A case of double outlet left ventricle
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Objective
Double outlet left ventricle (DOLV) is a very rare anomaly characterized by the aortic (Ao) and pulmonary artery originating entirely or predominantly from the left ventricle. Dual outlet ventricles with compatible atrioventricular (AV) connections account for 1% of congenital heart diseases, and DOLV accounts for <5% of them. The number of cases with prenatal diagnosis is very low. Thus, information about DOLV is limited in the literature. A case of DOLV detected in the early antenatal period, which was later confirmed by autopsy is presented.

Methods
26 years old, G2P1 patient was referred due to a suspicion of cardiac anomaly at the 17th week. On ultrasound, in addition to AVSD, we found that the pulmonary artery and the aorta were emerging from the left ventricle. The aorta was narrow (Figure 1). DOLV was diagnosed together with aortic coarctation. Amniocentesis (AS) was performed, and karyotype resulted as normal at the 20th week. The family was informed about the poor prognosis also by the pediatric cardiologist. The pregnancy was terminated due to the family’s request. Fetal heart was dissected in pathological examination; Aortic coarctation and AVSD, in which big arteries departed the left ventricle, were observed (Figure 2-3).

Results
DOLV is a cardiac morphology characterized by large arteries originating predominantly from the left ventricle. Although the exact incidence is unknown, it was reported as less than 1/200,000 live births. DOLV represents a heterogeneous patient group in terms of associated cardiac anomalies. In some studies, the most prominent associated cardiac morphologies were reported as atrial septal defect (ASD), ventricular septal defect (VSD), right ventricular hypoplasia and patent ductus arteriosus. The clinical presentation of DOLV is based on the underlying anatomy and emerging hemodynamics; the position of the associated ventricular or atrial septal defect, the size of the ventricles, and the configuration of the great arteries are important. There is no single hypothesis to explain the embryological formation of DOLV. In this context; while it was emphasized that abnormal differential conal growth and infundibular growth under the semilunar valves were important factors determining the normal and abnormal relationship between the great arteries, another view attributed this malformation to the differential absorption of the subpulmonic and subaortic conus during embryogenesis. DOLV is classified according to the location of the VSD in the great artery, such as the double outlet right ventricle. The extension of the VSD can be subaortic (most common), subpulmonic, bilateral, or distant. Our case had a subaortic VSD together with an ASD, but also with an aortic coarctation. Surgical treatment of DOLV has evolved over time. Various techniques such as biventricular repair, univentricular repair, and Rastelli procedure are applied. Because of the morphological heterogeneity of the disease, surgical options are determined by associated cardiac anomalies. The position of the great arteries plays a key role in deciding the surgical technique. The prenatal and postnatal diagnosis of DOLV type and associated additional cardiac anomalies such as atrial situs, aortic coarctation, and pulmonary stenosis are important for planning the type of surgery. The prenatal diagnosis of DOLV seems important in terms of directing the family to the center for pediatric cardiac surgery, but in our case the family opted for termination.

Conclusion
Although DOLV is a very rare pathology, it is also important to diagnose accompanying cardiac anomalies in the intrauterine period. This is important in the decision making whether to terminate or continue the pregnancy, and if the decision was to continue the pregnancy, to refer the patient to a center capable of doing the surgery.