Combined double chambered right ventricle and discrete subaortic stenosis
Abdel Latif S, Talaat S, Abd Alrahman B, Badr I
Kasr Alainy, Faculty of medicine, Cairo, Egypt

Objective
Double chambered right ventricle represents a very rare anomaly with reported incidence about 0.2% of all congenital heart defects. It may be associated with other cardiac malformations. Here, we present a case of combined double chambered right ventricle and discrete subaortic stenosis with no previous prenatal reports about such finding from which we could confirm that this association is of congenital origin.

Methods
A 32 year-old woman (G3P2) with unremarkable family history was referred to our office at 35 weeks gestation for suspected fetal cardiac anomaly.

Results
Ultrasound examination revealed four anomalous muscle bundles seen inside the right ventricle aligned in different planes (two are obliquely aligned and apically inserted and the rest are transversely aligned in a higher anatomical level) within its cavity. The higher one divides the right ventricle into proximal low chamber and distal high chamber (RVOT) without hemodynamic evidence of obstruction. A small ventricular septal defect is seen in the muscular septum showing left to right flow shunt. Unifocal attachment of short, thick tendineous cords is seen connecting both mitral valve leaflets to a dominant elongated papillary muscle causing posteromedial displacement of mitral valve orifice suggesting that the dominant papillary muscle is posteromedial in location while there is a small anterior and lateral rudimentary muscle connected to free wall of leftt ventricle not properly seen reaching mitral annulus. This strongly suggests parachute like asymmetric mitral valve. The ascending aorta is seen arising from the left ventricle through a fibro muscular tunnel (the posterior wall of the aorta not connected to anterior leaflet of mitral valve but rather to the free wall of the left ventricle) with a discrete membrane seen in subvalvular area. Both of which causes aortic stenosis clearly demonstrated on 2D, color and pulsed Doppler study (tunnel like and discrete form of subvalvular aortic stenosis). A final diagnosis of combined double chambered right ventricle (DCRV) and discrete subaortic stenosis (DSAS) was reached. A tunnel like subaortic stenosis with parachute like asymmetric mitral valve causing congenital mitral stenosis is also present without associated aortic coarctation. This picture of multiple left sided obstructive lesions represents an incomplete form of Shone syndrome.

Conclusion
To the best of our knowledge, this is the first case to be reported in prenatal literature about combined double chambered right ventricle (DCRV) and discrete subaortic stenosis (DSAS). This combination was first reported in1978 based upon cardiac catheterization study by Baumstark et al. Freedom and his colleagues suggested that the muscle bundles are congenital and the subaortic abnormality is acquired. However, an echocardiographic study was performed in 1988 by Vogel et al reported that one patient who was premature at the time of the study (35 weeks gestation) underwent echocardiographic assessment one week after delivery and showed this rare association leading to some support to the argument that this is possibly a congenital lesion. Our case gave us certainty to support that this association is of congenital origin.