THE SIGNIFICANCE OF ONE UMBILICAL ARTERY

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Absence of one umbilical artery is a rare finding, usually associated with other severe congenital malformations. The earliest recorded case is attributed to Casp. Bauhin in 1621, although Noortwyck (1743) quotes Vesalius' description '*unam tantum arteriam in fune Fallopius vidit*'. Otto (1830) collected 41 case reports from the literature, and Hyrtl (1870) described 14 cases of his own and found 16 more in the literature. Browne (1925) referred to one case with a single artery and numerous capillaries replacing the vein. Benirschke and Brown (1955) described 55 cases, and Richart and Benirschke (1958) described one further case with gonadal dysgenesis. Javert and Barton (1952) examined the umbilical cord of 297 aborted fetuses of four months' gestation and found one with a single artery. Of the 182 cases of symphodia collected by Ballantyne (1898), Kampmeier (1927) and Hendry and Kohler (1956), 95 had a single artery in the umbilical cord, and in the remainder the cord was not examined. Schatz (1900) found 11 cases among 46 acardiac monovular twins and three acardiac monovular triplets. There is therefore clearly a high incidence in acardiac fetuses and symphodia. In fetuses without acardia or symphodia the incidence appears to be relatively low, and only Javert and Barton (1952) provide data from which an assessment can be made.

This paper describes 11 cases encountered among routine autopsies performed at the Birmingham Children's Hospital from March, 1958, to February, 1959, in autopsies on 159 stillbirths and 252 live born babies of 8 weeks of age or less. Severe malformations were found in nine (81%) and in 23% of the controls.

Case Reports

Case 1. A female baby was stillborn at 36 weeks' gestation to a para 1 mother aged 20. There had been a threatened abortion at 3 months.

Autopsy showed rotation of the trunk through 90° so that the buttocks were facing the left side, and a large hernia into the body stalk. The sac was formed by

amnion and contained liver and loops of bowel. The lungs were hypoplastic and abnormally lobed. The heart showed a defect in the membranous part of the ventricular septum. The right renal artery arose from the proximal part of the superior mesenteric artery. The umbilical artery entered the right internal iliac artery and was wider than normal. The small intestine was short (97 cm.) and was strangulated distally in the hernial sac. The brain showed bilateral cirroid aneurysms of the Sylvian fissures with focal microgyria and encephalomalacia of the underlying cerebral tissue on the right side. The placenta weighed 237 g. and was normal. The umbilical cord lay in the wall of the hernial sac and was short (14.5 cm.). It contained one artery and one vein throughout its length.

Case 2. A male baby was born at full term after a normal pregnancy. On the third day he had a surgical repair for oesophageal atresia with tracheal fistula and died six days later with respiratory obstruction and infection.

Autopsy showed repair of the tracheo-oesophageal fistula with oesophageal anastomosis. There was a diaphragmatic urethral valve at 0.9 cm. from the bladder neck. The bladder was dilated and hypertrophied and there was bilateral hydronephrosis and hydroureter. The right umbilical artery was absent and the right common iliac artery was narrow.

Case 3. A male infant was born at full term after a difficult delivery and died six hours later. The antenatal history was not known.

At autopsy there was gross radial deviation of the left thumb and radius. The small intestine was incompletely rotated and unfixed, and there was a Meckel's diverticulum. The right kidney and ureter were absent. The left kidney had two cortical cysts and the renal pelvis and ureter were dilated. The bladder was grossly dilated and hypertrophied and filled with heavily blood-stained fluid. There were lacerations of the bladder wall 2.5 and 3 cm. in length. The internal urethral orifice was stenotic and partially valved by mucous membrane, and beyond it the urethra was atretic. The testes were intra-abdominal and normal. The right renal artery was absent, and there were two left renal arteries. The left common iliac vessels were small, and the left umbilical artery was absent.

Case 4. A female baby was born at 35 weeks' gestation to a gravida 3 mother after a normal pregnancy. The baby died after one hour.

Autopsy showed low set ears and bilateral pes cavus. The external genitalia were abnormal, the main feature resembling a scrotum with a median dimple on its anterior surface. On either side of the scrotum were two folds resembling labia. The anus was represented by a shallow depression. There was oesophageal atresia, tracheo-oesophageal fistula and atresia of the duodenum. The small intestine showed a Meckel's diverticulum and lack of mesenteric fixation. The colon ended blindly on the posterior wall of the bladder. The liver showed abnormal lobulation of its inferior surface. The gall bladder was absent. The larynx and left bronchus were atretic and the left lung was absent. The heart had a high ventricular septal defect and a common truncus. There were two renal arteries on each side. The right umbilical artery was absent, and the right common iliac artery was smaller than the left. There was a persistent left superior vena cava. Both kidneys were small and cystic. There was duplication of the right ureter and renal pelvis. The two ureters were thin cords uniting caudally and ending blindly in the bladder wall. The left ureter and ureteric orifice were moderately dilated. The bladder was grossly dilated and hypertrophied. The internal urethral orifice was stenotic, and from it a hypoplastic urethra passed to the scrotal dimple. There were two normal ovaries but no uterus nor vagina.

Case 5. A male baby was stillborn at 31 weeks to a primigravida mother aged 20. The antenatal history was not known.

Autopsy showed coarse facial features with a depressed nasal bridge and low-set ears. There was a left diaphragmatic hernia, and the left pleural cavity was also continuous with the pericardial cavity. The heart showed atresia of the tricuspid valve and pulmonary infundibulum. The right ventricle was a minute blind chamber. There was a large atrial septal defect. The pulmonary arteries and ductus arteriosus were hypoplastic. The left umbilical artery was absent and the left common iliac artery was narrow. The right kidney and ureter were absent. The left kidney was hypoplastic and cystic. The left ureter was a thin cord with a normal ureteric orifice. The bladder was much dilated and hypertrophied. There was stenosis of the bladder neck, atresia of the prostatic urethra and agenesis of the prostate. The testes were intra-abdominal and normal. The lower end of the colon was atretic, ending blindly on the bladder wall. The termination of the spinal cord was abnormal. The cord extended to the lower end of the sacral canal and passed into a small skin-covered cartilaginous peg on its external aspect.

The placenta weighed 240 g. and was normal. The umbilical cord was normal apart from absence of one artery.

Case 6. A female baby was born at 36 weeks to a para 1 mother aged 17. The delivery was normal but the baby had white asphyxia and died after eight hours.

At autopsy the ears were low-set and the external genitalia were abnormal, represented by a median mound of soft tissue covered by skin. There was a sinus at the junction of the anus and the perineal skin. There was bilateral renal hypoplasia and agenesis of the bladder. The ureters were dilated to cystic proportions distally, and at the lower end of the uterus they communicated with the vagina which was continuous with the perianal sinus. The uterus was bicornuate, the ovaries were normal. The right umbilical artery was absent, and the right common iliac artery was very small. The lower part of the left umbilical artery gave rise to a vessel which crossed over to the right and divided into the right external and internal iliac arteries.

The placenta weighed 606 g. and was normal. The cord was excessively long (83 cm.).

Case 7. The first of monovular male twins was stillborn at 32 weeks to a para 1 mother, aged 25, after prolapse of the cord. The antenatal history was normal. The second twin died after 56 hours and had no malformations.

At autopsy the nose was slightly beaked and the ears were normal in position but had a soft flat helix. The kidneys were enlarged and cystic, fused together and low in position. The renal arteries consisted of several minute vessels arising close to the aortic bifurcation. The two ureters were very narrow and passed to a hypoplastic bladder. The urethra and testicles were normal. There was hypoplasia of the stomach, liver and lungs. The right umbilical artery was absent and the right common iliac was smaller than the left.

There was a diamniotic-monochorionic placenta weighing 362 g. The umbilical cords were attached near the central double amniotic membrane, one being normal and one having a single artery and vein. There were no demonstrable vascular anastomoses.

Case 8. The second of twins was stillborn at 38 weeks to a para 2 mother aged 22, after a normal pregnancy. The first was a female infant with no malformations who thrived normally.

Autopsy examination showed a sirenomid foetus with a single curved lower limb ending in a foot with three broad digits. The trunk consisted of a featureless mass surmounted by a head process. There was no definable perineum, vertebral column, thorax or abdomen, and no upper limbs. The umbilical cord was attached at the upper end of the trunk. The head process was covered by skin and had two auricular appendages on the right with an adjacent red polypoid process. A smear from the process showed epithelial cells with the chromatin pattern associated with the female sex. Internal examination was not performed.

There was a bulky monochorionic placenta weighing 760 g. One umbilical cord was normal and attached marginally. The other was thinner, contained one artery and one vein, and had a velamentous insertion.

Case 9. A female infant was born at full term to a primigravida mother aged 23, after a normal pregnancy.

At three weeks she developed heart failure, which did not respond to treatment and she died at six weeks.

At autopsy the heart was grossly enlarged. There was hypoplasia of the ascending aorta and the aortic arch with a narrow elongated aortic isthmus and a wide ductus arteriosus. There was a small defect in the upper posterior part of the muscular ventricular septum. The aortic valve was bicuspid. The right umbilical artery was absent and the right common iliac artery was narrow.

Case 10. A male infant was born at 31 weeks gestation after surgical induction of labour. The mother was a primigravida aged 25 with fulminating pre-eclamptic toxæmia. The infant died 10 minutes after birth.

At autopsy there were no congenital malformations. Much amniotic cellular debris was found in the lungs. The placenta was normal. The umbilical cord contained one artery and one vein and was otherwise normal.

Case 11. A male baby was stillborn at 41 weeks to a para 3 mother aged 22. The antenatal history was normal.

Autopsy examination showed a macerated baby with only one umbilical artery but no other congenital malformation. The placenta had a marginal infarct. The umbilical cord was normal apart from absence of one artery.

Discussion

There were 11 cases of single umbilical artery among 411 autopsies, an incidence of 2.7%. There is no comparable perinatal investigation reported in the literature. Javert and Barton (1952) found a much lower incidence in fetuses aborted at four months.

Nine of the 11 babies had severe associated malformations, and in eight the malformations were multiple. This incidence of 81% is comparable with the 76% found by Benirschke and Brown (1955) and much higher than the incidence of 33% recorded by Hyrtl (1870), and the 56% of Otto (1830). There were malformations of the lower urinary tract in seven babies. One was a sirenoïd foetus with no perineal orifices, one had valvular obstruction of the posterior urethra, two had extreme hypoplasia or agenesis of the bladder, and three had atresia or severe stenosis of the bladder neck. In two cases with bladder neck obstruction there was also rectal atresia. There were four cases with renal hypoplasia or agenesis, four cases with malformations of the heart, and two with oesophageal atresia. Ballantyne (1904) stated that the commonest associated malformation was defective development of the bladder and sirenomelia. Benirschke and Brown (1955) found mainly malformations of the heart and central nervous system, while anencephaly was the commonest malformation found by Hyrtl (1870).

There were two monovular twins. In both cases the other twin had two umbilical arteries and no malformations. There was also a high incidence of twinning in Benirschke and Brown's (1955) cases. They found one monovular and four binovular twins, a total incidence of 9%. An abnormal cord was found in each pair. Hyrtl (1870) described female monovular twins (incidence 7%) and male triplets, one foetus in each set having the abnormality. Schatz (1900) found 11 cases in monovular twins and one in a single foetus. In each case only one of the twins had one artery.

There were seven placentas available for examination. One was infarcted and the remainder were normal. One umbilical cord was abnormally long, one was short and a third had a velamentous insertion. Benirschke and Brown (1955) found a high incidence of placental anomalies such as circumvallation, circum-margination, velamentous insertion of the cord and extensive infarction. Ballantyne (1898) stated that the placentas of symphodia tended to show abnormalities of size, shape, consistency and mode of insertion of the cord.

There were equal numbers of live and stillbirths. There was also a high incidence of stillbirths (76%) in Benirschke and Brown's (1955) cases. Hyrtl (1870) found only 7% were stillbirths, but this was probably due to autopsies being infrequently performed on stillbirths.

There were equal numbers of male and female foetuses, and this agrees with the findings of most authors. Kampmeier (1927), however, found a male preponderance in sirenoïds, and Hyrtl found only males in his series of 12 single foetuses.

No correlation was found between the abnormality and maternal factors. Of the mothers 60% were multiparous, and their ages ranged from 17 to 25 years. The pregnancies were normal in seven cases, in one there was a threatened abortion, and in one toxæmia of pregnancy. Benirschke and Brown (1955) had found a high incidence of toxæmia and hydramnios.

The condition can be simply diagnosed at birth by careful examination of the cut surface of the umbilical cord. This is of little practical value in the presence of obvious gross malformations. There were no gross external malformations in six of the cases, however: two of these, Cases 10 and 11, showed no associated malformations at autopsy; two, Cases 5 and 7, had malformations incompatible with life; two, Cases 2 and 9, had malformations which may be amenable to surgical treatment. Therefore a newborn infant without gross malformations visible externally, but with only one umbilical

artery, has a 2:1 risk of severe internal malformations, which may warrant early surgical correction.

Summary

Eleven cases of single umbilical artery are described. They were encountered among 411 autopsies on stillborn and live-born babies under 8 weeks of age. Nine had severe associated congenital malformations involving particularly the urinary tract. Absence of an umbilical artery can be diagnosed by simple examination of the cord at birth. In the absence of severe external malformations such an infant has a 2:1 risk of internal malformations which may require early surgical treatment.

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