

ORIGIN, COURSE AND ASSOCIATED CONGENITAL ANOMALIES OF TYPE 2 SINGLE UMBILICAL ARTERY: A FETAL ANATOMIC STUDY

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ABSTRACT

Introduction: Umbilical arteries normally originate from a pair of allantoic arteries. A failure of allantoic vascular system in early fetal life results in substitution by the vitelline vascular system, an inherent safety mechanism. This gives rise to anomalous course and origin of the umbilical artery. In these cases, the umbilical artery originates from the abdominal aorta and continues as a single umbilical artery.

Aim: The aim of this study is to elaborate upon our current understanding about the origin, course and associated anomalies of Type 2 single umbilical artery.

Material and Methods: Fifty five fetuses, terminated for severe congenital anomalies over a period of 10 years, were sent to the department of anatomy for academic evaluation of congenital anomalies. All the fetuses were dissected systematically to delineate the abnormalities.

Results: Thirty fetuses had two umbilical arteries with normal course on either side of allantois (urachus) and urinary bladder. Single umbilical artery was observed in 25 cases. Twenty had type 1 single umbilical artery and coursed normally. Five cases had single anomalous origin and course of umbilical artery, which was similar to type 2 single umbilical artery (SUA). After opening the abdominal cavity, the umbilical artery was not seen beside the urachus: instead it coursed posteriorly between the coils of intestine. When it was traced further, its origin was from the abdominal aorta. The aorta was hypoplastic below the origin of the single umbilical artery. All these cases were associated with cardiac, gastro-intestinal, vertebral, renal and limb abnormalities.

Conclusion: Very few cases of this abnormality have been described in literature. Only a few of these cases were diagnosed prenatally as a vitelline artery abnormality: our study will thus help refine prenatal diagnosis and management.

KEY WORDS: Single Umbilical Artery, Vitelline Origin Of Umbilical Artery, Vitelline Artery, Type 2 SUA, Congenital Anomalies.

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INTRODUCTION

The human umbilical cord usually consists of two umbilical arteries and one umbilical vein. The two umbilical arteries run one each on either side of the allantois (urachus) and urinary bladder and run downwards and posteriorly and appear to

be taking origin from the internal iliac arteries. However, in 0.5-1% of fetuses, one umbilical artery is either obliterated or simply does not form, giving rise to a single umbilical artery [1]. Occasionally, both umbilical arteries are missing and the one arterial vessel found in the umbilical

cord is in fact a persistent vitelline artery, which branches off the abdominal aorta [2]. A persistent vitelline artery can be identified mostly at necropsy and appears to be associated with serious developmental defects. This vitelline origin of umbilical artery was classified as Type II single umbilical artery (Type II SUA) by Blackburn and Cooley, and accounts for 1.5% cases of single umbilical artery [1]. Type II SUA is almost always associated with severe fetal malformations of the the caudal body wall including sirenomelia, caudal regression-dysgenesis, OEIS complex (omphalocele-exstrophy-imperforate anus-spinal defects), urorectal malformation sequence, anal atresia, exstrophy of bladder and complete urogenital agenesis; its origin and course should be determined accurately. In these cases, a single umbilical artery had originated either from the abdominal aorta or superior mesenteric artery and the abdominal aorta was hypoplastic distal to the origin of this umbilical artery [2, 3 and 4]. Normal foetus with type II SUA was reported in only a few cases which were diagnosed prenatally [5, 6]. Since little attention has so far been given to this variant of SUA in most of the published studies, we would like to elaborate upon the anatomical features of this variant of SUA and its associated anomalies.

MATERIALS AND METHODS

We evaluated 55 fetuses for congenital malformations, which were either medically terminated due to severe congenital malformations or aborted after IUFD at Bhaskar General Hospital over a period of 10 years. Detailed fetal and placental examinations were carried out for all the cases. The abdominal cavity was opened by a V-shaped incision extending from the umbilicus to both the anterior superior iliac spines and then a vertical incision extending from the umbilicus to xiphi-sternum. Both the umbilical arteries and the umbilical vein were traced. Any abnormalities in the origin and course of these vessels were noted. Then the abdominal cavity was examined for any associated abnormalities. Then the thoracic and cranial cavities were thoroughly examined and associated anomalies were noted.

RESULTS

Thirty fetuses had two umbilical arteries with normal course on either side of urachus and urinary bladder (Fig 1). Single umbilical artery was observed in 25 cases. Twenty cases had type 1 single umbilical artery which coursed normally (Fig 2).

We observed abnormal origin and course of umbilical artery in 5 cases. All five cases were of vitelline origin of umbilical artery (Type II SUA) showing similar course and termination of umbilical artery.

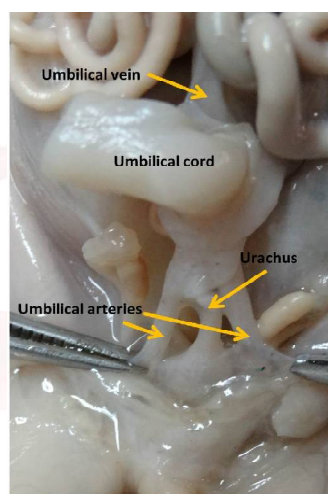


Fig. 1: Normal umbilical arteries.

Internal examination of abdomen in all the cases showed abnormal single umbilical artery (SUA) coursing upwards and posteriorly from the umbilical ring into the abdomen between the coils of intestine with an abnormal peritoneal fold (Fig 3). This vessel originated from the aorta instead of from the internal iliac arteries, and the aorta was narrow and hypoplastic distal to this origin. (Fig 4,5&6). All five cases were associated with other systemic abnormalities and are described in the Table 1 (Fig 7&8).

Fig. 2: Type 1 Single umbilical artery.

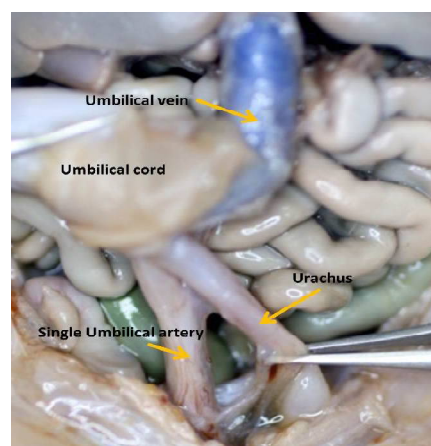


Table 1: Cases associated with other systemic abnormalities.

S.No	Gestational Age	Sex	Prenatal diagnosis	Fetal autopsy findings
Case 1	28 weeks	Male	Intra uterine fetal growth restriction (IUGR), Ventricular septal defect (VSD), Bilateral multi cystic dysplastic kidneys, Short curved lower limbs and Oligo hydramnios, Single umbilical artery	Short left lower limb, Scoliosis, Right sided aortic arch, VSD, Tracheo-Esophageal fistula with esophageal atresia, Anal canal agenesis, Common cloacal deformity (Uro-rectal septum malformation), Malrotated and unascended right kidney, malrotated left kidney, bilateral hydronephrosis, hypoplastic bladder opening into the rectum (cloacal defect), absent left testes, Single umbilical artery and hypoplastic aorta below the umbilical artery
Case 2	26 weeks	Indeterminate	Complex cardiac disease, polycystic kidneys, severe oligo hydramnios, Single umbilical artery	Lowset ears, high arched palate, absent labia and scrotum with a phallus, Bilateral absent radius and thumb, bilateral rocker bottom feet, post axial polydactyly of right foot, kypho-scoliosis, Right aortic arch with Tetralogy of Fallot, Anomalous pulmonary venous drainage, Tracheo-esophageal fistula with esophageal atresia, duodenal, ileal and anal atresia, Crossed renal ectopia (right kidney crossed to left and fused with lower pole of left kidney), bilateral hydronephrosis and hypoplastic bladder, Single umbilical artery and hypoplastic aorta below the umbilical artery
Case 3	32 weeks	Indeterminate	Severe oligo hydramnios, Single umbilical artery	Bilateral lowset ears, Single central lower limb without foot (Sirenomelia), right aortic arch with truncus arteriosus and VSD, Anomalous pulmonary venous drainage, Large intestine terminated as two blind pouches, anal agenesis, Bilateral renal agenesis, Single umbilical artery and hypoplastic aorta below the umbilical artery
Case 4	24 weeks	Indeterminate	Anhydramnios, bilateral renal agenesis	Bilateral lowset ears, Single caudal central limb without foot (Sirenomelia), Aberrant right subclavian artery, VSD, Large intestine ended as a blind sac with anal agenesis, Bilateral renal agenesis, Single umbilical artery and hypoplastic aorta below the umbilical artery
Case 5	20 weeks	Male	Gestational diabetes, Spine deformity, Short long bones, Single umbilical artery	Bilateral lowset ears, rocker bottom feet, hypoplastic abdominal and lower limb musculature, Hypoplastic single discoid kidney with single ureter, hypoplastic bladder, Hypoplastic lumbar vertebra and sacral agenesis, Single umbilical artery and hypoplastic aorta below the umbilical artery

Fig. 3: Abnormal peritoneal fold in Type 2 Single umbilical artery.

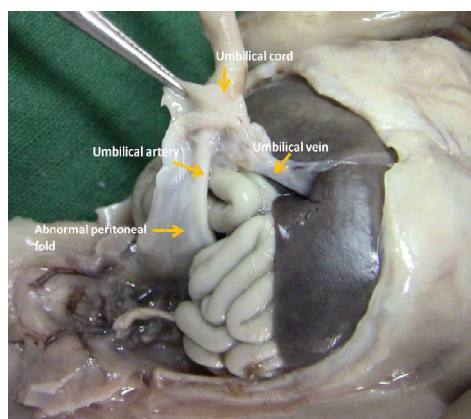


Fig 4: Hypoplastic abdominal aorta distal to the Type 2 Single umbilical artery.

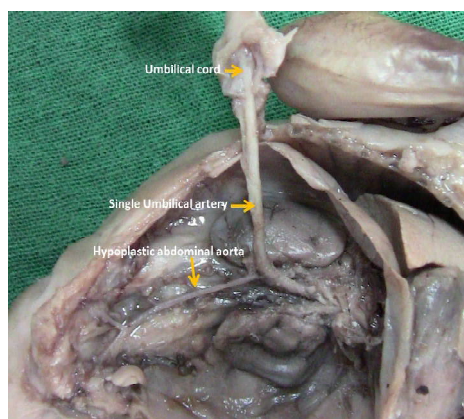


Fig 5: Right aortic arch with Type 2 Single umbilical artery.

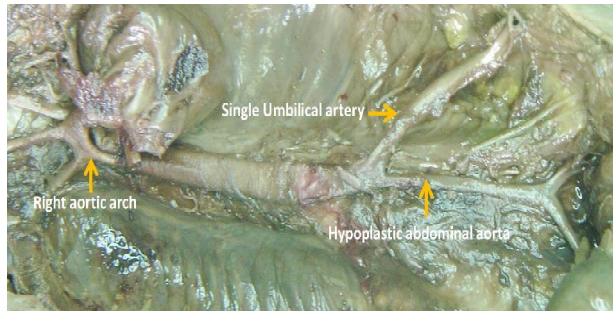


Fig 6: Hypoplastic abdominal aorta distal to the Type 2 Single umbilical artery.

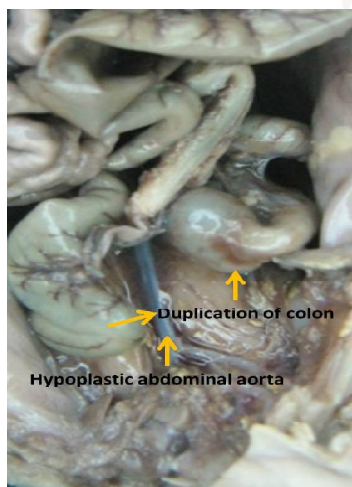
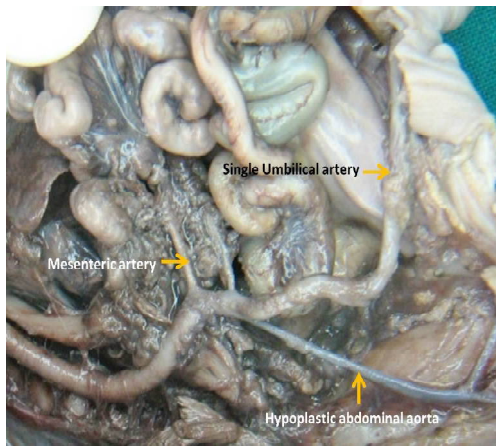
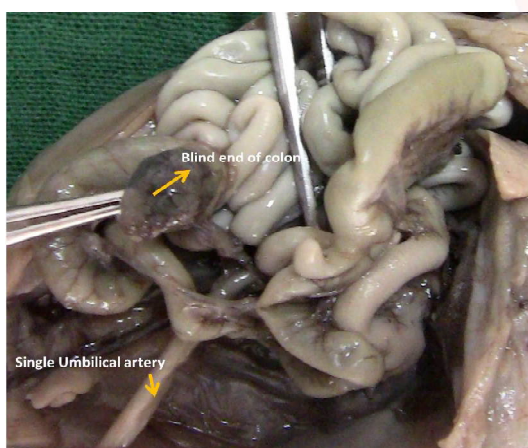


Fig 7: Duplication of colon.

Fig 8: Blind end of colon with rectal agenesis.



DISCUSSION

During the early period of umbilical cord organogenesis, blood vessels begin to appear within the connecting stalk from the angiogenic mesenchyme, accompanying the allantoic and vitelline ducts. Initially, the vitelline vessels are prominent and more numerous than those of allantoic origin. Later, due to regression of secondary yolk sac and the vitelline duct, the vitelline vessels also disappear. A few small thin walled vitelline vessels persist in about 5-8% of human umbilical cords at term gestation [7, 8]. In humans, allantoic vessels predominate in establishing the vascular system within the trophoblastic mass (Placenta) which ultimately constitutes the blood vessels in the umbilical cord. Normally, there are two arteries and one vein in the umbilical cord. The umbilical arteries course downwards on either side of urinary bladder towards the internal iliac arteries. When there is one umbilical artery instead of two in the umbilical cord, such a condition is called 'single umbilical artery' (SUA). SUA was classified into four types by Roger E Stevenson et al; type 1 as SUA allantoic origin and left single umbilical vein; type 2 as SUA vitelline origin and left single umbilical vein; type 3 as SUA of allantoic or vitelline origin, left umbilical vein and persistent right umbilical vein; type 4 as SUA of allantoic or vitelline origin and right single umbilical vein [7].

The first organ to form in human beings from the primary yolk sac is allantois. In conditions in which the allantois does not develop or is lost early in embryogenesis, the allantoic arteries do not develop within the connecting stalk. In this circumstance, the survival of the embryo depends on the persistence and further development of the vitelline arteries to provide arterial supply to the developing placenta (Choriovitelline placentation) [9, 10, and 11]. Monie W et al. experimentally induced persistence of vitelline arteries and failure of development of allantoic arteries in rats by giving retinoic acid (Vitamin A) [12]. This is an inherent "fail-safe mechanism" that provides vascular supply for the embryo via vitelline system when the allantoic vascular system fails. In this condition, there will be one umbilical artery in the umbilical cord and it will course

upwards and posteriorly from the umbilical ring into the abdomen. This artery appears to be originating either from the abdominal aorta or superior mesenteric artery instead of internal iliac artery. This was classified as Type II SUA.

Hoyme et al had implicated interruption of omphalo mesenteric artery (vitelline artery) as a possible causative factor/mechanism for gastroschisis, intestinal atresia, gallbladder agenesis, and other anomalies[13]. In the present series, one case had multiple intestinal atresias and renal anomalies, possibly explained by this type 2 SUA.

The relationship between the persistent vitelline artery and caudal body structure disruption was described by Stevenson et al [10]. They proposed "vascular steal" via persistent vitelline artery from abdominal aorta as the cause of sirenomelia. In the present study there were two cases of sirenomelia with bilateral renal agenesis and caudal single limb. Both cases showed the similar vascular pattern and the abdominal aorta appeared as if continuing with the single persistent vitelline artery causing the vascular steal.

Type II Single umbilical artery is rarely associated with a normal development of the fetus and presenting as an isolated finding [5, 7]. Clinical presentation of the neonate after birth could be normal or could be associated with acute intestinal obstruction, recurrent intestinal pain, or intra-abdominal haemorrhage. Even if the fetus shows no anomalies, this artery forms an abnormal peritoneal band from the posterior aspect of umbilical ring to the abdominal aorta or superior mesenteric artery. This abnormal peritoneal band traverses between the coils of intestine and may lead to intestinal volvulus and obstruction in neonates or in adults [14, 15]. Based on surgical findings, Postoloff described three types of persistent vitelline artery: a persistent band between the anterior abdominal wall & the ileal mesentery, a band in association with Meckel's diverticulum, or a free hanging cord [16]. Hence, whenever there is volvulus in a neonate, the paediatric surgeon must be mindful of these abnormal peritoneal bands.

Prenatal diagnosis of such a persistent vitelline artery can be done by its abnormal course; this

artery traverse abnormally upward and backward between the coils of intestine instead of downwards and backwards on either side of bladder. Doppler ultrasonography of umbilical arteries is the diagnostic tool for persistent vitelline artery. Because the type II SUA is associated with serious caudal body deformities, the prognosis is poor.

We proffer that in all these cases there will be vitelline origin of umbilical artery as a fail-safe mechanism where there is failure of umbilical artery origin from allantoic arteries. Hence, the entire blood from the abdominal aorta reaches placenta through the single umbilical artery and leads to vascular steal. As a minimal amount of blood circulates through the distal aorta, it becomes hypoplastic. This leads to the pathogenesis of caudal regression syndrome. Almost always such vitelline origin of umbilical artery would be associated with other systemic anomalies and a careful search for all such, therefore becomes mandatory.

Conflicts of Interests: None

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