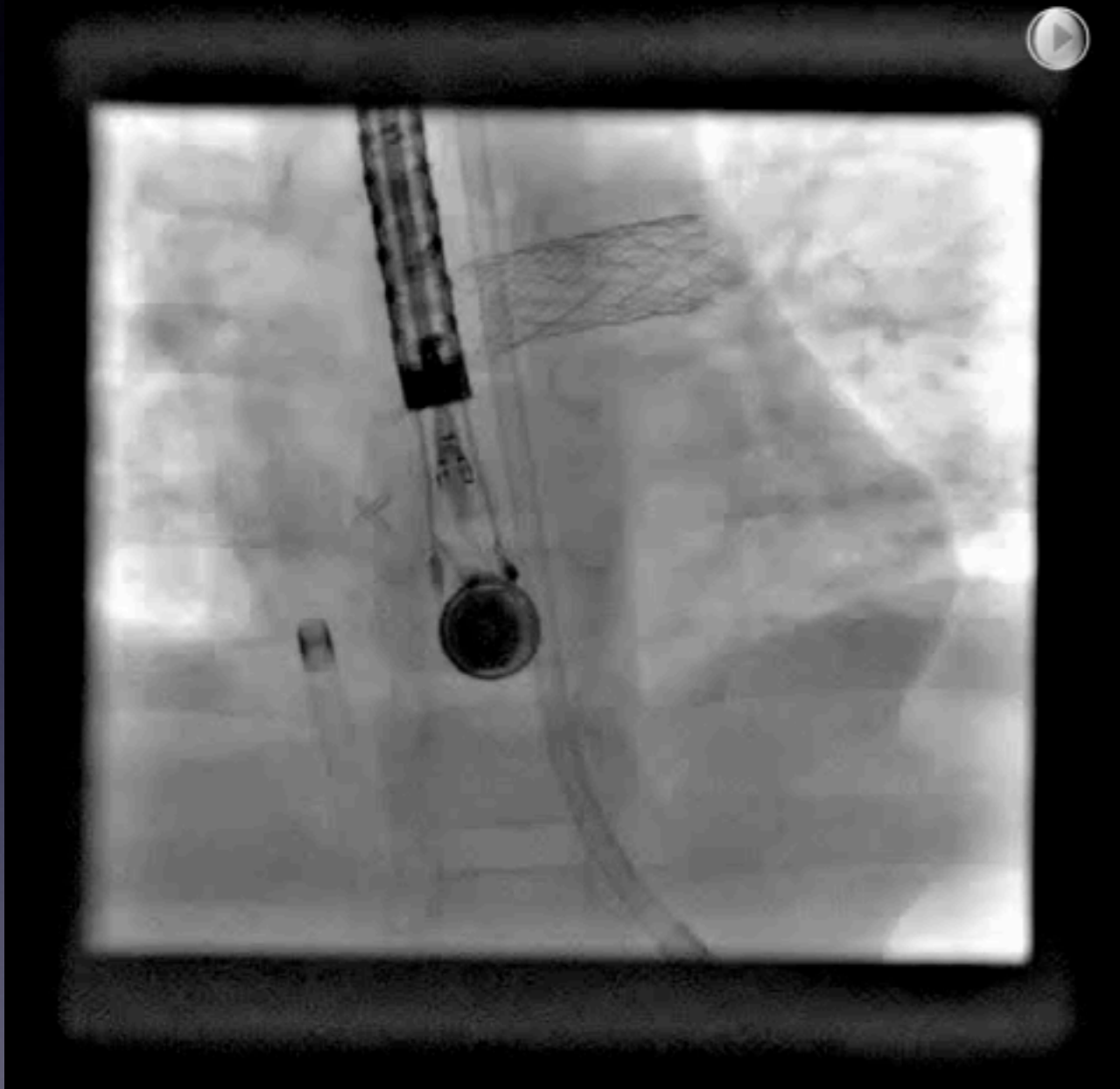
- 8 months after Fontan developed progressively increasing ascites
- Treated with diuretics including metolazone, lisinopril
- O2 saturation 95%
- MRI cardiac catheter: normal right sided pressures and fenestration spontaneously occluded

Failing Fontan

- Angiography showed mild LPA stenosis
- So LPA stenting performed 8 months after Fontan operation
- Ascites kept recurring
- So taken to catheter lab for creation of fenestration

Failing Fontan

Creation of fenestration

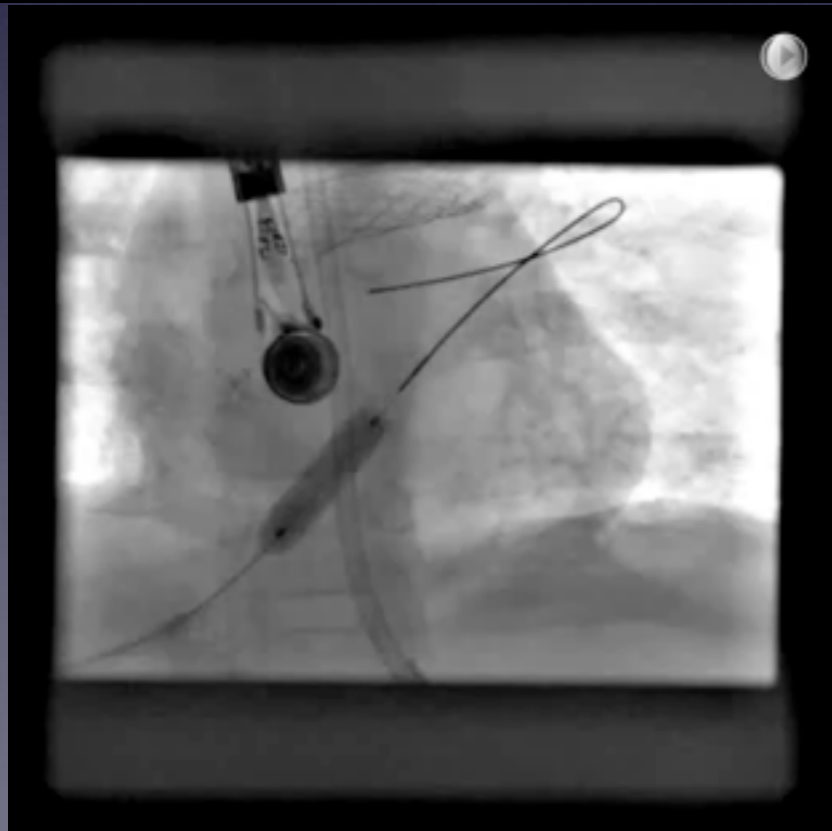
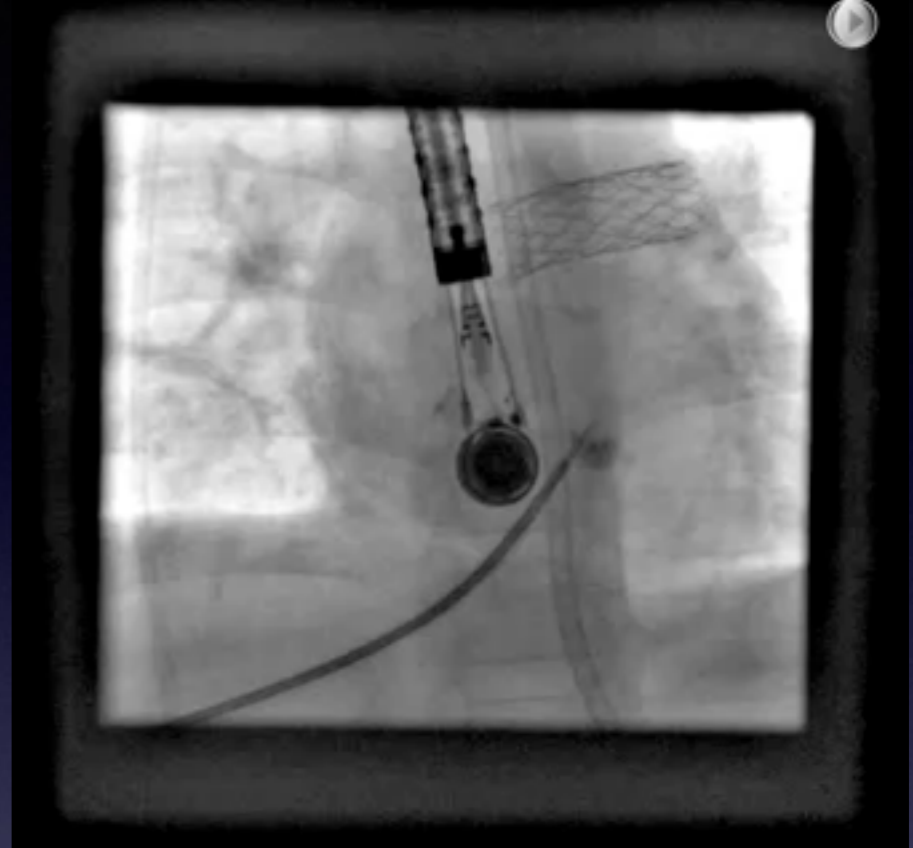
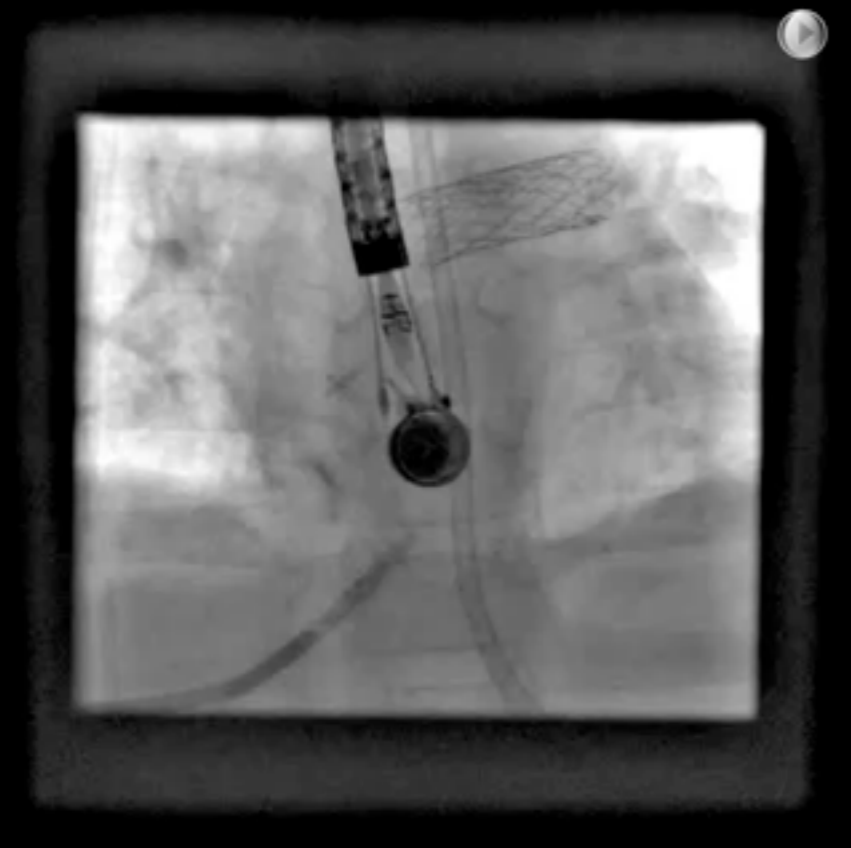


•Trans-septal approach from femoral vein failed

•So trans-hepatic approach attempted

Failing Fontan

Creation of fenestration



Failing Fontan

Creation of fenestration with premounted Genesis 5mm x 18 mm

- Saturations decreased from 90% to 75% immediately after the procedure
- Over 8 days, saturations improved to 80%
- Ascites resolved over 2 months
- Latest follow up 8 years later, no recurrence of ascites
- Saturation 85%
- Maintained on aspirin, diuretics and lisinopril

Failing Fontan

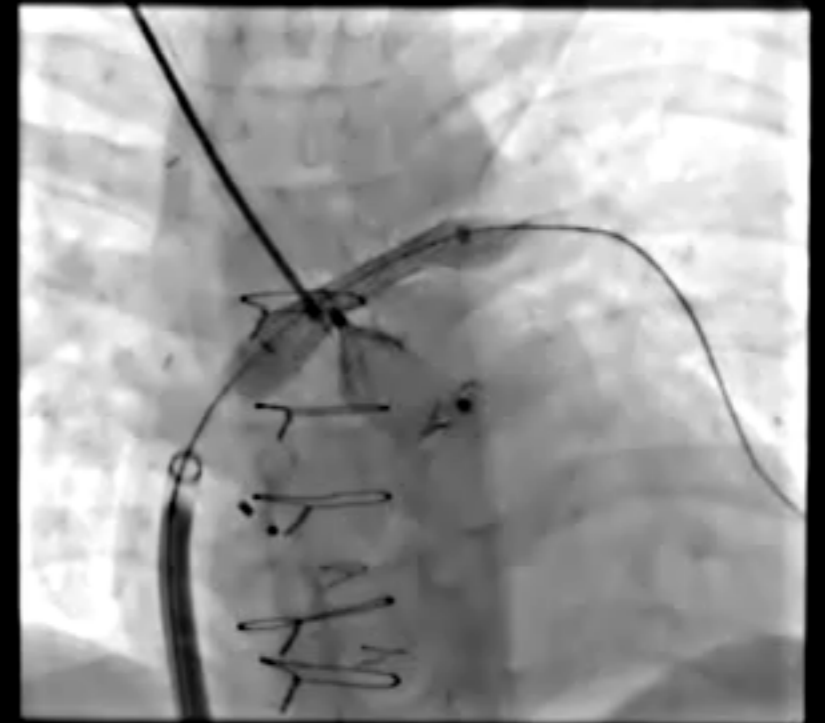
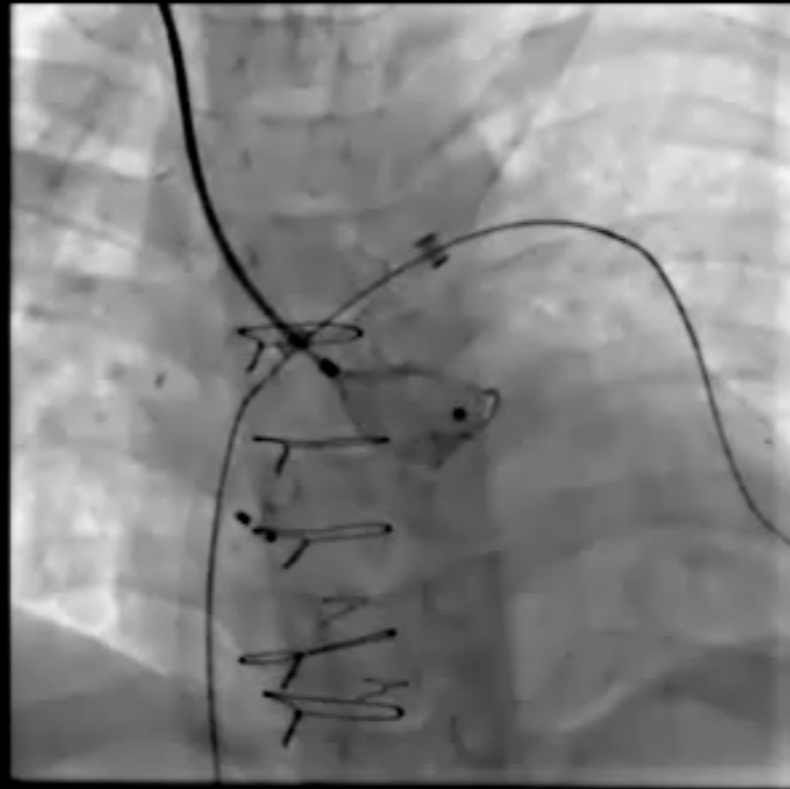
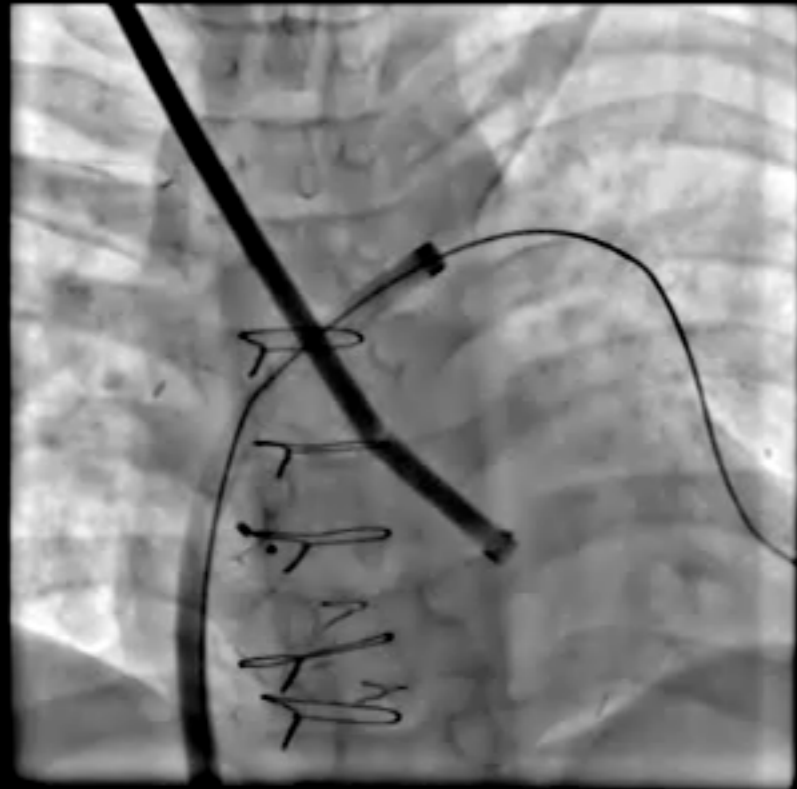
- 17 year old, original diagnosis of TGA, small VSD, straddling LAV valve, PVS
- Bidirectional Glenn shunt age 1 year
- Prior to Fontan, catheterisation showed PA pressure mean 18, ventricular EDP 12 mm Hg
- Had Fontan operation (+ MPA ligation) age 12 years
- Fontan fenestration closure 1 year later (2012)
- Presented with ascites 3 years later. Significant antegrade flow through MPA

Failing Fontan



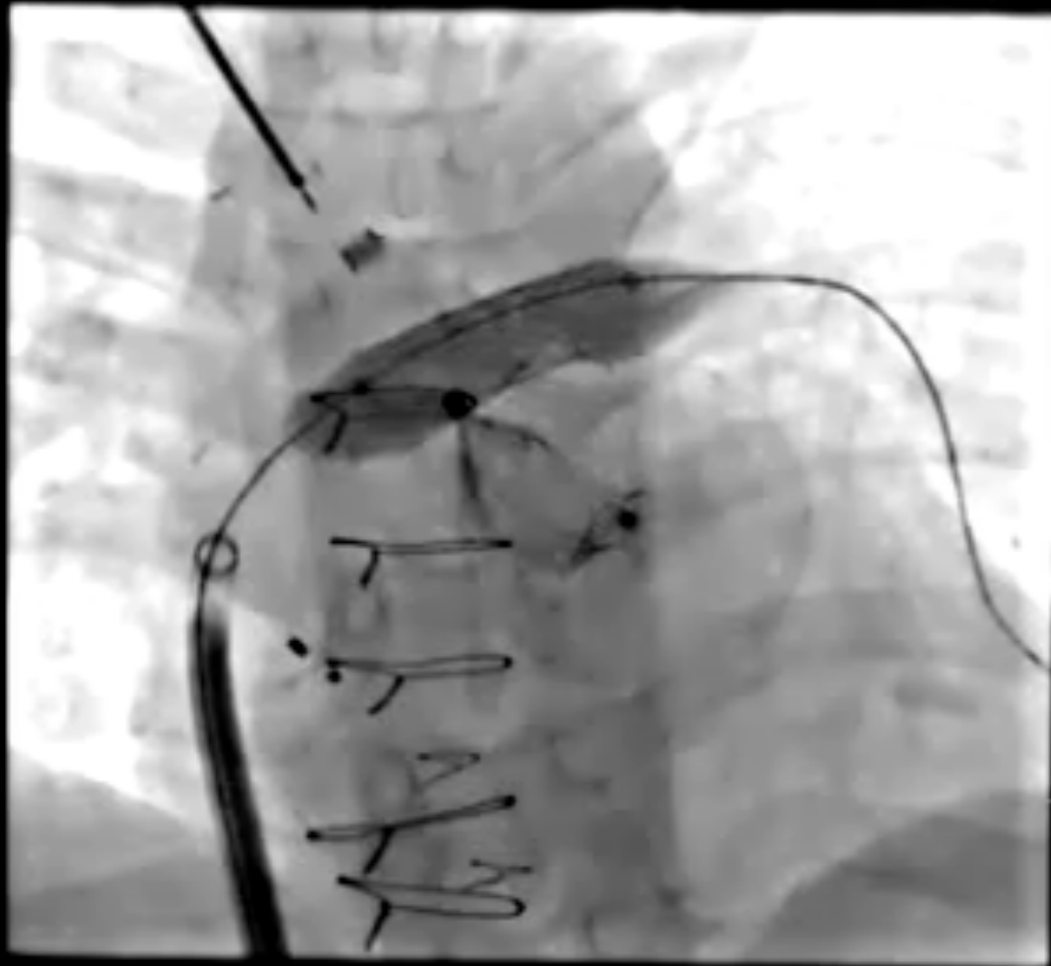
- SVC, PA pressure mean of 18mmHg

Failing Fontan



- 34 mm ASDO implanted combined with Palmaz 4010 stent on 18 mm balloon

Failing Fontan



- SVC/ PA pressure mean 15 mm Hg
- 3 months later, oedema disappeared, saturation 90%
- On diuretics, aspirin, enalapril and warfarin

Late Problems of Fontan circulation

Plastic bronchitis

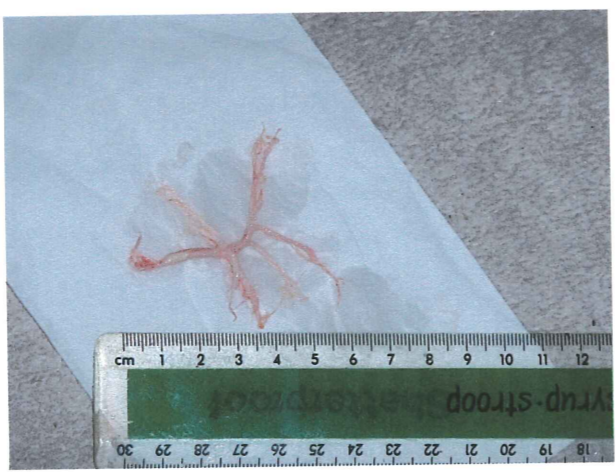
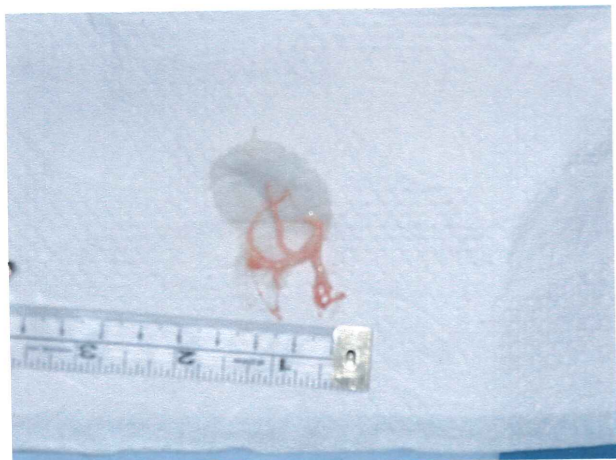
- Presentation: breathlessness, cough, wheezing, coughing up bronchial casts
- Early assessment and treatment: bronchial lavage, pulmonary vasodilators, cardiac transplantation

Failing Fontan

- Modified Norwood I age 2 days
- Hemi-Fontan with oversewing of left AV valve age 5 months
- Collateral occlusion: right and left sided collaterals age 1 year
- TCPC - 2 and 1/2 years
- Developed plastic bronchitis age 3 years (casts daily) - some improvement with medical treatment including steroids, physiotherapy, antibiotics, lisinopril, sildenafil
- MRI - RVEF 51%, unobstructed pathways
- Diagnostic catheter - normal pressures in Fontan circulation (PA mean 13 mmHg, LVEDP 8 mmHg)

Failing Fontan Plastic bronchitis - casts

7th
Jan
9pm
(in
hoop)

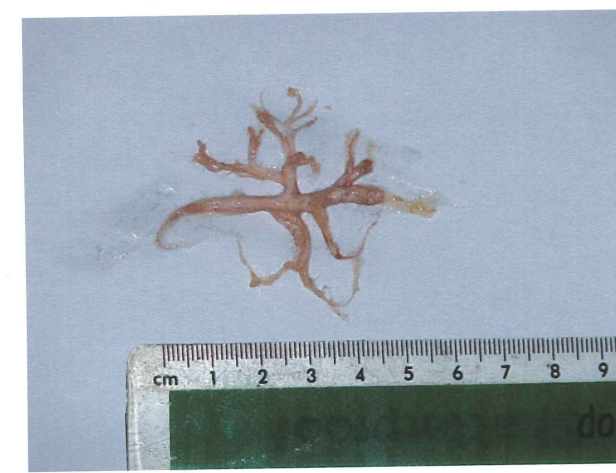


9th Jan
08h15

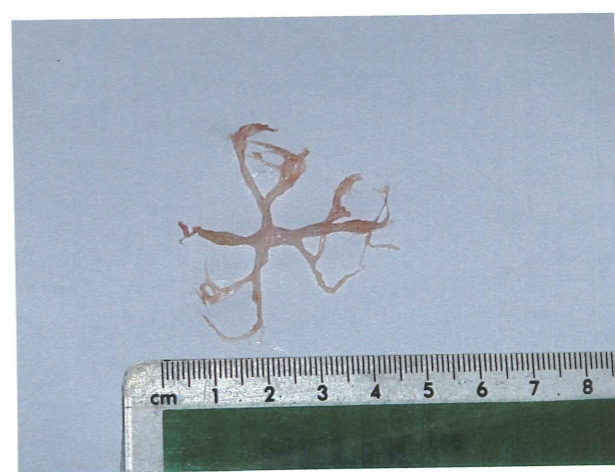
4th
Jan
2pm



8th Jan
12h00



6th Jan 6am

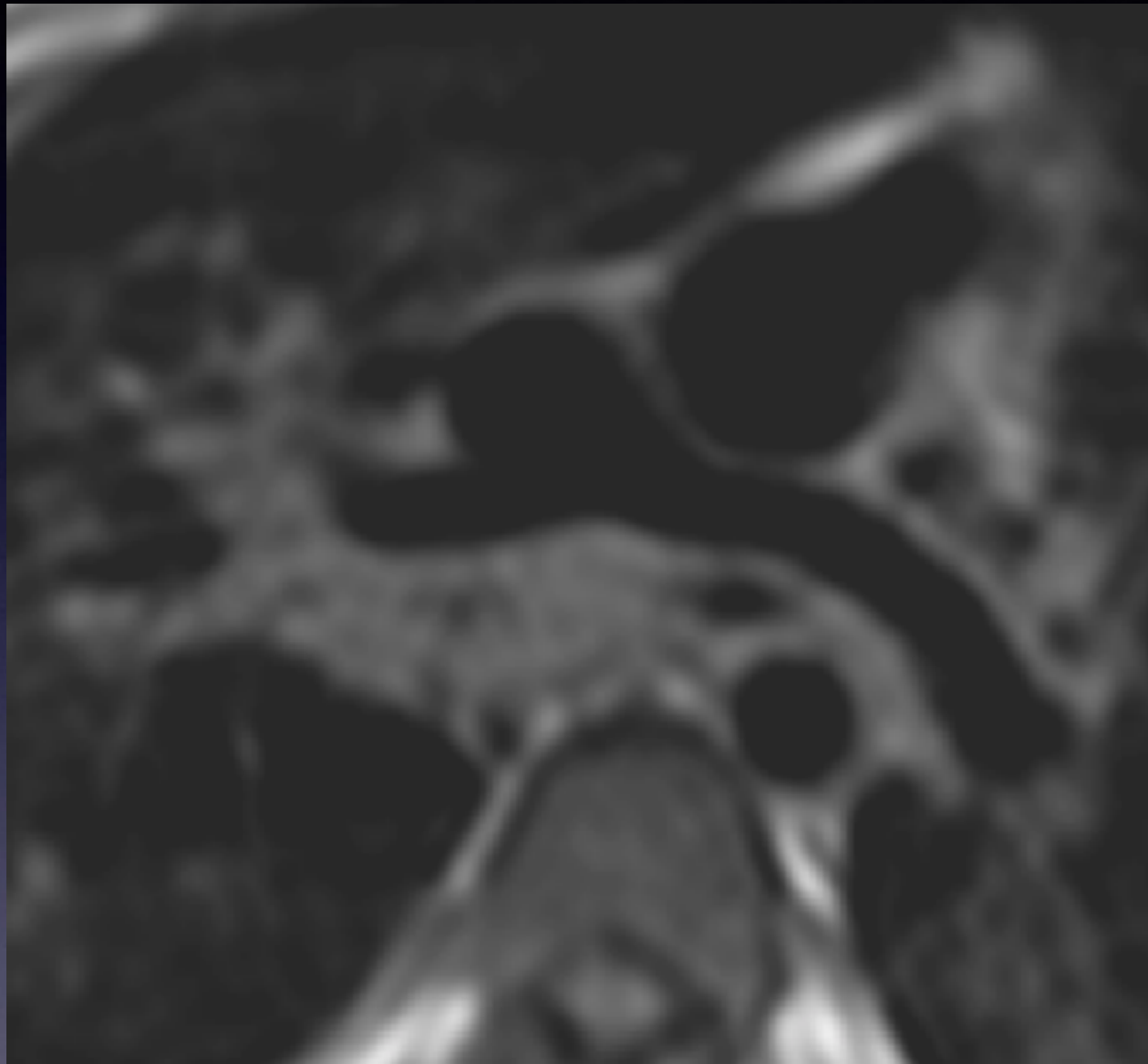


10 Jan
11am

11 Jan 2pm

Failing Fontan

Plastic bronchitis - Unobstructed pathways



- Proceeded to thoracic duct ligation

- Plastic bronchitis improved over 3 months

Evaluation of Failing Fontan

- Usually all modalities (echo, CT, MRI, cardiac catheterisation) are used for evaluation
- Management is never simple
- Interventions are often performed without haemodynamic basis in such low flow circulation
- A combination of treatments palliates patients to avoid need for transplantation (including pulmonary vasodilators, after-load reduction, PA stenting, creation of fenestration, occlusion of leaks or residual antegrade PA flow, baffle obstruction stenting, thoracic duct ligation (embolisation))