

CASE REPORT

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# Prenatal diagnosis of intrahepatic portosystemic shunt in intrauterine growth restricted fetus with transient fetal anemia and cardiomegaly: A case report

Babic I., Ferretti E., Jimenez-Rivera C., Gruslin A., Moretti F.

## ABSTRACT

**Introduction:** Congenital intrahepatic portosystemic shunts are abnormal communications between hepatic and portal vessels. We present a case of antenatally diagnosed portohepatic shunt in intrauterine growth restricted (IUGR) fetus with cardiomegaly and transient fetal anemia. **Case Report:** A 37-year-old primiparous woman was referred in a late second trimester for fetal growth assessment. Ultrasound revealed IUGR fetus at 9th centile with brain sparing and forward end-diastolic flow in umbilical artery. Middle cerebral artery (MCA) peak systolic velocity (PSV) Doppler was high for the gestational age, possibly representing fetal anemia. Fetal liver was enlarged with abnormal circulation consisting of tortuous dilated intrahepatic vessels originating from the umbilical vein and giving off several branches with no clear end point communication sites. Fetal echocardiography showed mild cardiomegaly with hyperdynamic flow state

possibly related to fetal anemia. A vigorous baby was delivery at 38-week, weighing 2880 grams. Postnatal abdominal ultrasound confirmed mild hepatomegaly with abnormally dilated middle hepatic vein branch communicating with left portal vein, representing intrahepatic portosystemic shunt. The neonatal course was uneventful and the child was discharged home at two weeks of life. One year follow-up ultrasound showed spontaneous resolution of intrahepatic portosystemic shunts. **Conclusion:** Our case of IUGR was probably related to intrahepatic shunt with increased preload causing cardiomegaly, and not driven by hypoxia. Transient fluctuating high MCA PSV Doppler was likely related to the hyperdynamic flow and not fetal or neonatal anemia, so transfusions were not required. IUGR fetuses without chromosomal or placental related pathologies should have thorough evaluation of the hepatic circulation to detect crucial congenital malformations and provide proper counseling.

**Keywords:** Cardiomegaly, Doppler, Fetal anemia, Growth restriction, Portosystemic shunt

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## INTRODUCTION

Congenital intrahepatic portosystemic shunts (IPSS) are abnormal communications between hepatic and portal vessels. The accurate diagnosis is usually made in the early neonatal life. As these abnormalities do not cause significant influence on fetal well-being, their recognition prenatally remains poor. Some case reports have described possible association between IPSS and fetal growth restriction. However, to date there are no published reports describing prenatal IPSS in IUGR fetus with fetal anemia and cardiomegaly. The spectrum of consequences, such as development of growth restriction, cerebral hyperperfusion and hyperdynamic heart function in association with IPSS as a rare anomaly, are potential prenatal characteristics. This may trigger timely diagnostic work up directed to reach early diagnosis and plan the optimal management. This is the first case report of prenatal IPSS in IUGR fetus with transient severe anemia and cardiomegaly.

## CASE REPORT

A 37-year-old healthy primiparous woman was referred to our tertiary care hospital at 27th week and 6 days of gestation for the evaluation of fetal growth. Ultrasound revealed IUGR fetus with overall growth at the 9th centile. Amniotic fluid was adequate for gestational age. Color and pulse Doppler were utilized according to standard application rules to assess the blood flow resistance in umbilical artery (UA) and middle cerebral artery (MCA) [1, 2]. Umbilical artery and MCA pulsatility indices (PI) were normal with low resistance to flow. However, MCA peak systolic velocity (PSV) showed high forward stream, 73 cm/sec (1.9 MoM), raising the suspicion of severe fetal anemia. Fetal liver was enlarged with abnormal circulation consisting of tortuous dilated intrahepatic vessels originating from the umbilical vein and giving off several branches with no clear end point communication sites (Figure 1). Possible presence of IPSS with unclear anatomical point of anastomosis was suspected. Amniocentesis for karyotype was offered but the woman declined. TORCH screen was negative. Fetal Echocardiography demonstrated mild cardiomegaly with hyperdynamic flow of unclear underlying cause. During the course of pregnancy, fetal surveillance was carried out with weekly ultrasound assessments as the overall fetal growth, stayed below the 10th centile with slow but steady decrease to below the 3rd centile by 37 weeks and 6 days. The UA, MCA and ductus venosus (DV) impedance flow remained normal; however MCA PSV was fluctuating between 1.07–1.96 MoM throughout gestation. Intrauterine fetal blood transfusion was not performed as there were no clear causes for development of fetal anemia. Intrahepatic vessels' diameters were stable with overall unchanged appearance throughout the course of the pregnancy. Labor was electively induced

at 38th week of gestation for IUGR and minimal growth interval.

## NEONATAL OUTCOME

The outcome was a vigorous female, symmetric IUGR weighing 2280 grams (3rd percentile), head circumference 31 cm (3rd percentile) and length 45 cm (3rd percentile). APGAR score was 7 and 8 in first and fifth minute and cord pH 7.15. The neonate was admitted to the neonatal intensive care unit for further assessment and investigations. There was no respiratory distress and O<sub>2</sub> saturations were 100% on room air. Baseline liver function tests and coagulation profile were within normal values. She had a small patent foramen ovale (PFO) that required follow-up at third month of life. Her CBC was reflective of IUGR with normal WBC at 11.7 x10<sup>9</sup>/L, hemoglobin at 189 g/l and a thrombocytopenia with platelets at 58x10<sup>9</sup>/L, which was transient and did not require transfusion. Phototherapy was implemented due to non-immune hyperbilirubinemia that resolved within 48 hours. Genetics testing failed to reveal any abnormalities including a normal karyotype; microarray 101 K oligonucleotide platform sequencing, deletion duplication analysis of ENG and ACVRL1 gene and 7-dehydrocholesterol excluded Smith-Lemli-Opitz. Postnatal abdominal ultrasound, as well as MRI scan, confirmed mild hepatomegaly with 4 vessels branching medially from the middle hepatic vein and communicating with the large left portal vein, representing IPSS (Figures 2 and 3).

Overall, the neonatal course was uneventful; breastfeeding was started ad lib from admission to discharge with no other relevant associated issues. Voiding and stooling were regular. The child was discharged home at second week of life. One year follow-up ultrasound showed spontaneous resolution of IPSS (Figure 4). The infant is currently 22-month-old, healthy with normal growth and development.



Figure 1: 2D color Doppler showing antenatal tortuous intrahepatic vessels with IPSS (Arrow pointing to the IPSS).

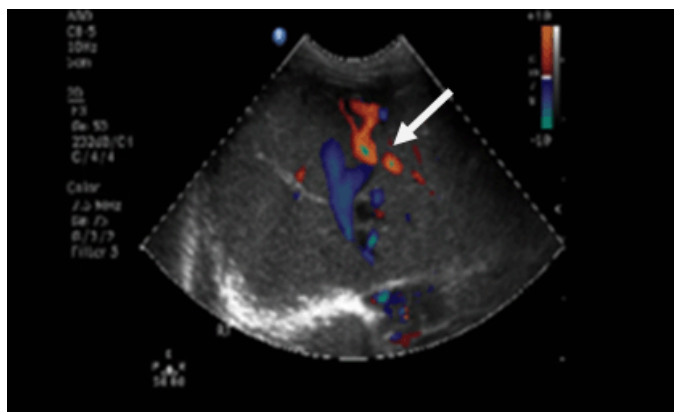


Figure 2: 2D color Doppler showing postnatal axial abdominal view of IPSS at second day of life (arrow pointing to the IPSS).



Figure 3: Magnetic resonance imaging scan at sixth day of life showing IPSS (arrow).

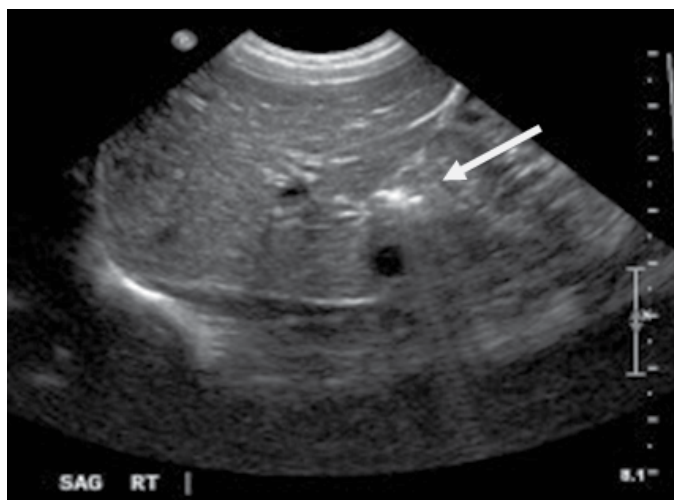


Figure 4: Normal ultrasound at one year of age (the arrow is pointing to the site of previously seen IPSS, currently resolved).

## DISCUSSION

Intrahepatic shunts are rarely recognised anomalies in utero. Their precise incidence is unknown and most of their pathophysiology is described through the

neonatal or infantile periods of life [3]. The IPSS may also represent one of the features of the multisystem morbidities among children, and adults. Luckily our case did not show any other comorbidities like hypoxemia or pulmonary hypertension at birth nor behavioral changes were noted, as it has been reported recently [4]. Literature review revealed two case reports of four cases with IPSS diagnosed prenatally [5, 6]. One case report described IPSS associated with IUGR and abnormal DV Doppler, whereas a second one, described three cases of various porto–hepatic anastomoses with spontaneous closure in the first few months of infants' lives.

To our knowledge, no published cases exist demonstrating prenatally diagnosed IPSS in IUGR fetus with concomitant development of fetal anemia and cardiomegaly. Application of colour Doppler was crucial to make the prenatal diagnosis [7]. The IPSS was classified as type 2, according to Park's classification [8].

The IUGR is commonly developed in fetuses with IPSS [3, 4]. Hypoperfusion and/or abnormal vessels' routes deviations, leave hepatocytes with decreased blood supply which ultimately leads to diminished cell proliferation and hypoxia [9–11]. Moreover, levels of liver enzymes, tested in neonatal period, show downregulation of hepatic cells which is probably one of the underlying causes that contributes to the smaller abdominal circumferences as a first abnormal feature in these fetuses, destined to remain small for the rest of the pregnancy [12]. Generally, IUGR fetuses are prone to develop polycythemia, thrombocytopenia and sometimes neutropenia due to decreased blood flow through the liver and bone marrow. Interestingly, blood flow remains normal which excludes placental insufficiency as a cause for IUGR. Although fetoplacental blood perfusion remained normal with low impedance to flow, in our case, the fluctuating levels of MCA PSV throughout the gestation were not quite understood. We hypothesised two possible explanations.

Firstly, the presence of a shunt may have caused shifting of the high stream blood flow to the heart, causing a hyperdynamic state which would have ultimately contributed to higher cerebral redistribution. Secondly, we can speculate that there was increased sequestration of red blood cells in the spleen, perhaps due to splenic stasis and poor venous flow as the baby was thrombocytopenic and hyperbilirubinemic after birth; in retrospect, the process may have started much earlier in utero. Regardless of the potential cause, the low count of blood cells was inconsistent and did not cause deterioration in fetal well-being.

We did not consider fetal blood transfusion at any point throughout the pregnancy as no obvious causes for the fetal anemia were found, and furthermore, there was no progression into fetal hydrops. This was confirmed later on, in the postnatal period as the neonate never required blood transfusion. In some cases a surgical or interventional radiology is needed to treat IPSS. In our case, no intervention was necessary as follow-up imaging revealed complete resolution of IPSS [13, 14].

## CONCLUSION

In conclusion, this case exemplifies that unexplained fetal growth restriction with hyperdynamic heart function state, should prompt a thorough evaluation of the hepatic circulation. This will allow the detection of crucial congenital vascular malformations and therefore facilitate antenatal and postnatal management. Furthermore, prenatal signs of increased cerebral perfusion along with hyperdynamic heart function may not necessarily require treatment with blood transfusion in utero, as described in our case, but rather close follow-up after birth.

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## Author Contributions

Babic I. – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Ferretti E. – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Jimenez-Rivera C. – Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Gruslin A. – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Passed away prior to drafting the manuscript, however provided a substantial input in choosing the case for a publishable manuscript

Moretti F. – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

## Guarantor

The corresponding author is the guarantor of submission.

## Conflict of Interest

Authors declare no conflict of interest.

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