Fetal Ebstein Anomaly

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What we know

- ***Strongly associated with fetal demise or neonatal death****: Lack of antegrade pulmonary blood flow, PR, Larger TV annulus (esp >z-score 6), Abnormal UA pulsatility, low UV velocity, Dx < 32weeks GA, pericardial effusion.*

- ***Fetal interventions*** *include NSAIDS, O2, MPA occlusion.*
  - *Only NSAID have data (data still sparse, possibly confounded by increased use of Starnes procedure. NSAIDS have both complications of renal dysfunction and ductal closure, but these seem uncommon*
- ***Postoperative management****:*
  - *Most critical factors: circular shunting, ability RV to support pulmonary circulation*
  - *The latter may be challenging to predict at times and teams are often torn between waiting on the PDA to close and going straight to Starnes or ECMO/Starnes.*

What we need

- ***Studies limited to fetuses with any degree of PR***
  - *What predicts mortality in this subset? Some are okay while others decompensate quickly.*
  - *Which pts in this cohort survive waiting for the duct to spontaneously close.*
  - *Does every single one need to go straight to Starnes?*
  - *Besides hemodynamic instability postnatally, are there other factors that are highly reliable prenatally in this subset?*
- ***More universal genetic testing and studies of association with outcome.*** *It is unknown if MYH7 variants are a risk factor for worse outcome because often not tested.*

- ***Further consideration of MPA occlusion in the fetus****, which would allow for adequate pulmonary blood flow but cessation of the circular shunt.*