

## **Fetal extra-abdominal umbilical vein varix: Two cases of a rare malformation**

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### **Abstract**

Fetal umbilical vein varix is a rare malformation of the umbilical cord. Most of the varices arise intraabdominally and there have been multiple studies about the diagnosis and management of fetal intra-abdominal umbilical vein varices. However fetal extra-abdominal umbilical vein varices are much rarer and up to date there are no clear criteria for the diagnosis or the clinical management of the condition. We saw two cases within a month in our ultrasound department in 2022 and had a favorable outcome in both pregnancies. In the first case the diagnosis was only established at 39 weeks of gestation, and we performed a cesarean section the next day. The second case was diagnosed at 31 weeks of gestation. The patient was admitted to hospital, and we administered Betamethasone for RDS prophylaxis. At 34 weeks of gestation there were fetal heartbeat abnormalities, and a cesarean section was performed. We also reviewed the literature of previous cases and our two cases included, so far only 19 cases of fetal extra-abdominal umbilical vein varix have been described.

**Keywords:** Umbilical cord; Diagnosis; Cases.

### **Introduction**

Fetal Umbilical Vein Varix (FUVV) is a rare malformation of the umbilical cord.

In most cases the varix is located intra abdominally arising between its entry into the abdomen and the portal system (fetal intra-abdominal umbilical vein varix FIUVV).

FIUVV accounts for approximately 4% of the malformation of the umbilical cord with an incidence of 0.4-1-1/1000 [1-4].

Fetal extra-abdominal umbilical vein varix (FEUVV) is much rarer and there are only few cases described in the literature.

The diameter of a normal umbilical vein increases during pregnancy from 2 mm at 15 weeks to about 7-8 mm at term [5-7]. Previous studies have proposed criteria for the diagnosis of FIUVV: umbilical vein diameter larger than 9mm and subhepatic umbilical vein diameter >50% of the intrahepatic umbilical vein diameter [1].

There are no such clear criteria for the diagnosis of FEUVV [8].

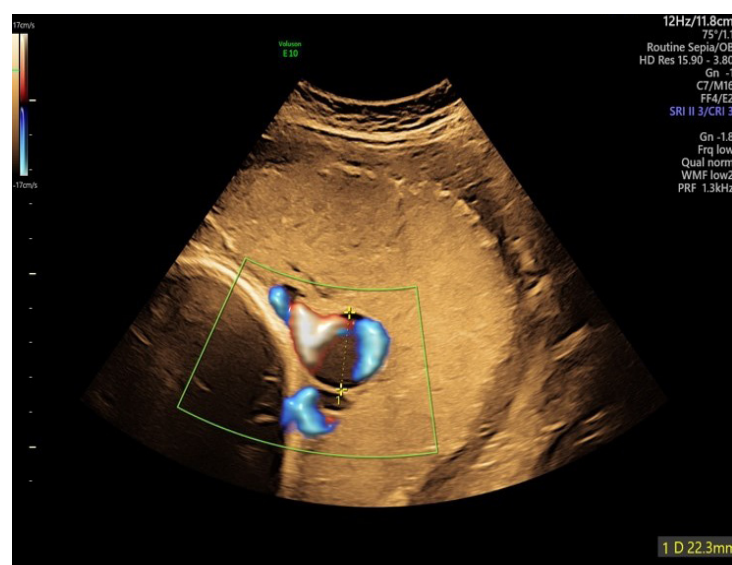
Fetal umbilical vein varix may have serious clinical consequences in the course of the pregnancy. Fetal Intrauterine Growth Restriction (IUGR) or even fetal death due to thrombosis of the umbilical vein have been previously described. Therefore, a detailed ultrasound to detect this anomaly is important. Close follow up of the varix is required and the option of a cesarean delivery to prevent dislodgment of the thrombus should be discussed with the patient.

We saw two cases of fetal extra-abdominal umbilical vein varix in our hospital in 2022. To our knowledge so far only 17 cases have been described in the literature [8-24].

## Case Presentation

The first case is a 28-year-old gravida 3 para 2. She was first referred to our unit because of breech position of the baby at 38 1/7 weeks of gestation. Up until she has received antenatal care and ultrasound examinations at a private practice nearby. She had a first trimester ultrasound but no combined screening for aneuploidies and the second trimester ultrasound with no observed anomalies was performed at 27 weeks of gestation.

On our first ultrasound examination we discovered the varix of the umbilical vein right at the placenta insertion site. The widest diameter was 22 mm (Figure 1).



**Figure 1:** Transabdominal 2D ultrasound using color doppler (transverse view) at 39 weeks showing a dilated umbilical vein on the surface of the placenta.

Apart from the extra-abdominal umbilical vein varix the female fetus was normally developed with an estimated weight of 2900 g (20<sup>th</sup> percentile), with a normal amount of amniotic fluid and physiological perfusion both in the umbilical artery and the Middle Cerebral Artery (MCA).

Due to the newly discovered varix of the umbilical vein, we didn't perform an external cephalic version but scheduled a cesarean section for the next day because of the increased risk of intrauterine fetal demise [1,3,25].

We delivered a healthy female fetus with a weight of 3020 g (32<sup>nd</sup> percentile), APGAR 9/10/10 and an umbilical cord arterial pH of 7.29.

We sent the placenta to pathology and our diagnosis of an extra-abdominal umbilical vein varix with a diameter of up to 30 mm was confirmed (Figure 2). There were no signs of thrombosis.



**Figure 2:** Placenta with a dilated umbilical vein at the insertion of the umbilical cord.

The second case is a 33-year-old gravida 1. She was referred to our unit at 27 weeks of gestation due to a newly diagnosed growth restriction and reduced amniotic fluid. So far, she had received her prenatal care including ultrasound examinations at a private practice.

The pregnancy was conceived via in vitro fertilization. Due to a beta thalassemia minor of both parents a preimplantation diagnostic was performed, and the fetus also tested positive for beta thalassemia minor.

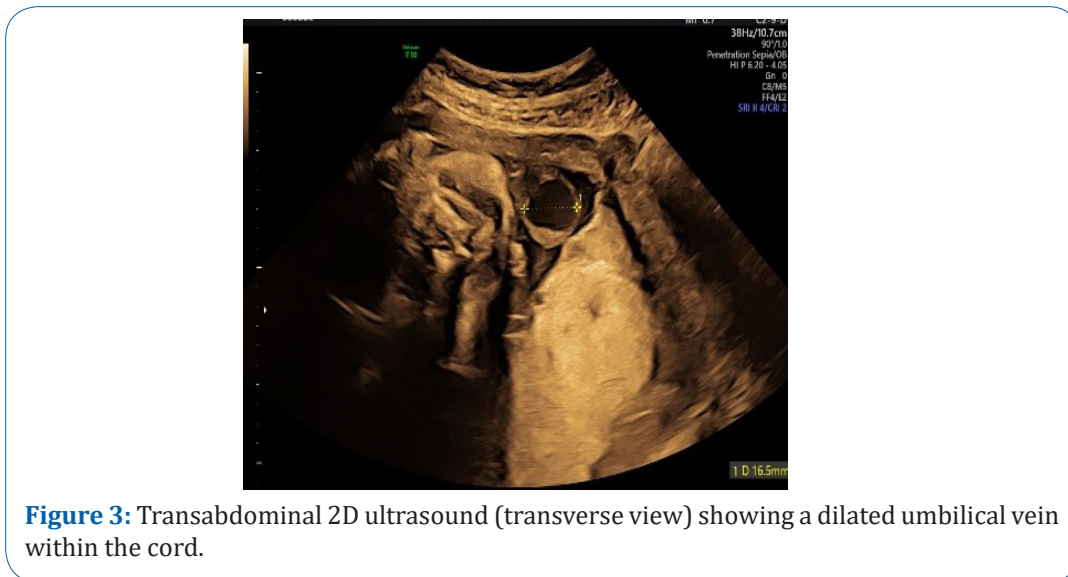
A combined first trimester screening showed a small risk for Trisomy 21,13 and 18 and the ultrasound at 22 weeks was normal as well.

On her first visit to our unit at 26 6/7 weeks of gestations we saw a female fetus with a severe early growth restriction (estimated fetal weight of 670 g, <3<sup>rd</sup> percentile) with no major anomalies. Amniotic fluid was at the lower end of the normal range. All long bones were shortened, furthestmost femur and humerus. There were no signs of thoracic hypoplasia, no cardiomegaly or signs of heart failure. The fetal perfusion including Ductus Venosus (DV) was normal and there were no signs of fetal anemia.

We interpreted these findings as an early placental insufficiency and recommended an amniocentesis. The couple declined the amniocentesis due to lacking consequences.

So, we scheduled weekly ultrasound checks with fetal hemodynamics and fetal biometry every two weeks. The fetus showed an accurate growth below the third percentile, amniotic fluid was stable at a low level and the perfusion in umbilical artery, MCA und DV remained normal.

At 30 3/7 weeks of gestation, we first discovered an extra-abdominal varix of the umbilical vein with a diameter of 16.5 mm (Figure 3).



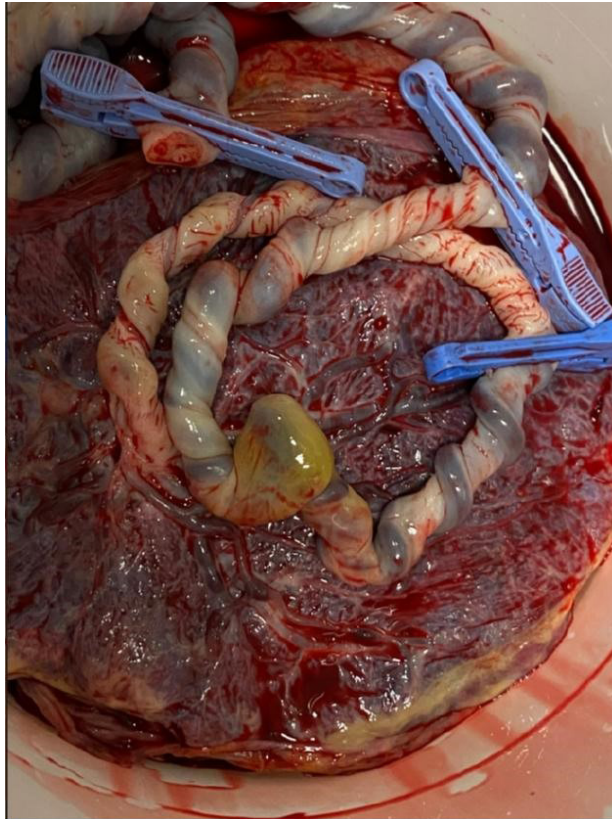
Due to the increased risk of intrauterine fetal demise in the presence of umbilical vein varix, combined with the growth restriction the patient was admitted to our hospital. She received RDS prophylaxis with Betamethasone 12 mg i.v. at 30 4/7 and 30 5/7 weeks of gestation, cardiotocography twice daily and doppler follow up every 2-3 days.

At 33 5/7 weeks of gestations cardiotocography showed signs of placental insufficiency with repeated decelerations so we decided to perform a cesarean section.

We delivered a female preterm fetus with a birthweight of 1365 g (<P3), APGAR of 8/10/10 and an umbilical cord arterial pH of 7.35.

She was immediately admitted to the NICU and needed CPAP support.

We sent the placenta to pathology and our diagnosis of an extra-abdominal umbilical vein varix was confirmed with no signs of a thrombus (Figure 4).



**Figure 4:** Placenta with a dilated umbilical vein in the central part of the cord.

## Discussion

We saw two cases of an extremely rare condition within a month and had a favorable outcome in both cases.

The diagnosis of an extra-abdominal umbilical vein varix prenatally is often difficult and there is only very little data on the accurate method of surveillance and management [21].

We reviewed the literature of the so far published 17 cases of extra abdominal umbilical vein varix.

The diagnosis was established between 20 and 39 weeks of gestation, in 5 cases the diagnosis was only established postdelivery. In the majority of the cases a thrombosis of the varix was found. The Diameter of the varices ranged for 1 cm to 5 cm.

Fetal heartbeat abnormalities were often the reason for delivery and in 12 of the cases a cesarean section was performed.

In 4 of the cases there was fetal demise, either pre- or postnatal. Further information is collected in table 1 (modified from Io et al) [23].

Studies showed that fetal extra-abdominal umbilical vein varix increases the risk of intrauterine fetal demise, fetal heart failure, fetal anemia, hydrops fetalis, thrombosis, umbilical cord twisting with

subsequent ulceration and blood loss [1,3,20,23-25].

There is also an increased risk of chromosomal abnormalities in the presence of intra-abdominal vein varix (Trisomy 21,18,) [5,25]. To our knowledge no chromosomal abnormality was detected in the presence of extra-abdominal umbilical vein varix.

**Table 1:** Clinical characteristics of 10 cases with extra umbilical vein varix (modified from Io et al).

Case No	Diagnosis (WG)	Location of varix	Diameter (cm)	Thrombosis	Non-reassuring fetal status	Delivery mode	Birth (WG)	Birth weight (g)	APGAR Score 1/5 min	Comment	Reference	Year of publication
1	Postdelivery	1 cm from Fetus	NA	+	Decreased fetal movement	VD	37	2250	NA	Long cord Fetal death	Ghosh et al [9]	1984
2	34	10 cm from Fetus	5	+	FHB abnormalities	CS	36	2420	4/6		Vesce et al [10]	1987
3	postdelivery	4 cm from placenta	3	+	Fetal death	-	39	3130	-	SUA	Schrocksadel et al [11]	1991
4	32	2 cm from the fetus	2	(+) artery	FHB abnormalities	CS	25	2041	NA/8		White and Kofinas [12]	1994
5	24	8 cm from placenta	4	+	-	VD	33	NA	NA	VSD	Shipp et al [13]	1995
6	Postdelivery	8 cm from fetus	5	+	-	VD	41	3920	9/10		Zachariah et al [14]	2004
7	31	3 cm from fetus	3	-	-	CS	Term	NA	NA		Akar et al [15]	2012
8	20	3 cm from fetus	4	+	FHB abnormalities	CS	35	1990	8/9	SUA	Trobs et al [16]	2012
9	35	10 cm from fetus	2	+	-	CS	35	2501	8/9		Kanenishi et al [17]	2013
10	postdelivery	Midportion	NA	-	FHB abnormalities	CS	37	3050	4/8		Soriano et al [18]	2015
11	24	Midportion	4	+	Fetal death	-	32	NA	-	Klippel-Trenaunay-Weber syndrome	Cruise and Rouse [19]	2016
12	postdelivery	Midportion	1.9	+	Fetal death	VD	38	NA	-	Cord hematoma	Al-Maghrabi et al [20]	2017
13	36	Attachementpoint of fetus	5	+	FHB abnormalities	VD	36	3647	8/9		Cassidy-Vu et al [8]	2019
14	23	Midportion	3	+	FHB abnormalities	CS	32	1744	8/9		Matsumoto et al [21]	2019
15	39	Fetal side	NA	-	FHB abnormalities	CS	39	NA	NA		Hayden et al [22]	2020
16	27	5 cm	1	-	FHB abnormalities	CS	33	1334	4/7	Neonatal Anemia	Io et al [23]	2021
17	24	On the placenta	10.5	+	FHB abnormalities	CS	24	610	4/5	Fetal Anemia, neonatal death	Jackson et al [24]	2021
18	39	On the placenta	3.3	-	-	CS	39	3020	9/10		This study	2022
19	31	Midportion	1.6	-	FHB abnormalities	CS	34	1365	8/10		This study	2022

If an umbilical vein varix is detected fetal echocardiogram should be performed to exclude cardiac malformations and a close surveillance of the pregnancy is recommended [21].

If fetal anemia is suspected a diagnostic cordocentesis can be offered to evaluate the need for fetal transfusion [23,24].

There are no clear recommendations of how to monitor a pregnancy with diagnosed fetal extra-abdominal umbilical vein varix. Expert opinions suggest weekly ultrasound prior to 28 weeks of gestation and bi-weekly examinations thereafter until delivery to evaluate for the presence of thrombosis in the umbilical vein varix [7].

The dislodgment of a thrombus is a great risk for the fetus and might be responsible for fetal loss [11,14,16,199,21].

Time of delivery should be planned individually but it seems appropriate to induce between 36 and 37 weeks gestation. In case of present thrombus in the varix a vaginal delivery increases the risk of thrombus dislodgement, and a cesarean section should be offered.

## Declarations

**Data availability:** Data supporting this case report are available from the corresponding author on reasonable request.

**Consent:** Written informed consent was obtained from the patients for the publication of this case report.

**Disclosure of interest:** The authors declare that they have no conflicts of interest.

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