ABSENT UMBILICAL ARTERY*

A REVIEW OF 113 CASES

BY

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The umbilical cord normally contains two arteries and one vein embedded within Wharton's jelly which is, in turn, enclosed by an epithelial membrane.

Hyrtil (1870) described the absence of one of the umbilical arteries in 12 instances (four infants and eight placentae) of which only two infants were malformed, one suffering from spina bifida and both having cleft palates. He also noted that male infants were affected more often than female infants and that females lacking one umbilical artery were more frequently malformed and often anencephalic. Neither the observation of male dominance nor the higher incidence of abnormalities in females have subsequently been confirmed, although there is a high incidence of anencephalic deformity in the female infants. He collected a total of 58 cases from the literature to add to the 12 of his own.

Browne (1925) reported a premature infant in whom the umbilical cord contained a single artery and the umbilical vein was replaced by a capillary network. Javert and Barton (1952) reported the results of the examination of 1,000 abortuses in which histological sections from 104 umbilical cords were examined. Only one was found to contain a single artery.

Benirschke and Brown (1955) described a retrospective study of 55 infants in whom one artery was missing from the entire length of the umbilical cord. Of these 55 only 13 (24%) were considered to be normal children and of the remaining 42 there were 27 (64%) who suffered from a wide range of congenital abnormalities, many of whom had multiple defects. Placental abnormalities were common (39%). Little (1958) examined the umbilical cords of 1,200 consecutive deliveries, finding

that 12 (1%) contained only one artery throughout, and in four (0.3%) the arteries fused locally near the placenta; congenital abnormalities were present in 25%. Recently Benirschke and Bourne (1960) reported the incidence of absent umbilical artery to be 15 (1%) in 1,500 consecutive deliveries. Eight (53%) were considered to be developmentally normal, although one was stillborn, whilst seven (47%) were found to possess major or minor congenital abnormalities. Only 12 (80%) remain alive, one of whom has survived operative treatment for oesophageal atresia and tracheo-oesophageal fistula.

It seems fairly certain that the overall incidence of this abnormality is in the region of 1% of all deliveries. Furthermore, they report that absent umbilical artery occurred seven times (7%) in a series of 100 consecutive unselected twin pregnancies. This is much higher than has hitherto been supposed.

Materials and Method

A total of 113 infants in whom one umbilical artery was absent (Fig. 1) has been studied at the Boston Lying-in Hospital and the Children's Medical Centre, Boston, Mass. The series is composed of 55 cases first observed in a mainly retrospective study, 15 cases reported to be present in the examination of 1,500 consecutive placentae, together with a further 43 instances observed during the past three years in the Pathology Department of the Boston Lying-in Hospital. With the exception of some of the 55 instances which were retrospective, the examination consisted of a gross evaluation of the placenta together with a microscopic study of at least three blocks of tissue. These included sections from the middle portion of the umbilical cord, a segment of rolled membrane and a section of full thickness placental tissue. In the event of absence of an umbilical artery from the histological section of the cord, the secundines, which had been

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FIG. 1.—Absence of one umbilical artery. Section of the umbilical cord showing the dilated vein and a single artery. Note the muscular wall and small lumen of the artery. There are no residual ducts or vessels present. (H. and E. $\times 10$.)

stored awaiting 'histological clearance', were re-examined to confirm the absence of an artery from the entire length of the cord. In instances of foetal death confirmation of the absence of one of the intra-abdominal vessels was sought at necropsy. Owing to the high incidence of associated congenital abnormalities, it was the usual practice to inform the paediatrician immediately upon the finding of a single umbilical artery. This assisted in the early diagnosis of oesophageal atresia in at least one infant.

A total of approximately 6,000 deliveries occur each year in the Boston Lying-in Hospital. With the exception of the consecutive series mentioned above the placentae are examined in the laboratory only when indicated by such conditions as maternal illness or disease; foetal distress, abnormality or prematurity; plural pregnancy; hydramnios and other complications of pregnancy as well as patients of clinical or pathological interest.

It is realized that 98 of the total 113 cases are derived from selected material from which it might be erroneous to draw definite and final conclusions. The reasons for the selection of the material from which these cases are derived, however, are so diverse that there is ample justification for the analysis of certain factors.

Maternal Factors

Age. An analysis of the ages of the 113 mothers who were delivered of infants lacking one umbilical

artery is shown in Table 1. It is noticeable that 26 (62%) of the 42 primigravidae were under the age of 25, whereas only 13 (22%) of the 59 multigravidae were under the same age. Compared with the average ages of a control series of patients, this series shows an increased incidence of the anomaly among the younger primigravidae and the older multigravidae. The age groups of the total series, however, conform fairly closely to those of the controls, because the increased incidence at the younger end of the primigravidae scale balances the similar, but less well marked, rise at the older end of the multigravidae scale. These findings are in broad agreement with Baird and Walker (1959), who state 'The incidence of foetal deformity . . . is highest in the 15-19 age group and decreases with age till thirty-five; a slight rise occurs in the age group 35 and over'.

The average age of the 19 mothers of infants with central nervous system anomalies, 28.3 years compared with 28.5 years, failed to confirm that these, in particular, occur in a high age group (Hegnauer, 1951).

Parity. Forty-two of the mothers were primigravidae and 59 were multigravidae whose parity varied from one to 15. These are shown in detail in Table 2 and agree essentially with the control series except for the high number pregnant for the

TABLE 1
ANALYSIS OF AGES OF MOTHERS DELIVERED OF INFANTS LACKING ONE UMBILICAL ARTERY

Age (years)	Primigravida	Multigravida	Total
20	5	1	6
21-25	21	12	33
26-30	8	15	23
31-35	6	17	23
36-40	2	12	14
41	—	2	2
Total . . .	42	59	101
Average age . .	25.3	30.7	28.5

TABLE 2
GRAVIDITY

Gravidity	No.
1	42
2	21
3	15
4	11
5	3
6	5
7	1
8	2
15	1
Total . .	101

sixth time, an observation that is not considered to have any significance.

The 59 women who had been pregnant previously had produced a total of 111 live born infants, 12 stillbirths (a rate of 103 per 1,000) and 32 abortions. Four of the 111 live born infants died in the newborn period so that only 107 (69%) infants survived the newborn period out of a total of 155 previous pregnancies. This incidence (31%) of foetal loss in the past history of these patients is surprisingly high and much greater than the 11% of the control series.

Maternal Diseases. A high incidence of pre-eclampsia (30%) was described by Benirschke and Brown (1955). This was not confirmed by Little (1958). Both authors agree that the large number of pre-eclamptic patients in Benirschke's series was the result of material selection and that it did not reflect the true incidence of this complication of pregnancy. This series of 113 cases includes the 55 described by Benirschke, together with a further 58 additional cases in which pre-eclampsia was diagnosed in only five (9%) instances. This indicates that pre-eclampsia is not associated with the absence of an umbilical artery either as an aetiological factor or as a result of the abnormality. Altogether a total of 19 patients (16%) were found to be suffering from pre-eclampsia in the total of 113.

The association, however, of pre-eclampsia and a single artery in a total of 19 patients resulted in seven stillbirths, three neonatal deaths and nine living infants, which illustrates that this combination, even if it is fortuitous, is particularly lethal to the foetus.

A careful study of the general health of the mothers has not disclosed any factor that might influence the pregnancy so as to predispose the foetus to this particular anomaly. The high incidence of foetal wastage in the previous pregnancies of multigravida has been described; a high proportion (8%) of patients suffered from antepartum haemorrhage.

Two otherwise normal infants suffered from erythroblastosis foetalis due to rhesus incompatibility, one being stillborn and the other dying on the first day. Four of the mothers suffered from epilepsy (*grand mal*).

Hydramnios. The incidence of foetal abnormality is particularly high in association with hydramnios. Macafee (1950) stated that the incidence of foetal abnormality was 46% in a series of hydramniotic pregnancies. It is not surprising, therefore, that there should be a fairly high incidence of hydramnios in association with absence of an

umbilical artery. Nineteen (19%) patients were considered to suffer from hydramnios in the 101 in whom adequate information was available and this figure is probably a conservative estimation, because in the original study of 55 cases only four were present, 15 occurring in the last 58 patients. This high incidence of hydramnios was not confirmed by Little (1958). It is interesting that all 19 (100%) of these infants suffered from congenital abnormalities and that 12 were stillborn; five died in the neonatal period and the two who survive do so because of successful surgical treatment of oesophageal atresia. The association between hydramnios and oesophageal atresia is now accepted (Scott and Wilson, 1957), and in one of the surviving patients the early diagnosis of oesophageal atresia was hastened by the finding of umbilical artery aplasia within a few hours of the birth of the infant.

Heredity. There is little or no evidence to show that this condition is of an hereditary nature. Only one mother was known to suffer from any major congenital abnormality (pulmonary stenosis) and one had congenital syphilis. No abnormalities were known to be present in the male parents but they were, necessarily, not subjected to the same scrutiny as the mother. Furthermore, the wide range of associated congenital abnormalities seems to mitigate against a single hereditary factor. Four mothers (4%) suffered from *grand mal*, which might be significant, but the numbers are too small to justify the assumption that it was in any way a causative factor.

There is, however, some evidence that the anomaly forms part of a diathesis in which abnormalities are frequently multiple and may be recurrent. These assumptions are based upon the past history of 59 multigravid patients in which 48 (31%) out of a total 155 pregnancies failed to produce a living child. It is unfortunate that more detail was not available concerning the loss of these infants, but such information as is available does not reveal any definite recurrence of single umbilical artery abnormality, although several are known to have produced abnormal infants previously. Several mothers have subsequently produced normal children, all of whom possessed two umbilical arteries. That these patients experience a greater than average difficulty in producing living normal children is definite and it is possible that the finding of arterial abnormalities in the umbilical cord may lead to the recognition of such 'high-risk' patients even in their first pregnancy. The parents of one infant were known to be related cousins; there was no other recorded evidence of consanguinity.

TABLE 3
DETAILS OF 15 (1%) AFFECTED INFANTS FROM 1500 CONSECUTIVE DELIVERIES

Number	Sex	Twins	Gestational Age (weeks)	Complication of Pregnancy	Foetal Anomalies	Follow-up
1	M	—	35	Chorioamnionitis	None found	Not traced
2	F	—	39	Placental infarct	None found	Normal
3	F	F/F monochorial	38	None	Forked xiphoid	Normal development
4	M	—	42	Irregular heart 0-130; discharge meconium	None found	Normal
5	M	M/M monochorial	38	650 g. lighter than other twin	Hydrocele, abnormal cardiac rhythm	Developing normally; bigger than twin
6	M	—	38	None	Slight club foot—right	Normal development
7	F	—	40	None	None found	Normal
8	F	F/M dichorial	38	None	Hyperteiorism, short fingers	No problem
9	F	—	37	Hydramnios	Oesophageal atresia and fistula, sacral anomaly	Operated, survived
10	F	—	37	Hydramnios	Anencephaly	Stillborn
11	F	—	40	None	None found	Normal
12	F	—	40	None	None found	Normal
13	M	—	37	None	Renal agenesis, atresia an ⁱ , accessory pancreas, equinovarus	Died
14	F	—	37	Maternal floor infarction; long cord	None found	Stillborn
15	F	—	40	Long cord	None found	Not traced

(After Benirschke and Bourne, 1960.)

The Infants

Incidence. In the examination of a series of 1,500 consecutive placentae a total of 15 (1%) of the umbilical cords were found to contain only one artery throughout their length. Seven (46%) of these 15 infants suffered from congenital abnormalities, of which three were of major significance (Table 3). One was a stillborn anencephalic and one died from renal agenesis. There were eight (53%) normally developed infants of which one was a macerated stillbirth in whom no congenital abnormality could be found.

The overall mortality in this short series is three out of 15 (20%) with one further child surviving operative repair of an oesophageal fistula and atresia.

The relatively high incidence of twin infants in this consecutive series (20%) is confirmed to some extent by the fact that in a series of 100 consecutive twin deliveries an absent umbilical artery was present seven (7%) times. The overall incidence of one in 100 is confirmed by Little (1958) and by Bourne (1961) who recognized the condition five times in 475 (one in 95) consecutive umbilical cords. A similar incidence is also reported by Emig (personal communication).

The Fate of the Infants. It is realized that the total series of 113 cases is partially selected, but nevertheless an analysis of these patients is very informative. Table 4 shows the fate of these infants.

Live Infants. Only 48 (42%) are alive, of whom 14 (29%) are known to suffer from congenital

TABLE 4
FATE OF THE INFANTS

Infants	Nos.	%
Total	113	100
Alive	48	42
Normal infants	34	
Known congenital abnormality	14	
Dead	65	58
Congenital anomaly incompatible with life	39	
Congenital anomaly compatible with life in infants dead of other causes	5	
Possible anomaly in unexamined infants	15	
No anomaly	6	

abnormalities; 27 infants were either seen or were considered by their medical attendant to be both physically and mentally normal. It was impossible to trace seven infants, all of whom, according to their records, were normal when last examined. In the absence of any information to the contrary these infants are considered to be living and normal.

It has been possible to trace all 14 infants who are alive and who suffer from congenital abnormalities of varying severity. Several of these infants (Table 5) are not expected to reach adult life and two survive only because of successful surgical repair of oesophageal atresia.

Dead Infants. Sixty-five (58%) of the infants died or were born dead, of whom 59 (91%) were suffering from definite or probable congenital abnormalities (Table 4), and only six (9%), who died of known causes, were considered to be normally developed for their maturity.

TABLE 5
KNOWN ABNORMALITY COMPATIBLE WITH LIFE IN 14 SURVIVORS

Maturity (weeks)	Weight (lb.)	Sex	Abnormality	Follow-up (years)	Remarks
40	7.1	F	Congenital heart disease	17	Twin
41	5.1	M	Oesophageal atresia	6	Successful operation
36	7.1	M	Brain cysts	4	Very poor prognosis
37	5.11	M	Hydrocele, umbilical hernia, inguinal hernia	4	
32	3.6	M	Mental defect; optic fundi abnormal	3	Poor prognosis
42	4.15	F	Multiple haemangiomas	2	
42	6.2	M	Arthrogryphosis multiplex cystica, cryptorchid	2	
32	2.5	M	Cryptorchid	2	Twin
39	8.0	F	Marfan's syndrome: arachnodactyle	1	
38	5.13	M	Sternal abnormality	1	Twin
38	3.2	M	Cardiomegaly (?) cause	1	Twin
38	5.8	M	Talipes equinovarus	0.6	
38	5.1	F	Hyperteiorism	0.3	Twin
36	4.6	F	Oesophageal atresia	0.5	Successful operation

Congenital abnormality of such a major degree as to be incompatible with life was present in 39 (60%) of these children, whilst five (8%) who died from other causes were suffering from anomalies that probably contributed to their death, and 15 (22%) were considered to be suffering from a congenital abnormality, although severe maceration or lack of a post mortem examination prevented exact diagnosis.

Absence of one umbilical artery seems, in itself, to be a simple and innocuous congenital abnormality, but it is associated with a perinatal mortality involving 58% of the affected infants, together with a further 12% who are alive but abnormal in some way. Such a relationship is considered to be significant.

Congenital Abnormalities. The types of congenital abnormality from which the affected infants suffered have been classified into systems. These are listed in Table 6 and they are detailed in Table 7.

The majority of the babies suffered from multiple defects which, if serious, are listed separately so that the total is greater than the number of infants concerned. No system or peculiar abnormality is particularly prominent, although aplastic lesions of

TABLE 6
CONGENITAL ABNORMALITIES

Abnormalities	Nos.	%
Known congenital abnormalities (mostly multiple)	58	51
Gastro-intestinal	31	
Skeletal	28	
Genito-urinary	27	
Cardiovascular	21	
Neurological	19	
Others	10	
Possible abnormality (unexamined, dead)	15	14
No known abnormality	40	35
Total number of cases	113	100

TABLE 7
DETAILS OF CONGENITAL ABNORMALITIES

Abnormality	Nos.
Gastro-intestinal	31
Oesophageal atresia	8
Imperforate anus	6
Exomphalos	5
Malrotation of gut	4
Herniae	4
Cleft palate	3
Cysts of larynx	1
Skeletal	28
Talipes equinovarus	9
Poly and syndactyle	6
Spine (excluding spina bifida)	5
Phocomelia	2
Arthrogryphosis	2
Achondroplasia	1
Arachnodactyle	1
Congenital dislocation hip	1
Microphthalmia	1
Genito-urinary	27
Sexual aplasia	5
Renal aplasia	5
Polycystic kidneys	4
Undescended testes	3
Megaloureters	3
Hypospadias	2
Cryptorchid	2
Recto-vaginal fistula	1
Clitoral hyperplasia	1
Double uterus	1
Cardiovascular	21
Interventricular defect	5
'Congenital heart disease'	5
Fallot tetralogy	4
Triloculare	3
Cardiomegaly	2
Coarctation of aorta	1
Absent valves	1
Neurological	19
Anencephaly	5
Hydrocephaly	4
Spina bifida	4
Mental defect	2
Angioma	1
Brain cysts	1
Cyclops	1
Nerve paralysis	1
Others	10
Turner's syndrome	2
Hyperteiorism	1
Haemangiomas	1
Pulmonary aplasia	1
Splenomegaly	1
Asplenia	1
Abnormal optic fundi	1
Cystic pancreas	1
Cystic liver	1

TABLE 8
INFANTS DEAD OF ABNORMALITIES INCOMPATIBLE WITH LIFE

Abnormality	Stillborn	Neo-natal Death	Total
Anencephaly	5	—	5
Hydrocephaly	2	2	4
Exomphalos	1	1	2
Congenital heart disease .. .	4	11	15
Cyclops	1	—	1
Cranial nerve paralysis .. .	—	1	1
Spina bifida	—	4	4
Renal aplasia	1	2	3
Gastro-intestinal aplasia .. .	—	4	4
Total	14	25	39

TABLE 9
KNOWN ABNORMALITIES IN INFANTS DEAD OF OTHER CAUSES

Cause of Death	Abnormality
Broncho-pneumonia	Syndactyle
Hyaline membrane	Tracheo-oesophageal fistula
Stillborn macerated	Unilateral renal hypoplasia
Stillborn macerated	Cleft palate
Stillborn macerated	Talipes equinovarus

the alimentary and genito-urinary tracts are more frequent, and lesions of the central nervous system less frequent than might be expected.

Thirty-nine infants died of congenital abnormalities incompatible with life (Table 8). Congenital heart disease is the most common abnormality considered incompatible with life. Table 9 shows the anomalies present in those five infants whose death was considered to be due mainly to other reasons.

The congenital abnormalities from which the surviving infants suffer together with their weight and maturity at birth, sex, period of follow-up and comments are shown in Table 5. Several of these children will not survive to reach adult life. Eight (57%) were premature by weight although only two (14%) were born before the 36th week of gestation. This is not surprising if the absence of an umbilical artery slows the actual growth of a foetus *in utero*, but it does not necessarily prevent the foetus from maturing. It is upon maturation rather than actual body weight that the infant subsequently depends for its survival.

Trivial abnormalities such as cutaneous naevi and minor haemangiomas have not been included in this series. The number of uncomplicated or single malformations is very low. There are only 14 (24%) whilst the number of multiple abnormalities, 44 (76%) is much higher than expected. The great increase in the incidence of congenital abnormalities in this series is due mainly, but not entirely, to a rise in the number of multiple abnormalities. Neel

(1958) found that only 14.9% of malformed children had multiple defects; Schull (1958) reported that the percentage of infants with multiple abnormalities rose to 34.3% if the parents were related.

Sex. It was suggested by Hyrtl (1870) that there is a male preponderance associated with this abnormality. This was not supported by Benirschke and Brown (1955) or Little (1958), nor is it supported in the present series, which contains 55 female and 49 male infants. There is one hermaphrodite and the sex of eight infants is not recorded. An analysis of the various abnormalities, the stillbirths, surviving infants or maturity also fails to show any sex domination (other than those which are expected, such as the anencephalic infants, all five of whom were female). There is, in this series, a total of 16 twin infants, whose sex is predominantly female. There are, in fact, 13 female and three male infants. No reason is offered for this apparent female dominance, which despite the small number of cases, does appear to be significant.

Side of Absence. It is only possible to define the absent side at necropsy. Unfortunately adequate information is available upon only 22 infants in whom the artery was absent, 12 times on the right side and 10 times on the left. The intra-abdominal part of the umbilical artery was easily recognizable in more than half the babies, although in some it existed as a very thin threadlike structure. Fig. 2 demonstrates absence of the right artery. The dissection displays the branches of the aorta as seen from the dorsal aspect and shows marked aplasia of all the aortic branches on the right side. A similar developmental abnormality of these vessels was frequently noted at necropsy, but the corresponding lower limb was never smaller than the contralateral limb. When present, deformities of the lower limbs were usually more severe upon the affected side.

Maturity and Weight. The maturity of 109 infants is known out of a total of 113. Six were non-viable, i.e. born before the 28th week of pregnancy, and 103 were viable. Table 10 shows the maturity, sex, average birth weight and average placental weight of the infants. The average birth weight is lower than normal, especially in the more mature age groups. The average placental weight is also less than normal. It seems reasonable to suppose that the absence of an umbilical artery could cause a diminution of oxygen supply to the foetus and this theory receives some support from the small average foetal weight. There is, however, no compensating enlargement of the placenta.

FIG. 2.—The viscera of a stillborn infant dissected and photographed from the dorsal aspect. The aorta is normal. The left iliac, umbilical and femoral arteries are normal. The right common iliac artery, which is about one quarter of its normal size, is distributed mainly along its internal iliac branches. The right umbilical and femoral arteries have no visible lumen and exist as threads in connective tissue.

Infants suffering from congenital abnormalities tend to be born prematurely and in this series 56 (59%) of the 95 viable infants of known weight were premature by weight at birth.

TABLE 10
MATURITY, SEX AND AVERAGE BIRTH WEIGHT OF
INFANTS WITH AVERAGE PLACENTAL WEIGHTS

Maturity (weeks)	Number	Sex (if known)		Average Birth Weight		Average Placental Weight (oz.)
		M	F	lb.	oz.	
<28	6	2	1	—	—	—
28	2	1	1	1	8	8
29	2	1	1	1	4	10
30	2	1	1	2	15	12
31	1	—	1	2	12	9
32	5	2	3	2	12	13
33	4	2	2	3	11	14
34	3	2	1	2	14	12
35	12	5	7	3	13	15
36	9	5	3	5	4	18
37	11	5	6	5	2	14
38	11	6	5	6	5	15
39	7	3	4	6	13	17
40	20	8	12	6	5	22
41	6	3	2	6	7	19
42	6	3	3	5	12	14
>42	2	—	2	5	0	17
Not known	4	—	—	—	—	—
Total ..	113	48	55	—	—	—

Placentae. A relatively high incidence of placental abnormality was recorded by Benirschke and Brown (1955), and Little (1958) reported an increased frequency of velamentous insertion of the umbilical cord. In this series there was a total of 32 (29%) placentae that showed gross abnormality including, velamentous insertion of the cord (12); circum-margination (five); circumvallation (five); lobed (four); placenta praevia (three) and maternal floor infarct (three). Massive infarction of the maternal surface of the placenta with resulting intra-uterine asphyxia has been noted by Benirschke and Bourne (1960) to occur in successive pregnancies and to be associated with absence of an umbilical artery.

Accidental ante-partum haemorrhage occurred in six pregnancies from which only one infant survived; four were stillborn and one died after 24 hours. Placenta praevia was present on three occasions. A history of threatened abortion was given by eight patients, an episode which did not appear to affect adversely the infant's chance of survival.

Twins. The incidence of absent umbilical artery is greatly increased in plural pregnancy. In the present series of 113 babies the abnormality occurred 16 times (14%) in twin pregnancies; only five (31%) of these 16 infants are alive and well.

In a consecutive series of 100 twin pregnancies the abnormality was present on seven occasions. This incidence of 7% of twin pregnancies, or 3.5% of twin infants, as compared with an overall incidence of 1% appears to be a significant increase. A similar unexplained observation (Stevenson, Worcester and Rice, 1950) is the finding of a doubled incidence of foetal malformation in twin pregnancies.

Benirschke and Bourne (1960) considered that the increase of this anomaly might be caused by a relatively higher proportion of monozygous twins. This appeared to be correct in a smaller number, but the present larger series of 16 infants does not support the theory. Six (37%) infants were monozygous because of monochorial placentation, while 10 (63%) were dizygous because of dichorial placentation or heterosexual twins, so that they obey the general rule regarding the zygosity of twins, which states that nearly one-third are monozygous and two-thirds dizygous.

Discussion

Variations in the number of vessels in the umbilical cord are known to occur, but the available information upon their origin and significance is sparse. The

most important vascular aberration is absence of one of the two umbilical arteries through the length of the umbilical cord and is associated with atresia of the corresponding intra-abdominal portion of the artery. This abnormality is present in 1% of pregnancies and in 7% of twin pregnancies. Anastomoses between the umbilical arteries normally occur at the placental end of the umbilical cord either in the cord itself or in the adjacent placenta. These anastomoses presumably serve to equate the intra-arterial pressures and ensure equal distribution of blood throughout the placenta. They may also act as an arterial shunt when a portion of the foetal placental circulation is closed for physiological or pathological reasons and could, therefore, exert a profound influence upon the ability of the foetus to maintain its placental reserve or to resist the anoxic effects of large placental infarcts.

Foetal Abnormalities. The incidence of congenital abnormality amongst an average population is between 1% and 2.5% of all live and stillbirths (Malpas, 1937; Logan, 1951; Lewis, 1956) of which approximately half are gross foetal abnormalities. Absence of one umbilical artery appears to be the commonest major congenital abnormality of the human foetus.

There is a significant rise in the incidence of congenital abnormalities associated with foetuses having only one umbilical artery. The width of the range of the congenital abnormalities suggests that the absence of an umbilical artery is not a secondary effect but is probably a primary lesion to which, some at least, of the associated anomalies are secondary.

It is possible that these abnormalities are the result of the hypoxia to which the foetus is subjected during the organo-genetic period. The efficiency of a single artery or the date of closure of a second artery, are not standard so that both the time and degree of relative oxygen lack would vary widely. There is no suggestion that all the anomalies in this series are caused by a single agent, as some are considered to be hereditary (Potter, 1952) but, if a factor such as anoxia were playing a part in their production, it would have to be capable of acting over a long period in order to account for the wide variety of abnormalities concerned.

However, the majority of anencephalic infants possess two normal umbilical arteries, which indicates that either absence of an umbilical artery has no teratogenic influence, or, if it does, that there are other agents which are equally effective. Similar arguments can be forwarded concerning the other abnormalities of this series, in that they also are

considered to occur in the presence of two normal umbilical arteries so that the precise embryological and developmental significance of absence of one umbilical artery is unknown.

The high incidence of associated foetal abnormality occurring in babies with only one umbilical artery suggests that the 'anomalies' are in some way connected. The presence of only one artery in the umbilical cord may constitute a severe impediment to normal foetal development by causing a resistance to the blood flow and possible lack of oxygen to the foetus. This, together with an absence of the juxtaplacental umbilical arterial anastomosis, may also be a cause of intra-uterine death. This series includes 16 twin infants and the fact that 14 (88%) of these infants were smaller than their normal siblings does support the theory that a single umbilical artery impairs normal growth.

Minor congenital abnormalities are conspicuous by their absence in this series. When two factors that are known to be associated with major congenital abnormalities are combined, such as hydramnios and absent umbilical artery, the foetal loss is particularly high.

Cardiac Malformation. There are several reasons why a high incidence of cardiac abnormalities might be associated with absent umbilical artery. In itself the absence is an anomaly of the cardiovascular system and other changes may occur because the artery normally develops during the period of rapid formation of the circulatory system. A heavy strain would certainly be imposed upon the heart if a functioning artery underwent degeneration in early foetal life. It has been shown that a single artery can support an apparently normal foetus but the resistance to blood flow and degree of cardiac overload must, at times, be considerable and may predispose to abnormalities within the heart. One might assume that measuring the diameter of the remaining artery would offer some means by which one could determine if an increased resistance to blood flow had existed in intra-uterine life. Unfortunately, the thick muscular wall causes their lumina to vary greatly after delivery, so that there is no reliable method whereby their functional efficiency within the uterus can be assessed (Reynolds, 1952).

Aetiology. It is disappointing that even such a large series does not give any definite indication of the cause of this abnormality. There is no definite evidence that the condition is familial or recurrent, although it does occur in the children of women who have a poor reproductive ability. It is more

frequent in the younger primigravida and the older multigravida. The fact that four mothers suffered from epilepsy is interesting but the number is too small to be significant.

We have considered the possibility that the vessel which was formed in a normal manner has subsequently ceased to function, and atrophied. Such a cessation of function would cause some anoxia, however transient, which could easily disturb the embryological development of the foetus and cause congenital abnormalities. The time at which the artery ceased to function would be the teratogenetic termination period and as such would determine which foetal organs would be most severely affected. If the time varied widely, so would the malformation.

The theory that the artery once was present and functioning is supported to some extent by the invariable presence of an intra-abdominal umbilical artery, albeit often attenuated, on the affected side. It is, however, difficult to accept that a properly functioning umbilical artery could entirely disappear from the umbilical cord without leaving some histological evidence of its presence. In four instances, however, the residual arterial structure was present and, having a diameter much greater than that of the vitelline vessels, it is assumed that these remnants are aplastic umbilical arteries.

It is our opinion that the second umbilical artery was never satisfactorily formed within the umbilical cord in the majority of the infants in this series but, that in a few instances, a functioning vessel was present at some stage of foetal development.

Clinical Importance. Absence of an umbilical artery, occurring in 1% of patients, is associated with a high incidence of foetal abnormality (65%). Many of these abnormalities are not obvious, nor are they readily apparent upon simple general examination of the newborn. In most maternity units the placenta, membranes and umbilical cord are examined, weighed and measured and it would be a simple procedure to examine routinely the cut end of the cord. The vessels are easily identified and the finding of a single artery should stimulate a further and more detailed examination of the infant. Such an observation might be of great benefit if the presence of unsuspected abnormality were to be considered in those cases of single umbilical artery encountered. Benirschke and Bourne (1960) report that the diagnosis of oesophageal atresia was made six hours after delivery in an infant who was re-examined following the reported finding of a single umbilical artery. The child is well one year after surgical treatment for the condition. The importance of hydramnios in association with absent

umbilical artery is stressed, for, in this series, all of the 19 infants in whom this combination was present suffered from a major congenital abnormality.

Prognosis. Details of the congenital abnormalities suffered by the affected children have been recorded together with the prognosis both before and after delivery. Absence of one umbilical artery does in itself diminish a child's chance of survival. The incidence of intra-uterine asphyxia, stillbirth and prematurity is higher than normal. The anomaly appears to cause a slowing of intra-uterine growth so that the average birth weight is less than the expected normal average. It is interesting to note that these infants, when healthy, are similar to twin infants in this respect because their ability to survive depends more upon their maturity than weight. This series produces further confirmation that this principle applies also to the single pregnancies.

When the infants are born alive their prognosis is identical to that of their associated abnormality, if present, together with superimposed prematurity. As stated above, they withstand prematurity fairly well and, having survived the newborn period, they develop physically and mentally into normal children, provided, of course, that they are not encumbered by the frequently associated congenital abnormalities.

Summary

A total of 113 cases of absence of one umbilical artery are presented and analysed, together with a review of the literature.

The abnormality is slightly increased amongst the younger primigravidae and the older multigravidae, but does not increase in incidence as parity rises. A total of 59 multiparous women had had 155 previous pregnancies from which only 107 (69%) infants survived. A total foetal loss of 48 (31%) is considered excessive and indicates that these women, as far as reproduction is concerned, belong to a high risk group. Hydramnios occurred 19 times; all (100%) the infants suffered from major congenital abnormalities and only two survive as a result of surgical treatment for oesophageal atresia. There is, at present, no evidence to show that the condition is hereditary.

The incidence of absent umbilical artery is 1% of all births and 7% of twin pregnancies.

The foetal loss in a consecutive series was 20% but in the total of 113 instances the foetal loss was 65 (58%). Fourteen of the 48 infants who survive are known to suffer from congenital abnormalities.

Major congenital abnormalities were present in 58 (51%) of the infants; only 40 (35%) were developmentally normal and a further 15 (14%) were thought to suffer from a congenital anomaly. The abnormalities were mostly multiple and not confined to any particular system. Obstructive lesions or atresia of the gastro-intestinal and urinary tracts were relatively common.

Fifty-six (59%) of the 95 viable infants of known weight were premature by weight at birth, but the average weight of all the infants at birth was less than normal. The placenta was considered to be abnormal in 32 (29%) of the pregnancies.

It is suggested that routine examination of the cut end of the umbilical cord at delivery might result in early diagnosis of some of the associated congenital abnormalities.

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Addendum

Since writing this article it has been discovered that one of the infants considered to be developmentally normal is, in fact, suffering from a marked degree of mental retardation.