REVIEW / Pediatric imaging

Umbilical vein varix: Importance of ante- and post-natal monitoring by ultrasound

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Abstract Foetal intra-abdominal umbilical vein varix is rare. Colour Doppler ultrasonography helps distinguish this vascular anomaly. A detailed anatomic scan must be performed to exclude associated anomalies: forms associated with additional complications are found in 29 to 35% of the cases. Intra-uterine foetal demise (IUFD) is a complication of umbilical vein varix. However, recent studies are more reassuring. When foetal intra-abdominal umbilical vein varix is isolated, there is no reason to change the management of the pregnancy. Foetal sonographic follow-up is recommended, focusing on an increase in the size of the varix and the appearance of a clot. A particular clinical form, connecting the umbilicus to the extra-hepatic portal vein should be known, because of a high risk of thrombosis. On the basis of this finding, postnatal monitoring by ultrasound is necessary.

Introduction

FIUUV is characterised by the dilatation of the foetal umbilical vein between its entry in the abdomen and its ending in the portal system. The incidence is low, ranging from 0.4 to 1.1/1000 [1–3]. It accounts for about 4% of the malformations of the umbilical cord in the foetus [4–6]. The diagnosis is based on colour Doppler sonography. It justifies a full foetal assessment, in a reference centre, to search for other anomalies that are associated in one third of the cases [2]. To date, over 150 cases of the isolated form of FIUUV have been reported in the literature. It was initially thought to be a serious anomaly, with a mortality of up to 44% due to IUFD [6–8], making certain authors propose inducing labour as of 34 weeks of amenorrhoea (WA), in spite of the morbidity generated by prematurity. The foetal

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Abbreviations: FIUUV, Foetal intra-abdominal umbilical vein varix; IUFD, Intra-uterine foetal demise; WA, Weeks of amenorrhoea; IUGR, Intra-uterine growth restriction.
risk, in recent publications including a larger number of isolated forms of FIUVV, appears to be lower, leading to a re-evaluation of the obstetric care [1]. Antenatal monitoring by sonography is indispensable in particular, in the search for a thrombus of the FIUVV. A specific clinical form, associated with abnormal anastomosis of the umbilical vein, should be known, because of the sonography characteristics and the frequent complications.

**Sonography of the foetal umbilical vein**

A sagittal section centered on the umbilical opening is used to analyse the sub-hepatic intra-abdominal segment of the umbilical vein. After its point of entry, a first 90° angle orients the vein under the abdominal wall, in the direction of the liver (Fig. 1). A second angle directs it to the rear according to an ascending sub-hepatic trajectory. The transverse section used to measure the abdominal perimeter is used to visualise the intra-hepatic portion of the umbilical vein that connects to the left portal vein opposite the origin of the lower left portal vein. The unit takes a horizontal trajectory to the right, forming the portal sinus (Fig. 2). The latter appears in the shape of an "L" established from the end of the umbilical vein and connecting the right and left portal branches. These two sonography sections do not allow to visualize the main portal vein. From the section of the abdominal perimeter, scanning to the bottom and to the right reveals the junction of the main portal vein with the portal sinus at the place of division between the right portal vein and the left portal vein [9]. The normal diameter of the vein, measured at its intra-hepatic segment, increases in a linear manner during the pregnancy, passing from 2 to 8 mm between the fifteenth week of amenorrhoea and term [7,10].

**Positive diagnosis**

FIUVV is detected as an anechoic, oval-shaped or rounded mass, located between the abdominal wall and the lower edge of the liver [11,12]. It is in continuity with the umbilical vascular axis on sagittal sections [13]. The pulsed and colour Doppler modes confirm the vascular nature of the abnormality and reveal a venous type flow (Fig. 3a and b). It allows to rule out other fluid images that may be seen in this space: liver cyst, cyst of the bile ducts, cyst of the mesentery, gastric duplication. FIUVV is defined according to two criteria: either a diameter exceeding 9 mm [14] or a diameter of the sub-hepatic segment of the upper umbilical vein exceeding 50% the diameter of the intra-hepatic segment [15]. These criteria have been used separately or most often together, as in all of the series published over the last 10 years [1–3,5,10,15].

The diagnosis of umbilical vein varix justifies a detailed foetal anatomical assessment in a reference centre to look for other abnormalities. These associated forms account for 29 to 35% of FIUVV [2,5]. The disorders most often seen involve the cardiovascular system and the uro-genital tract [2,8], but it may also consist of excess amniotic fluid. No specific association has been found. The severity of the lesions varies. Certain minor anomalies such as pylectasis or a single umbilical artery are usually not used to classify the observation among the associated forms [3,10]. Chromosome abnormalities are found in 6% of the cases of FIUVV, most often trisomy 21, 18 and 9 and triploidy [8]. They occur in 28% of the associated forms and under 2% of the isolated forms [2]. For most authors, abdominal umbilical vein varix does not justify the systematic use of a caryotype in this context of an isolated anomaly [1,14].

The mean age of gestation, at the time of the diagnosis of FIUVV, is between 27.5 and 30.5 WA ranging from 18 to 41 WA [1,3,8,10,15]. There is no difference between the isolated forms and the associated forms [5]. In two-thirds of the cases, the venous anomaly is detected after 28 WA with a normal first sonogram, supporting the hypothesis of a disease acquired during the pregnancy [2].

**Evolution during the pregnancy**

When the anomaly is recognised, the umbilical vein diameter is between 9.7 and 13 mm with extremes of 5.6 and 20 mm [1,3,6,8,10,16]. In 61% of the cases, the dilation does not evolve in the follow-up sonograms. The diameter remains identical that initially measured or increases by 1 to 3 mm, parallel to the linear increase in the diameter of the

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**Image 1.** Ultrasound image in a 22-week foetus showing the longitudinal course of the umbilical vein. UV: umbilical vein; B: bladder; H: heart; S: spine; arrow: umbilical vein varix.

**Image 2.** Ultrasound image in a 32-week foetus showing the normal intra-hepatic umbilical vein (UV) connection to the left portal vein (LPV), creating the portal sinus, at the level at which the abdominal circumference is usually measured. ILPV: inferior branch of left portal vein; RPV: right portal vein; S: stomach; AG: adrenal gland.
umbilical vein during the pregnancy. A 4 to 9 mm increase in the dilation is only observed in 28% of the foetuses and does not seem to be related to the precocity of the diagnosis \[1,6,8,10,17,18\]. The disappearance of FIUVV is only reported in 4 cases \[1,6\]. According to Mankuta et al., it probably involves an error in the interpretation of the initial sonogram, incorrectly measured at the non-linear segment of the vein, and not a real regression in the dilation \[1\]. A turbulent flow, defined in colour Doppler sonography by a bi-directional flow, is reported in 28 to 50% of the cases at the level of the dilated segment of the umbilical vein \[1,3,10\]. According to Weissmann-Brenner et al., it is in part related to the size of the lesion, since the diameter of the dilation is greater in foetuses presenting turbulences \[10\].

**FIUVV complications during pregnancy**

The potential gravity of the isolated forms of FIUVV is due to complications arising during the pregnancy. They are mainly represented by IUFD, thrombosis and intra-uterine growth restriction (IUGR). The overall frequency is assessed at 10% (Table 1). In the work first published by Mahony et al. in 1992, three IUFD were observed in seven cases of isolated forms of FIUVV, representing a very high rate of mortality of 43% \[7\]. Since, 5 deaths have been reported between 29 and 38 WA \[6,8,15\]. Among all of the cases, the occurrence of IUFD is assessed at 4.8%, and is inferior to the occurrence reported in associated forms of FIUVV \[8,15\] although superior to the 0.7% rate generally reported during pregnancy \[19\]. The occurrence is not related to the precocity of the appearance or the extent of the dilation \[10\]. Two mechanisms have been proposed: the formation of a thrombus at the level of the dilation creating an obstacle for the venous return or an increase in the cardiac pre-load that may be responsible for heart failure \[1,8\]. The histopathological examination of the in utero deceased foetuses has not confirmed these hypotheses. The appearance of a thrombus at the level of the umbilical vein varix has to be searched for systematically. The diagnosis is based on the detection of

<table>
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<tr>
<th>Authors</th>
<th>Year</th>
<th>FIUVV</th>
<th>Ante-natal thrombosis</th>
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FIUVV: foetal intra-abdominal umbilical vein varix; IUGR: intra-uterine growth restriction; IUFD: intra-uterine foetal demise.
incomplete filling of the vascular lumen in colour Doppler sonography, although the clot may also be detected in the form of an intravascular echogenic image [20]. Only two cases of thrombosis have been observed, one at 22 and the other at 32 weeks [7,20]. In once case, the foetus died at 29 weeks without the death being formally attributed to the thrombosis [7]. The risk of seeing a thrombus appear is higher with major dilation of the umbilical vein and the presence of turbulences. IUGR is defined by a foetal biometry inferior to the tenth percentile for the term. While only one observation has been reported before 2005, the occurrence, in recent works, is estimated at between 4 and 10%, higher than the 3% occurrence of IUGR usually reported [1,3]. To account for this difference, the hypothesis of a reduction in the oxygen supply to the foetus related to the presence of FIUVV is unlikely, because of the lack of correlation between the size of the dilation and the presence of turbulences [15,21]. Heart failure has often been suggested as a potential complication of the venous anomaly. However, in the isolated forms, no correlation has been found between the presence of anasarca and FIUVV [2]. According to Bas-Lando et al., there is currently no reason to perform a systematic foetal echocardiogram [3], even though the majority of teams recommend it [1].

Recently published studies modulate the global occurrence of complications and reconsider their relative importance. Only taking into account the last five publications, accounting for two-thirds of the cases, the complications only arise in 6% of the isolated forms of FIUVV and are essentially represented by IUGR without cases of IUFD.

**Prenatal monitoring**

Due to potential complications and the still limited number of case reports published, attentive monitoring is recommended, in particular during the third trimester of pregnancy. The frequency of the sonograms varies, depending on the teams, from one examination every two weeks to two examinations per week. The main goal is to detect a thrombus [3,10]. The existence of major dilation, superior to the mean diameter of a FIUVV (10–12 mm) and the detection of turbulences, are risk factors justifying obstetric monitoring and repeated sonograms.

**Birth**

The obstetric care of the delivery is not the same. Certain teams recommend inducing labour, as of 34 weeks, mainly due to the risk of IUFD [1,6]. The mean age of gestation at the time of delivery ranges from 36 to 38 weeks [1,3,5,10]. Vaginal delivery accounts for 64 to 83% of births, after induction motivated by the existence of FIUVV in more than half of them. The frequency of inducing labour, when not systematic, is high, between 62 and 89% of the deliveries, and superior to the rate of induction usually observed [3,10]. In 54% of cases, it is induced between 34 and 36 weeks, either due to the size of the dilation or the presence of turbulences [10] or de facto for some teams [1]. The frequency of caesarean-section births is 17 to 20% [1,3]. According to Bas-Lando et al., it is significantly higher than the 10% frequency usually observed by this team, the difference accounted for by the failures in inducing labour [3]. The indications for a caesarean-section correspond to the usual obstetrical criteria, FIUVV only justifying two cases, of which one case of antenatally diagnosed thrombosis [20]. No obstetric complications were reported in the literature, independently from the type of delivery, including deliveries at term.

The birth weight reflects the obstetric care. The mean is 2850 grams in centers proposing induced labour and 3200 grams in the others [3,5,10]. Induced labour also accounts for the lower weight at birth in cases of FIUVV associated with turbulences [10] and the higher frequency of intensive care hospitalisation in the series published by Bas-Lando et al. [3]. In view of these results, several authors do not recommend inducing labour in the isolated forms of FIUVV [1,3], others propose inducing labour at 36–37 weeks in case of major umbilical vein varix, especially if there are turbulences [10]. Delivery by caesarean section is proposed in case of complications, in particular in case of thrombus of the umbilical vein.

Because of the risks for the newborn, delivery in a level three establishment should be considered.

**Specific form: the umbilical vein ending in the extra-hepatic portal system**

Although the sonographic presentation is identical that of FIUVV, this form should be distinguished due to its anatomical particularity, its evolution during the pregnancy and especially the frequent complications. Six cases have been identified in the literature, to which we can add a case monitored in our centre [22–27].

**Anatomical particularity**

The description is based on the post-natal imaging but above all, on the intra-operative findings. In this form, the venous dilation is associated with a malformation of the umbilical-portal system, the dilated venous segment not ending at the portal sinus but at the caudal part of the superior mesenteric vein, just opposite the confluence with the splenic vein. There is no round ligament and the falciform ligament is short. Embryologically, the portal system arises from a double venous system: the umbilical veins from the placenta and the vitelline veins from the yolk sac. Under the influence, in particular, of the hepatic cords, these two systems undergo major modifications. The right vitelline vein gives rise to the definitive portal vein, the proximal segment of the left umbilical vein becomes the umbilical vein itself. According to Benoist et al., this anomaly may be the result of early and proximal anastomosis between the left umbilical vein and the right vitelline vein [22].

**Diagnosis and evolution**

As in the previous form, venous dilation presents as an anechoic formation located between the lower side of the liver and the umbilicus. As opposed to FIUVV, venous dilation is always diagnosed before 28 weeks (mean: 23 weeks), the initial diameter is about twice as wide (mean: 20 mm), and there is always the presence of turbulences (Table 2). The evolution,
during the pregnancy, is usually marked by an increase in the dilation that may reach 23 mm. Our case report is the only one in which the vein was moderately dilated at the time of the diagnosis and stable during the follow-up controls.

### Thrombosis

Found in all case reports, thrombosis is initially found in the distal segment of the superior mesenteric vein and/or the origin of the portal vein. The diagnosis may be antenatal, as observed in two case reports [23,25], although in five out of seven cases, it was post-natal during the first three days of life [22,24,26,27]. The evolution may be favourable [23,25], although the thrombosis is often extensive towards the intra-hepatic portal system (Fig. 4), justifying a laparotomy in five case reports [22,24,26,27]. In spite of the resection of the aneurismal lesion and thrombectomy, it evolved towards full portal thrombosis in three cases.

### Value of early post natal sonography

Confronted with FIUVV, it is important to recognise an abnormal anastomosis of the umbilical cord in order to screen and quickly treat a thrombosis of the portal system. The diagnosis may be suspected, as in one of the case reports, when the dilated segment of the umbilical vein has, under the bile duct, a trajectory directed towards the rear [25]. When the malformation is not suggested during the pregnancy, only the post-natal sonography can distinguish the ending of the umbilical vein at the level of the distal segment of the superior mesenteric vein from its usual ending at the left portal branch. However, this examination is not systematic [3]. In the case reports published in the literature, besides one case of thrombocytopenia, sonographs have been prescribed only after the FIUVV has been diagnosed antenatally.

### Conclusion

FIUVV is a rare foetal vascular anomaly. After reviewing the studies recently published, the evolution may be considered favourable when isolated. It is necessary, at the time of the diagnosis, to obtain a reference obstetric sonogram. Without an associated lesion, the probability of a chromosome abnormality is low and the systematic obtention of a caryotype is not justified. In view of the still limited number of cases, sonography monitoring is indicated during the third trimester of the pregnancy in order to search for a distinct increase in the dilatation or the appearance of a thrombus. Without any complications and considering that no cases of IUFD have been reported over the last years, the induction of labour before term is not justified. A sonographic assessment should be obtained during the first days of life in order to search for an abnormal anastomosis of the umbilical vein in the extra-hepatic portal system, requiring the immediate care of the infant in a specialised paediatrics unit.

### Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.
References


