Absent pulmonary valve: a prenatal case series
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Objective
To analyse the proportion of absent pulmonary valve syndrome (APVS) amongst a cohort of prenatally diagnosed fetal cardiac lesions, and to describe selected cases (including clips), depicting the variable presentations of this rare cardiac anomaly.

Methods
Search of departmental database including case records, from January 2012 to December 2016. Cases with adequate volumes and cine clips were only included.

Results
A total of eight cases were confirmed as APVS (Total CHD = 422, TOF = 65). The evaluation of the three vessel view, with and without, is critical in making this diagnosis, as some of these cases may not show dilatation of PA and its branches, especially early on in gestation. Case 1 presented at 14 weeks following referral for fetal hydrops. An early targeted scan revealed hydrops, alobar holoprosencephaly, abnormal facial profile, overlapping digits, findings of dilated right atrium (RA) and severe pulmonary regurgitation (PR) during systole [a to - and fro pattern of flow across the pulmonary valve (PV)]. Fetal Karyotype (KT) done via chorionic villus sampling (CVS) showed Trisomy 13. Case 2 presented at 11+3 weeks following referral for hydrops. An early target scan revealed hydrops, enlarged RA and RV and same findings as Case 1. Fetal KT done via CVS demonstrated a Robertsonian translocation - 47, ###, rob (13; 13) (q10; 10) +13. (CLIPS). Case 3 presented at 23+2 weeks, after referral for suspected cardiac anomaly. Target scan revealed enlarged RA, dilated branch pulmonary arteries with alternating to - and fro flow across PV. Aliasing was noted during systole and severe PR during diastole. Fetal KT revealed Trisomy 18. Case 4 was a 24 year old antenatal woman, a school dropout, with h/o surgery for cleft palate in childhood. She was referred for cystic lesion behind fetal heart. Target scan revealed cardiac findings same as Case 3 (STIC volume of this case is described via images and cine clips). Couple refused further workup.

Conclusion
A high index of suspicion needs to be maintained based on the characteristic color Doppler findings and RA dilatation. Prompt prenatal diagnosis of APVS should pave the way to additional diagnostic studies.