A case of ventriculo-coronary arterial communication in pulmonary atresia with intact ventricular septum
Kamel R, Abdel Salam SA, Badr I
Fetal Medicine Unit, Cairo University, Cairo, Egypt

Objective
To report a case report of ventriculo-coronary arterial communication in pulmonary atresia with intact ventricular septum.

Methods
A 23-year old woman (G2P1) with unremarkable family history was referred to our institution at 25 weeks of gestational age due to abnormal fetal heart. Fetal cardiac examination was done using 2 dimensional and 3 dimensional ultrasound (Glass body and color STIC mode) as well as color Doppler using an abdominal volume probe (Voluson E10, GE Healthcare, Zipf, Austria).

Results
Our ultrasound examination revealed severe hypoplasia of the right ventricle showing mural overgrowth with complete obliteration of the outlet part (infundibulum) and the apical trabecular part with only residual small inlet part (unipartite right ventricle), small sized, severely hypoplastic tricuspid valve annulus, yet, with patent tricuspid valve leaflets showing antegrade flow across with trivial early systolic tricuspid regurgitation, vigorous right to left flow shunt across foramen ovale between the small right atrium and the large left atrium, totally disconnected main pulmonary artery from the infundibular portion of the RV with non visualized pulmonary valve leaflets, ductal dependent pulmonary circulation. The small main pulmonary artery and its confluent central branches are seen with complete retrograde filling from the tortuous arterial duct, situs solitus with concordant atrioventricular and ventriculo arterial connections, the right coronary artery is seen dilated (ectatic) at its proximal and mid segments with a fistulous communication between the RV branch of the RCA and the right ventricular free wall. A bidirectional flow pattern in the RCA is seen with significant systolic retrograde flow inside.

Conclusion
Our final diagnosis was pulmonary atresia with intact ventricular septum associated with ventriculo-coronary communication (RCA to RV fistula). A well defined vascular structure was seen just cranial to the ectatic proximal RCA with a suspicious fistulous communication between the ascending aorta and left atrium. This finding was strikingly found in glass body rendering of an offline color STIC volume. Reviewing the 2D and color Doppler exam, we found that this vascular structure is the left atrial appendage having a chicken wing appearance. For unknown apparent reason, the follow up study at 28 weeks gestation revealed intra uterine fetal death. Parents refused pathological examination due to religious aspects.