Objective
To study the incidence of PRUV and its clinical implications in a South Asian population attending the feto-maternal unit of a tertiary level private health institution for routine antenatal scanning. To the best of our knowledge, this is the first case series report from our ethnic population.

Methods
A retrospective, descriptive, observational study conducted in the department of feto-maternal Medicine, Edappal hospital, Kerala, India. The database of ultrasonography was searched for an ultrasound finding of PRUV in all the consecutive pregnancies during the period Jan 1st 2009 to Sept 30th 2014 after observing the inclusion and exclusion criteria. The feto-maternal department of our institution provides routine prenatal scanning services including early viability scans; first and second trimester genetic sonograms; target scans along with specialized examinations like fetal echo and neurosonogram; and growth scans along with doppler examination for the obstetric and the Assisted reproductive technique (ART) unit of the hospital. The routine protocol for an antenatal client of this institution is an early viability scan followed by a combined screening for abnormalities at 11 – 13 + 6 weeks, a target scan at 18 – 20 weeks and a growth scan with doppler examination in third trimester. The department also extends the prenatal diagnostic service for antenatal clients referred to it from other hospitals for second opinion in view of variety of reasons. The client base and henceforth the study cohort had mixed low to high risk women and all were south Indian. The inclusion criteria included all women with gestational age greater than 14 weeks and availability of complete follow up including postnatal details. The exclusion criteria were fetuses with situs abnormalities. In case a woman had a scan late in her pregnancy with us, all her previous scan images were reviewed if they were available though it was not mandatory for recruitment in the study. A total of 20, 450 consecutive antenatal women underwent ultrasonic scans during the defined period of the study satisfying the above criteria. All scans were performed using Voluson 730 expert (GE Healthcare, Vienna, Austria ) with 2Dimensional (2D) curvilinear transducer of frequency 2 - 5 MHz by trained Fetal medicine consultants. Reporting and image archiving is done using a dedicated software installed in computer workstations and hard drives. A diagnosis of PRUV was done based on following criteria - 1. 2D and colour Doppler demonstration of curving of the umbilical vein towards the stomach (or fetal left) in the standard fetal abdominal circumference plane 2. Slight inclination of the ultrasound transducer to demonstrate the location of gall bladder medial to the umbilical vein (between PRUV and stomach) 3. Status of Ductus venosus (DV) was noted in all cases using colour Doppler by its aliasing and characteristic waveform All fetuses with PRUV had full evaluation of fetal anatomy. Fetal echocardiography, neurosonogram or 3Dimensional evaluation was done depending on associated findings. Fetal Karyotype was done in individual case basis based on associated malformations and obstetric indications rather than the finding of PRUV alone. Complete obstetric history and postnatal fetal outcomes was collected in each case from the hospital records. Data was organized and statistically analysed using Microsoft Excel.

Results
From January 1st 2009 to September 30th 2014, a total of 20, 450 consecutive women underwent prenatal scanning at our centre satisfying the criteria of inclusion and exclusion. A total of 23 cases of PRUV were identified amongst them thus yielding an incidence of 0. 11% (1 in 889). The mean and standard deviation for maternal age was 24 ± 4. 6 years while that for the gestational age at diagnosis was 22 ± 2. 9 weeks. Earliest diagnosis was made at 14 weeks of gestation. Two cases were diagnosed from their first scans in our department in the third trimester. Two cases were twin pregnancy, one dichorionic diamniotic and one monochorionic monoamniotic (one fetus from each pair). Twelve fetuses were female and rest male. The essential details of the individual cases are outlined in table I and III. Out of the 23 cases of PRUV, 21 cases were of in utero type while two cases were of extrahypophyseal type. Both the extrahypophyseal cases were associated with absent Ductus venosus and direct drainage of PRUV into the right atrium. 11 of the 23 cases had undergone a Nuchal translucency (NT) scan while 5 out of them gave consent for combined genetic screening. All the NT values were normal so were the combined screen results. 22 cases had not undergone either a first trimester or a second trimester genetic screen by individual choices. A. Major abnormalities (12 out of 23 cases, 52. 2 %)Abnormality Number of cases Cardiovascular ( 9 out of 23, 39. 1 % of cases) Isolated Ventricular septal defect 1 Interrupted Inferior Vena Cava 1 Tricuspid Atresia 1 Tetralogy of Fallot 2 Atrialventricular septal defect 2 Double outlet right ventricle 1 Aortic stenosis 1 Other major abnormalities Congenital Diaphragmatic hernia 3 Unilateral renal agenesis 1 Bilateral renal agenesis 1 Fetal growth restriction 1 Sirenomelia 1 B. Minor abnormalities (4 out of 23 cases, 17. 4 %) Single umbilical artery 3 Absent Nasal bone 2 Umbilical cord varix 1 Club foot 1 Polydactyly 1 Hyperechoic bowel 1 Table I lists the major abnormalities that were associated with the 23 cases of PRUV. 16 out of 23 cases (69. 6%) had additional abnormalities. Major abnormalities were present in 12 cases (52. 2 %). Most common affected system was cardiovascular (9 out of 23, 39. 1 %) followed by renal system. Single umbilical artery was noted in three cases and was the most common associated minor marker. There were two cases of renal agenesis. Case Source of client E/R * Age in years Parity** & gestational age(weeks + days) Type of PRUV Additional findings Karyotype Fetal/Postnatal outcome 1 R 24 G1, 20 + 5 EH Mid muscular VSD Not done *** Required surgical closure postnatally, baby well after surgery 2 E 36 G1, 21 +1 EH Isolated interrupted IVC 46 XX Findings confirmed by postnatal Echo. Infant thriving well 3 E 23 G1, 23 +1 EH Right sided CDH, FGR 46 XY Termination of pregnancy 4 E 25 G2 P1, 24+1 EH AVSD, Absent Nasal bone 46 XX Termination of pregnancy 5 E 30 G3 P1 A1, 20 +4 EH Tricuspid atresia Not done Termination of pregnancy 6 E 23 G1, 20 +4 EH TOF, SUA in one fetus(MCMA twin ) Not done Termination of pregnancy 7 E 26 G2 P1, 21+4 EH Unilateral renal agenesis Not done Findings confirmed postnatally, normal development 8 E 18 G2 P1, 14 EH Pericardial effusion Absent nasal bone 46 XX Developed hydrops, fetal demise at 26 weeks 9 R 23 G1 1 EH TOF Absent DV, Post axial polydactyly, Overlapping fingers, Bilateral echogenic kidneys, Small umbilical cord hernia 46 XX Termination of pregnancy 10. E 23 G1, 22+6 IH Absent DV, Direct drainage of PRUV into right atrium Not done Normal outcome 11. R 28 G1, 17+2 IH Direct drainage of PRUV into right atrium Sirenomelia Bilateral renal agenesis XY 46 XX Termination of pregnancy 12. E 22 G3 P2 L2, 28 EH DORV, AVSD, AS Not done Termination of pregnancy Table II depicts the fetal outcome in 12 cases of major abnormalities. 7 cases opted for termination for the pregnancy. One case had still birth following development of hydrops. One case of extrahypophysial PRUV had good outcome aided by absence of any of associated malformations. Fetal karyotype was counselled in 10 out of 23 cases based on the priori risk of the mother (Table II, case 2) and nature of associated fetal abnormalities (all the other nine cases). Six women accepted fetal karyotyping. Karyotype was normal in all six cases. S. No Case No Source of client E/R * Age Years Gest age (weeks + days) and Parity ** Type of PRUV Marker Antenatal course Birth weight in grams Outcome 1 11 E 26 20 +6 G1 IH Isolated Unventilent 2930 Favourable 2 14 E 22 22+3 G2P1L1 IH Isolated Unventilent 2560 Favourable 3 15 E 22 G2A1 IH Isolated Unventilent 2600 Favourable 4 16 E 29 23+3 G1 IH Isolated Unventilent 3100 Favourable 5 17 E 24 21+6 GEP2L2 IH Isolated Unventilent 3000 Favourable 6 18 E 18 20+6 G2P1L1 IH Isolated Unventilent 2750 Favourable 7 19 E 22 23 GI IH Isolated Unventilent 2900 Favourable 8 20 E 25 23 GI IH Twins (DCDA)& with CTEV# in one fetus (B) along with PRUV Preterm labour at 35 weeks A - 1900 B - 1800 B required conservative management for CTEV. Favourable for both 9 21 E 23 24+3 G2P1L1 IH Umbilical cord varix Unventilent 3200 Favourable 10 22 E 34 22+6 G2A1 IH Hyperechoic bowel Unventilent 2500 Favourable 11 23 E 18 29+5 G1 IH Polyhydramnios PPROM 5 at 35 + 5 2700 Favourable Table III highlights the fetal outcomes in seven cases of isolated PRUV and four cases with minor markers. All had normal outcomes including normal birth weights and normal postnatal growth. Review of maternal history did not reveal any preconceptional risk factors or development of any antenatal complications.

Conclusion
The incidence of PRUV in our study based on a South Asian population was determined and found to be in the reported range in the literature. No risk factors could be identified. After a diagnosis of PRUV a detailed survey of fetal anatomy should be done. Fetal karyotype should be reserved for cases with additional findings or obstetric indications. The determinants of prognosis were type (intra or extrahyphseal) and associated findings. Isolated extrahyphseal PRUV carries good prognosis.