

Case 2 for UK Cardiac Network

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Congenital heart disease -

- 27 year old male, fit (given previous cardiac history)
- Born with Transposition of Great vessels at birth, with an associated VSD, and allegedly had a Mustard operation at 2 days of life (no arterial switch, as there was a single coronary artery). No immediate complications.
- In 2017, developed bradycardia and had a pacemaker fitted.
- Recently in 2018, collapsed in street with out of hospital cardiac arrest and died.
- No recent ardiograms

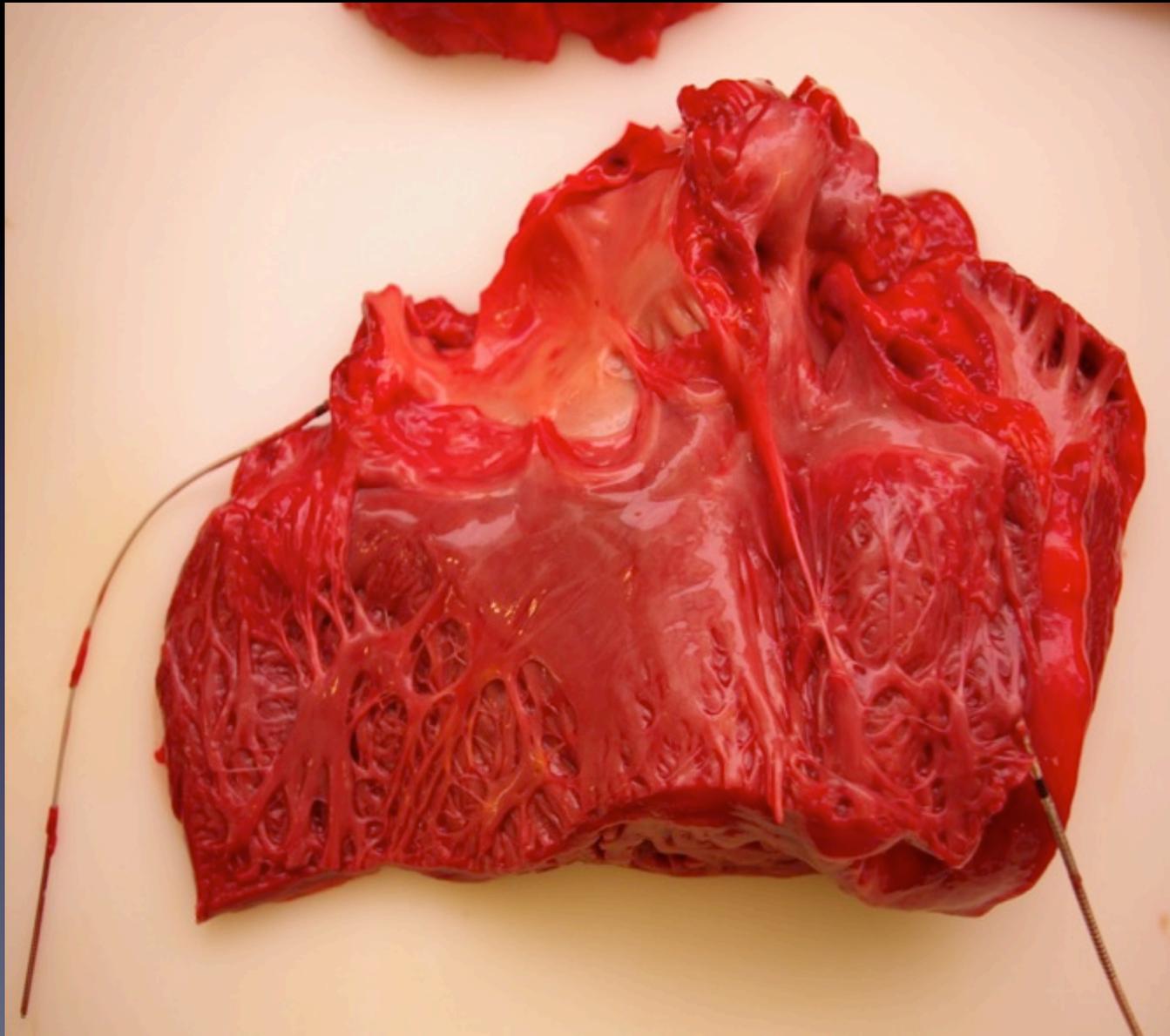
Internal examination

- Heart 818g (RV 55mm LV 45mm.)
- 'Grossly left ventricle looked like a right ventricle and the right ventricle looked like a left ventricle. (There was much coarser trabeculation of what should have been the left ventricle, and what should have been the right ventricle was much smoother and more compact, and looked like left ventricular myocardium.
- There was an 'aorta' leading from the ventricle on the left side, but it was in a position which corresponded with the position of the pulmonary artery in a normal right ventricle. Similarly the position of the pulmonary artery in the right sided ventricle resembled the position of the aorta in the left ventricle.
- There was a single coronary artery as was already known about.'
- VSD repaired but could not see the repair macroscopically.
- No evidence of complications with the conduits (SVC/IVC to left atria , and pulmonary veins to right atria) e.g. leaky, stenotic, thrombi?.
- The liver was acutely congested, and there was pulmonary oedema, in keeping with LVF.

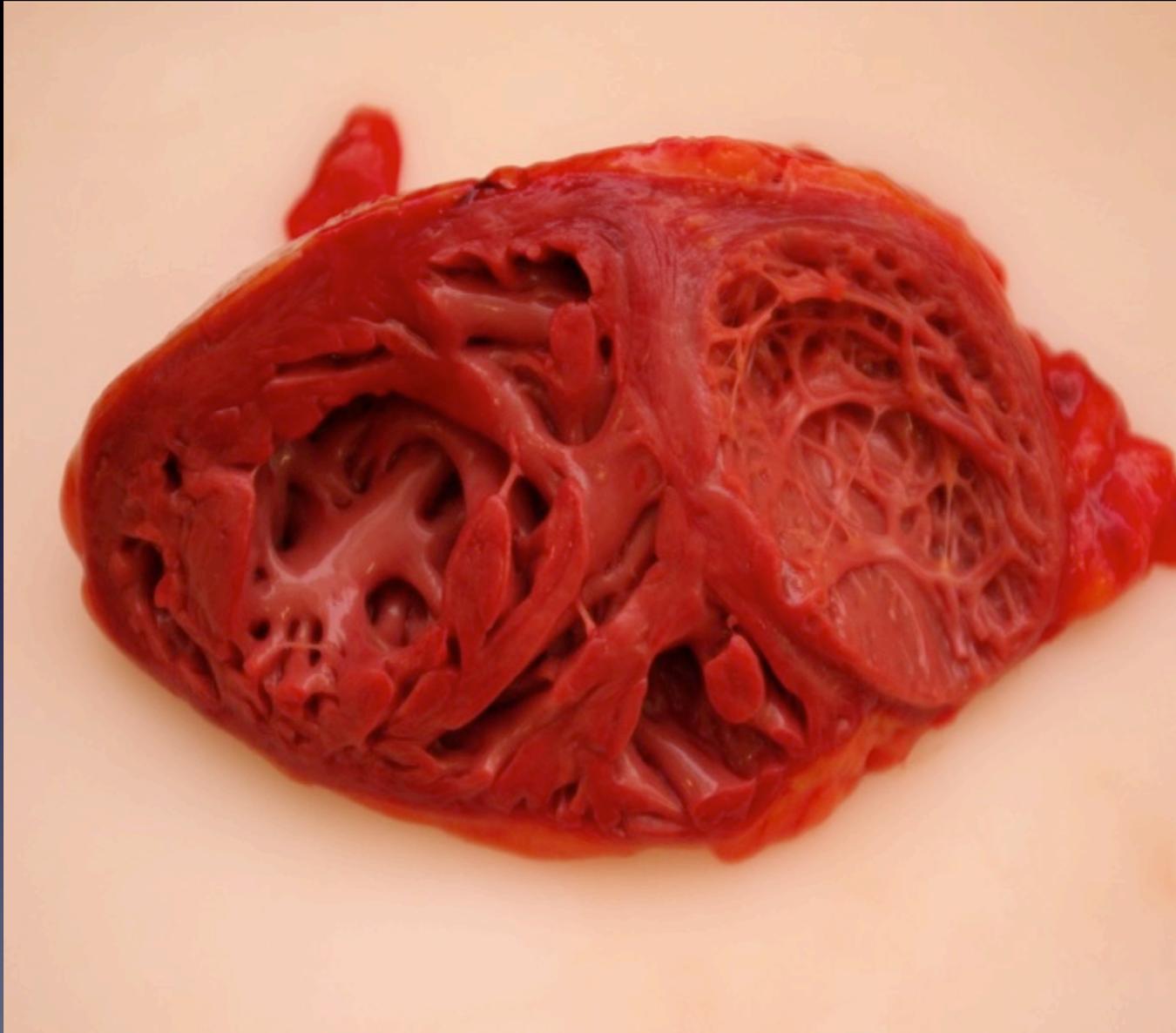
Histology

- Liver: Acute sinusoidal congestion. Normal architecture. No significant fatty deposition or inflammation.
- Lung: Focal haemorrhage. Generalised alveolar wall capillary congestion. Some pulmonary oedema and increased alveolar macrophages. No evidence of pneumonia
- Heart: Focal subendocardial myocardial fibrosis, seen in lateral left ventricular wall.

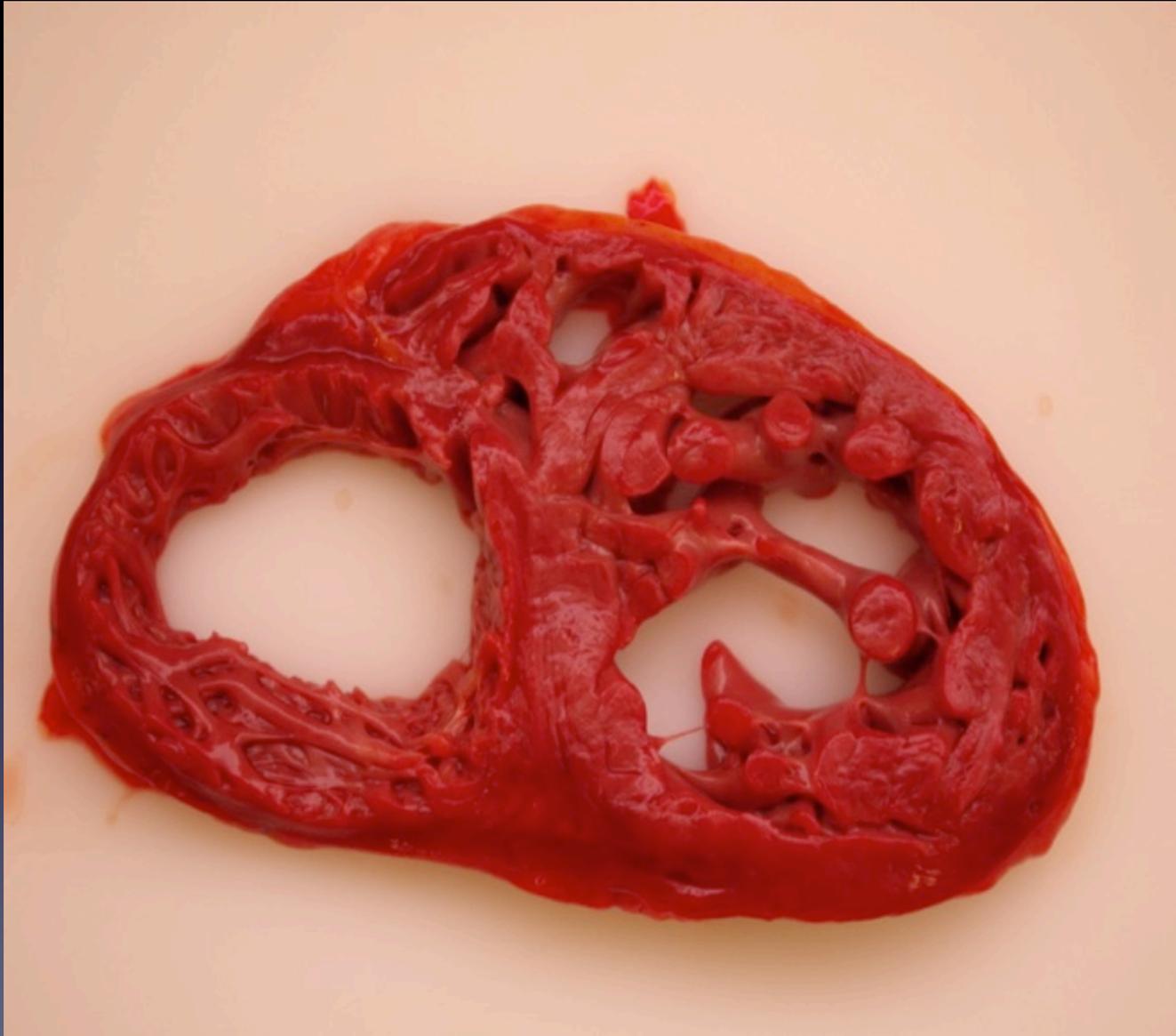
Right ventricle



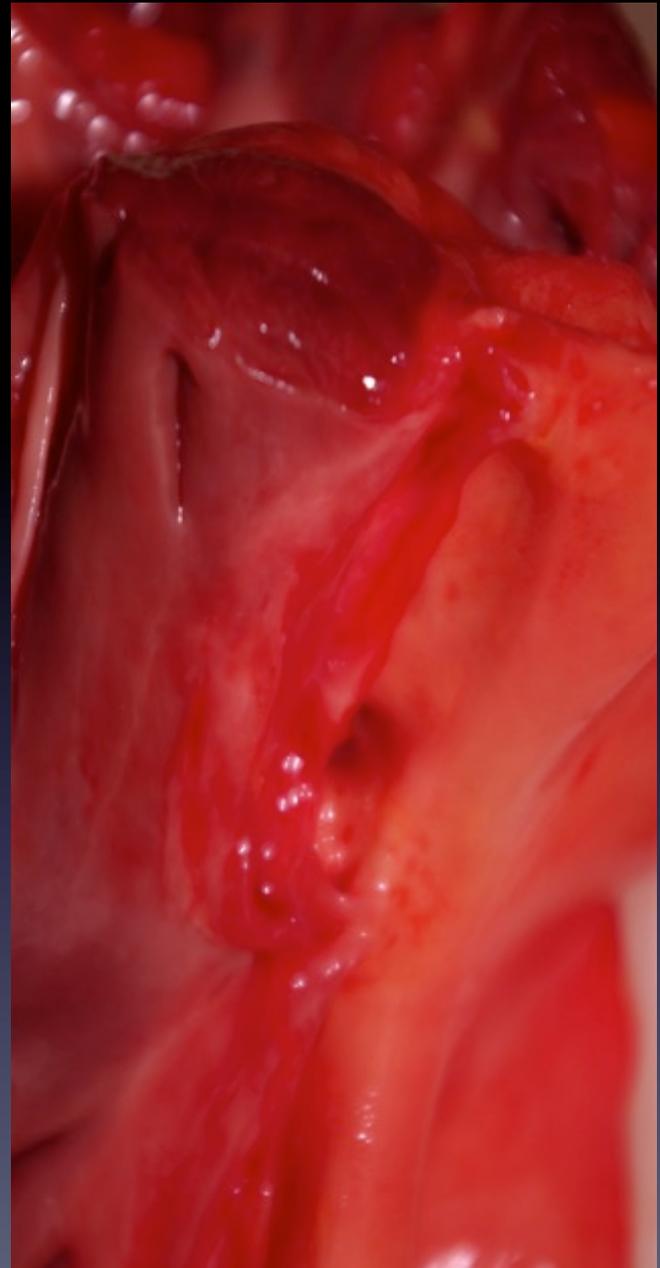
Apex



Mid-ventricular slice



One
coronary
artery



Thoughts?

- Recognised reality for complex congenital heart disease
- Often die (relatively acutely) in their 20-40 age range from dysrhythmia
- Cause poorly understood but likely reflect interstitial fibrosis around conduction system, myocyte drop-out and the surgery effects.
- Likely volume/pressure mismatch (ie non-physiological)
- Problems with the conduits (e.g. stenosis) and pump failure.