



Anomalous origin of a Shepherd Hook ductus arteriosus: an obstructive kink should be of concern

Abdel Latif S, Sabry N, Badr I
Kasr Alainy, faculty of medicine, Cairo, Egypt

Objective

We present a case of anomalous origin of an abnormal shepherd hook ductus arteriosus from the left innominate artery in case of TOF with pulmonary atresia and high thoracic right aortic arch.

Methods

A 33-year-old woman, G3P2 presented at Kasr Alainy hospital at 24 weeks' gestation because of a suspected cardiac anomaly. Detailed fetal echocardiography was performed with application of 4D STIC by using high definition color flow angiography to evaluate the anatomy of various vascular structures.

Results

Our evaluation revealed dilated aortic root overriding a large outlet sub aortic VSD. The main pulmonary artery was seen totally disconnected from the infundibulum of the right ventricle (valvular pulmonary atresia) with hypoplastic both central confluent branch pulmonary arteries. The aortic arch was right sided with mirror image branching and in a high thoracic location (opposite to the sternoclavicular joint) (Figure 1, video 1). There was a retrograde filling of the main and central pulmonary arteries through a tortuous ductus arteriosus arising from the left innominate artery in a trifurcation pattern (cranially the left common carotid artery, horizontally the left subclavian artery and caudally the ductus arteriosus). This was concurrent with marked ductal folding and excessive lengthening on its way to reach the base of left pulmonary artery, mimicking a shepherd hook. Despite being at mid gestation, there was 2D sonographically evident annular constriction of the ductus arteriosus at its kink that was confirmed on high definition color Doppler study with spatiotemporal image correlation technique (Figure 2, video 1, 2). The thymus gland was also severely hypoplastic. A tetralogy of Fallot with pulmonary atresia associated with high thoracic right aortic arch, left ductus arteriosus and severe thymic hypoplasia was confirmed, which raised our suspicion of 22q11 chromosomal micro deletion for which amniocentesis and fluorescent in situ hybridization were performed and confirmed the presence of this chromosomal micro deletion. Parents opted for termination of pregnancy.

Conclusion

In our case, more than one imaging finding has important consideration. The ductus arteriosus was left sided while the aortic arch was right sided with mirror image branching with lack of vascular ring. The ductus arteriosus arised from the left innominate artery. No prenatal reports present about this anomalous origin despite being a well known very rare finding in postnatal life. Moreover, the ductus arteriosus showed excessive lengthening and severe kinking nearly 180 degrees mimicking a shepherd hook. At 2010, Constantine and his colleagues reported this abnormal ductal configuration in autopsy specimens of two cases, both showed segmental annular constriction by an obstructive kink in one case and intimal proliferative ridges in the other one. High definition flow with 4D STIC clearly defined this obstructive kink in the ductus arteriosus in our case. Despite complete ductal dependant pulmonary circulation which requires early post natal prostaglandin infusion prior to urgent surgical BT shunt (arterial duct stenting is not practical due to its marked tortuosity), this abnormal ductal configuration with evident obstructive kink should raise concern about close follow up for fear of premature ductal constriction with consequent in utero fetal death or immediate sudden post partum neonatal death.