HIGH-IMPACT CLINICAL CASES IN FETAL CMR

This section highlights clinical cases that demonstrate the transformative impact of fetal CMR in prenatal care. Captured using different scanner vendors, each case demonstrates the versatility of this technology in addressing complex diagnostic challenges and optimizing prenatal management.

PHILIPS scanner, 3T

ABSENT PULMONARY VALVE SYNDROME WITH TETRALOGY OF FALLOT (APVS/TOF)

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CASE PRESENTATION

- A 31-year-old woman from Bangladesh (gravida III, para II) with a BMI of 26 at MRI presented with suspected APVS/TOF during second trimester anatomy scan and was referred for detailed diagnostic evaluation.
- Pregnancy history indicated early postnatal death of the first child due to suspected CHD.
- Prior to referral, fetal DiGeorge syndrome was diagnosed via amniocentesis.

INVESTIGATION

- Fetal echocardiography confirmed APVS/TOF with associated agenesis of the ductus arteriosus, aneurysmal branch pulmonary arteries, and right ventricular hypertrophy; also, a left persistent SVC was suspected.
- A fetal CMR was performed at gestational week (GW) 37+6 to validate the diagnosis and evaluate the size and extent of pulmonary artery aneurysms.

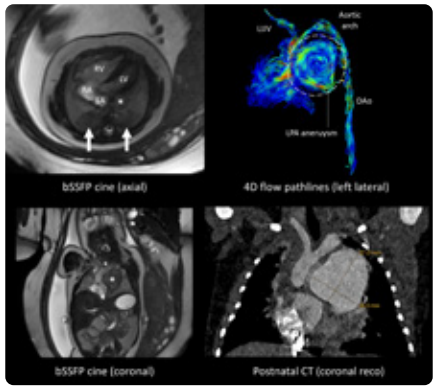

Figure 4: SbSSFP cine images reveal mesocardia/rightward shift of the heart, along with RV enlargement and hypertrophy. Characteristic of APVS, aneurysmal branch pulmonary arteries were observed, including a giant LPA aneurysm (asterisks), measuring 27 x 26 mm on postnatal CT, showing vortical flow at 4D flow pathline visualization. Prenatal axial cine MRI also revealed hypointense changes in both lower lung lobes, indicative of early pulmonary involvement (arrows).

- MRI scan protocol included T2w TSE images, axial and coronal bSSFP cine images, and 4D PC flow imaging.
- MRI findings were: APVS/TOF with agenesis of the ductus arteriosus; mesocardia with right ventricular enlargement and hypertrophy; aneurysmal branch pulmonary arteries with a giant LPA; absence of LSVC; structural lung changes in both lower lobes, likely due to compression of the hilar structures.

DIFFERENTIAL DIAGNOSIS

- Fetal CMR confirmed the previously suspected diagnosis of APVS/TOF, including agenesis of the ductus arteriosus.
- In addition to previous echocardiography, fetal CMR revealed mesocardiac/rightward shift of the heart, which affected the right hilar structures in addition to the RV enlargement; also, the extent of the giant LPA aneurysm could be assessed. The presence of a LSVC could be excluded *(Figure 4)*.

OUTCOME AND FOLLOW-UP

- The child survived the initial corrective surgery and subsequent follow-up operations but suffers from severe failure to thrive, attributed to recurrent pneumonia with respiratory insufficiency and right heart failure caused by concomitant tracheobronchomalacia.

TAKE HOME MESSAGES

- In addition to evaluating intra-cardiac and cardiovascular anatomy, fetal CMR offers the distinct advantage of visualizing the anatomical relationships between the heart and lungs, as well as between the thoracic vessels and bronchi. Therefore, the clinical report should emphasize a detailed description of these anatomical relationships and assess potential adverse postnatal effects.

GE scanner, 1.5T

DORV OR TOF IN A FETUS WITH VSD, ABNORMAL ORIGIN OF THE AORTA AND HYPOPLASTIC MAIN PULMONARY ARTERY?

Authors: *Julia Geiger*, Emanuela Valsangiacomo Buechel***
Institution: *University Children's Hospital Zurich, *Imaging Department, **Department of Ped Cardiology*

CASE PRESENTATION

- A 32-year-old G1P0 pregnant woman was referred for fetal CMR at GW 34 after fetal echocardiography suggested TOF with ventricular septal defect (VSD) and right ventricular outflow tract (RVOT) stenosis. The exact morphology of the pulmonary bifurcation and side branches could not be defined.
- Fetal CMR was requested because of limited echocardiographic image quality due to maternal obesity.

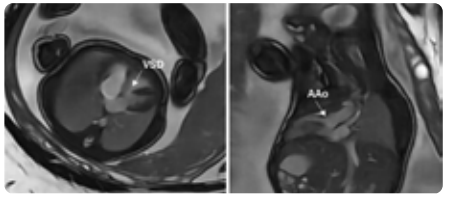

Figure 5: The axial and sagittal oblique DUS-gated cine bSSFP images depict the large VSD with overriding AAo.

INVESTIGATION

- Echocardiography at GW 28 showed balanced ventricles connected by a large VSD with overriding of the aorta. The RVOT, main and branch pulmonary arteries appeared hypoplastic and the pulmonary artery bifurcation was not visible. In addition,

double outlet right ventricle (DORV) was expressed as a possible differential diagnosis for TOF.
- Genetic assessment revealed no abnormalities, in particular no suspicion of DiGeorge syndrome.
- Fetal CMR at GW 34 showed symmetric ventricular size and a large VSD with overriding aorta arising from both ventricles *(Figure 5)*. The RVOT and main pulmonary artery (MPA) could be visualized showing a smaller size compared to the ascending aorta. The PA bifurcation and both PAs could be identified, whereas the arterial duct was hardly visible *(Figure 6)*.

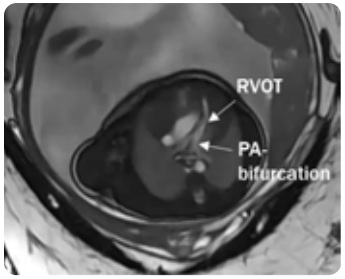

Figure 6: The axial DUS-gated cine bSSFP image shows the hypoplastic RVOT and MPA, but a non-stenotic pulmonary bifurcation and pulmonary arteries.

DIFFERENTIAL DIAGNOSIS

- Fetal CMR demonstrated the origin of the ascending aorta overriding the VSD. The differential diagnosis of DORV could be excluded and the diagnosis of TOF was favoured.
- In contrast to echocardiography, the anatomy of the RVOT, pulmonary bifurcation and pulmonary arteries was clearly visualized.

OUTCOME AND FOLLOW-UP

- Postnatal echocardiography confirmed the diagnosis of TOF with malalignment VSD, overriding aorta, and mild hypoplasia of the MPA and pulmonary arteries. Transcutaneous oxygen values were within the desired range and the newborn did not require any cardiac neonatal interventions.

TAKE HOME MESSAGES

- Fetal CMR overcame echocardiography limitations from maternal obesity, accurately diagnosing TOF with VSD, overriding aorta, RVOT/MPA stenosis, and hypoplasia but patent pulmonary artery bifurcation and side branches. Combined with echocardiography, it enabled optimized perinatal management, avoiding prostaglandin use.

SIEMENS scanner, 1.5T

FETAL CMR FOR POSTNATAL INTERVENTION PLANNING IN OBSTRUCTED INFRADIAPHRAGMATIC TOTAL ANOMALOUS PULMONARY VENOUS RETURN

Authors: *Eleanor Schuchardt and Team*
Institution: *Rady Children's Hospital Heart Institute, University of California San Diego*

CASE PRESENTATION

- 28-year-old gravida-3 obese (BMI 47) female with a fetus, prenatally diagnosed with complex CHD including DORV, subaortic ventricular septal defect, pulmonary stenosis and infradiaphragmatic total anomalous pulmonary venous return (TAPVR).
- Fetal echocardiography suggested pulmonary venous obstruction with loss of pulsatility. The vertical vein insertion site was presumed to be near the ductus venosus but could not be definitively identified.
- The postnatal care plan considered a staged interventional approach, with percutaneous stenting of pulmonary venous obstruction, followed by surgical repair.

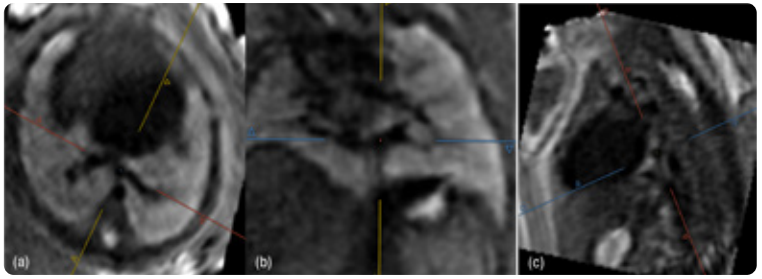

Figure 7: After motion correction using the slice to volume reconstruction tool kit, the multiplanar reconstruction was utilized to systematically assess the anatomy. Transverse (a), coronal (b) and sagittal (c) views demonstrate the inferior pulmonary veins meeting the confluence.

INVESTIGATION

- Three echocardiography scans at GW 25, 29 and 34. Imaging limited due to maternal body habitus and fetal movement.
- Fetal CMR with *smart-sync* gating was performed and showed the pulmonary venous drainage. T2 black blood imaging was found to be most useful for the intrathoracic anatomy.
- Slice-to-volume (SVR) reconstruction with Northh Medical's post processing platform was performed to define intrathoracic anatomy *(Figure 7)*. Below the diaphragm, SVR was less effective due to reduced vessel-liver contrast. Cine imaging clarified pulmonary venous drainage, showing the vertical vein joining a left-sided vein before reaching the ductus venosus.

DIFFERENTIAL DIAGNOSIS

- Fetal echocardiography identified the key elements (complex intracardiac anatomy and infradiaphragmatic TAPVR), however, the vertical vein's connection to the systemic vein was unclear. Fetal CMR clarified this, offering a 3D reconstruction as a backup in the event of no postnatal CT, boosting provider confidence in postnatal planning *(Figure 8)*.

OUTCOME AND FOLLOW-UP

- After stenting of the ductus venous, the infant was allowed to recover, transition and grow.
- Complete intracardiac repair and TAPVR repair was successfully performed at 3 months of age.
- Now at 4 months, the infant is doing well with no pulmonary venous obstruction and has mild pulmonary hypertension, improving on sildenafil.

TAKE HOME MESSAGES

- Fetal CMR provided a clear characterization of pulmonary venous drainage, enhancing confidence in postnatal care planning.

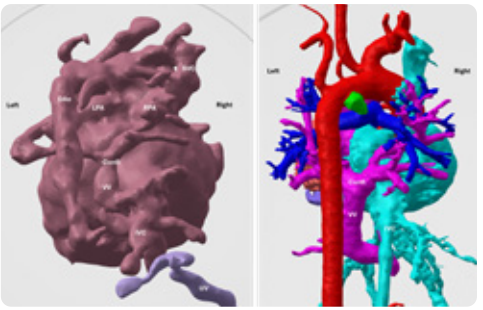

Figure 8: A 3D reconstruction was generated using Mimics. Here the posterior view shows the pulmonary venous confluence (Confl) to the vertical vein (VV), which ultimately crosses rightwards and drains to the superior aspect of the IVC.