# Vasa praevia – ultrasonographic diagnosis and clinical management

#### Abstract

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Introduction. Vasa praevia is a high-risk, uncommon disorder of placentation. The possible fetal complications of ruptured vasa praevia include asphyxia, hemorrhage, rapid exsanguination and fetal or neonatal death. Materials and method. We searched PubMed, MEDLINE and the Cochrane Library using appropriate variables (e.g., "vasa praevia", "placenta praevia", "velamentous cord insertion", "succenturiate", "bilobed placenta") and medical literature pertaining to vasa praevia up to January 2021. We selected randomized control trials, observational studies, systematic reviews and four national guidelines. The diagnostic and management strategies recommended in medical literature were compared and reviewed. Results. A risk-adapted approach has been widely used to facilitate the ultrasonographic diagnosis of vasa praevia. The use of transvaginal color and pulse wave Doppler has not proven cost-effective in the general population, however it is recommended in the evaluation of patients in high-risk groups. Not all cases of vasa praevia can be diagnosed antenatally. Conclusions. Vasa praevia is a rare obstetrical condition that has been associated with a high perinatal mortality rate if undiagnosed antenatally. Standardized prenatal targeted screening protocols for vasa praevia in both singleton and multifetal pregnancies are needed in order to improve the diagnosis of vasa praevia, the neonatal survival and to lower the rate of intra- and postpartum complications. Keywords: vasa praevia, placental anomalies, velamentous umbilical cord insertion, bilobed placenta, ultrasound diagnosis, vasa praevia management

Introducere. Vasa praevia este o entitate clinică rară, cu un proanostic fetal rezervat. Printre posibilele complicatii fetale și neonatale șe numără: deceșul antepartum, asfixia, hemoragia, exsanguinarea rapidă si decesul imediat post-partum. Materiale si metodă. Am studiat bazele de date PubMed, MEDLINE si Cochrane prin folosirea cuvintelor-cheie "vasa praevia", "placenta praevia", "inserție velamentoasa", "placentă bilobată", "succenturiată" și literatura medicală referitoare la vasa praevia, până în ianuarie 2021. Am selectat studii randomizate și patru ghiduri naționale. Strategiile de diagnostic și management recomandate în literatura medicală au fost comparate si revizuite. Rezultate. O abordare adaptată la factorii de risc a fost utilizată pe scară largă pentru a facilita diagnosticul ultrasonografic al vasa praevia. Utilizarea ecografiei transvaginale Doppler nu s-a dovedit rentabilă la populația aenerală, însă este recomandată în screeninaul pacientelor din grupurile cu risc crescut. Nu toate cazurile pot fi diagnosticate prenatal. Concluzii. Vasa praevia este o afecțiune obstetricală rară, care, nediagnosticată antenatal, a fost asociată cu o rată ridicată a mortalității perinatale. Sunt necesare protocoale standardizate de screening prenatal, atât în cazul sarcinilor unice, cât și în cazul celor multifetale, cu scopul de a îmbunătăți rata de diagnosticare a vasa praevia, supraviețuirea neonatală și pentru a reduce riscul complicațiilor intra- și postpartum.

**Cuvinte-cheie:** vasa praevia, placenta praevia, inserție velamentoasă, placentă bilobată, diagnostic ecografic, conduita în vasa praevia

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#### Introduction

*Vasa praevia* is a rare disorder of placentation associated with an estimated prevalence between 1 in 2000 and 1 in 5000 pregnancies<sup>(1-7)</sup>. The reported incidence of *vasa praevia* varies, however, based on the method of conception (between 1 in 513 and 1 in 6000 cases in naturally conceived pregnancies, to 1 in 293 cases in artificially conceived pregnancies)<sup>(8,9)</sup>. It has been associated with a high perinatal mortality rate of approximately 60% if undiagnosed prenatally and managed appropriately<sup>(3)</sup>, carrying additional maternal risks associated with undergoing emergency caesarean section<sup>(10-12)</sup>.

Antenatal diagnosis of *vasa praevia via* ultrasound scanning has allowed for a significant drop in perinatal mortality (most recent case series suggesting a mortality rate of less than 10%), compared with intrapartum or postpartum diagnosis (which have reported survival rates of 43.6%). Fewer cases of blood transfusions in newborn infants with prenatally diagnosed *vasa praevia* have been reported (approximately 3%), compared with newborns diagnosed intra- or postnatally (approximately 60%)<sup>(12)</sup>.

#### **Objectives**

The focus of this paper is to evaluate the role of ultrasound imaging in the diagnosis of *vasa praevia*, as

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**Figure 1.** Ultrasonographic examination of a patient at 37 weeks of gestation, with vasa praevia and bilobed placenta (type II vasa praevia). The succenturiate lobe and the main placental mass are connected by the fetal unprotected vessel (a). Color and pulsed wave Doppler indicate that the heart rate is in keeping with the fetal heart rate, therefore this is a fetal vessel (b, c) (arrow)

well as to describe the etiology, the various clinical presentations, and the management strategies for this condition.

#### Materials and method

We searched PubMed, MEDLINE and the Cochrane Library using appropriate variables (e.g., "vasa praevia", "placenta praevia", "velamentous cord insertion", "succenturiate", "bilobed placenta"), along with the medical literature related to *vasa praevia* up to January 2021. We selected randomized control trials, observational studies, systematic reviews and four national guidelines developed by the Society for Maternal-Fetal Medicine, the Royal College of Obstetrician and Gynecologists, the Royal Australian and New Zealand College of Obstetricians and Gynecologists and by the Society of Obstetricians and Gynecologists of Canada. The diagnostic and management strategies recommended in medical literature were compared and reviewed.

#### **Risk factors and complications**

*Vasa praevia* is an uncommon obstetric condition in which the fetal umbilical vessels, unsupported by the umbilical cord or placental tissue, cross the amniotic membranes above the internal os of the cervix and below the fetal presenting part<sup>(10,12,13)</sup>. Abnormal insertion of the umbilical cord (velamentous cord insertion or insertion in the lower third uterine part diagnosed at the first-trimester ultrasound<sup>(14-16)</sup>), placental anomalies (*placenta praevia*, bilobed or succenturiate), multiple pregnancies or pregnancies obtained *via* assisted reproduction methods are just a few of the potential risk factors for *vasa praevia*<sup>(9,17-19,21,22)</sup>.

Vasa praevia has a high risk of rupture in both active and augmented labor (especially when amniotomy is performed), leading to fetal hemorrhage, rapid exsanguination, or even death. During active labor, fetal blood vessels may be compressed, leading to fetal asphyxia<sup>(17)</sup>.

#### Vasa praevia classification

*Vasa praevia* has been classified into two main types<sup>(13)</sup>. In type I *vasa praevia*, the umbilical cord has a velamentous insertion and fetal blood vessels overlie the internal os, or are located near it. Thus, patients with low-lying placenta or resolved *placenta praevia* are at risk. Type II occurs when succenturiate or additional placental lobes, connected by the fetal umbilical vessels located over or near the cervix, have been identified<sup>(23)</sup>.

In order to facilitate the diagnosis of *vasa praevia* using ultrasound examination, a consensus has been reached regarding the distance of the fetal vessels from the internal os, with a proposed threshold of no more than  $2 \text{ cm}^{(24-26)}$ .

#### Antenatal diagnosis

The antenatal diagnosis of *vasa praevia* is generally made around 18 to 24 weeks of gestation by routine

ultrasound scanning<sup>(27,28)</sup>, but needs to be confirmed by a follow-up ultrasound evaluation during the third trimester (at 30 to 32 weeks of gestation)<sup>(3,24,29)</sup>. The ultrasound diagnosis of vasa praevia during the second trimester has a reported estimated detection rate of 93%, 99% specificity<sup>(30)</sup> and a 20% rate of cases resolved before delivery<sup>(25,31)</sup>. If vasa praevia is first discovered in the third trimester, ultrasound examination has proven to be much less effective<sup>(22)</sup>. Zang et al., in 2019, proved that accurate antenatal diagnosis of vasa prae*via* can be achieved by a two-stage screening strategy. In the first trimester, patients with velamentous cord insertion at the inferior pole of the placenta and second trimester patients with low-lying placenta must be classified as high-risk for vasa praevia and transvaginal ultrasound offered, specifically searching for vasa praevia at the time of the mid-trimester anomaly scan. The recommended strategy is to use color Doppler to diagnose or exclude vasa praevia by identifying vessels within 5 cm of the internal  $os^{(32)}$ .

The screening for vasa praevia can also be stratified according to risk factors: velamentous umbilical cord insertion, succenturiate or placenta with accessory lobes, IVF pregnancy, multifetal pregnancies<sup>(9,17-19,21,22)</sup>. If, during the transabdominal scan, vasa praevia is suspected, transvaginal ultrasound evaluation using color and pulsed wave Doppler should be performed to confirm or to exclude the diagnosis. The presence of an arterial vessel in close proximity or directly over the internal cervical os, with a blood flow rate matching the fetal heart rate, confirms the existence of vasa prae $via^{(33-35)}$ . When technically possible, it is important to establish the course and the existence of the fetal vessels within the membranes, in order to rule out other conditions (such as venous sinus, funic presentation or marginal vein) that may explain the presence of a vessel near the cervix (Figure 1)<sup>(36)</sup>. The importance of cervical-length ultrasound screening is still unknown<sup>(37)</sup>.

#### Intrapartum diagnosis

The detection of the pulsating fetal vessels in close proximity to the internal os *via* vaginal examination during early labor, vaginal bleeding or signs of acute fetal compromise following the spontaneous rupture of the membranes or amniotomy (e.g., sinusoidal fetal heart rate tracing or sudden onset fetal bradycardia) facilitate the intrapartum diagnosis of *vasa praevia*<sup>(38)</sup>.

#### Management

The prenatal diagnostic rate of *vasa praevia* is approximately 98%<sup>(30,39)</sup> and it provides the improvement of neonatal and maternal outcomes due to the administration of corticosteroids timed between 28 to 32 weeks of gestation for fetal lung maturation, and elective preterm hospitalization timed between 30 to 34 weeks, based on individual factors, such as: history of preterm birth, preterm contractions, vaginal bleeding, distance from hospital and planned delivery *via* elective caesarean section prior to spontaneous rupture of membranes<sup>(27,28)</sup> timed between 34 to 37 weeks of gestation (in asymptomatic women)<sup>(40,41)</sup>, in a center capable of providing immediate neonatal blood transfusion (using type O negative blood) and pediatric support.

The American College of Obstetricians and Gynecologists and the American Institute of Ultrasound in Medicine have recommended an algorithm for the management of *vasa praevia*<sup>(42)</sup>. Placental location, its distance from the internal cervical os and the placenta cord insertion site should be examined in the mid-trimester ultrasonography<sup>(30,43,44)</sup>. If no anomalies have been detected, routine care should be applied<sup>(38)</sup>.

If placental anomalies have been detected (lowlying placenta or placenta praevia identified during the middle of the second trimester) in asymptomatic patients, a follow-up ultrasound scan is recommended around 30 to 32 weeks of gestation $^{(3,24,29)}$ , since these conditions, even though resolved, are often associated with vasa praevia<sup>(38)</sup>. In order to rule out vasa praevia, transabdominal, coupled with transvaginal ultrasonography with color and pulsed wave Doppler are recommended. If patients are symptomatic or present with vaginal bleeding, ultrasound evaluation may be indicated earlier. If vasa praevia and a cervical length of less than 25 mm are identified, hospitalization and corticosteroid administration should be conducted with planned delivery via elective caesarean section at 35 to 36 weeks of gestation. If placenta praevia is diagnosed, without vasa praevia, elective delivery shall be carried out at 37 weeks of gestation<sup>(4,36)</sup>. In cases when vaginal bleeding or premature rupture of membranes occur in patients antenatally diagnosed with vasa praevia, the admission to the birthing unit and continuous electronic fetal heart rate monitorization are recommended. When technically possible and if time permits, rapid tests for fetal hemoglobin should be conducted. Urgent caesarean section is recommended if abnormalities are detected during the fetal heart rate monitorization or in the fetal biochemical test<sup>(38,45-51)</sup>.

There is no conclusive data regarding the role of transvaginal ultrasound cervical length measurements and cerclage<sup>(37)</sup>. The 2017 prospective population-based cohort study suggests that outpatient care is feasible and associated with excellent outcomes if vaginal bleeding or preterm contractions are absent and if there is no evidence of cervical shortening<sup>(52)</sup>. The weekly measurement of cervical length is recommended.

The preoperative evaluation of the course of the fetal vessels using color Doppler ultrasonography lowers the risk of intraoperative vessel laceration. If, however, laceration has occurred, immediate cord clamping should be performed, in order to limit the fetal/neonatal blood loss<sup>(38,49)</sup>.

No consensus has been reached regarding the optimal time for performing the caesarean section<sup>(53)</sup>. The delivery between 34 to 37 weeks of gestation limits the risks of iatrogenic preterm delivery and complications regarding the onset of preterm labor<sup>(12,54)</sup>.

#### Discussion

Prenatal screening for placental cord insertion, placenta praevia or low-lying placenta, multifetal or IVF pregnancies, succenturiate or bilobed placenta or low cord insertion (in the first trimester) facilitate the timely diagnosis of vasa praevia<sup>(12,24)</sup>. The screening for vasa praevia is recommended during the mid-trimester ultrasound<sup>(27,28)</sup> (99.8% specificity, and 100% sensitivity)<sup>(30)</sup>. The novel strategy on a two-stage protocol of screening for vasa praevia suggests that the antenatal diagnosis of vasa praevia and the appropriate monitoring and delivery can potentially reduce the overall rate of stillbirth by about 10%<sup>(32)</sup>. According to medical literature, the universal transvaginal screening with color and pulse Doppler is not recommended or cost-effective. False positives may occur when membrane separation or marginal placental sinuses are present, or when there is an umbilical cord loop covering the cervix. False negatives may result from improper angle of insonation during the color Doppler examination<sup>(19)</sup>.

When risk factors are present, targeted ultrasound screenings are recommended. Even so, undiagnosed cases of vasa praevia still occur<sup>(6,55)</sup>.

The role of cervical length ultrasound screening in the management of vasa praevia and the outcome of outpatient follow-up versus hospitalization at 30-32 weeks of gestation must still be established by prospective multicentre studies. The optimal timing of delivery in pregnancies with vasa praevia must also be further studied in ensuing randomized controlled trials. Due to insufficient data being provided regarding the management of *vasa praevia*, the available guidelines are inconsistent in their recommendations and management strategies. Most often, vasa praevia is managed according to individual institutional policy.

#### Conclusions

Vasa praevia is a relatively rare obstetric condition associated with a high perinatal mortality rate. When diagnosed antenatally (around 18 to 24 weeks of gestation), excellent outcomes can be expected. Being a relatively rare and underreported disorder, screening strategies for prenatally detectable risk factors should be further developed in both singleton and multiple pregnancies. It is necessary to establish if cervical-length ultrasound screening in pregnancies diagnosed with vasa praevia is useful. The routine screening for the umbilical cord insertion site is strongly recommended and should be performed when technically possible<sup>(42,56)</sup>.

Based on national guidelines, observational data and expert opinion, the current recommendations include ultrasound screening for risk factors, the administration of prenatal corticosteroids at 28 to 32 weeks of gestation and the delivery by elective caesarean section at 34 to 37 weeks of gestation in asymptomatic women, earlier (34 to 35 weeks of gestation) if the patients are at high risk of preterm delivery, with a further recommendation of delivery around 32 to 34 weeks of gestation in twin pregnancies.

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