

Total anomalous pulmonary venous connection: Challenges with prenatal diagnosis



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Objectives:

Total anomalous pulmonary venous connection (TAPVC) is rarely detected prenatally. We reviewed all cases of TAPVC who underwent fetal echocardiography (FE) with a focus on indication for referral and time of diagnosis, to improve prenatal recognition.

Results:

- Various experienced operators scanned 18 cases over 21y
- Most women (n=15, 83%) were referred in last 10y when the 3VT view was added to the national cardiac screening protocol (FASP; Fetal Anomaly Screening Programme, **Fig 1**) and over half in the last 5y (n=11, 61%).
- Indications for FE: Most were abnormal screening views (n=13, 72%) of which one was for suspected TAPVC (**Fig 2**)
- Median gestational age at 1st scan = 22.1w (range:15.3 to 34) and at diagnosis = 23.4w (range: 20.4 to 31.9).
- Seven cases (39%) were diagnosed on 1st scan, eight on follow up (44%) and three (17%) were overlooked antenatally and diagnosed postnatally.
- Seven (39%) had intracardiac drainage, six (33%) were supracardiac (**Fig 3**), four (22%) infracardiac and one (6%) was of mixed type.
- TAPVC was isolated in nine cases (50%). Six had VSDs, one had BSVCs. An aberrant right subclavian artery was found in five cases (28%)
- Extracardiac associations: fetal growth restriction (n=2), cleft lip and palate (n=1), short long bones and hyperechogenic bowel (n=1)
- Genetic associations: Two neonates had spinal muscular atrophy and one had Cat-Eye syndrome

Methods:

- Retrospective study of cases scanned antenatally in three tertiary hospitals in London (UK) from 2003-2024, including those overlooked on FE
- Patient and pregnancy characteristics, available prenatal sonographic data and postnatal outcomes were reviewed.
- Cases with abnormal situs or major congenital heart disease were excluded
- Cases with ventricular septal defect (VSD) or bilateral superior vena cava (BSVC) were included

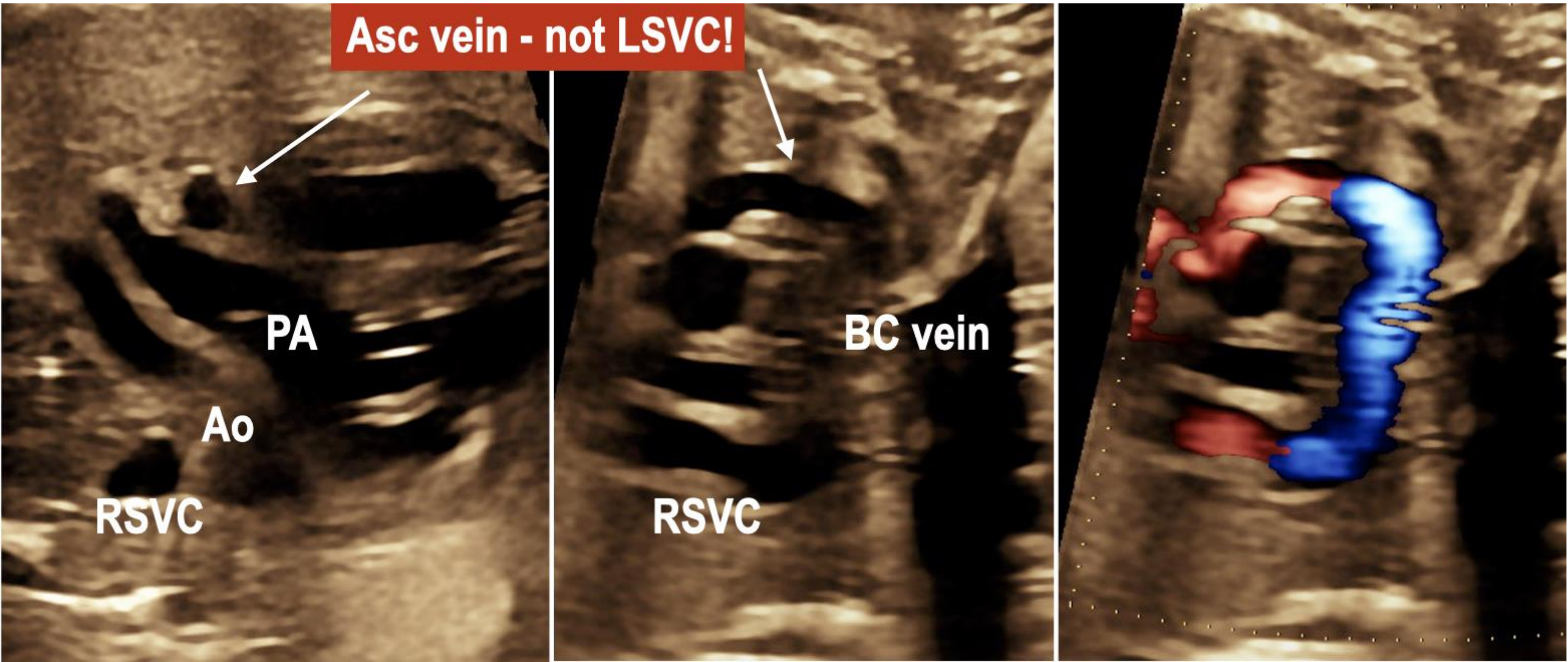


Figure 3. Case referred for suspected BSVCs. There was supracardiac TAPVC to the brachiocephalic vein (BC). Note the ascending (asc vein) simulates a left SVC

TAPVC cases (2003 - 2024)

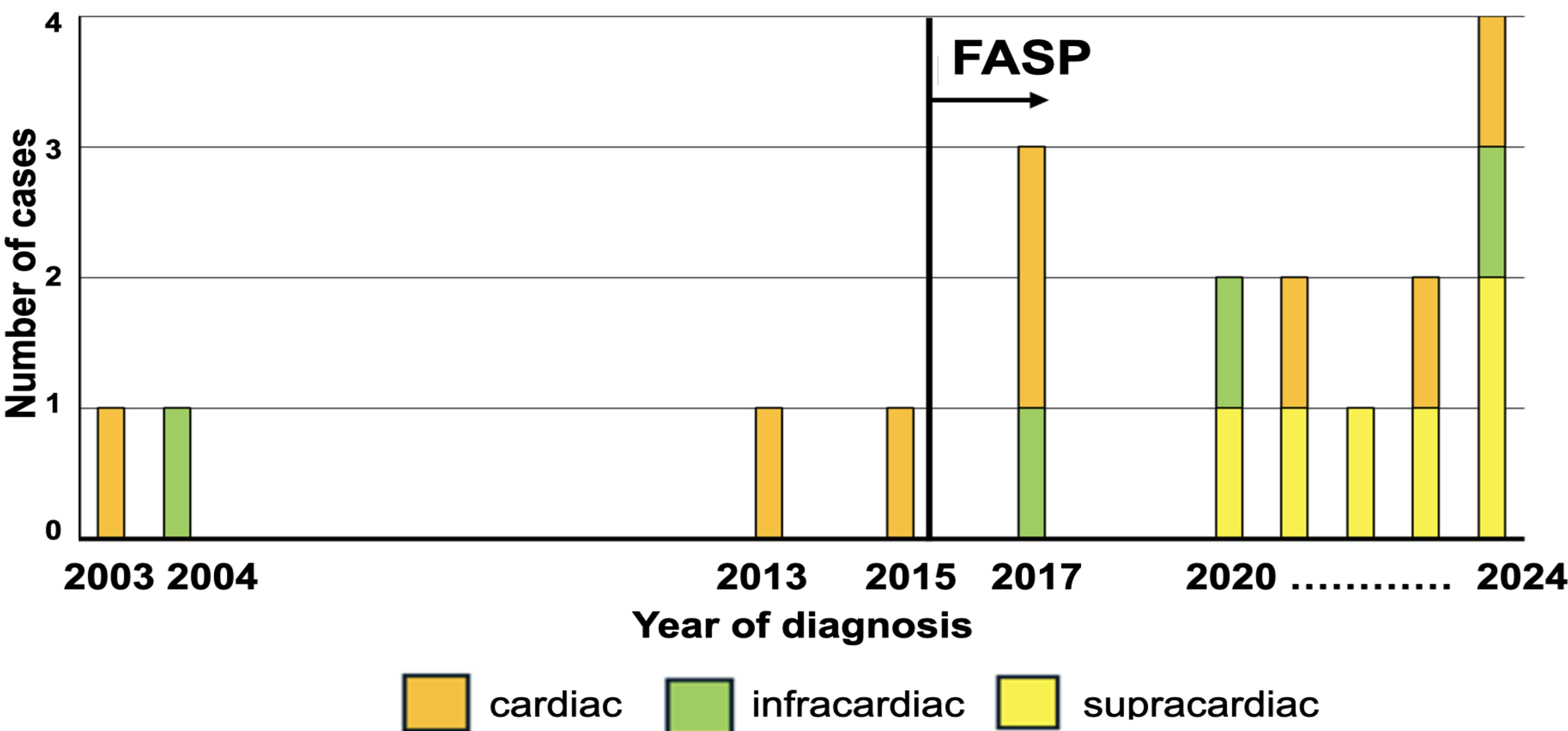


Figure 1. FASP impact on prenatal diagnosis

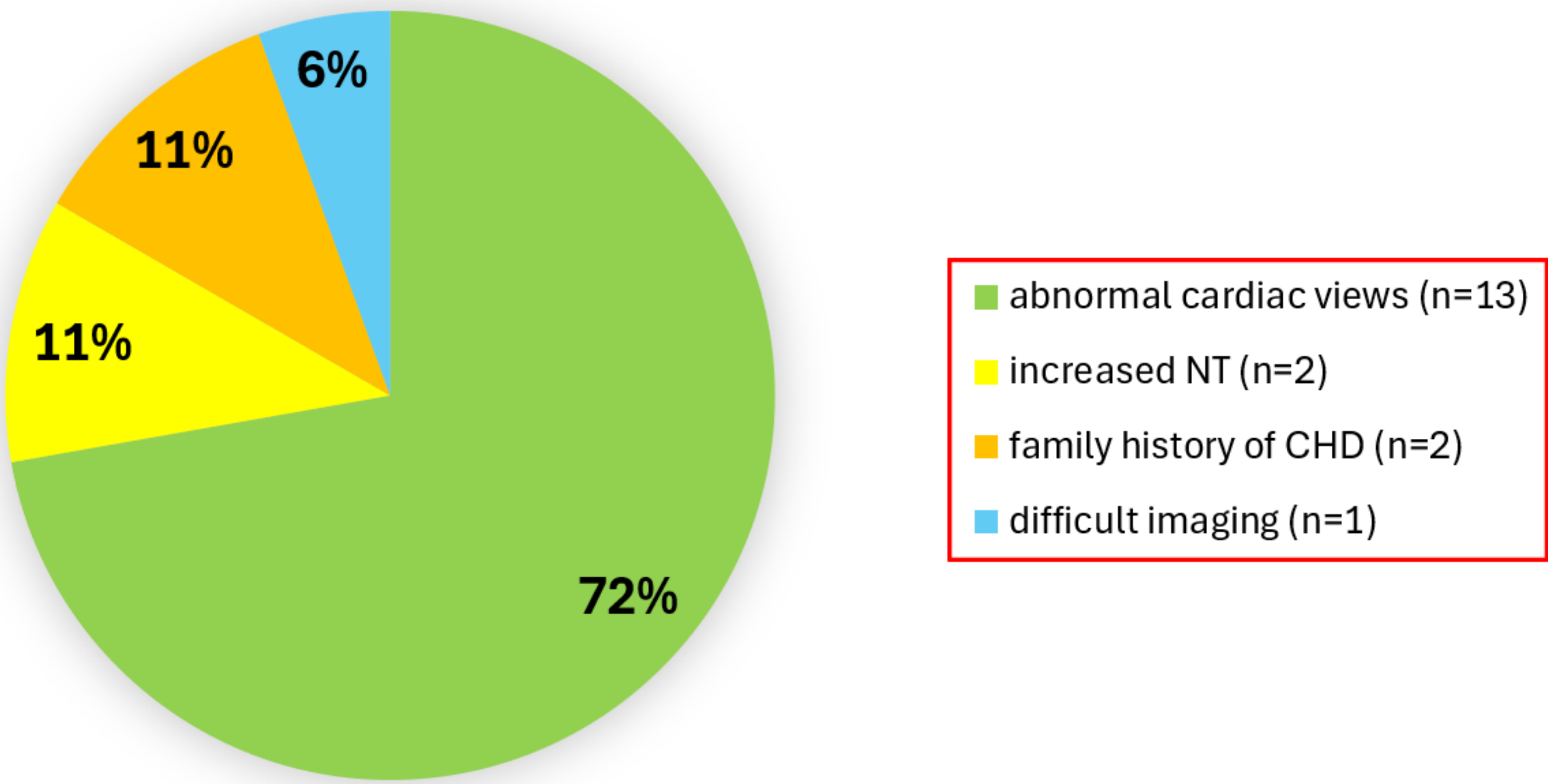


Figure 2. Indications for FE referrals

Conclusion:

- Prenatal diagnosis of TAPVC remains challenging. It requires a high degree of suspicion even for experienced operators.
- Most cases are referred for abnormal cardiac views (unrelated to pulmonary veins).
- A low threshold for referral and a high degree of suspicion during FE may improve fetal diagnosis.