THE YEAR BOOK *of* PEDIATRICS

(1961-1962 YEAR BOOK *Series)*

EDITED BY

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Sirenomelia and Monomelia with Renal Agenesis and Amnion Nodosum. Interest has recently been directed toward sympodia, a fetal malformation better known as sirenomelia, sympus dipus or mermaid fetus. This malformation consists of more or less complete fusion of the lower limbs to form a single extremity. Monomelia, an abnormality in which there is a single lower extremity that shows no indication that it has been derived from fusion of two lower limbs, has been reported less often.

Severe dysplasia of the fetal urinary tract is apparently an invariable accompaniment of sirenomelia and monomelia. There has been only an occasional reference to the liquor amnii in reported cases, and there are no previous histologic studies of the placental membranes.

A. D. Bain, M. M. Beath and W. F. Flint[1] (Univ. of Edinburgh) report 2 cases of sirenomelia and 2 of monomelia. Potter facies, large spadelike hands (Fig. 1) and pulmonary hypoplasia were present in all 4 cases, and kidneys were entirely absent. Illustrations of sirenomelia and monomelia in reports by several authors show what is now regarded as the characteristic facial appearance first described by Potter in association with renal agenesis. These illustrations also show the large flattened and clumsy appearance of the hands described by Bain and Scott in connection with severe urinary tract dysplasia. These features, however, have not been generally recognized as assocated with sirenomelia and monomelia.

In 3 of the 4 present cases, the placenta was examined histologically and amnion nodosum was found. As shown by Scott and Bain, this lesion is found only in association with oligohydramnios. Oligohydramnios was noted clinically in 2 of the cases.

Severe urinary tract dysplasia, such as renal agenesis or complete urethral atresia, and its associated fetal and placental changes are constant findings in sirenomelia and monomelia.

(1) Arch. Dis. Childhood 35:250-253, June, 1960.

Little is known concerning the cause of sirenomelia and monomelia, but several hypotheses have been put forward, including the suggestion that oligohydramnios is one cause. The oligohydramnios is obviously secondary to the kidney or urinary tract defect, however, and although it can ac-

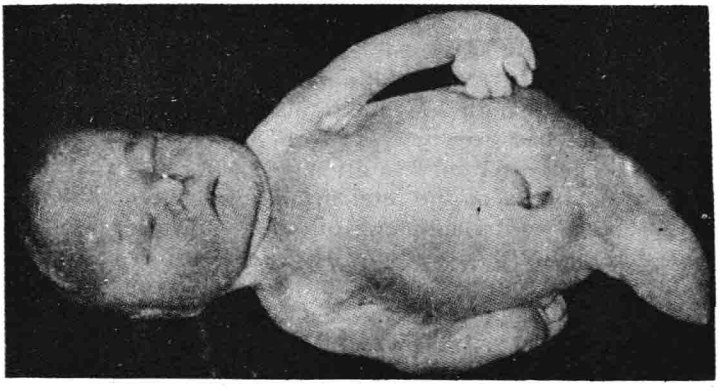

Fig. 1.—Monomelic fetus showing large spadelike hands. (Courtesy of Bain, A. D., *et al.*: Arch. Dis. Childhood 35:250-253, June, 1960.)

count for the facies and hand malformation, it is unlikely to produce fusion of the lower limbs or monomelia. Absence of one umbilical artery has been cited as an etiologic factor, on the assumption that it impairs blood supply to the lower limbs. However, absence of one umbilical artery is often seen in babies with varied congenital malformations and occasionally in apparently normal infants.

Association of Sirenomelia with Potter's Syndrome. J. G. Bearn[2] (Middlesex Hosp., London) reports a case in which the features of Potter's syndrome, the typical facies, aplasia of the lungs and renal agenesis, occurred in a fetus with sympodia showing a second-degree deformity. Although the typical facies described by Potter has not been reported in papers on sympodia, it is likely that most sympodial fetuses show the facies, in view of the usual absence of renal tissue in this deformity. The peculiar facies may be explained by compression of the head by the uterine wall, secondary to the oligohydramnios usually present both in sympodia and in renal agenesis without gross abnormalities of the lower limbs. The flattening of the nose, receding chin and flat-

(2) Arch. Dis. Childhood 35:254-258, June, 1960.

tened malpositioned ears could all be due to mechanical pressure on the developing head.

That all sympodial monsters have but one umbilical artery is curious. In the present case the right umbilical artery persisted, although both Ballantyne and Kampmeier found, in their cases, that the artery within the umbilical cord was a persistent vitelline artery.

► [The author states that "the observation that all sympodial monsters have but one umbilical artery is a curious finding." That this observation does not hold for all cases is illustrated in the preceding article by Bain *et al.*; in Case 1 of their series the umbilical cord contained three vessels. Nothing is stated regarding the vessels in the other 3 cases. In the article by Faierman that follows, the author states that in 95 of the 182 cases of sirenomelia collected by Ballantyne (1898), Kampmeier (1927) and Hendry and Kohler (1956), there was but one umbilical artery. In the remaining cases the cord had not been examined. One must conclude that sirenomelia, or sympodia (we prefer the former term because it seems more interestingly descriptive of the condition) may be associated with a single umbilical artery but that the latter is not necessarily the cause of the abnormality.

Whether amnion nodosum will be found in all cases also remains to be determined.—Ed.]

Significance of One Umbilical Artery. Edith Faierman[3] (Birmingham, England) reports 11 cases of single umbilical artery among 411 autopsies on stillborn and live babies under age 8 weeks, an incidence of 2.7%. Nine infants had severe associated malformations, and in 8, the malformations were multiple. Seven had malformations of the lower urinary tract. One was a sirenoid fetus with no perineal orifices, 1 had valvular obstruction of the posterior urethra, 2 had extreme hypoplasia or agenesis of the bladder and 3 had atresia or severe stenosis of the bladder neck. Two infants with bladder neck obstruction also had rectal atresia. Four infants had renal hypoplasia or agenesis, 4 malformations of the heart and 2 esophageal atresia. Ballantyne stated that the commonest associated malformation was defective development of the bladder and sirenomelia. Benirschke and Brown found mainly malformations of the heart and central nervous system; anencephaly was the commonest malformation found by Hyrtl.

There were 2 monovular twins. In both cases, the other twin had two umbilical arteries and no malformations. Benirschke and Brown also found a high incidence of twinning.

Seven placentas were available for examination. One was

(3) Arch. Dis. Childhood 35:285-288, June, 1960.

infarcted and the others were normal. One umbilical cord was abnormally long, one was short and one had a velamentous insertion. Benirschke and Brown found a high incidence of placental anomalies, such as circumvallation, circummargination, velamentous insertion of the cord and extensive infarction.

In the present series there were 5 stilllbirths and 6 live births. Benirschke and Brown also reported a high incidence of stillbirths (76%). Hyrtl, in 1870, found only 7% stillbirths, but this was probably due to the fact that autopsies were infrequently performed on stillbirths.

There were equal numbers of male and female fetuses with one umbilical artery. This agrees with the findings of most authors. Kampmeier, however, found a male preponderance in sirenoids, and Hyrtl found only males in his series of 12 fetuses.

No correlation was found between the abnormality of the umbilical artery and maternal factors. Of the mothers, 60% were multiparous, and they were aged 17-25. Pregnancies were normal in 7 cases; in 1 there was a threatened abortion and in 1, toxemia of pregnancy. Benirschke and Brown found a high incidence of toxemia and hydramnios.

Absence of one umbilical artery can be diagnosed at birth by careful examination of the cut surface of the umbilical cord. This is of little practical value in the presence of gross malformations. There were no gross external malformations in 6 of the present patients, however. Two of these showed no associated malformations at autopsy, 2 had malformations incompatible with life and 2 had malformations which may be amenable to operation. Because of these findings, Faierman concludes that a newborn with no gross external malformation but with only one umbilical artery has a 2:1 risk of severe internal malformation, which may warrant early surgical correction.

► [The frequency of major congenital abnormalities in association with absence of one of the umbilical arteries and the fact that there were no gross external malformations in 6 of the 11 cases reported in this series makes it imperative that the obstetrician routinely examine the umbilical cord after cutting and tying it. He should record in every case whether two umbilical arteries are present. All we know is the incidence of this abnormality in stillbirths or newborns coming to autopsy. How often this finding may occur in infants whose associated defects may not be fatal is unknown and remains to be determined. The fact that in 2 cases in this series there were malformations amenable to surgery indicates the importance of a thorough study of infants with a single umbilical artery

if they survive the immediate newborn period, although they may have no gross external defects.

The obstetrician will have to learn not to be an old shut-mouth. Everything he has learned about abnormality or disease in the mother, both in previous pregnancies and the present one—drugs administered, problems of labor and delivery, abnormalities of placenta, membranes and cord—must be passed on immediately to the physician responsible for the care of the newborn. In turn, the physician who looks after the infant must keep the obstetrician informed about the findings in relation to the infant, for they may have bearing on future pregnancies. In the past the physician who took care of both mother and infant was in a good position to give excellent care; however, he usually didn't know what to do with his information.

Since writing the foregoing comment, we encountered the papers that follow and that report the incidence of absence of one umbilical artery in consecutive deliveries. We asked Dr. Kurt Benirschke if he could account for the high incidence of cardiac anomalies reported in his early "retrospective" study, compared with the large number of genitourinary abnormalities noted in the Faierman study. He commented:

"The obstetrician does not examine the cord very often for the absence of one umbilical artery, but he should, since it occurs in 1% of all deliveries. At times the absence of one umbilical artery is a fairly good clue to look further for abnormalities in the baby. The obstetrician should also examine the cord because it is so very simple. The fact that he hasn't done so is emphasized by the lack of knowledge existing about this abnormality. In fact, it has not been commented on since it appeared in the old German literature, although it must have occurred in every hundredth baby at least.

"I think the discrepancy between reported genitourinary anomalies and cardiac and central nervous system abnormalities is none too surprising. The material is not very extensive in either case, and as far as we are concerned, most of our cases came from a retrospective study and from patients who were autopsied at Boston Lying-in Hospital rather than at Children's Hospital. I suspect that one reason for our preponderance of central nervous system anomalies is that many babies with genitourinary abnormalities at Boston Lying-in Hospital would have been transferred to Children's Hospital had the abnormalities been recognized at birth and had the babies not died at the Boston Lying-in Hospital but at Children's Hospital subsequently. Although I did look at the records at Children's Hospital during my study, I feel that no attention had been paid there to the possibility of such an abnormality and that therefore it was not listed in as many cases as it might have been.

"This anomaly not only has been detected in the past by pathologists but originally it was described by an obstetrician. In fact, one of the earlier observers, Schatz, found it to be quite frequently present in acardiac twins. Sometimes one of identical twins turns to some sort of a monstrous teratoma-like formation and is nourished by reverse circulation through anastomosis from the other twin. Most often in such acardiac monsters the umbilical cord has only one umbilical artery. This has been described not only by Schatz but by other German obstetricians, and I have seen it repeatedly in our twin material as well."—Ed.]

Absent Umbilical Artery: Review of 113 Cases is presented by Gordon L. Bourne and Kurt Benirschke[4] (Boston). Previously, the authors had reported that one umbili-

(4) Arch. Dis. Childhood 35:534-543, December, 1960.

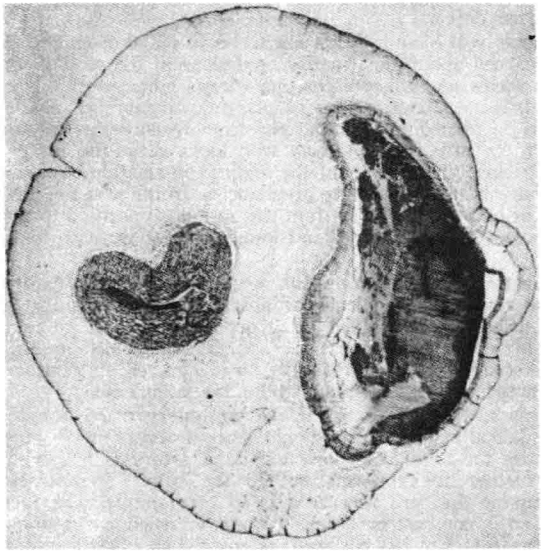

Fig. 2.—Absence of one umbilical artery. Section of umbilical cord showing dilated vein and single artery. Note muscular wall and small lumen of artery. No residual ducts or vessels are present. Hematoxylin-eosin; ×16. (Courtesy of Bourne, G. L., and Benirschke, K.: Arch. Dis. Childhood 35:534-543, December, 1960.)

cal artery was lacking in 15 (1%) of 1,500 consecutive placentas and cords examined. In 100 consecutive twin deliveries, an absent umbilical artery was present 7 (7%) times.

In the present study, of 113 infants who lacked one umbilical artery (Fig. 2), the abnormality was slightly more common in younger primigravidas and older multigravidas. Hydramnios occurred 19 times. There is no evidence that the condition is hereditary.

Only 48 infants survived and 14 (29%) of these are known to have congenital abnormalities (Table 1). Of 65 (58%) infants who were born dead or died after birth, 59 (91%) had definite or probable congenital abnormalities. The abnormalities associated with the absence of one umbilical artery were usually multiple and were not confined to any particular system (Table 2).

The width of the range of congenital abnormalities in infants lacking one umbilical artery suggests that absence of the artery is not a secondary effect, but is probably a primary lesion to which, some at least, of the associated anom-

TABLE 1.—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 haemangiomata	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	Hyperteleorism	0·5	Twin
36	4·6	F	Oesophageal atresia	0·5	Successful operation

TABLE 2.—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
Hyperteleorism	1
Haemangiomata	1
Pulmonary aplasia	1
Splenomegaly	1
Asplenia	1
Abnormal optic fundi	1
Cystic pancreas	1
Cystic liver	1

alies are secondary. The abnormalities may possibly result from the hypoxia to which the fetus is subjected. If this is true, the hypoxia would have to act over a long period to account for the wide variety of abnormalities observed.

The authors suggest 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.

Nature of Neonatal Pulmonary Hyaline Membrane. R. A. Barter and T. G. Maddison[5] (Royal Women's Hosp., Melbourne) state that detailed microscopic study of the lungs of 133 infants affected by hyaline membrane disease showed

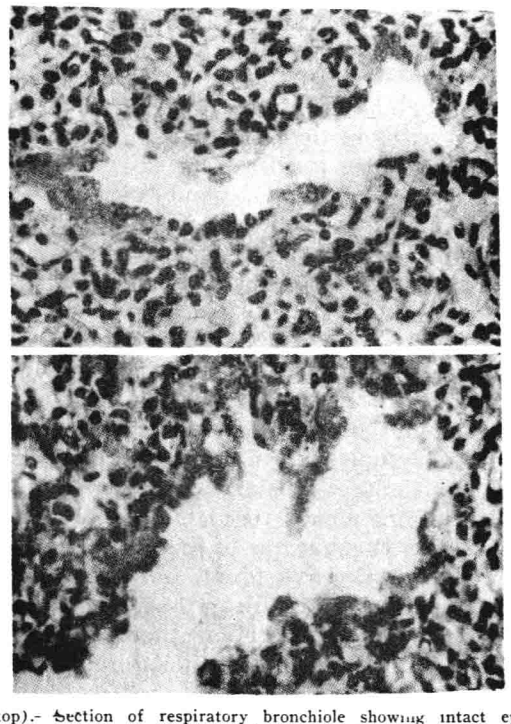

Fig. 3 (top).- Section of respiratory bronchiole showing intact epithelial cells flanked by hyaline membrane. Some nuclei are pyknotic and others in membrane are fragmented. Hematoxylin-eosin; ×228.

Fig. 4 (bottom).—Section of respiratory bronchiole showing typical hyaline membrane and characteristic irregular edge. Hematoxylin-eosin; ×228.

(Courtesy of Barter, R. A., and Maddison, T. G.: Arch. Dis. Childhood 35:460-464, October, 1960.)

that the membranes result from epithelial cell necrosis in respiratory bronchioles and form in situ. In sections stained to show elastic fibers, the characteristic pattern for respiratory bronchioles is seen around all hyaline membrane affected spaces. Spaces lined by membranes usually have a

(5) Arch. Dis. Childhood 35:460-464, October, 1960.