

A second vestigial umbilical vein: a case report

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Abstract

A healthy female infant was found to have two umbilical arteries and two umbilical veins at the cut surface of the cord at birth. Detailed inspection of the cord showed the second vein to represent a short segment vestigial vessel that, moreover, was not associated with any other congenital anomaly as is often found in infants with umbilical vein anomalies.

Introduction

The primitive umbilical cord starts to develop at around 12 days after implantation, forms a connecting stalk by 20 days and, by 5 weeks, contains the yolk sac stalk (vitelline duct), allantois remnant and umbilical vessels including two arteries and a vein. By 10 weeks the vitelline duct disappears and, by 3 months all barring the umbilical vessels and Wharton's jelly remain. Initially, two umbilical veins originate in the chorionic villi from where they carry oxygenated blood to the embryo. They first connect with the hepatic venous plexus but, by the 6th week *in utero*, the proximal segment of the left vein and the entire right vein involute, leaving the left vein as the only venous contact with the placenta.¹

Hence, the vast majority of newborns have three vessels in their umbilical cord: characteristically two arteries and a single vein. Anomalies in this vascular arrangement are relatively common (0.5-1% of live births), and may be associated with other anomalies including renal (the majority, including duplex and ectopic kidneys), as well as cardiac (mostly septal defects) and chromosomal abnormalities (especially trisomy 18), intrauterine growth retardation and stillbirth, amongst others.²⁻⁴ However, these anomalies almost always involve the umbilical arteries and, in most cases, amount to a single rather than two arteries in 0.5-2.5% of pregnancies.² In contrast, anomalies of the umbilical vein(s) are extremely rare and, although they have been characterised into four groups (Table 1),⁵ many amount to a very small number of anecdotal reports.⁶ Most involve persistence of abnormal umbilical venous structures (Group I), persistence of the right umbilical vein with one or two arteries (Groups II, III, respectively) and absence of the left vein (Group IV). These anomalies are separate from distinct vascular loops within the cord and intra-abdominal varices and are often associated with significant congenital abnormalities. This report describes the presence of two umbilical veins, only one of which was complete, the second short segment representing a vestigial vein and, we assume, is a *forme fruste* of the persistent right umbilical vein anomaly seen in Group II.⁵ No previous report of such an anomaly exists in the current literature and, in this case, was not associated with other congenital anomalies.

Keywords

Vestigial umbilical vein

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Case report

A healthy Caucasian infant was born at term weighing 3.7kg by elective, uncomplicated caesarean section following a



Figure 1: Cut section of umbilical cord. Cut surface of neonatal-end of umbilical stump showing two smaller umbilical arteries (lower midline and right) and two veins oozing blood (lower left and upper midline)

previous caesarean section. All routine antenatal assessments and ultrasound scanning at 16 and 24 weeks gestation were entirely normal. Initial examination at birth confirmed a healthy female infant without any dysmorphic features, abnormalities of the external ears and pre-auricular areas. However, inspection of the neonatal end of the umbilical cord clearly showed two typical protuberant, thick-walled umbilical arteries in spasm and, in addition, two larger, thin-walled flaccid umbilical veins oozing blood in characteristic fashion (Figure 1). The latter were probe-patent to one centimetre at the fetal end, but further, detailed inspection with sequential dissection of the remainder of the umbilical cord and its insertion into a macroscopically-normal placenta could not confirm four vessels (Figure 2). Detailed histological sections of several segments of cord confirmed two arteries and a single vein. Similarly, no vascular loops could be detected both macroscopically and after histological dissection. Ultrasound scanning of the infant at four days of age showed normal internal abdominal organs and, in particular, no extension of the urachus, no renal or vascular anomalies involving the inferior vena cava and portal systems and excluded the presence of any vitelline structures,⁶ persistence of the right sided umbilical vein^{5,7} or other abnormal intra-abdominal vessels or varix.⁸

Table 1: Classification of umbilical vessel anomalies

Group	Vitelline cord	UA	LUV	RUV	Associated anomalies
Group I	present	2	1	0	usually
Group II	absent	2	1	1	sometimes
Group III	absent	1	1	1	common
Group IV	absent	1	0	1	common

UA=umbilical artery; UV=umbilical vein; LUV=left UV; RUV=right UV. Modified from Stevenson and Hall, 2005.⁵

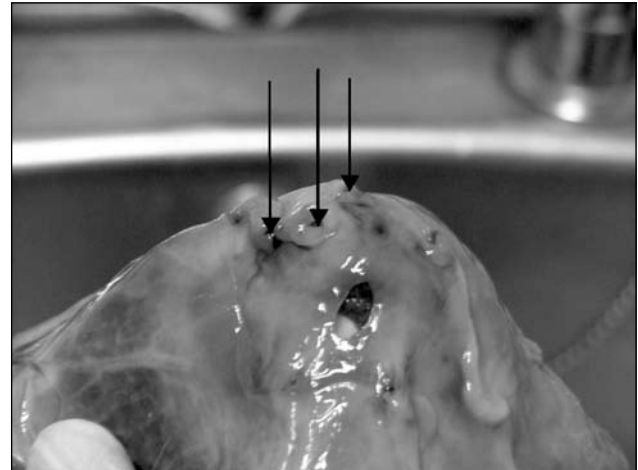


Figure 2: Insertion site of umbilical cord into placenta. Transected insertion of umbilical cord into placenta showing three vessels indicated by arrows

Discussion

This infant had two umbilical veins at the insertion into the infant's abdomen but not rostrally throughout the remainder of the umbilical cord or at its origin at the placenta. A venous loop could not have accounted for this extra vessel as a particular tortuous *but single* umbilical vein looping on itself would, on cross-section, have resulted in three (not two) apparent umbilical veins or just one long tangential vein in cross section depending on the angle of transection. Macroscopically the extra vessel had all the characteristics of a vein (Figure 1) and, histologically, the complete vessels were clearly made up of two normal arteries and one vein. Therefore, this second of the 'two' vessels would appear to represent a vestigial umbilical vein or an isolated, short-segment duplication of a single umbilical vein. A blind-ending branch coming off the complete vein was a possibility but no connection to this vessel could be demonstrated, even during dissection.

Review of the literature confirms that the persistence of the left umbilical vein (LUV) alone constitutes normality, whilst the right umbilical vein (RUV) is destined to atrophy at six weeks gestation. Persistence of the RUV together with the LUV (with two or a single umbilical artery (SUA), or a solitary RUV alone is documented in anomalies classified into Groups II, II and IV, respectively.⁵ Hence, it is probable that the vestigial vein in this case represents an abnormal vessel and, therefore, is more likely to be a vestigial right umbilical vein.

Interestingly, this infant had no associated congenital anomalies, a feature present in 30-60% of infants with a SUA² and most cases of abnormal umbilical veins (UVs) comprising anomaly Groups I, III and IV.⁵ The latter anomalies are very rare amounting to a few anecdotal reports in some cases (e.g. <5 for Group I and III). Group II anomalies (2UAs + 2UVs) would appear to be more common, and have been estimated to occur in

approximately 1 in 400 pregnancies,² yet may also be associated with congenital renal, cardiac and chromosomal malformations. This association would result in significant fetal loss in utero and explain the paucity of cases with Group II anomalies in the literature (approximately 19 in a comprehensive review in 2005)⁵. However, of those who complete pregnancy and survive, many do not manifest any other malformations and, indeed, several authors recommend that further investigation may not be necessary in this subgroup.⁷

This case appears to represent a variant of the Group II persistent RUV anomaly where the two UAs and single LUV are associated with a persistent, vestigial RUV. However, this anecdotal variant has not been reported previously and, although apparently benign and not associated with any other abnormalities in this patient, it would still be prudent to investigate any similar cases, at least by ultrasonography.

Parental consent was obtained to publish this report.

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