

Vasa Praevia; a Preventable Tragedy

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Vasa praevia can have the catastrophic consequences of a fetal or neonatal death. There are few antenatal diagnoses whereby ultrasound can have such a direct impact on outcome and dramatically increase the chances of survival in pregnancies affected by this condition. The exclusion of vasa praevia can take under a minute and its inclusion within the routine anomaly scan should be prioritised. Until this occurs, healthy babies will continue to die due to this condition.

Keywords: Vasa Praevia, Pregnancy Complication, Prenatal Diagnosis, Ultrasound Technique, Ultrasound Best Practice

'The condition of vasa praevia has robbed us of our beautiful baby boy, our first child and we will never be the same again. We just wish one of the medical staff had had the foresight to scan for this condition'

Natalie Samat, whose child Henry Cameron Samat died from un-diagnosed vasa praevia, November 2005

Introduction

The aim of this paper is to persuade all those undertaking the detailed anomaly scan that excluding vasa praevia (VP) is a worthwhile endeavour and one that is easily achievable within the confines of the second-trimester anomaly scan. In this paper, we review the mechanisms leading to VP as well as the incidence, clinical implications, and risk factors associated with this condition. A brief review of the literature will help further to build a convincing case for routine ultrasound screening for this condition. Images will enable the reader to understand the ultrasound views required to exclude VP and clear guidance on the technique required to confirm the diagnosis will be provided. Best practice guidance will be given in the form of an algorithm and information regarding internet access to educational support materials will also be made available to assist the reader. It is hoped that the devastation felt by numerous parents at the preventable death of a healthy baby, as expressed so clearly by Natalie Samat in the given quotation, will then be confined to the past.

What is Vasa Praevia?

Vasa praevia is a condition whereby fetal or placental vessels course over the cervix beneath the presenting part.¹ These vessels run unprotected by either the Wharton's jelly or placental tissue and are, therefore, vulnerable to laceration and compression at any stage of pregnancy, though these complications most commonly arise at the time of delivery.¹⁻³ This condition may occur as the result of either velamentous cord insertion (VCI), when the umbilical cord inserts into the membranes away from the placental mass and the velamentous vessels cross through the lower uterine segment (VP type 1 – Fig. 1), or from fetal vessels running between lobes of a

placenta with one or more accessory lobes (VP type 2 – Fig. 2).^{4,5} The reason why VP has such a poor outcome is due to the potential for the exposed vulnerable fetal vessels which course close to the cervix to rupture when the membranes break, leading to fetal exsanguination.¹⁻⁶ The presenting fetal part may also exert direct pressure on the unprotected vessels and fetal death can occur due to asphyxiation.⁷

What is the Incidence of Vasa Praevia?

The reported incidence of VP is approximately 1 per 2500 deliveries.³ The latest yearly UK birth rate figures show that there were 734 000 live births⁸ which would account for approximately 293 babies being affected by this condition. The literature however supports the possibility that this figure is likely to be an underestimation in view of the fact that the condition of VP in asymptomatic pregnancies may be unrecognisable at the time of delivery, and therefore not reported in the literature.⁵ Furthermore, the incidence of VP in IVF pregnancies has recently been reported to be as high as 1 in 300 pregnancies,^{9,10} which has been attributed to the increased proportion of placental morphologic alterations seen in pregnancies achieved by using assisted reproductive techniques.^{11,12}

What are the Implications of Vasa Praevia?

The impact of VP on the pregnancy can be devastating with a high reported fetal and neonatal mortality rate.¹⁻³ An alarming complication of VP is fetal bleeding which often presents unexpectedly leading to rapid exsanguination, shock and fetal death.⁶ In the pre-ultrasound era, the reported mortality for this complication was at least 75%.¹³ Traditionally, VP presents with painless vaginal bleeding at the time of spontaneous or artificial rupture of the membranes. Abnormalities of the fetal heart rate and fetal distress quickly follow, leading to an emergency caesarean section and the delivery of a severely anaemic, hypovolemic neonate, which requires rapid reposition of volume and blood transfusion as a life saving manoeuvre.¹⁻³ Reported methods for the diagnosis in symptomatic cases include the palpation of velamentous vessels through the dilated cervix, direct visualisation of the vessels

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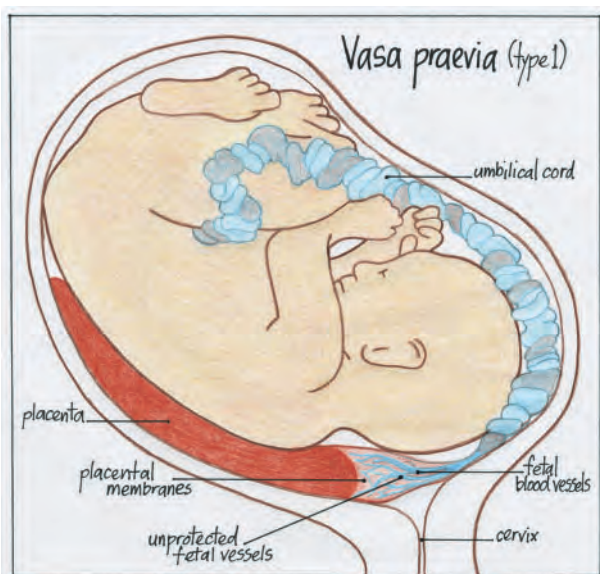


Figure 1. Illustration of vasa praevia type 1.

with an amnioscope or testing the origin of blood looking for fetal haemoglobin.¹³ Unfortunately, the emergent and often chaotic situation faced by the perinatology team often precludes the use of any of these diagnostic techniques.

In the last two decades, prenatal ultrasound has played a critical role in the detection of VP well in advance of the development of clinical complications. Indeed, a recently published large multi-centre study of VP reports on the significant improvement in the perinatal outcome when an ultrasound antenatal diagnosis of VP is made and a caesarean section is appropriately scheduled.¹⁴ The study involved 155 cases of VP, of which only 61 (39%) were diagnosed prenatally, and demonstrated that in 97% of cases where a prenatal ultrasound diagnosis was available the infant survived, compared with a 44% survival rate when the condition was undiagnosed. Even when the infant survives an undiagnosed VP, this study clearly demonstrated that the post-natal course is often complicated, with over half of the infants requiring a blood transfusion and registering poor Apgar scores.¹⁴ Numerous authorities now support the view that the only effective strategy to reduce the high mortality rate associated with VP is a reliable antenatal ultrasound diagnosis and timely delivery by caesarean section.

The Role of Ultrasound in Excluding Vasa Praevia

It would seem appropriate to discuss the literature in light of any concerns that the ultrasound community might have about incorporating the exclusion of VP into their departmental scanning protocols. The prevalence of this condition, impact on scanning time, equipment, optimum gestational age for screening and any additional skill levels required are important points to be considered.

VP has been discussed in the literature over many years with Lobstein reporting the first case of a ruptured VP back in 1801.⁵ In 1987, Gianopoulos *et al.*¹⁵ first described the diagnosis of VP using ultrasound in a woman with a history of a low-lying placenta and a succenturiate lobe. In 1990, Nelson *et al.*¹⁶ reported the use of colour flow Doppler to diagnose VP at 26 weeks gestation. It was only in 1996 when the first case of VP was diagnosed at the time of the routine second-trimester anomaly scan. This was identified within the confines of a 20-minute scan and resulted in the subsequent

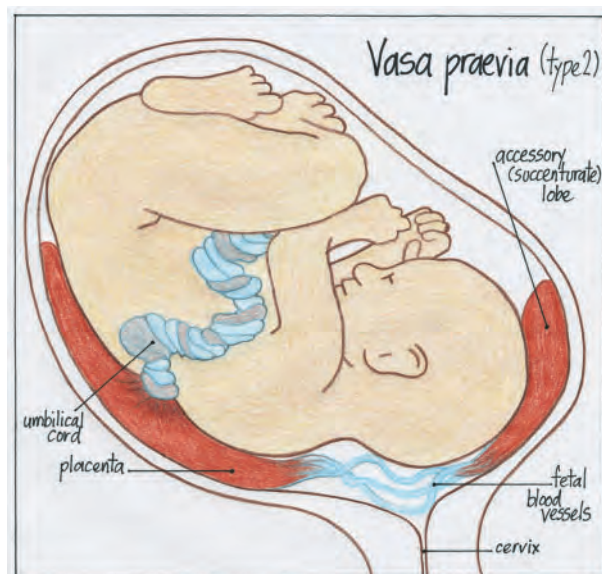


Figure 2. Illustration of a vasa praevia type 2.

delivery at 35 weeks of a healthy baby by Caesarean section.¹⁷

Although VP is a rare condition and 1 in every 2500 deliveries³ might not seem a very high incidence rate, it has to be remembered that these babies are at risk of death. In IVF pregnancies the chance of a VP rises considerably to a reported 1:300.^{9,10} As the risk of an isolated facial cleft is 1:1000¹⁸ and many ultrasound departments routinely assess the fetal upper lip as part of their anomaly scan as recommended by the Royal College of Obstetricians and Gynaecologists (RCOG),⁹ it seems appropriate that exclusion of VP, where the outcome is possibly a fetal or neonatal death, should be prioritised by the scanning community.

The chances of VP being present are minimal when the umbilical cord inserts into the placental mass or in the absence of a succenturiate or bilobed placenta.⁵ As the assessment of the placenta is already included within departmental ultrasound protocols,¹⁹ the priority should be for the placental cord insertion to be evaluated as velamentous insertion of the cord is a pre-requisite for the most common VP, type 1. The confident identification of a normal placental cord insertion is in itself a worthwhile pursuit during the second-trimester anomaly scan, with an incidence of VCI of 1% in the unselected population.²⁰ There are a number of associated complications accompanying a velamentous insertion; apart from the most catastrophic VP, there is also increased risk of placental abruption, fetal growth restriction, congenital anomaly, low Apgar score, prematurity and retained placenta.²⁰⁻²²

Several studies have remarked that the investigation of the cord insertion when undertaken in the first or second trimester takes less than one minute and can easily be incorporated into the allotted scanning time requiring no additional skills.^{20,21} Accurate assessment of the placental cord insertion prior to 11 weeks is not possible due to the trophoblast normally covering a large area of the uterine cavity.²³ Examination of the cord insertion if the patient presents for a booking scan or nuchal translucency thickness measurements between 11 and 14 weeks of gestation is advantageous as the umbilical cord and its insertion site into the placenta can be readily identified at this stage.²³ Transvaginal (TV) examination in spontaneous multiple pregnancies and those obtained with assisted reproductive techniques should routinely be undertaken to exclude VP if a transabdominal (TA) scan using colour Doppler does not clearly demonstrate the area around the internal os. This then allows, where there is a high index of suspicion, for close surveillance of the pregnancy and



Figure 3. TA scan demonstrating a grey scale image of the placental cord insertion.

confirmation of VP if present, at the second-trimester scan. It is important to highlight that the diagnosis of velamentous insertion should be specifically looked for, if a specific search for velamentous insertion is not undertaken then the diagnosis is invariably missed.²⁴

Consideration should also be given to the equipment necessary to exclude VP; colour flow mapping and pulsed-wave Doppler facilities considerably aid the diagnosis and numerous authors remark on the improvement in visualisation of the abnormally located vessels as a result of these facilities.^{1,2,5,25} Indeed some authors suggest a quick sweep over the cervix with colour flow mapping is a helpful additional measure to exclude VP during routine obstetric scans.⁵ However, the common policy of performing obstetric scans with an empty maternal bladder can occasionally make it difficult to visualise the lower segment. In this regard, the use of transvaginal probes for those cases at high risk of VP can be extremely helpful in establishing the definitive diagnosis by enhancing the visualisation of the entire lower uterine segment. This can also be easily incorporated in those departments who routinely assess the uterine cervix to screen for short cervix and risk of preterm delivery. With guidance from the RCOG¹⁹ suggesting all ultrasound equipment should be less than five years old, it is likely that many units undertaking detailed anomaly scans will have these facilities in place. Other technologies such as colour Doppler three-dimensional ultrasound and magnetic resonance imaging (MRI) have been reported in the prenatal diagnosis of VP, but these methods are cumbersome and have limited practical utility, so will not be discussed in this review (Editor's note: see accompanying case study in this issue).

When evaluating the literature regarding VP it is clear that there are few large studies due to the low overall prevalence of this potentially devastating condition. However, all of the case studies reported state how important an antenatal diagnosis is to improving the outcome of this condition⁵ and that it should be included in the routine ultrasound evaluation of the pregnancy. Oyelese and Smulian succinctly remarked in 2006: 'We can think of no other condition in which prenatal diagnosis... makes such a dramatic impact on the difference between survival and death for an otherwise healthy infant'.²⁵

Ultrasound Technique for Excluding Vasa Praevia

The ultrasound technique for ruling out VP is based on:

- assessment of placental cord insertion site;

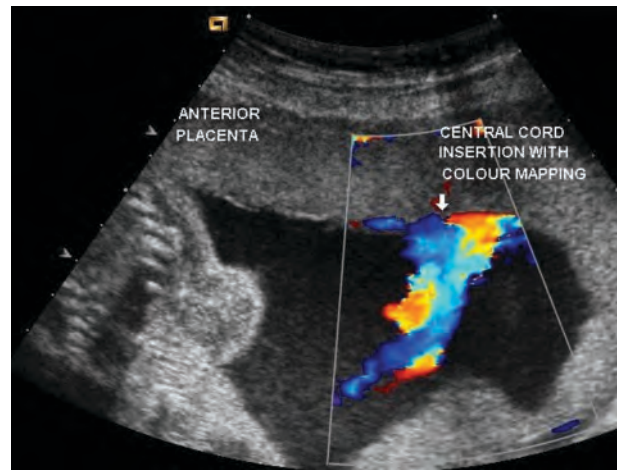


Figure 4. TA scan using colour Doppler ultrasound demonstrating the branching vessels into the placenta.

- assessment of placental appearance and location;
- consideration of other risk factors.

Assessment of Placental Cord Insertion Site

The umbilical cord inserts in the placental mass, either in the centre or paracentrally, in about 90% of pregnancies. In about 10% the insertion occurs at the margins of the placenta (marginal insertion), and in about 1% of pregnancies it inserts in the membranes away from the placental edge (velamentous cord insertion). The placental cord insertion site can be readily determined by ultrasound as part of the standard 20-minute second-trimester anomaly scan. Once the placental site has been determined attempts should be made to image the placental cord insertion site. This is done by searching the fetal surface of the placenta with high resolution grey-scale ultrasound at the appropriate magnification until this is identified (Fig. 3). Care must be taken not to mistake the true insertion site with loops of umbilical cord overlying the placental surface by demonstrating the entry of the main branches of the umbilical vessels into the chorionic plate with colour flow imaging confirming the diagnosis (Fig. 4). Also note that when the site of cord insertion into the placental mass is clearly identified, the possibility of VP type 1 is extremely low and no further action is necessary.⁵

In contrast, when the insertion site cannot be identified, efforts must be made to exclude VCI as this group of patients have a higher risk of having VP (6%).²⁶ Where a VCI cannot clearly be demonstrated in the upper segment of the uterus with a transabdominal scan, it is imperative that the area above the internal os is evaluated for velamentous insertion. This should be undertaken using a transvaginal approach and incorporating colour Doppler for a reliable diagnosis of VP (Figs. 5 and 6). Some authors have proposed a transperineal approach,²⁷ but the use of colour Doppler ultrasound is critical.

Assessment of Placental Appearance and Location

Consideration should also be given to the overall appearance and location of the placenta, as fetal vessels running across the cervix between lobes of placenta with one or more accessory (succenturate) lobes is a pre-requisite for VP type 2 (Fig. 7). It should also be noted that some pregnancies complicated by VP may have a combination of type 1 and 2 (Fig. 8).⁵

Placenta praevia (where the placenta partially or totally covers the internal os) and marginal placenta (where the margin of the placenta reaches the internal os) are considered

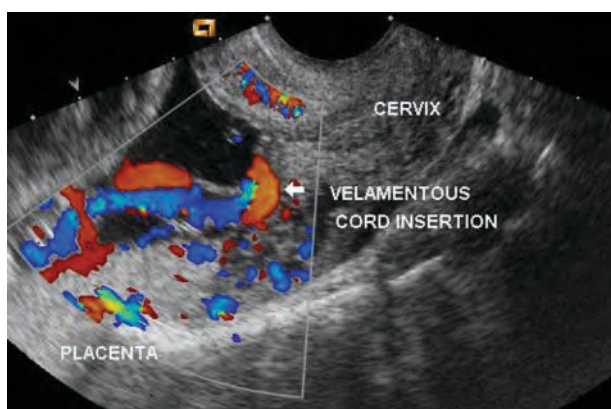


Figure 5. Colour Doppler demonstrates a VP type 1.



Figure 8. Pathological specimen shows the fetal side of a bilobed placenta with a velamentous cord insertion between the lobes (© Francois Manson and www.TheFetus.net; reproduced with permission).

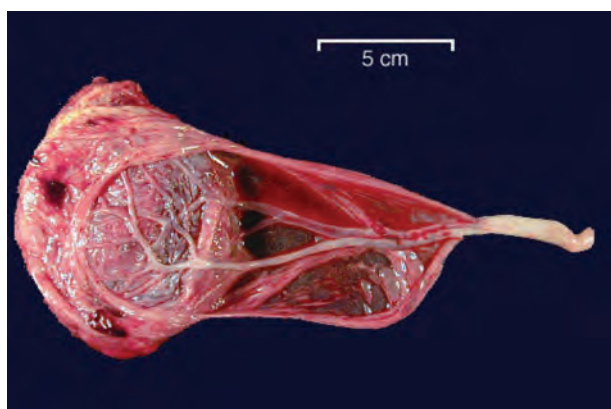


Figure 6. Pathological specimen of a velamentous cord insertion showing the umbilical cord inserting into the placental membranes and the unprotected fetal vessels (Courtesy of Edward C. Klatt MD).

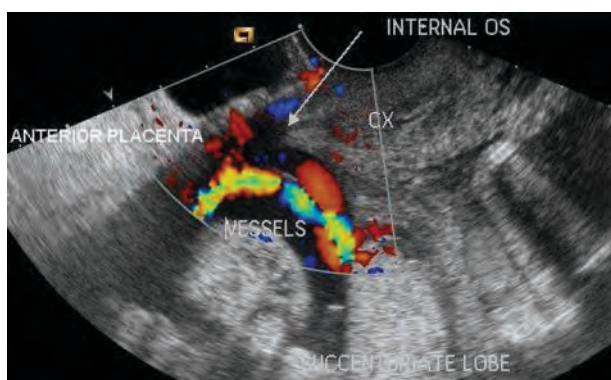


Figure 7. Vasa praevia type 2 demonstrates fetal vessels crossing the cervix from the main anterior placenta mass to a posterior succenturiate lobe.

to be risk factors for VP.⁵ Where present it is important to check for vessels near the cervix and if clearly normal then no further action is necessary (Fig. 9). As assessment of the placental site and its relationship to the internal os is already undertaken within most ultrasound departments at the time of the anomaly scan, the addition of colour Doppler over the area around the cervix is easily achievable. If optimum visualisation of the cervix cannot be achieved with a transabdominal scan, then a transvaginal approach should be undertaken.

Consideration of Other Risk Factors

Other risk factors for VP include pregnancies achieved by *in vitro* fertilisation (incidence of VP 1:300)⁹ and multi-fetal gestations (10% of VP occurs in twins).⁵ As these pregnancies are often referred for an 11–14 week scan, a high index of

suspicion should be maintained, observing the placental cord insertion and excluding placental variants. If normal appearances are demonstrated, no further action is needed. If however appearances are suspicious or overtly abnormal further assessment should occur at the routine anomaly scan. Patients presenting with vaginal bleeding should also be treated with caution and a higher index of suspicion for VP maintained.

Difficulties in Diagnosing Vasa Praevia

Obese patients, abdominal scarring and awkward fetal position can make diagnosis of VP difficult. In cases where the cord is seen close to the cervix, shifting the cord by moving the patient, or waiting and reassessing a few minutes later, can be helpful. Colour flow mapping should enable a cord insertion remote from the cervix to be demonstrated thus excluding VP (Fig. 10).

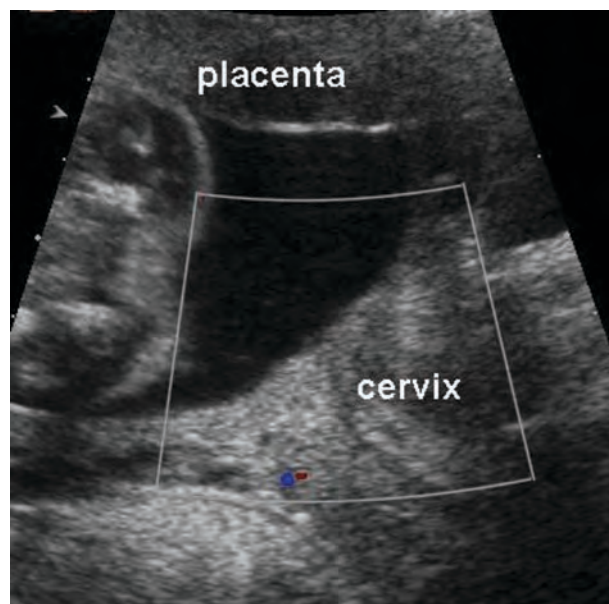


Figure 9. Transabdominal view of the internal os using colour Doppler demonstrating the absence of VP.

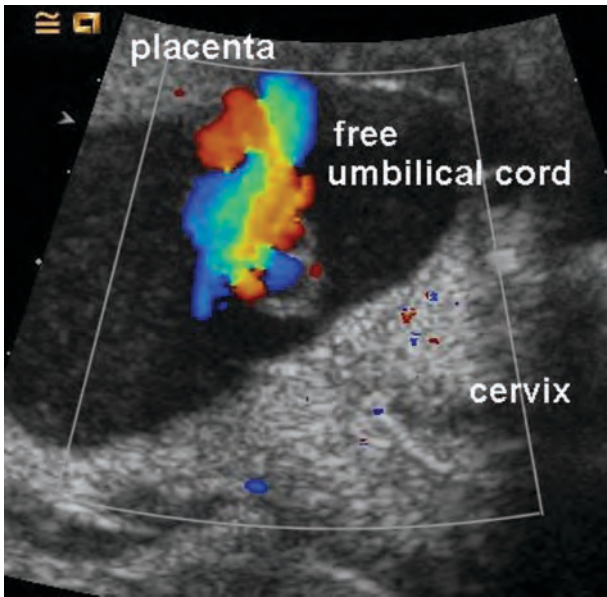


Figure 10. Transabdominal scan showing umbilical cord close to the internal os. A few minutes later after movement of the patient, the umbilical cord moved away from the cervix.

In the second trimester visualisation of the placental cord insertion site is easier to achieve than later on in pregnancy as the amount of amniotic fluid allows for a proper visualisation of the placental surface in almost all cases. In the third trimester, however, the relative decrease in amniotic fluid volume can make visualisation quite difficult. In pregnancies with a posterior placenta, gentle manipulation of the fetus through the maternal abdomen can allow proper placental surface visualisation which may be assisted by placing the mother in the lateral position. It is important where visualisation is poor and the cord insertion has not been identified previously, that a transvaginal examination of the lower segment is undertaken.

Flash artefact, which can occur as a result of amniotic fluid movement due to fetal activity, can also be a problem as this can mimic a vessel near the internal os. Repeating the evaluation once movement has subsided will usually eliminate this artefact (Figs. 11 and 12).

Any persistent structures near the cervix should always raise the suspicion of VP (Figs. 13 and 14). The use of colour Doppler will enable the operator to determine if these are vascular in nature. Alternatively a spectral trace obtained from

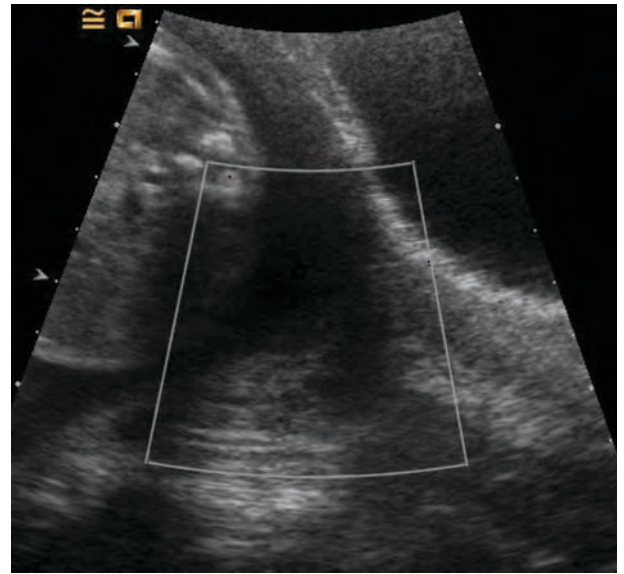


Figure 12. Further evaluation and a repeat image confirmed this to be flash artefact.



Figure 13. A second-trimester vaginal ultrasound showing a sagittal section through the cervix with suspicious soft tissue (arrow) abutting the internal os.

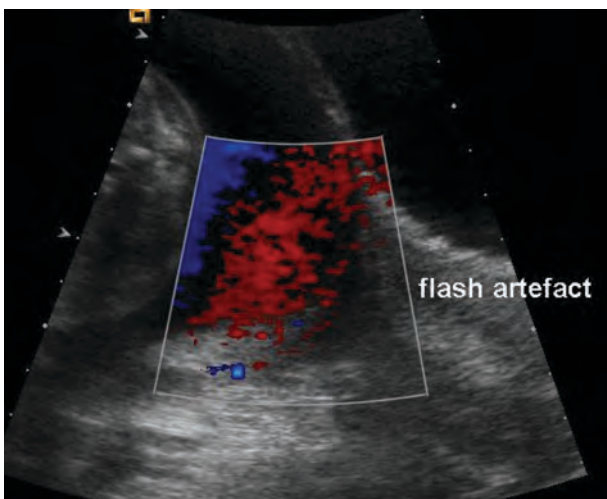


Figure 11. A transabdominal scan raising the possibility of suspicious vessels near the cervix.

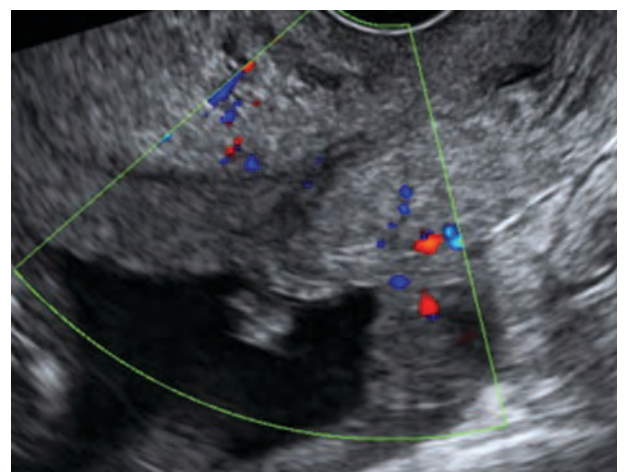


Figure 14. The use of colour Doppler shows the suspicious structure is without Doppler signal and thus is not a vessel (© Philippe Jeanty and www.TheFetus.net; reproduced with permission).

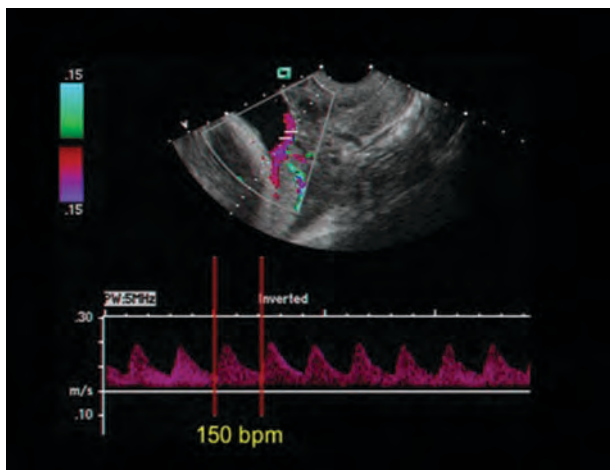


Figure 15. Second-trimester vaginal Doppler image shows a high frequency heart rate at the level of the VP. This helps to distinguish VP from maternal cervical vessels (© Philippe Jeanty and www.TheFetus.net; reproduced with permission).

the suspicious region will show pulsations at the fetal heart rate where the appearances are due to VP (Fig. 15).

Best Practice Guidelines

We recommend the algorithm proposed by Philippe Jeanty's team⁵ (Fig. 16) as a guide during the routine second-trimester

anomaly ultrasound scan and believe the guidance can also be followed in the late first trimester and for those pregnancies presenting in the third trimester.

Departmental Software Support for Vasa Praevia

To ensure assessment of the placental cord insertion site is incorporated into all routine scans it is prudent to include this in departmental guidelines and protocols. For those departments currently using the Astraia fetal database (Astraia software GmbH, München, Germany), it is possible to have a list of cord anomalies and variants in the 'placenta box' of the 'drop down' menu. This could include for example, 'three vessels with a normal cord insertion' or 'velamentous cord insertion'. The manufacturers of the Astraia database have been in discussion with the UK Vasa Praevia Awareness Group and agreed that an additional field box 'cord insertion' will be integrated in the new version of Astraia (due for release Autumn 2007). This will only refer to the placental cord insertion and can be changed to a mandatory field by the department administrator ensuring staff will not be able to bypass this field.

Online Educational Support

The UK Vasa Praevia Awareness group (www.vasapraevia.co.uk) provide excellent comprehensive training materials on their website. These can be downloaded free of charge.

Conclusions

Routine scanning of obstetric patients has changed dramatically over the last few decades. Advances in scanning

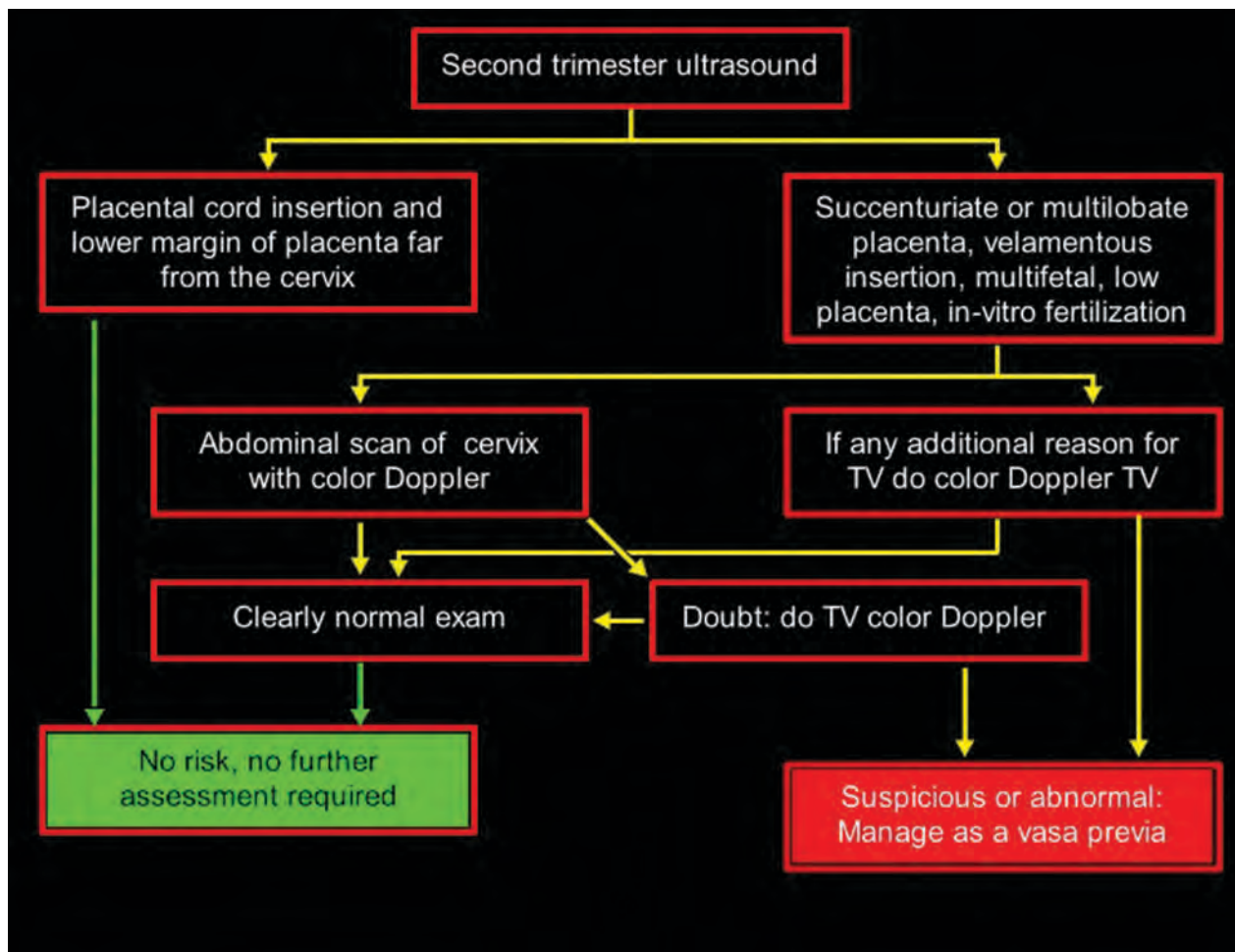


Figure 16. Proposed diagnostic algorithm for the second-trimester detection of VP (Reproduced from Derbala Y, Grochal F and Jeanty P (2007) with permission).⁵

equipment have meant that more and more subtle fetal anomalies and pregnancy complications can now be identified with confidence. It is the duty of ultrasound professionals to keep pace with these advances, to incorporate them into a standard routine scan and, on identifying any anomaly, to refer patients for appropriate counselling and/or interventional procedures.

Unfortunately, there is often a time lag between innovation and its incorporation into standard practice. New methodology has to be tried and tested, new skills have to be acquired by practitioners, and additional scanning time may have to be incorporated into an already time-pressured environment.

In relation to identifying VP on a routine scan there is little excuse. The method for excluding VP is a simple colour Doppler technique, uses skills that trained practitioners already possess, and takes approximately a minute of extra examination time. Vasa praevia is life threatening to a healthy baby; a proper diagnosis and an elective Caesarean section easily prevent death. Mothers should have confidence in the ultrasound practitioner's ability to assess not only fetal wellbeing, but also any other pregnancy-related anomalies. Routine evaluation to exclude VP should be adopted in all routine obstetric scans as a matter of urgency. Until this occurs, babies will die unnecessarily and families will continue to suffer the avoidable loss of a healthy baby.

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