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CONJOINED TWINS—FURTHER OBSERVATIONS

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The occurrence of conjoined twins is so rare that few single individuals have an opportunity of studying more than one example, but since the conjoined twins of Kano (Aird, 1954) were reported it has happened that we have had an opportunity of collecting a small series, and it seems desirable that such additional information as has emerged from it should be briefly put on record.

The title "conjoined twins" seems preferable to such alternatives as "craniopagus," "thoracopagus," "ischiopagus," and so on, because it standardizes the index of the titles of papers and ensures that no titles will be misplaced by those who prepare the great medical indexes.

Since the conjoined twins of Kano, which are referred to above, I have had an opportunity of examining two sets of living twins. The first of these was the Keighley twins, who were so extensively joined at the vertex that operative separation proved impossible, though the brains were separate. Franklin, Tomkinson, and Rohan Williams (1958) report that six craniopagus twins have been recorded in the available literature since 1949, but to these can be added the Keighley twins (which were not recorded in medical literature), and a craniopagus born in Homburg in the early 1950's and illustrated at the age of 3 years in the weekly journal *Neue Illustrierte*. In these German twins there is very extensive bilateral parietal conjunction, and the heads are rotated on each other in such a way that the two left eyes are virtually in line. An unsuccessful attempt was made in the case of these twins to narrow the area of conjunction by increasing pressure of a circular nylon band. In the case of the Keighley twins the sagittal axes diverged also, but only slightly, and in the craniopagus recorded by Franklin *et al.* (1958) the cranial axes are said to be at right angles.

At the kind invitation of Dr. Wijthoff, who delivered the babies, and Dr. Straat, who performed the operative separation, I was permitted to examine and to see the operation performed upon the Dutch twins of Leeuwarden. They were joined between umbilicus and xiphoid process by a tube of skin and a narrow bridge between the two livers.

Particulars of Four Stillborn Pairs

In addition to these we have had an opportunity of examining four pairs of stillborn twins. What was clear in these was that the degree of conjunction of internal organs varies fairly accurately with the extent of external union if the junction is median.

The first specimen, very kindly provided by Mr. N. Alders and Dr. T. K. Owen, showed externally a conjunction from the umbilicus to the lower part of the

sternum. In these twins the thoracic organs showed no conjunction, nor did the stomachs, but the enteric canal was single from the middle of the duodenum to a point on the ileum where one might have expected to find a Meckel's diverticulum—in fact, to the level of the vitelline duct and therefore to the umbilicus. It is of interest that in this case an antenatal diagnosis of conjunction was made by Dr. R. McK. Crichton on the grounds that the x-ray films showed the twins facing each other, the chests closely approximated, and the vertebral columns in a position of exaggerated lordosis.

The second example, kindly sent to us by Mr. J. F. Foulkes, showed external conjunction from the manubrium sterni on each side to a common umbilicus. The external organs manifested a complexity that would have been quite incompatible with surgical separation. Each half of the thoracopagus was provided with a pharynx, larynx, and upper trachea. In the highest point of external conjunction the trachea and oesophagus on one side joined with that on the other to form a single tube, proceeding through a joined diaphragm to reach a single stomach, which in turn emptied into a single jejunum, but the ileum again branching at the site of a single Meckel's diverticulum to pass separately on each side into a normal caecum. The liver was extensively joined, the plane of conjunction running obliquely, so that surgical division of the liver, had it been attempted, would have transected on both sides biliary and venous channels. A single heart was situated in the thorax of one of the twins, and from it two separate aortas proceeded asymmetrically to provide a circulation, one aorta for each side. The arrangement of the cardiac chambers and of the great veins was extremely complex and has not yet been fully analysed. A curious feature, when one remembers that lung alveoli bud out from growing bronchi, was the appearance in the thoracic cavities of a number of separate and isolated patches of collapsed lung tissue which might have been taken for bronchial pulmonary segments.

The third case, provided by Dr. Vivienne Croxford with a prenatal diagnosis by Dr. Ethel Pepper, was that of a conjunction that extended from the mouth to the umbilicus. The whole gastro-intestinal tract was single, and in other respects the organs followed the pattern just described.

The fourth example was provided by Professor J. M. Browne in this hospital. It closely resembled the first of the specimens mentioned above.

In the first and third cases the prenatal diagnosis was not communicated to the parents, the presumption being that the twins would be born dead. This seems a humane rule to follow.

Origin of the Condition

There are very strong reasons for assuming that most, if not all, conjoined twins are uniovular in origin. The grounds for this belief are as follows: (1) Conjoined twins nearly always unite at nearly identical parts of their external surfaces. Probably the great majority of deviations from this rule are relatively minor degrees of torsion or of stretching of the twins *vis-à-vis* each other. This would, for example, explain the slight asymmetry of the axes of craniopagus twins. (2) Conjoined twins are nearly always obviously of the same sex, and are never of opposite sexes. (3) A fairly full genealogical table is available of the descendants of the original "Siamese" twins, Eng and Chang Bunker. These twins are estimated to have had more than 1,000 descendants in the United States of America and twins did not appear again in the family until a few years ago, four generations after the original brothers. These last twins were apparently uniovular, but separate and normal. This very low incidence of twinning in the family very nearly excludes a binovular origin.

The nature of the initial fault is not yet certain. As Hamilton (1954) first suggested, the phenomenon could be explained by the assumption that monovular twins develop, as they do in some other genera, by what is for these genera a normal method of propagation, symmetrical division of the embryonic area of the blastocyst, separation remaining incomplete when conjunction occurs. Fairly strong evidence that conjoined twins arise in an abnormality in one germinal area is the fact that very often one or other of the twins, or both, bear supernumerary rudimentary limbs.

The alternative explanation may be that uniovular twinning is due to the development of two separate embryonic axes in the same blastocyst. Conjunction would then depend upon the overlapping to a varying extent and the fusion of the two axes over an area of varying extent.

The possibility, however, that in at least a few cases conjunction is determined by the fusion of two separate blastomeres cannot be entirely ruled out, though it would require two separate abnormal embryological events: first, rupture of the amniotic partition (nearly all, or perhaps all, conjoined twins lie in a single amniotic sac), and, secondly, a fusion of the embryos. Such a fusion of genetically different embryological tissues would theoretically not be impossible. There are one or two exceptional circumstances which may indeed point to this kind of origin.

1. Conjoined twins are not identical in the same degree as separate uniovular twins. Differences of feature, of stature, and of temperament are nearly always detected. Indeed, the differences between the original "Siamese" twins, Chang and Eng Bunker, were actually caught by a contemporary English political cartoonist, who used a drawing he made of them to illustrate his conception of the bloated capitalist inescapably tied by circumstances to his inebriate wage-slave brother.

2. Conjunction of extensive degree is very commonly associated with asymmetry in the internal organs. We have had one example of a single asymmetrical heart and one example of dextrorotation of the heart, and other misplacements of viscera are frequent.

3. Endocrine asymmetry sometimes occurs too. In the case of conjoined twins previously reported (Aird, 1954) the twin who died after operation had less than her share of adrenal tissue, and her sudden death was suggestive of acute adrenocortical insufficiency. In the case of the Leeuwarden twins the pulse rate after operation showed a

rapid and rather anxious rise with a fall in blood pressure which was corrected by hydrocortisone.

4. An example has been brought to our notice of an even more remarkable endocrine disturbance. The late Major Southwell Oliver provided me with a photograph of adult Chinese twins joined at the pelvis. One of these twins is a normal-looking Chinese adult male of rather handsome appearance, while the other has the slight obesity, the chubby hands, and the features of a eunuch. This twin is taller and heavier than his brother, with characteristically broad shoulders. The precise below- and above-waist ratios in these twins are difficult to estimate, for they are dressed, but it is clear that this ratio in the case of the eunuchoid twins is of characteristic eunuchoid type with long legs and a relatively shorter body. I am informed that the eunuchoid twin had no detectable testes. This seems a very remarkable example of conjunction, and strongly suggests a freemartin phenomenon. I understand that a true freemartin has not been described in man, and this photograph would thus seem to be of historic importance. If my interpretation of the photo-

The Chinese conjoined twins.

graph is correct this must be an example of the development of conjoined twins by the union of initially separate blastomeres. The rarity of this situation, and the almost invariable equality of sexes in conjoined twins, strongly suggest that all but the very smallest possible minority of conjoined twins are uniovular.

5. I have said that in the great majority of cases conjoined twins are united by similar points of their anatomy. Nevertheless, occasionally conjunction is duplicated. The original Biddendon twins are said to have been joined at the hip and at the shoulder, and in pictorial representation they are usually delineated as having precisely that double junction. We have recently had an opportunity of treating surgically a baby patient whose case will be reported in great detail later (Aird and Gavourin, unpublished data). To this child was attached a teratoma which was provided with a peritoneal cavity in continuity with his own, pelvic colon and rectum, and two lower limbs. The teratoma was attached to the host at two separate points—lower abdomen and peritoneum. Other examples of duplicated "conjunction" are on record. It is conceivable, of course, that germinal areas in a single blastomere might overlap and fuse at two separate points, but other explanations are nevertheless possible. Twins fused primarily at one area might conceivably obtain a secondary fusion later, or separate blastomeres might conceivably become attached to each other at two different points.

Discussion

While it is admitted that the weight of evidence points to the great majority of conjoined twins deriving from a single blastomere, the considerations which I have enumerated above should at least keep our minds open to the possibility of a union between separate blastomeres. It would seem desirable to test the blood of conjoined twins for chimerism (the blood changes consequent upon the possession of blood-forming tissue of different types, such as binovular twins of different blood group have sometimes if their placental circulations have communicated). The demonstration

of a chimera would very strongly suggest, though its absence would not of course disprove, an origin from two separate ova and its presence might depend upon a third separate (and binovular) twin.

There have been several recent interesting reports of conjoined twins. Franklin *et al.* (1958) have recorded a craniopagus, and mention that this is the sixth recorded since 1949. This figure obviously does not include the Keighley twins, for these authors state that the last case known to have been born in England was that of Cameron (1928), and it is unlikely to include the Homburg twins, for I do not think these have been described in the literature. Stiggelbout (1958) records stillborn macerated conjoined twins in Kaduna, Nigeria, and notes the relative prominence of that country in the history of conjoined twins. He mentions the twins which I reported earlier (Aird, 1954) and the Port Harcourt twins reported by Roddie in 1957. In fact, Nigeria has been even more prolific of surviving conjoined twins. McLaren (1936) recorded successful separation of conjoined twins born to a Hausa woman in Sokoto, Nigeria, and they are still surviving. In addition to these were the stillborn twins recorded by Bowesman (1940). The survivor of the Nigerian twins that I separated in 1953 is alive and well, a very bright child, normal physically and mentally. Unfortunately her mother died of a placenta praevia in 1957.

The history of the separation of twins can now be fairly clearly documented. The first successful operation was that performed by Professor Doyen in 1902 in Paris for the separation of the Radica-Doodica sisters, one of whom had developed abdominal tuberculosis. The tuberculous sister died but the other survived. It is notable that Professor Doyen had a cinematograph film made of the operation, and by courtesy of the editor of the *Nursing Mirror* I have a copy of a print of two of the frames from the film. This film preceded the opening of the first public cinema theatre by about two years, though it was not the first example of the use of a motion-picture camera in the operating theatre. I am told that Professor Doyen was accused of having shown this film to the general public for profit, and felt impelled to vindicate his honour in a duel.

The second successful separation was done in October in 1912 in the Military Families Hospital at Portsmouth by an unnamed R.A.M.C. officer; one of these twins died four months later, but the other survived and was well in 1954, though we have since lost sight of her.

The third successful operation was that done by Dr. Holm, of Minnesota, on January 8, 1927, and the fourth was by Dr. D. W. McLaren (1936) in Nigeria.

Summary

Some account is given of three sets of living conjoined twins and four stillborn pairs of conjoined twins that have come to our attention.

A possible instance of a freemartin effect in human conjoined twins is illustrated.

The arguments for a monovular origin for the very large majority of conjoined twins are presented, but it is suggested that conjunction may really be due to the union of binovular germinal areas, and it is suggested that chimerism should be sought in conjoined twins.

I am grateful to all my colleagues who have provided me with the material and data on which this report is based. To the names of those to whom we are indebted mentioned in the text must be added Dr. C. Bowesman,

Professor J. McC. Browne, Brigadier J. Houston, Dr. A. D. J. Farquharson, Captain Gavourin, Dr. G. F. Longbotham, Mr. Hujohn Ripman, Dr. J. Rooth, and Dr. A. O. Sasegbon.

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BREAST CANCER

A NEW APPROACH TO THERAPY

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[WITH SPECIAL PLATE]

The best results of surgery and radiation in the treatment of breast cancer show no grounds for complacency, and it is difficult to foresee that any spectacular improvement will come from existing methods which assume that the disease in the early stages is localized to the breast and adjacent lymph nodes. The condition, even when apparently early, may already be beyond effective reach of contemporary treatment, since Cade type 1 patients with a carcinoma in an outer quadrant commonly show involvement of the internal mammary lymph-node chain and presumably more distant sites, and in types 1 and 2 tumour-cell emboli are already being disseminated, particularly during surgery. Goodall (1958) draws attention to the frequency with which the ovary is involved in patients supposedly type 2.

At present the prognosis is largely one of conjecture. A woman under 30 is almost certainly doomed within a year, especially if her breast cancer arises in association with pregnancy or lactation. In the woman of 65 or more prognosis is relatively favourable, but between these ages the outlook is completely uncertain save in the known advanced case. Recent work involving endocrine ablation offers further prospects in the late breast cancer, but even with such radical measures as pituitary ablation or bilateral adrenalectomy with oophorectomy, positive palliative results are obtained only in approximately 50% of cases, and a good remission lasting for a year or longer in rather less than 25% (Illingworth, 1958).

A similar remission in a minority of women in any age group may be obtained with androgen therapy, especially when the symptoms are due to bony metastases (Martin and Hynes, 1954). Eventually, irrespective of the nature of the endocrine therapy employed, the tumour ceases to respond and death of the patient inevitably follows. This is due to an apparent change in the growth pattern of the neoplasm from one of endocrine, presumably oestrogen, dependency to a truly autonomous type, and may arise from a selective breeding out of the oestrogen-dependent cells under endocrine therapy. It occurred to us that it should be possible to prevent the emergence of the