

Absence of the ductus venosus: a case report

The ductus venosus connects the umbilical vein with the inferior vena cava permitting oxygenated blood to return from the placenta to the fetal heart. Its absence has been recorded infrequently and has been associated with fetal demise. A case in which the umbilical vein joined tributaries of the hepatic vein and was associated with cardiomyopathy and early death from respiratory failure, is presented.

Jacqueline Smith

RSCN, MSc, Neonatal Nurse Practitioner,
The Townsville Hospital, Neonatal Unit, and
Doctoral Student at James Cook University,
Queensland, Australia
Jacquelinej_smith@health.qld.gov.au

John Whitehall

FRACP, Director of Neonatology,
The Townsville Hospital,
Neonatal Unit, Queensland, Australia
john_whitehall@health.qld.gov.au

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Key points

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1. It was not until a decade ago that the first cases of absent ductus venosus (ADV) were detected *in utero*.
2. The ductus venosus plays a key role in the distribution of the umbilical venous return.
3. In fetuses with ADV umbilical venous occurs either by extrahepatic venous drainage or intrahepatic venous drainage.
4. The prognosis appears to be better in ADV cases with intrahepatic venous drainage.
5. Genetic counselling should be made available to parents of ADV infants in view of the high incidence of associated anomalies.

The ductus venosus (DV) is a blood vessel unique to the fetal circulation. Before birth the DV functions as a direct connection between the umbilical vein and inferior vena cava, bypassing the hepatic circulation and liver and shunting up to 40% of the oxygenated blood to the fetal brain and myocardium. In conjunction with other fetal shunts – the foramen ovale and ductus arteriosus – it plays a critical role in preferentially shunting oxygenated blood to the fetal brain.

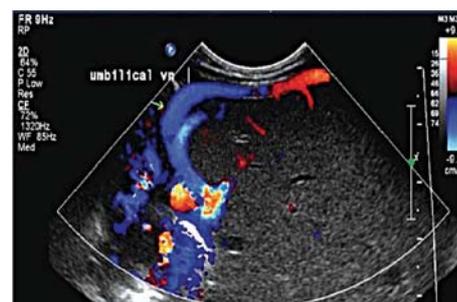
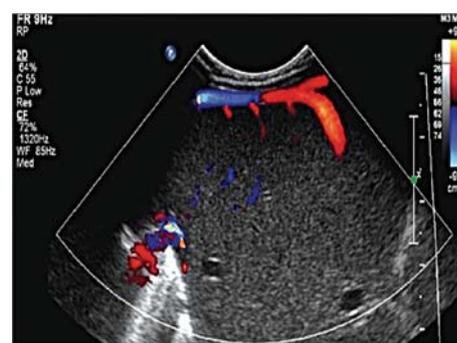
Ultrasound is an invaluable diagnostic tool in maternal-fetal medicine and with modern techniques the DV is easily identified. A case history with a rare finding of the absence of the DV, which was diagnosed during a routine ultrasound examination, is reported.

Case report

Before birth, a male infant was diagnosed with an absent DV, thick myocardium, tricuspid regurgitation and hydrops, together with an absent left kidney and large cysts in the right kidney. The umbilical vein coursed anteriorly over the surface of the liver towards the heart (FIGURE 1).



FIGURE 1 The umbilical vein coursing anteriorly over the surface of the liver towards the heart.



FIGURES 2 and 3 A confluence of veins which lead to the right atrium.

The infant was delivered by caesarean section at 29 weeks and six days for polyhydramnios and worsening hydrops fetalis. After birth he needed prolonged ventilation due to difficulties in oxygenation, associated with pulmonary hypertension as revealed by echocardiography and lung disease by X-ray.

Ultrasonography confirmed the umbilical vein coursing anteriorly to join a confluence of veins inferior to the right atrium. Multiple branches of the portal system were revealed to join the hepatic veins in a porto-systemic shunt which also drained into the confluence. The inferior vena cava was interrupted and though not visualised was concluded to also drain into the confluence (FIGURE 2 and 3). Head ultrasonography initially revealed a grade 3

haemorrhage and subsequently atrophy.

Echocardiography revealed bi-directional shunting through a patent foramen ovale, multiple small ventricular septal defects and a very thick myocardium which may not have compacted. There was also persistent pulmonary hypertension.

The infant remained in need of high concentrations of oxygen delivered under high pressure by various forms of ventilation, did not respond to nitric oxide or sildenafil and was unable to be resuscitated on day 98.

Literature review

Paired umbilical veins appear at the end of the second embryonic week. They enter the cardinal veins (major systemic venous channels), which then join the sinus venosus. The umbilical veins then fuse with the veins of the primitive gut and drain into the sinus venosus. At around the fourth to fifth week of embryonic development the liver bud extends swiftly and integrates the cranial portion of the vitelline vein (which drains blood from the yolk sac) into the hepatic sinusoids. The right umbilical vein regresses, and all the blood then flows through the left umbilical vein. Connections between the left umbilical vein and the intrahepatic sinus venosus are progressively lost. To ensure normal blood flow, the ductus venosus is formed by the coalescence of several hepatic sinusoids. This phase of development has been called critical anastomosis. If this normal development is disturbed, the embryology of the sinus venosus is disorganised and the ductus venosus fails to form. If the ductus venosus is absent other vascular connections must develop to allow proper oxygenation and development of the fetus¹.

It was not until a decade ago that the first cases of absent ductus venosus (ADV) were detected *in utero* using modern ultrasound techniques²⁻⁵.

The fetal DV connects the intra-abdominal umbilical vein to the inferior vena cava (IVC) at its inlet to the heart. The pressure gradient causes well-oxygenated blood in the DV to accelerate towards the left lateral wall of the IVC enabling its preferential streaming through the foramen ovale and ultimately to the cephalic and coronary circulation⁶. However the significance of the absence of the DV with direct communication of the umbilical vein to the heart is still very unclear. A study by Kiserud et al⁶ noted that in normal fetuses the amount of umbilical

venous blood streaming through the DV to the left heart decreased from 30% to less than 20% during the second half of pregnancy, ultimately resulting in an increase in percentage of blood flow to the liver compared to the brain. This data supports the hypothesis that the DV shunt plays less of an important role in supplying well-oxygenated blood to the brain and the myocardium in late gestation⁷.

In the absence of the DV the normal streaming of highly oxygenated umbilical venous blood through the foramen ovale to the left atrium is absent. According to Jaeggi et al⁸ the reason why we do not see any growth restriction in these infants is because the entire oxygenated blood from the placenta returns directly to the heart via the umbilical vein so that fetal arterial blood oxygen concentration may not be affected.

The DV however plays a key role in the distribution of the umbilical venous return, even more so because the fraction of the umbilical blood streaming through the DV increases significantly in hypoxaemia and decreased return^{6,9}.

There are two different routes for umbilical venous return that have been described in fetuses with ADV:

- extrahepatic venous drainage bypassing the liver, where the umbilical vein directly connects to the iliac vein, the IVC, the renal vein, the right atrium or exceptionally the left atrium of the coronary sinus^{8,10-12}
- intrahepatic drainage, without liver bypass, where the umbilical vein connects to the portal sinus in its usual way without giving rise to the DV^{6,11,13}.

In a cohort, studied by Berg et al¹⁴, a total of 63 prenatally diagnosed cases with ADV were reported over the ten year study period. A significant association was demonstrated between extrahepatic umbilical venous drainage, portal vein agenesis and cardiomegaly, which has been linked to severe postnatal complications. In infants with extrahepatic umbilical venous drainage who have no additional fetal anomalies, the prognosis seems to depend on the presence or extent of any fetal congestive heart failure^{8,12}. The prognosis of isolated ADV seems to be more favourable in the presence of intrahepatic venous drainage¹⁴.

Among the fetuses with no or minor associated anomalies the outcome was significantly better in the group without liver bypass. None of the 13 fetuses without

liver bypass died or had long term sequelae attributed to ADV, whereas in the group who were diagnosed with liver bypass, 20 out of 29 died¹⁴.

According to some reviews previous cases of ADV with extrahepatic umbilical venous drainage were associated with fetal malformations which included aneuploidies (varying numbers of chromosomes), high output cardiac failure and significant agenesis of the fetal portal system. Postnatal complications in this group can include pulmonary oedema, hepatic tumours, congestive heart failure and focal nodular hyperplasia^{8,11-13}. It has also been noted that cardiomegaly seems to occur frequently when there is direct drainage of the umbilical vein to the heart, which suggests a high central venous pressure. This can be due to volume overload because of the loss of the DV regulatory mechanism, which may result in high output failure and fetal hydrops^{10,12}.

Conclusion

Absence of the ductus venosus has been reported infrequently. If the umbilical vein joins the portal vein within the liver the prognosis is more favourable but, as in this case, if it courses externally, the outcome is poor. It is often associated with other abnormalities and heart failure. This case appears unique with the widespread abnormalities of systemic and portal vasculature.

Careful evaluation of the DV should be done routinely on fetuses that show evidence of cardiomegaly and polyhydramnios. In view of the limited knowledge of the mechanisms involved in the genesis of this anomaly and its associations with congenital malformations, fetal karyotyping should be considered. Genetic counselling should also be made available to the parents of an ADV fetus in view of the high incidence of associated anomalies.

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