Objective
Persistent hyperplastic primary vitreous (PHPV) is a very rare developmental malformation of the eye due to the persistence of the posterior portion of the tunica vasculosa lentis and hyaloid artery. In most cases, PHPV is not an isolated finding, frequently occurring in conjunction with cataract. In addition, it is usually associated with intracranial abnormalities such as hydrocephalus. We report a case of PHPV with congenital cataract and hydrocephalus diagnosed at 29 weeks’ gestation by ultrasound and prenatal magnetic resonance imaging.

Methods
A 20-year-old primigravida was referred to our prenatal diagnosis unit at the 29th week of gestation because of the fetal hydrocephalus detected on routine ultrasound scan. Detailed ultrasound examination revealed a male fetus with triventricular severe hydrocephalus where lateral ventricular atria was measured 39 mm on the right, and 34 mm on the left side. Bilateral lenses were echogenic representing cataract. The cataract was more prominent in the right side and axial plane of the right eye demonstrated prominent thickness of the hyaloid artery. Fetal MRI performed at 30 week’ of pregnancy confirmed the ocular sonographic findings and showed bilateral detachment of neural retina.

Results
This case is, to our knowledge, is the fourth description in the literature of prenatal ultrasonographic diagnosis of PHPV.

Conclusion
PHPV is a very rare anomaly which can be diagnosed during the prenatal period. Detailed fetal eye evaluation should be performed, especially when there is abnormal brain findings.