Introduction
This case describes preoperative and intraoperative imaging used to diagnose and assist repair of a right coronary artery (RCA) to right atrial (RA) fistula in a twelve day old neonate.

History
A female infant of 34 + 4 weeks postconceptional age presents for ligation of a large RCA to RA fistula. The patient was asymptomatic from birth and serum troponin T remained low. Echocardiography on the first day of life demonstrated a proximal RCA diameter of 0.33 cm and confirmed the fistula with shunting from the RCA origin to the RA with peak velocity 1.0 m/s, mild RA and right ventricular (RV) dilation, mildly depressed RV systolic function and low normal left ventricular (LV) systolic function.

Operative Course
Intraoperative transesophageal echo (TEE) showed a dilated proximal RCA that bifurcated 1 cm beyond the aortic origin, giving rise to the fistula which emptied into the RA at the base of the RA appendage. The fistula was mobilized, encircled and temporarily occluded with TEE employed to demonstrate absence of flow from the fistula into the RA. Normal RV function on TEE and unchanged ST segments during test occlusion confirmed that the distal RCA was unaffected. The fistula was thus ligated. Postoperatively, the patient was quickly weaned off all cardiac medications and to room air following extubation on POD2. Her postoperative transthoracic echo demonstrated no residual fistula, normal appearing distal RCA, mildly dilated RA with normal RV and LV systolic function.

Discussion
Coronary artery fistulae (CAF) are abnormal direct communications between any coronary artery and any cardiac chamber or vessel. They can be iatrogenic, but are more commonly congenital and thought to arise from persistent embryonic intratrabecular spaces and coronary sinusoids. CAF is a rare defect, occurring in 1 in 50,000 live births and representing 0.2-0.4% of congenital heart defects.

Fistulas drain predominantly into low pressure structures with 92% draining to the right side of the heart. The risk of developing complications from a CAD increases with age. These include aneurysm formation, myocardial ischemia and infarction, congestive heart failure, arrhythmias, bacterial endocarditis and rupture. As such, the current treatment philosophy is to intervene early. However, the need to treat asymptomatic children has been debated because of case reports and series documenting spontaneous closure of congenital CAF, as well as a lack of information regarding long-term complications of intervention.

Here we describe a case of a large RCA to RA fistula initially diagnosed in fetal life and confirmed on postnatal echo. In this case echo was sufficient to define the lesion preoperatively such that coronary angiography was not needed. Intraoperative TEE was integral to successful surgical repair.

Legend
Image 1: Intraoperative midesophageal aortic valve short-axis TEE view with and without color-flow Doppler demonstrating RCA to RA fistula.
Image 2: Intraoperative TEE with color-flow Doppler demonstrating RCA to RA fistula before (A) and after (B) test occlusion.

References
2. Ismail et al. BMJ Case Reports. 2012.