CONJOINED TWINS

A REVIEW WITH A REPORT OF A CASE

S. CYWES, M.B., CH.B., M.MED.(SURG.) (CAPE TOWN), AND C. E. BLOCH, M.B., CH.B. (CAPE TOWN); *Department of Surgery, University of Cape Town and Red Cross War Memorial Children's Hospital, Cape Town*

The term 'conjoined twins' denotes twins who are physically joined together. They vary from two symmetrical individuals joined by minor superficial connections, to monsters represented only by portions of the body attached to each other or to a more completely developed host. Rarely one twin is incorporated in the body of the other as an 'included foetus'.[1]

Potter[2] provides a comprehensive classification, but from a surgical aspect the classification used by Benson *et al.*[3] is considered the most practical.

CLASSIFICATION

A. *Symmetrical*

1. *Thoracopagus, xiphopagus or sternopagus.* The twins are joined at the chest, or sternum, and face each other. The liver is often fused and the wider the attachment, the greater the possibility of a common heart or gastro-intestinal tract.

2. *Omphalopagus.* The twins are joined at the umbilicus and may have fusion of livers or gastro-intestinal tracts.

3. *Craniopagus.* The twins are joined at the heads. Usually the fusion is median but it may be vertical, occipital or parietal. The two brains may be connected by a bridge of neural tissue although they are usually separate.

4. *Pygopagus.* The twins are joined at the sacrum and coccyx, and face away from each other. They may have a single spinal cord at this level and there is often fusion in the perineal region with a single common anus.

5. *Ischiopagus.* These twins are joined by their pelves. The urogenital organs are usually shared.

B. *Asymmetrical*

One twin is normal but the other incomplete and attached as a parasite.

In reviewing the literature on this subject, we found that reports of cases of conjoined twins appear under numerous different titles. We support Aird's[4] plea for the common use of the title 'conjoined twins' in preference to 'craniopagus, thoracopagus, ischiopagus', etc.

INCIDENCE

Conjoined twins are rare. Potter[2] from the Chicago Lying-in-Hospital estimates the incidence as 1 in 60,000 births, and Robertson[5] assessed it at 1 in 50,000. According to Bland and Hammar,[6] the estimated incidence in Central Africa is considerably higher.

The commonest type is thoracopagus—the incidence being 1 in 82,000 births. The rarest type is craniopagus, with an estimated incidence of 1 in 2-4 million births. From the analysis of the records of 117 cases, Robertson[5] determined the incidence of the sites of fusion. 73% of conjoined twins were thoracopagus; 19% pygopagus; 6% ischiopagus; and 2% craniopagus.

Female conjoined twins occur 2 to 3 times more commonly than males in reported series.[7, 8]

According to Aird[9] 'We may expect each year over the world as a whole, the birth of half a dozen or more conjoined twins capable of separation.'

AETIOLOGY

Ambrose Paré[10] in has famous book, *Of Monsters and Prodigies,* written in 1560, states: 'We call Monsters what things soever are brought forth contrary to the common decree and order of nature. So we terme that infant Monstrous, which is borne with one arme alone, or with two heads.' He believed that God punished man's wickedness in this way.

When Leeuwenbeck discovered spermatozoa it was believed that double monsters were due to the fertilization of a single ovum by two spermatozoa.

By the middle of the 19th century, it was fully realized that conjoined twins and other duplicate monsters originated from one ovum. However, it was incorrectly supposed that uniovular twins started to develop separately and fused later. An important finding which renders the 'fusion' theory most unlikely is the observation that conjoined twins are always joined at identical points.[11]

The most widely accepted explanation of conjoined twins postulates that they are the result of fission of a single embryo. If fission occurs at the stage of the inner cell mass, before the embryonic disc has developed, separate twins will develop. If twinning occurs after separation of the embryonic disc, there will be only one amniotic sac, and if the centres of the axial growth are far enough apart, separate twins will develop in the single amniotic sac. If, however, the centres of growth are close together, the intermediate part might be shared and result in conjoined twins.[2, 8, 12]

Petersen[13] claims that there is a defect in the primitive streak or a degree of propinquity between two developing axes on a single embryonic disc which may give rise to conjoined twins.

Withci[14] blames ageing of the ovum. His conclusions were based on the observation that over-ripe frog's eggs give rise to a larger number of monsters.

Dragstedt[15] postulates that environmental factors may play a part, e.g. deficient blood supply or infection. It is known from experimental work that this must play a part, e.g. Stockard[16] in 1921 was able to produce monsters in fish by lowering the temperature at a critical period. The case reports on conjoined twins sadly lack details of possible environmental aetiological factors especially during the first trimester of pregnancy.

It is interesting to note that conjoined twins always differ slightly from each other in size and appearance, much more so than separate monovular twins. Mirror imaging, e.g. *situs inversus,* occurs in 73% of conjoined twins, whereas it is found in only 22% of separate monovular twins.[9]

DIAGNOSIS

While the diagnosis of twins is usually made during the antenatal period, the diagnosis of conjoined twins is rarely

made before labour. Most commonly the problem is recognized either after delivery or when there is obstruction to delivery in the second stage. Dystocia, although rare, has been reported.[17] Conjoined twins are often delivered by the unskilled, under primitive conditions. The Kano twins[9] were delivered by a student midwife. Most have been delivered *per vaginam* and some by caesarean section.

On only two occasions have conjoined twins been recognized during the antenatal period.[18, 19] Gray, Nix and Wallace[20] suggest the following radiological criteria: (*a*) The heads at the same level; (*b*) unusual backward flexion of the spines; (*c*) unusual proximity of the spines; (*d*) no change in the relative positions after movement, manipulation and time.

HISTORICAL REVIEW[3, 9, 21-23]

The earliest recorded case is that of the Biddendon maids born in 1100 in England. They lived for 34 years with a single pair of upper and lower limbs, and a common rectum and vagina. During the 14th century, the Florentine twins were born. According to the bas-relief in the Church of La Scala they had three arms and three legs. During the 15th century, the Scottish brothers lived at St. James' Court for 28 years. They were joined from the waist downwards.

Other instances of conjoined twins, historically famous, are well documented by Aird.[4, 9]

Luckhardt[24] in 1941 gave a full report of the most famous conjoined twins, Chang and Eng Bunker (1811-1874). They were born in Bangkok, Siam, in 1811 as xiphopagus twins, of a half Chinese mother and a Chinese father. At home they were known as the 'Chinese twins'. In 1824 they were taken to North America by Barnum and exhibited to the general public. Thereafter they were known as the 'Siamese twins'. Both married. Chang had 10 children and Eng 9. Illness of one did not affect the other. Chang drank heavily, but Eng never suffered from his brother's debauches. They died at the age of 63 years. It was found that they were joined by a band of liver with communication of the portal circulation.

Surgical Separation

This has been attempted on several occasions but survival of one twin is unusual and survival of both a rarity.[25] To date there have been reports of only 12 successful separations with survival of both twins.

The first recorded attempt at separation was made in 1495—craniopagus twins—in an attempt to save the one twin after the other had died. This proved unsuccessful.

In 1689 Farius at Basle made another unsuccessful attempt at separation. In 1690 Konig gradually tightened a ligature around the connecting band of skin, cartilage and muscle of xiphopagus twins in infancy. This finally cut through and both twins lived.

In 1902 Professor Doyen in Paris performed the first successful operation for the separation of thoracopagus twins. They were the Radica-Doodica sisters, one of whom had developed tuberculosis. The tuberculotic sister died soon after separation, but the other survived.

In 1912 an unnamed RAC officer at Portsmouth, performed the second successful separation—ischiopagus sisters. The one died at 4 years, but the other lived.

In 1927 Holm[26] in Minnesota successfully separated xiphopagus twins. In 1953 Grossman *et al.*[27] reported the first successful separation of craniopagus twins. One however, died one month after separation.

Other successes have been reported by McLaren,[28] Reitman *et al.*,[29] Ochsner,[30] Dragstedt,[15] Wilson and Storer,[23] Petersen,[13] Koop,[25] and in *Time* magazine.[31, 32]

Most successes have been obtained in xiphopagus twins and least in the craniopagus type, although Baldwin and Dekaban[33] successfully separated twins joined by the brow with a single right anterior cerebral artery, and Voris *et al.*[34] reported another success.

The success of surgical separation depends on the extent of the union. Twins joined by skin, subcutaneous tissue and cartilage, are more likely to be successfully separated. The success rate drops markedly if a vital organ is shared. In very extensive union there may be no prospect of separation, e.g. those who share a single pelvis, a length of trunk or one pair of limbs.

Aird[9] states 'However great the risk of the death of one or both children, it would seem that operation should nearly always be undertaken if the children are known each to have a full complement of the organs and tissues necessary for separate life'.

Thus pre-operative assessment and planning are of the utmost importance. Except in emergencies such as the case reported below, a series of special investigations should be undertaken to detect which organs are being shared and whether the separation is feasible.

Detailed descriptions of the investigations and preparation before separation, and of the actual separation, are outlined by Aird[9] for thoracopagus twins; by Koop[25] for pygopagus twins; and in a symposium by O'Connell *et al.*[22] for craniopagus twins.

However, not too much time should be wasted in doing these investigations because, not only may the babies die in the interim, but as a leading article in the *British Medical Journal*[21] so aptly states, 'This delay must affect the psychological development of the children, and in addition the parents must be upset by the long period of uncertainty whether the children will live or die, coupled with seeing them in hospital in their pitiful condition'. With the advances in paediatric surgery in all its spheres to its present state all the necessary investigations can be carried out soon after birth, and separation performed early in life.

Two anaesthetic and two surgical teams are required for the operation. Special attention should be given to the draping and towelling.

Postoperatively it is important to bear in mind the possibility of the development of adrenal insufficiency in one twin—probably the cause of death in one of Aird's Kano twins.[9]

In the following case report we shall describe the surgical separation of omphalopagus twins who presented as an emergency with ruptured omphalocoeles.

CASE REPORT

On 16 March 1964 Mrs. S.A., aged 28 years, was delivered vaginally of conjoined twins at Groote Schuur Hospital, Observatory, Cape Town. This was her first confinement.

Obstetrical History

Her last menstrual period was on 29 June 1963 with the expected date of delivery on 6 April 1964. Her blood group was B Rh negative and that of her husband O Rh positive. At no stage during the pregnancy were any antibodies exhibited. Her Wassermann reaction was negative and her haemoglobin 11.5 G/100 ml.

During the early months of pregnancy she suffered from nausea and vomiting for which she was given 5 mg. of 'Stelazine' (trifluoperazine) twice daily and 'ancoloxin' (25 mg. of meclozine HCl plus 50 mg. of pyridoxine HCl) twice daily. These drugs were taken for a period of 3 months.

At no stage during her antenatal attendances were twins suspected, although she was considered to be at full-term on 11 March 1964.

Past History

In 1951 she was treated for pulmonary tuberculosis. The radiographs of her chest during pregnancy showed no evidence of residual disease. In 1959 she had a nephropexy for a ptosed right kidney. In 1961 a benign osteochondroma of her left iliac crest was removed.

There was no history of twins in the husband's family, but unknown to Mrs. S.A., her mother had had a miscarriage of a 2½-month twin pregnancy many years ago.

On 15 March 1964 her membranes ruptured and she was admitted to the labour ward. The liquor was persistently stained with meconium. The following day there appeared to be no progress in the delivery, despite a fully dilated cervix, and forceps were applied under pudendal block. It was then noted that instead of an umbilical cord, small bowel protruded from the umbilical area of the delivered live baby. The breech of an unsuspected twin then presented and a stillborn baby was delivered. A single placenta was delivered.

The babies and the placenta were immediately transferred to the operating theatre of the Red Cross War Memorial Children's Hospital.

The stillborn baby was found to have a large ruptured omphalocoele with evisceration of the liver and most of the gastro-intestinal tract. The live baby also had a ruptured omphalocoele with evisceration of a few coils of small bowel. Neither baby had an anus. The live twin had a dislocated right hip.

The two babies, together with the mass of eviscerated bowel and the common placenta were placed on the operating table and the live baby was intubated and anaesthetized (Fig. 1). After the coils of bowel had been disentangled, it was established that the terminal ileum of the live baby was joined to the terminal ileum of the stillborn baby about 7 cm. from the ileocaecal junction (Fig. 2). The bowel of the stillborn baby had a dusky appearance.

The live twin was detached from the other by dividing the ileum at its junction with the ileum of the stillborn baby. The placenta and stillborn baby were removed and the live baby was cleansed and draped. After the defect in the anterior abdominal wall had been enlarged by an incision upwards in the midline, it was found that the liver, spleen, pancreas, stomach, duodenum and small bowel as far as the terminal ileum were normal. There was no colon. Both kidneys appeared normal.

At the lower end of the abdominal wall defect, a small opening was found, leading into a large cloaca. A diagrammatic representation of the structures which entered the cloaca is shown in Fig. 3. Two uteri, each with a fallopian tube and ovary, entered separately. A large, cystic swelling which occupied the left side of the abdomen entered the cloaca on the left. The left ureter entered into this bladder-like structure. The right ureter opened into the cloaca on the right. The cloaca had one opening onto the perineum.

The bladder-like structure was excised after division of the left ureter, which was found to be duplicated up to the pelvis of the kidney.

The right ureter was divided at the point where it entered the cloaca. The distal 5 cm. of ileum was detached with its blood supply and a conduit fashioned into which the ureters were implanted. The distal end of the conduit was brought out through an incision in the right iliac fossa. The terminal ileum was brought out through an incision in the left iliac fossa. The cloacal defect was reconstructed to serve as a genital tract. The defect in the anterior abdominal wall was repaired (Fig. 4). 50 ml. of blood were lost during the operation.

The possibility did arise, during the operation, of utilizing the colon of the stillborn twin. However, the colon already had a dusky appearance of non-viability.

The live baby weighed 5 lb. 9 oz.

The postoperative course was surprisingly uneventful. The ileal bladder functioned well. On the second postoperative day the ileostomy started to function and then oral fluids were started. At first there was considerable delay in gastric emptying but this gradually improved. Intravenous supplementation was required to cope with the excessive ileostomy losses during the first 3 postoperative weeks, and excoriation of the skin around the ileostomy required energetic treatment. X-rays confirmed the presence of a dislocated right hip and showed some separation of the symphysis pubis. The histology of the cyst-like structure confirmed the impression that it was a urinary bladder.

Postmortem Findings on the Stillborn Twin

The body was that of a stillborn infant weighing 6 lb. 1 oz. Low slung ears, a huge omphalocoele with ruptured sac and attached umbilical cord, evisceration of the liver and most of the gastro-intestinal tract, congenital dislocation of the hips

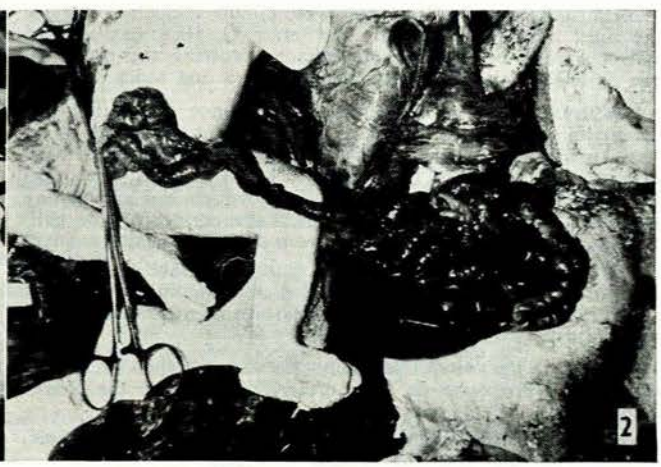

Fig. 1. Twins still covered with vernix on operating table with the live one anaesthetized. The mass of eviscerated bowel and liver plus the single placenta can be seen lying between the two babies. The clamps are applied to the umbilical vessels of the live twin and to the cord going to the placenta.
Fig. 2. The ileum of the live twin joins the ileum of the stillborn infant. Note the dusky appearance of the bowel of the stillborn baby.

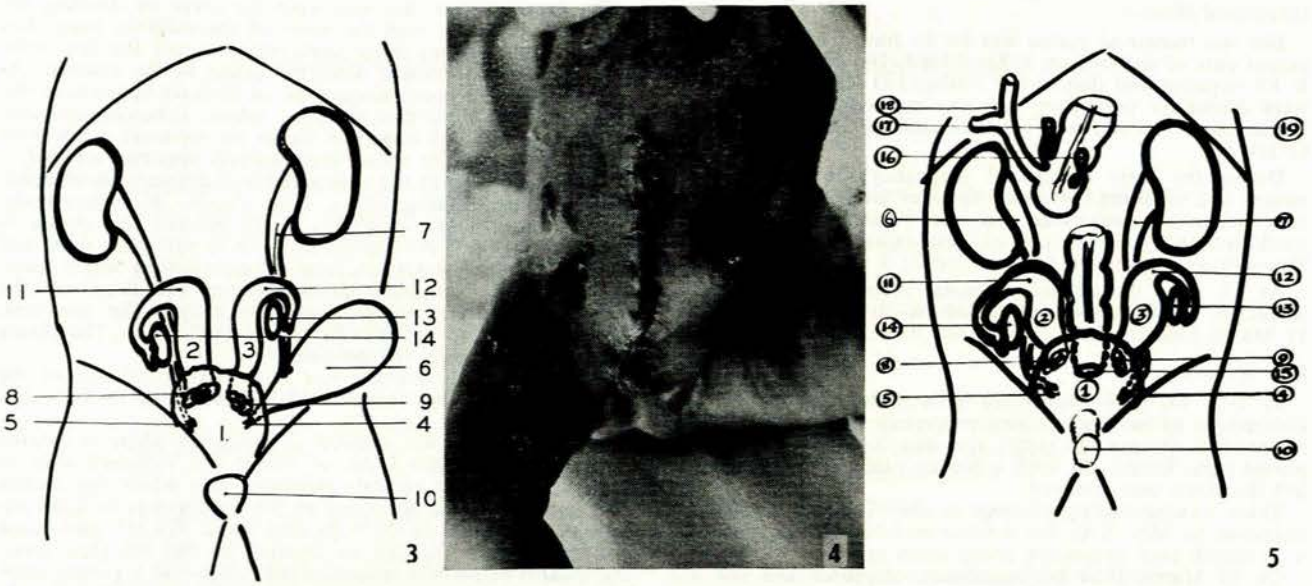

Fig. 3. Diagrammatic illustration of the genito-urinary findings at operation on the live twin: 1. Cloaca; 2 and 3. Uteri; 4. Bladder opening into cloaca; 5. Right ureter opening into cloaca; 6. Bladder; 7. Duplicated left ureter; 8 and 9. Uteri opening into cloaca; 10. External single opening of cloaca onto perineum; 11 and 12. Fallopian tubes; 13 and 14. Ovaries.
Fig. 4. The ileal urinary conduit can be seen in the right iliac fossa and the ileostomy in the left iliac fossa. The defect in the anterior abdominal wall has been repaired.
Fig. 5. Diagrammatic illustration of autopsy findings in the stillborn twin: 1. Cloaca; 2 and 3. Uteri; 4 and 5. Ureteric openings into cloaca; 6 and 7. Ureters; 8 and 9. Uteri opening into cloaca; 10. External single opening of cloaca onto perineum; 11 and 12. Fallopian tubes; 13 and 14. Ovaries; 15. Rectal opening into cloaca; 16 and 17. Two appendices; 18. Ileal junction to ileum of live twin: 19. Colon.

and failure of fusion of the symphysis pubis, were noted. There was significant scoliosis of the spine with convexity to the right and associated deformity of the chest. Just below the defect in the abdominal wall a fistula was found which led to a cloaca. The external genitalia were female with only one external opening in the perineum. No anal opening could be detected.

A large, complete placenta, with its membranes, was attached to the umbilical cord. There was only one chorion and one amnion and the umbilical cord showed 2 umbilical arteries. Histology of the placenta showed budding of syncytiotrophoblasts, excessive fibrosis of villi and numerous placental infarcts in greater than normal amounts.

Cardiovascular system. The heart and its arterial and venous connections were normal but for a patent foramen ovale and widely patent ductus arteriosus. The heart weighed 18 G and the histology was normal.

Respiratory system. The lungs were minute atelectatic organs, the right weighing 10 G and the left 5 G. Histology showed atelectasis and extensive haemorrhage involving the bronchial tree as well as the interstitial tissue, pleura and septa.

Gastro-intestinal tract. The site of severance of the ileum of the living infant from the ileum of this infant was noted to be 7 cm. from the ileo-caecal valve (Fig. 5). Two appendices, each with its own mesentery, were attached to the caecum. The colon ended by way of a fistula into a cloaca, which had an opening into the abdominal wall and one onto the perineum. The gall-bladder, bile ducts and pancreas were macroscopically normal.

A small cyst was present on the anterior surface of the right lobe of the liver. Detailed histological examination of the gastro-intestinal tract was not undertaken, since the organs were kept as pathological specimens.

Genito-urinary tract. Externally the kidneys showed a mild degree of hydronephrosis. The ureters opened into the cloaca on each side. No bladder was found (Fig. 5).

Two uteri, each with its own fallopian tube and ovary, opened separately into the cloaca.

Reticulo-endothelial system. The thymus was large and histologically normal. The other viscera showed no macroscopic abnormality.

DISCUSSION

To the best of our knowledge only 2 pairs of conjoined twins have been encountered in South Africa previously to the case reported here. The first, the Edendale twins, was reported by Tibbet in 1951.[11] No separation was attempted. The second was a pair investigated in Johannesburg in 1962, but they died before separation could be attempted.

We have been unable to find a report of a case similar to ours, presenting with ruptured omphalocoeles necessitating emergency surgery. However, 2 cases of conjoined twins born with unruptured omphalocoeles have been reported. In 1957 Wilson and Storer[23] reported on conjoined twins with omphalocoeles which were repaired, but the babies died before they could be separated. The other report by Roddie[35] was also in 1957. These twins died.

Two pairs of conjoined twins who had the ileum of one joining the ileum of the other and but a single colon, have been reported—by Tibbet[11] in 1951 and by Spencer in 1956.[36]

SUMMARY

The literature on conjoined twins is reviewed. A practical classification is given and the aetiology is discussed. A brief historical sketch of conjoined twins and their separation by surgery is given.

A case of omphalopagus twins with ruptured omphalocoeles necessitating emergency surgery is reported and discussed.

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