The missing umbilical artery

II. Paediatric follow-up

ELIZABETH M. BRYAN and H. G. KOHLER

From the Department of Paediatrics and Child Health, University of Leeds, and Department of Pathology, The Maternity Hospital at Leeds

Bryan, E. M., and Kohler, H. G. (1975). Archives of Disease in Childhood, 50, 714. The missing umbilical artery. II. Paediatric follow-up. Of 143 infants with single umbilical artery detected by routine examination of the placenta, 25 had major malformations at birth; 3 of these survive. Another 6 were stillborn and 2 died during the first year of life. At follow-up 14 children could not be traced. 18 were assessed on the basis of reports by family doctors or parents ('report group'); 14 of these were considered normal. The remaining 78 infants and children were given a clinical examination ('examination group'); 64 were found to be normal.

Malformations found in 10 children (6 from the examination group and 4 from the report group) are discussed. Most of the abnormalities detected were less severe and less conspicuous than those revealed at birth, and in a few instances only might have been diagnosed by a more thorough examination in the perinatal period. Failure to detect these 'less severe and less conspicuous' malformations is generally unlikely to be detrimental to the infant, with the exception of urinary tract anomalies which are known to predispose to infection. Included in the examination group were 16 children (out of an original 22) who had been 'normal' but small-for-dates at birth; 14 of these had now caught up. The remaining 2 were found to have abnormalities that had not been manifest at birth.

The finding of single umbilical artery at birth commits the paediatrician to an intensive search for malformations which are not immediately apparent, but prolonged surveillance for this reason alone is not advocated.

Recently we reported a study of 143 infants with a single umbilical artery (SUA) born at Leeds Maternity Hospital over a 7-year period (Bryan and Kohler, 1974). 25 of them had major congenital malformations, of whom 21 died within the first 4 weeks of life, 1 died aged 3 months, and 3 are still alive. In addition, there were 6 stillborn infants in whom no malformation was found at necropsy.

In the remaining 112 infants no major abnormalities were detected in the neonatal period and these will be referred to as 'normal'. It is these children with whom we are now concerned. The parents were not told of the absence of an umbilical vessel and no child was kept under surveillance on this account. However, some were seen again in paediatric follow-up clinics for other reasons such as prematurity or low birthweight.

In 1973 it was decided to review these children in order to see whether their development was in any way abnormal, and in particular, whether any unsuspected congenital abnormality had come to light.

Method

The mother's obstetric case notes were first studied. A letter was then sent to the family doctor asking for a report on the child and permission to carry out an examination. Though a large number of families had moved, we were able to account for 98 of the 112 children, being unable to trace the remaining 14 (Fig. 1).

Of the 98 children, 2 had died—a girl aged 3 months...
The missing umbilical artery

FIG. 1.—Fate of 143 infants with a single umbilical artery. (Collected from a population of approximately 20 000 births.)

Results

In 64 instances the parents were satisfied with the child's progress and nothing abnormal was found on examination. The heights and weights of these children were all above the 3rd centile for their age and are shown in Fig. 2. Head circumferences are shown in Fig. 3. The remaining 14 children are listed in the Table.

A girl (Case 1) aged 5 years, who had been small-for-dates at birth, was thought by her mother to be mentally slower than her two sibs, though she was attending a normal primary school. Her weight was on the 10th centile. She had somewhat abnormal facies, with wide-set eyes and a short, webbed neck. Chromosome studies showed a mosaic XO/XX karyotype with 20% of the cells containing 45 chromosomes. Another girl (Case 2) had webbing of the neck but normal chromosomes. A girl (Case 3) aged 6 years, who suffered from primary enuresis, had a heavy Esch. coli in-
fection of her urine, but no abnormality of the renal tract was shown on intravenous pyelography.

Case 6 was the second and smaller of dizygotic twins. At 38 weeks’ gestation he weighed 1990 g with a head circumference of 33 cm (25th-50th centile). Apart from mild haemolytic disease due to Rhesus incompatibility, there were no neonatal problems. At 6 months the head circumference had increased to above the 97th centile and thereafter the rate of growth continued parallel to the 97th centile, being 50-8 cm at 20 months of age. He was otherwise well with normal psychomotor development but still below the 3rd centile for height and weight.

Four children (Cases 9, 12, 13, and 14), were below the 3rd centile for weight, and one (Case 11) was for both height and weight. He was well proportioned and no cause was found for his poor growth. Case 9 had bird-like facies, with a bone age of 3 months (Greulich and Pyle, 1956) at a chronological age of 14 months. Karotype was normal and all other investigations were unhelpful. Case 21 was a severely mentally retarded boy of 5 years with short stature, single palmar creases, and curved 5th fingers. Again karotype was normal but this may be a familial condition as the only sib, a girl, is mentally retarded. In neither case has a causative factor been found.

Of the 22 ‘normal’ infants who were small-for-dates at birth, 16 were reviewed at ages ranging from 4 months to 6 years 6 months. 14 of these children have caught up in weight to the 10th centile or above, and one girl, at 6 years of age, has reached the 97th centile (Fig. 4). The 2 who failed to gain weight satisfactorily have been discussed previously (Table I, Cases 9 and 6) and both have abnormalities in addition to their poor weight gain.

Discussion

In the introduction to our previous paper (Bryan and Kohler, 1974) we outlined the practical problems arising from SUA. In the light of our findings we can now focus on two of these, namely (1) the detection of hidden abnormalities that may manifest themselves later on in childhood, and (2) the fate of infants who are small-for-dates at birth.

In any sizeable group of children examined during the first few years of life some occult congenital malformations such as lesions of the cardiovascular system, are likely to be detected for the first time. We believe, however, that the diagnosis of 10 cases of developmental anomaly in 98 children who were ‘normal’ at birth is more than would be expected in a random group. (Single palmar creases hydroceles, and inguinal herniae were not classified as developmental anomalies). For effective comparison a control series would be required, but is not available.

The abnormalities detected perinatally were varied and there was no evidence of any predominant type of malformation. Similarly, the abnormalities discovered at follow-up were of diverse nature, but not surprisingly tended to differ in type and severity from those that were manifest at birth; they were morphologically less conspicuous and less likely to give rise to symptoms. However, some of them such as hemivertebra or webbing of the neck could have been recognized at birth, thus reducing the discrepancy that exists between our results and a hypothetical control series.

In analysing our findings we have also considered certain observations and suggestions made by other authors. Froehlich and Fujikura (1973) who reported on the follow-up examination of 266 infants with SUA draw attention to inguinal herniae, the only abnormality found with a significantly raised incidence. Only one instance was found in our
The missing umbilical artery

Girls

Boys

FIG. 2.—Heights and weights of girls and boys with SUA at follow-up.

HEAD CIRCUMFERENCE

GIRLS

Boys

FIG. 3.—Head circumference of SUA children at follow-up (Tanner, Whitehouse, and Takaishi, 1966).

series. Large angiomatous naevi (i.e., >3 x 1 cm) were found in 4 out of 18 cases (22%) by Vlietinck et al. (1972). None of our cases had such large lesions and only 6 children (8%) had smaller ones. A greater susceptibility to recurrent infections in SUA children has been claimed by Kristoffersen (1969), but it does not appear that this finding has general validity, as it has not been put forward by any other authors. In our series there was no evidence of it.

Retarded motor development at the age of 1 year was found in 3 out of 16 infants with SUA by Fujikura (1964). However, all except 4 children in our series reached their motor milestones as expected. Of these 4, one (Case 21) was mentally retarded and another (Case 17) had transitory benign hypotonia. The other 2 did not walk until near their second birthday, but have subsequently progressed normally.

A fairly widespread belief has existed—probably
as a result of various publications in the 19th and early 20th centuries—that abnormalities of the urogenital tract are selectively associated with SUA. This belief was strengthened by a report of 8 cases of renal tract abnormality found by intravenous pyelography (IVP) in 24 children with SUA (Feingold, Fine, and Ingall, 1964). This remarkably high incidence has not been confirmed in subsequent studies (Van Leuwen, Behringer, and Glenn, 1967; Vlietinck et al., 1972). Only one of our cases, a girl with an Esch. coli urinary infection had an IVP and this proved to be normal. All the other urine specimens were free from protein, cells, and significant bacterial growth. Thus, even if some minor renal tract anomalies did exist, they were not, apparently, causing any trouble and are unlikely to require surgical intervention; we are not convinced of the need for routine IVP in healthy children with SUA.

In the study of small-for-dates infants with SUA we face the question as to whether retarded intrauterine growth is due solely to impaired nutrition or, alternatively, to some teratogenic influence on cellular development. Apart from 2 children who had previously undetected malformations, all the children we reviewed had gained weight well and this suggests that they had suffered only a reduction in cytoplasmic mass due to nutritional deprivation. Had there been a reduction in number of cells, as one might expect if a teratogenic factor were operative, then the poor weight gain might have continued in extrauterine life.

Finally, we must consider the management of the infant who, after thorough examination, appears to have no abnormality other than SUA. Should this child be subjected to extensive investigations or prolonged medical surveillance? Such a follow-up programme would inevitably involve considerable expense in time, money, and, not least, parental anxiety. In agreement with Hnat (1967), we feel that this is not justified. The onus must still lie, principally, on the doctor responsible for the initial examination of the infant: provided that she or he is sufficiently aware of the significance of SUA, few congenital anomalies need escape detection at this time. Of the small number that will not be apparent at birth, delay in detection is unlikely to have a serious effect on the outcome. An exception to this rule is renal tract malformation, as prompt recognition of urinary infection as a presenting sign of underlying anomaly may prevent irreversible renal damage. We would suggest that the family doctor be advised of the significance of SUA and of the importance of early referral of any child with suspected urinary tract infection.

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Correspondence to Dr. H. G. Kohler, The Maternity Hospital, Hyde Terrace, Leeds 2.
The missing umbilical artery. II. Paediatric follow-up.

E M Bryan and H G Kohler

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doi: 10.1136/adc.50.9.714

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