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**Case Report**

# Fetal umbilical vein aneurysm

**Raisa Samuel[1]*, Preethi Navaneethan[2]**

[1]Department of Obstetrics and Gynaecology, St Stephen's Hospital, Delhi, India
[2]Department of Obstetrics and Gynaecology, Christian Medical College, Vellore, Tamil Nadu, India

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***Correspondence:**
Dr. Raisa Samuel,
E-mail: raisasamuel@gmail.com



## ABSTRACT

Fetal umbilical vein aneurysm is an uncommon anomaly also known as umbilical vein varix. It accounts for 4% of umbilical cord abnormalities with an incidence of 0.4-1.1/1000. The etiology of fetal vein aneurysm has not been well established. The diagnosis is made by colour doppler sonography. The management of umbilical vein aneurysm is yet in process of establishment. The possibility of sudden hemodynamic change caused by aneurysm or thrombosis needs to be screened on a regular basis and early delivery can be considered in need.

**Keywords:** Fetal umbilical vein aneurysm, Umbilical vein varix, Colour doppler, Umbilical cord, Thrombus, Funisitis

## INTRODUCTION

Fetal umbilical vein aneurysm is an uncommon anomaly also known as umbilical vein varix. It accounts for 4% of umbilical cord abnormalities with an incidence of 0.4-1.1/1000.[1]

It is defined as focal dilatation of the umbilical vein at least 50% wider than the nondilated part of the umbilical vein or more than 9 mm.[2] They are more common in external than the intra-abdominal portion of the umbilical cord.[3]

The etiology of fetal vein aneurysm has not been well established.[4] It was first described in 1985.[5] There are over 150 cases of isolated fetal umbilical vein aneurysm reported till date.[4] Earlier it was believed to be a serious anomaly which can cause 44% of intrauterine fetal death.[6] Due to the risk of mortality, induction of labor was done at 34 weeks in spite of the morbidity of prematurity.[7] Recent studies have shown that fetal risk due to isolated umbilical vein aneurysm is much lower and thereby leading to reevaluation of obstetric care.[7,8]

The diagnosis is made by colour doppler sonography. Due to the recent advances in the ultrasonography, more cases are diagnosed and its vital for these patients to have antenatal close monitoring to look for a thrombus.[7]

The management of umbilical vein aneurysm is yet in process of establishment. The possibility of sudden hemodynamic change caused by aneurysm or thrombosis needs to be screened on a regular basis and early delivery can be considered in need.[9]

## CASE REPORT

A booked outside G2A1 patient was referred to our hospital for antenatal checkup at 33 weeks of gestational age for further management of umbilical cord abnormality. She had termination of pregnancy 3 months back for anomalous fetus suggestive of open neural tube defect at 20 weeks of gestational age. She had done her antenatal checkup elsewhere and was diagnosed to have umbilical vein defect at 32 weeks of gestational age.

Her first trimester screening was normal and morphology scan done at 20 weeks was also normal. The ultrasound done at 32+5 weeks of gestational age showed focal dilatation of umbilical vein measuring 2.1 cm of diameter for a length of 4.6 cm in the free loop of the cord. The rest of the umbilical vein shows a diameter of 0.8cms. There was no turbulent flow in the dilated part of the umbilical vein. The umbilical cord had two arteries and one vein. The placenta had no abnormalities. The doppler studies also showed normal flow.

She had a repeat ultrasonogram at 36 weeks of gestation which showed focal dilatation of umbilical vein measuring 2.5cms of diameter for a length of 6cms in the free loop of the cord. (Figure 1.) There was no turbulent flow in the dilated part of the umbilical vein. She had regular antenatal visits and was monitored closely.

She was admitted at 39+1 weeks in early labour and was augmented with oxytocin. She had normal vaginal delivery and delivered a boy baby weighing 3.560kg with APGAR of 9 at 1 minute and 10 at 5 minutes. Placenta with attached membranes and umbilical cord weighing 694gm. The umbilical cord measured 60cm in length. There was a central dilatation, 17 cm from the proximal end (Figure 2) The diameter ranging from 0.7 cm (near the placental disc) to 4.1 cm (dilated segment). Rest of 43 cm of umbilical cord was not dilated. (Figure 3). The adjacent areas were edematous. Sectioning the dilated segment of umbilical cord revealed three blood vessels, one of which was grossly dilated with diameter of 3 cm filled with clot 2.3x1.4 cm covering about half of the circumference of the vessel wall. The rest of the two blood vessels appeared unremarkable. There were no knots or strictures seen. There was no calcification, hematoma formation or hydropic change. Cross sections of the dilated portion of the umbilical cord showed markedly dilated and thinned out umbilical vein with loss of muscle fibers. The lumen displays acute laminated thrombosis. The wall showed ischemic infarction with extravasation of inflammatory infiltrates and RBCs. The adjacent umbilical arteries were normal with focal areas showing acute thrombosis. The surrounding mucoid connective tissue (Wharton's Jelly) appeared edematous. Cross section of distal and proximal normal looking umbilical cord showed vein with congestion and dense acute inflammatory infiltrates involving the entire thickness of the wall of umbilical vein and arteries with mild spillage into surrounding connective tissue. Final report of histopathology was umbilical vein aneurysm with secondary umbilical vessel thrombosis and funisitis. She had an uneventful postnatal period and both mother and neonate were discharged in stable condition.

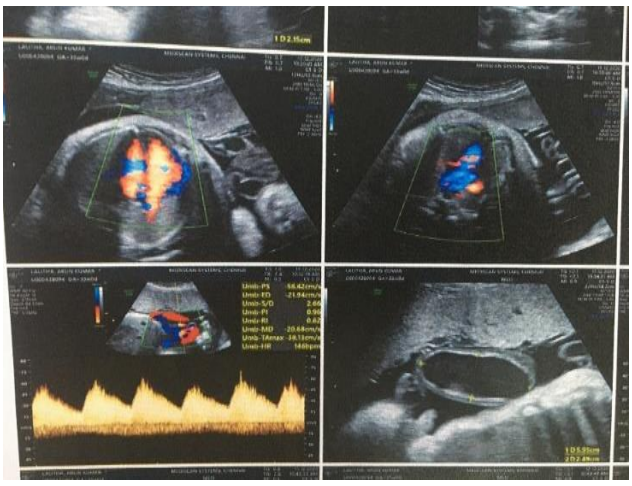

**Figure 1: Colour doppler imaging of turbulent blood flow within the aneurysm.**

**Figure 2: Gross specimen of the placenta with the umbilical vein aneurysm.**

**Figure 3: Umbilical vein aneurysm with diameter of 0.7 to 4.1 cm with rest of the umbilical cord not dilated.**

## DISCUSSION

Fetal umbilical vein aneurysm is a rare fetal vascular anomaly. It is diagnosed with the help of high-resolution ultrasound. It's seen as an anechoic, oval-shaped or rounded mass in continuity with umbilical vascular axis on sagittal sections.[7] There are two ultrasound criteria for its diagnosis- 1) diameter exceeds 9 mm and 2) diameter of the sub-hepatic segment of the upper umbilical vein exceeds 50% of the diameter of the intra-hepatic segment.[9]

The diagnosis of umbilical vein aneurysm warrants detailed anatomical assessment for other associated anomalies. The most often associated anomalies are cardiovascular and uro-genital. They are seen in 29-35%.[10] They are also seen to have associated polyhydramnios.[11] Chromosomal abnormalities are seen in 6% of fetal umbilical vein aneurysms, mostly trisomy 21, 18 and 9.[6] Hence when we suspect other anomalies or chromosomal

abnormalities, karyotyping by amniocentesis should be offered. The complications that may arise during pregnancy are intrauterine fetal death, intrauterine growth restriction and thrombosis. Since this condition is very rare, there are no clear guidelines regarding the management of these pregnancies.[6] From the various case reports and case series it indicates close monitoring of fetal growth and delivery is indicated only at term for the obstetric indications.[12]

## CONCLUSION

Fetal umbilical vein aneurysm is a rare entity. There is no clear understanding of its etiology and its significance in pregnancy. Older studies have described poor outcomes whereas recent studies have shown favourable outcomes. The prognosis is mainly determined by associated anomalies. Isolated umbilical vein aneurysms have good prognosis. There are no clear guidelines for management. Once we have diagnosed an umbilical vein aneurysm, we have to perform fetal echocardiography and detailed anatomical scan to rule out other associated anomalies. They will also require close fetal monitoring for fetal growth restriction and can have delivery at term for obstetric indications.

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