A case of malposition of the great arteries with intraventricular septal defect

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Objective
Congenital heart disease (CHD) is one of the major health problems in neonates worldwide causing high morbidity and mortality with incidence ranging from 3.7 to 17.5/1000 live births in the world. Anatomically corrected malposition of the great arteries is a rare form of congenital heart disease in which the great arteries are abnormally related to each other and to the ventricles, but arise nonetheless above the anatomically correct ventricles. The abnormally related aorta originates above the anatomically left ventricle and the abnormally related pulmonary artery arises above the anatomically right ventricle.

Methods
A 22-year-old healthy woman (gravida 2, para 1) without risk factors for congenital heart disease presented at 21 weeks gestation for routine screening of fetal cardiac malformations.

Results
Biometry was consistent with fetal age. No other abnormalities were detected. In the 2D scan, the fetal stomach was, as it is normal, on the left side; however, the cardiac axis pointed to the right. By the acquisition of 4-dimensional (4D) sonography with spatiotemporal image correlation (STIC) and tomographic ultrasound imaging (TUI), we realized the abnormal four chamber view with right apex and a discrepancy between larger left and smaller right ventricles was seen. Furthermore, the right atrium was near to the fetal spine and the left ventricle was anterior. A intraventricular defect of about 5.1 mm was noted. The enlarged aorta arose from the left ventricle and the small pulmonary artery arose from the right ventricle. On the three vessels view, the main pulmonary artery trunk (2.0 mm) was thinner than the aorta (4.7 mm). After genetic counseling, the patient opted for termination of pregnancy. It was terminated at 22 week of gestation, and the parents declined the autopsy.

Conclusion
Fetal echocardiography is an important part of ultrasound screening in second trimester. The four chamber and three-vessel view are essential and indispensable in assessing the heart structure. Four-dimensional sonography with STIC-TUI facilitates visualization of the anatomic situs, thus giving additional information compared to 2D echocardiography in the prenatal diagnosis of abnormal great vessels connections. Early prenatal diagnosis is important for pregnancy counseling and planning of postnatal management, which is of particular relevance in fetuses with malposition of the great arteries.