SINGLE UMBILICAL ARTERY—ITS ASSOCIATION WITH CONGENITAL MALFORMATIONS

by

V. K. MITAL,* M.S., F.R.C.S., BRIJENDRA K. GARG,** M.D., M.R.C.P., D.C.H.

and

URMILA GUPTA,*** M.S.

The umbilical cord normally has two arteries and one vein. Whittaker (1870) mentions that in early gestation there are two arteries and two veins. Monie (1952) states that one vein disappears at the time the congenital herniation returns to abdominal cavity, around the 10th week of gestation. One of the umbilical arteries may be absent and although such an occurrence was described in 1621, it was Benirschke and Brown who in 1955 reported a significantly higher incidence of congenital malformations in babies who had a single umbilical artery (SUA).

Since this first publication several reports have appeared on this topic. The largest study to-date has been published by Froehlich and Fujikura (1966) who reviewed all single births (26,539) included in a collaborative study from twelve different institutions in U.S.A. conducted during 1959-63. Two reports have been published from India (Rohatgi, 1967; Saigal and Srivastava, 1968), and the present report is on the largest number of cases and also includes the controls.

*Reader in Surgery.

**Reader in Pediatrics.

***Reader in Obstetrics and Gynecology, G.S.V.M. Medical College, Kanpur.

Received for publication on 14-2-1969.

Observations

A total of 4,612 consecutive neonates (4,530 single births and 41 twin pairs) were examined for the presence of congenital malformations over a two year period. The umbilical cords of all the babies were sectioned to see the number of blood vessels. If a cord was found to have a single umbilical artery further sections were made at different levels to confirm this finding.

Of the 4,530 cords of single births, SUA was seen in 37 (0.81%), and 15 (40.5%) of these babies had congenital malformations. In the remaining 4,493 single births with normal cord vessels, congenital malformations were noted in 92 (2.05%) babies. There were 4 instances (4.8%) of sua in 82 cords of 41 twin births and one of these was associated with a grossly malformed (acardiac, acephalic monster) and stillborn foetus (25%). While in the remaining 78 twin members there were only 2 (2.56%) instances of malformations (table 1).

TABLE I

Malformations in babies with SUA and controls

	Infants with SUA			Infants with normal umb. vessels		
	Total No.	No. malf.	Per cent malf.	Total No.	No. malf.	Per cent malf.
Single births	37	15	40.5	4493	92	2.05
Twins	4	1	25.0	78	2	2.56
Total	41	16	39.0	4571	94	2.05

Table II shows the congenital anomalies seen in babies with SUA. The malformations have been divided into two categories: A, if fatal or major, and B, if non-fatal.

TABLE II

Individual malformation in single births with SUA

Malformations	Number	Per cent	Type	Outcome
Gastro-intestinal system (4)				
Exomphalos	1	2.7	A	Neonatal death*
Tracheo-oesophageal fistula	1	2.7	A	Neonatal death
Cleft palate	1	2.7	B	Alive
Imperforate anus	1	2.7	B	Alive
Central nervous system (4)				
Anencephaly	2	5.4	A	Stillborn
Iniencephaly	1	2.7	A	Stillborn
Dermal sinus	1	2.7	B	Alive
Skeletal system (3)				
Talipes equinovarus	2	5.4	B	Alive
Polydactily	1	2.7	B	Alive
Integumentary system (2)				
Haemangioma face	1	2.7	B	Alive
Skin tags in nostril	1	2.7	B	Alive
Others (2)				
Hypospadias	1	2.7	B	Alive
Dextrocardia	1	2.7	A	Neonatal death

*refers to early neonatal (first 7 days) death.

Discussion

In our series the incidence of SUA was 0.81% in single births and 4.8% in twin births. An incidence of about 1% is reported by various workers (table III). A higher incidence of 2.9% has been reported in retrospective series by Seki and Strauss (1964). This high incidence is due to the fact that the placentae were examined because either the neonates had died or some abnormality was suspected. The drawbacks of a retrospective series are obvious in the report by Lenoski and Medovy (1962) who found a low incidence of 0.2% in a prospective series, while this was 5.2% in retrospective series where the umbilical cords of 229 perinatal autopsy cases were examined. Froehlich and Fujikura (1966) found an overall incidence of SUA to be 0.76%, and this was 1.22% in whites and 0.44% in Negroes. A high incidence (7%) of SUA has been reported in twins by Bourne and Benirschke (1960).

TABLE III

Reported incidence of SUA and congenital malformations in single births

Author (year)	No. of neonates	% SUA	% congenital malformations
Benirschke and Brown (1955)	..	55 cases	64.0
Little (1958)	1200	1.10	25.0
Benirschke and Bourne (1960)	1500	1.00	47.0
Lenoski and Medovy (1962)	2500	0.20	20.0
Papadalos and Paschos (1965)	7866	0.40	31.2
Seki and Strauss (1966)	3353	2.90	55.0
Cairns and Mckee (1964)	2000	1.00	10.0
Froehlich and Fujikura (1966)	25539	0.76	28.6
Rohatgi (1967)	1430	0.94	50.0
Saigal and Srivastava (1968)	1000	0.60	50.0
Present series	4530	0.81	40.5

We noted congenital malformations in 40.5% single births with SUA, while in the control series the incidence of malformations was only 2.05%—a highly significant difference. There was only one instance (25%) of congenital malformation (acephalic acardiac monster) among 4 new borns with SUA in twin pregnancies. Thus, although SUA was found more frequently in twin births, the incidence of malformations was not more. This is in agreement with the observations of Benirschke (1962). The incidence of congenital malformations in neonates with SUA reported by other workers is shown in table III. Froehlich and Fujikura (1966) found that although the incidence of SUA in whites was 1.22% and in Negroes only 0.44%, the reverse was true for associated malformations as these were found in 42.1% Negroes and 23% whites. The highest incidence of malformations (64%) has been reported by Benirschke and Brown in 1955 in their retrospective and highly selective series. Cairns and Mckee (1964) detected congenital malformations in 10% cases with SUA and stated that the association of SUA with congenital malformations has been overemphasized.

Bourne and Benirschke (1960) found 21 cases of cardiovascular malformations among 113 neonates with SUA. They attributed this high incidence of cardiac anomalies to the fact that the artery normally develops during the period of rapid development of the circulatory system. In fact, the absence of one umbilical artery is in itself an anomaly of the cardiovascular system. Froehlich and Fujikura (1966) found only 8 cases of cardiovascular malformation among 203 neonates with SUA.

In a preliminary report, Feingold *et al* (1964) reported abnormal pyelograms in 33% of 23 infants with SUA and concluded that routine intravenous pyelography is indicated for all newborns with SUA to permit early detection of surgically correctible abnormalities. But the addition of more newborns with SUA to the study has shown a decline in the incidence of genito-urinary abnormalities. The true incidence of such anomalies and the value of routine pyelography will be evident from the study of a very large series of cases (Gellis, 1965-66).

The association of SUA with trisomy-18 (Uchida *et al*, 1962) and Turner's gonadal dysgenesis (Richert and Benirschke, 1958) has also been reported.

Among twin births, we found one instance of acephalic, acardiac monster, the other twin being normal, Acardiac or hemi-acardiac monsters always have one umbilical vein and one umbilical artery. This aspect will be discussed by us in a separate communication (Mital *et al*).

In this series of 37 cases of SUA, 6 (16.2%) were stillborn, of which 3 (50%) had associated malformations. There were 5 (13.5%) early neonatal (first week) deaths of which 3 (60%) had associated congenital malformations. Alternatively, if we took into account only 15 cases of SUA who had congenital malformations it was seen that 3 (20%) were stillborn and 3 (20%) died in the early neonatal period. Seki and Strauss (1964) reported that of their 60 cases with SUA, 27 (45%) were stillborn (including 13 pre-viable foetuses under 1000 g.) and 15 died during the neonatal period. Froehlich and Fujikura (1966) reported 9.8% stillbirths and 3.3% neonatal deaths in infants with SUA, Rohatgi (1967) reported that of 7 cases of SUA associated with congenital malformations, 4 were stillborn and 2 died in the neonatal period. Saigal and Srivastava (1968) reported a mortality of 50% among malformed infants with SUA (they did not include stillbirths in their series).

It is not known whether SUA predisposes to malformations and if so what the underlying mechanism is. SUA is not considered to be a teratogenic agent *per se* (Seki and Strauss, 1964), but the presence of SUA may constitute a severe impediment to the normal foetal development by causing a resistance to the blood flow and possible lack of oxygen to the foetus. This fact may account for foetal abnormalities and for the average low birth weight seen in these cases (Bourne and Benirschke, 1960). Whatever may be the cause for malformations, it is obvious that SUA is associated with malformations in significantly larger number of neonates. It is suggested that the placentae of each newborn should be examined for the presence of SUA and in its presence a careful search must be made for congenital malformations. Even if no malformation is seen this fact must be noted on the case-records so that it may be helpful in future follow-up studies.

Summary

A total of 4612 consecutive neonates and placentae (4530 single births, 41 twin pairs) were examined for the presence of SUA and congenital malformations. Of 4530 single births SUA was seen in 37 (0.81%) neonates and 15 (40.5%) of these SUA had associated malformation, while among the remaining 4493 single births with normal cord vessels, congenital malformations were seen in 92 (2.05%) babies SUA was found in 4 (4.8%) of the 82 cords of 41 twin births and one (25%) was associated with congenital malformation, while in the remaining 78 twin members there were 2 (2.50%) instances of malformation. Stillbirth and early neonatal death were recorded in 3 (20%) instances

each of the 15 malformed babies with SUA. It is concluded that congenital malformations are seen in a significant number of cases with sua and that the placentae of all neonates should be routinely examined for the presence of SUA.

References

1. Benirschke, K. and Brown, W. H.: Obst. and Gynec., **6**: 399, 1955.
2. Benirschke, K. and Bourne, G. L.: Am. J. Obst. and Gynec., **79**: 251, 1960.
3. Benirschke, K.: Am. J. Obst. and Gynec., **84**: 1959, 1962.
4. Bourne, G. L. and Benirschke, K.: Arch. Dis. Childh., **35**: 534, 1960.
5. Cairns, J. D. and Mckee, J.: Canad. M.A.J., **91**: 1071, 1964.
6. Feingold, M., Fine, R. N. and Ingall, D.: New England J. Med., **270**: 1178, 1964.
7. Froehlich, L. A. and Fujikura, T.: Am. J. Obst. and Gynec., **94**: 274, 1966.
8. Gellis, S. S.: Editorial comment in Year Book of Pediatrics, 1965-66 series, Year Book Publishers: Chicago, p. 223.
9. Lenosky, E. F. and Medovy, H.: Canad. M.A.J., **87**: 1229, 1962.
10. Little, W. A.: Bull. Sloane Hosp. Wom. N.Y., **4**: 127, 1958.
11. Mital, V. K., Rohatgi, P. and Garg, B. K.: to be published in J. Obst. and Gynec. India.
12. Monie, I. W.: Manitoba M. Rev., **32**: 491, 1952.
13. Papadalos, C. and Paschos, A.: Obst. and Gynec., **26**: 367, 1965.
14. Richert, R. and Benirschke, K.: New England J. Med., **258**: 974, 1958.
15. Rohatgi, P.: J. Obst. and Gynec. India, **17**: 718, 1967.
16. Saigal, S. and Srivastava, J. R.: Indian Pediat., **5**: 202, 1968.
17. Seki, M. and Strauss, L.: Arch. Path., **78**: 449, 1962.
18. Uchida, I. A., Bowman, J. M. and Wong, H. C.: New England J. Med., **266**: 1198, 1962.
19. Whittaker, J. T.: Am. J. Obst. and Gynec., **3**: 193, 1870, cited by Javert, C. T., in Spontaneous and Habitual Abortion, New York, 1957, The Blackistan Division, McGraw-Hill Book Co. p. 143.