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Xiphopagus Twins

REPORT OF OBSTETRIC AND SURGICAL MANAGEMENT OF A CASE

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The term conjoined twins is applied to twins who exhibit some degree of fusion of their bodies. This fusion occurs at various sites and may involve the sharing of some vital internal organ or organs. Surgical separation has been attempted on numerous occasions, but up to the present time only about 12 cases have resulted in the survival of both twins (in other cases one of the twins has survived separation). The twins are not infrequently stillborn or die in the neonatal period, possibly due to prematurity or associated congenital abnormalities. In some the fusion is such that separation would be impossible and it may be necessary to sacrifice one twin; this is especially the case in those joined at the head, as in the cases of Wilson and Storer (1957) and of Spencer (1956). The term "Siamese twins" is used as a synonym for this condition and originated with the famous brothers Chang and Eng, who were born in Bangkok in 1811 and later travelled around America with a sideshow.

The incidence is difficult to ascertain, as not all cases have been recorded, although more are being reported in the literature. Robertson (1953), who analysed Tartuffi's figures, assessed it at about one in 50,000 births. The incidence in Central Africa is probably higher. This is the third known case to have been born in Harare maternity hospital since its opening in

March, 1950, during which period there have been 41,826 deliveries—an incidence of approximately one in 14,000. The previous case was reported by Sladden (1959) and the first one was not reported and was stillborn (Husband). The higher frequency is not altogether surprising, since the incidence of all twins in African women is higher (one in every 25 deliveries in this hospital, compared with the usually quoted figure of one in 80). A large number of reported cases have occurred in the coloured races, African and Asiatic, the highest proportion coming from Nigeria (Nvule and Timmis, 1945; Bowesman, 1940).

FORMATION

Conjoined twins are monozygotic and occur as a result of division and incomplete separation of the inner cell mass. Why this should occur is unknown (Hamilton, 1951). Aird (1959) suggests the possibility that in a few cases conjunction may be determined by the fusion of two separate blastomeres and puts forward certain circumstances which may point to this kind of origin. Various types occur and are classified according to the site of fusion of the two bodies. Sheares (1941) classified them in the following manner:

(1) *Diplopagi*—double monsters in which the components or component parts are equal to and symmetrical equivalents of one another.

- (a) Divided at their cephalic poles only.
- (b) Divided at their caudal poles only. (This group includes *craniopagus*.)
- (c) Fused twins divided approximately at both poles with more or less fusion of the thorax, abdomen and pelvis.
 - (i) *Thoracopagus*: joined only in the thoracic and upper mid-abdominal region.
 - (ii) *Xiphopagus*: joined only in the region of the umbilicus and up to the xiphoid cartilage.
 - (iii) *Pygopagus*: joined by the postero-external aspects of the sacrum.
 - (iv) *Ischiopagus*: joined by fusion of the pelvis end to end.
 - (v) *Rachipagus*: joined by fusion of the vertebral columns.

- (vi) Ectopagus: connected along the sides of the bodies.
- (vii) Prosopothoracopagus: joined by the faces, necks and chests.

(2) Unequal and asymmetrical monsters, one component of which is smaller than and dependent upon the other (autosite and parasite).

Most of the successful separations have been in xiphopagus twins, where the junction has been of skin, subcutaneous tissue and cartilage and possibly a small amount of liver tissue. Cases have been reported by McLaren (1936), Reitman *et al.* (1953) and Wilson and Storer (1957). Koop (1961) has reported the successful separation of pygopagous twins, who are now nearly four years of age. (Ochsner (1957) mentioned a similar case.) Full investigations of the twins should be carried out, if possible, to determine the presence of shared organs, and this involves radiological and chemical tests; the efficacy of these was well shown by Aird (1954). Wilson and Storer (1957) reported three cases of their own and reviewed the literature of surgically treated cases up to that time.

CASE REPORT

The patient was a primiparous African female, aged about 30 years (Hosp. No. 3402 Mat.). Her last menstrual period was early in October, 1960, giving an expected date of confinement in mid-July, 1961.

She first attended the ante-natal clinic on 23rd March, 1961, when the duration of pregnancy was about 24 weeks. She had had no previous pregnancies and there was no relevant previous medical history.

Physical examination and laboratory estimations revealed no abnormality. Her height was 5 ft. 4in., weight 149½ lb.; blood pressure was 120/60. There was a trace of albumin in the urine, but no demonstrable oedema. The height of the fundus was equivalent to a pregnancy of 24 weeks. The presentation was a breech and the foetal heart was heard. Vaginal examination revealed an adequate pelvis with no abnormality.

Subsequent attendances were made at monthly intervals. On 26th May, 1961, she was admitted to the maternity hospital with moderate oedema but no albuminuria and a blood pressure of 160/110. The height of the fundus was that of a 34-week pregnancy. She was treated with bed rest, sedation with phenobarbitone (65 mg. three times daily), Reserpine tablets (0.25 mg.

three times daily) and a low-salt diet. The oedema rapidly subsided and the blood pressure was reduced to a resting level of 150/90.

On 5th June, 1960, at 6 a.m., labour pains commenced. At 11.45 a.m. she was showing signs of bearing down, and a vaginal examination was performed. The pelvis was found to be adequate, the cervix was fully dilated and four limbs were present in the vagina. A general anaesthetic was given and a further vaginal examination was performed. This confirmed the presence of four lower limbs, not covered by membrane, presenting through the cervix. A diagnosis of locked twins was made and an attempt was made to disimpact the twins by pulling gently on one pair of legs while pushing up the other. This was found impossible and an attempt was now made to pull on all four feet and deliver the infants together. The manoeuvre also failed. A hand was now passed up into the uterus between the ventral surfaces of the twins. This led to the discovery of a bridge of tissue joining the twins above the level of the umbilicus. One foetal heart was felt to be beating.

A diagnosis of conjoined twins was made and it was decided to deliver the mother by caesarean section. After a lower segment incision had been made one breech was extracted through the incision and delivery was again prevented by the junction with the other twin. Delivery was finally accomplished (both as breeches) by making a further vertical midline incision through the upper cut edge, making a T-shaped incision. The uterine wounds were then closed in layers. The total blood loss was 15 ounces (430 ml.). The placenta was normal in size and weight and had a single cord.

After delivery, the first infant was found to be stillborn. The second infant cried within five minutes and needed only routine resuscitative measures (administration of oxygen and extraction of mucus). The infants were found to be joined from the xiphisternum to within 1 cm. of the umbilicus. There was a single common umbilical cord.

The mother's post-operative progress was uneventful and she was discharged on the fourteenth post-operative day.

THE TWINS (Fig. 1)

Xiphopagus twins were delivered, the one being stillborn. The live twin was pink and

breathed and cried normally, whilst the dead one was cold and blue; the cyanosis and mottling did not extend over the skin bridge.

They were joined by a bridge of tissue extending from the xiphisternum down to the single umbilicus present; the length of fusion was approximately $3\frac{1}{2}$ inches. There was a single umbilical cord entering the umbilicus. There appeared to be slight cartilaginous union on palpation of the upper end of the fused area, but no other tissues or organs, apart from skin and subcutaneous tissue, could be felt.

The conjoined weight was 7 lb. 10 oz. (3.5 kg.) and the crown-heel lengths were 16 and 17 inches respectively. They were both males and lay facing each other. Both appeared normally formed, with no evidence of any other obvious abnormalities. An X-ray showed no bony junction or obvious tissues in the bridge. Gas was present in the bowel of the live twin, but not in that of the dead one, who appeared to have a pneumothorax (incurred during attempts at resuscitation) (Fig. 2).



Fig. 1—This shows the junction quite clearly with the single umbilical cord entering the "common" umbilicus. The infant on the right is the common one.

Approximately $1\frac{1}{2}$ hours after delivery separation was undertaken. A general anaesthetic was given by Dr. L. L. Theron. An elliptical incision was made around the skin bridge, slightly over to the "dead" side; there was minimal bleeding. The umbilical cord entered the skin at the caudal end of the bridge and divided just under the skin into two groups of vessels, one running to the liver of each twin. The peritoneal cavities were quite distinct and separated by a layer of subcutaneous tissue. The liver of the live twin was normal, whilst the other was blue and mottled. The organs were quite normal and in their correct positions (no evidence of mirror image). At the upper end of the bridge there was union between the xiphisternal cartilages and this was divided and the dead twin removed. Peritoneum, subcutaneous tissue and skin were sutured. There was minimal blood loss and the whole procedure took less than 30 minutes.

The weight of the twin was 3 lb. 15 oz. (1.8 kg.). The infant was put in an incubator and seemed in reasonable condition. The fol-

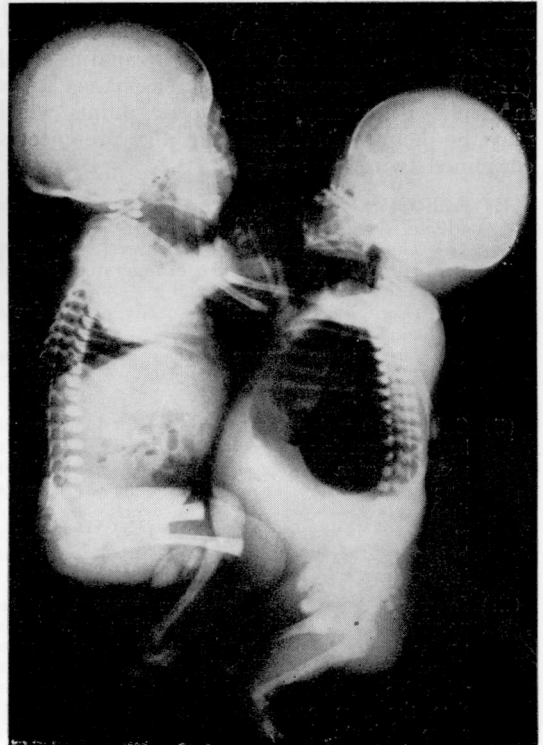


Fig. 2—X-ray of the twins, showing the presence of gas in the bowel of the live infant. This also shows how superficial the junction appears.

lowing day it had passed urine and meconium; X-ray of its chest appeared normal. It appeared, however, to have a cerebral cry. That evening (6th June, 1961) it collapsed for a while, but recovered and appeared to be progressing satisfactorily. Unfortunately it collapsed again in the early hours of 7th June, 1961, and died approximately 36 hours after birth.

AUTOPSY

The stillborn infant had unexpanded lungs and no other abnormality apart from a slightly enlarged cystic right kidney.

The second infant showed atelectasis and no other abnormalities were demonstrable. There was no evidence of gross intracerebral damage.

DISCUSSION

Diagnosis

The diagnosis is usually made during labour at the time of delivery. However, it is occasionally made during pregnancy from radiological studies. This is easier when there is some bony fusion, as with Wilson and Storer's (1957) case and also that of Franklin *et al.* (1958), who demonstrated craniopagus twins in triplet pregnancy. Diagnostic points which may be helpful in suggesting the presence of conjoined twins are:

- (i) Radiological evidence. Gray, Nix and Wallace (1950) suggest the following criteria:
 - (a) The heads are at the same level and plane.
 - (b) There is an unusually exaggerated lordosis of the foetal spines.
 - (c) There is an unusual proximity of the foetal spines.
 - (d) There is no change in the relative positions of the foeti after movement, manipulation or time.
- (ii) Only one amniotic sac is present. If a second bag of membranes can be felt vaginally the presence of a double monster can be dismissed. The presence on vaginal examination of more than one pair of limbs uncovered by intact membrane is suggestive. (Rupture of the second bag of waters before delivery of the first infant is very rare.)
- (iii) In cases where two breeches present in the pelvis, the impossibility of moving one foetus up or down relative to the other suggests some degree of fusion. This is to be distinguished from locked twins (double breech) by the fact that the sign can be elicited long before the stage of impaction of the breeches in the pelvis is reached.
- (iv) Further vaginal examination, usually necessitating a general anaesthetic, may demonstrate the bridge of tissue between the twins.

Obstetric Management

Many cases have been delivered *per via naturales*, but there is frequent difficulty. Roxburgh (1946) reported obstructed labour from this cause; and although Aird (1954) states that dystocia is uncommon, he mentions another case, and we have found, on reviewing the literature, that many cases have caused difficulty and have necessitated either destructive procedures (Sladden, 1959; Husband, 1961) or caesarian section, as in the present case. Quite frequently premature labour occurs, so facilitating delivery due to the small size of the twins. Roddie (1957) points out the advantage of breech delivery. This is most favourable since all four legs are accessible and traction on these may result in delivery of both trunks simultaneously, provided the conjoined bulk is not too large nor the mother's pelvis too small. If there is sufficient independent mobility of the heads they may then be delivered one after the other, with or without the help of forceps.

In the event of difficulty in delivery of the conjoined trunks, particularly if the monster is large or the mother's condition does not permit intrauterine manipulation for fear of rupture of the uterus, perforation of the abdomen and evisceration of one foetus will reduce the conjoined bulk. Dystocia during delivery of the heads may be dealt with by perforation of one skull or craniotomy.

Intrauterine separation of the twins by division of the band of tissue has been described by Dwyer. Ripman and Williams (1959).

The most satisfactory method of delivery, however, is probably by the abdominal route, since it leaves the mother with an undamaged birth canal and gives the best chance for survival of one or both twins following surgical separation. With a large monster or a wide area of fusion it may be necessary to extend the uterine incision in order to extract the infants through it.

This pair of xiphopagus twins presented the interesting situation of having one dead and one alive. Under these circumstances, of course, it was impossible to carry out any involved investigations, as it was necessary to separate them at the earliest time. It was obvious that there was virtually no cross circulation and examination suggested no sharing of major organs or tissues. Actual separation was an extremely simple procedure and it was unfortunate that the second twin failed to survive, as we had high hopes for its continued survival.

Aird (1954) stated that however great the risk of 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." This applies more urgently, of course, when one twin appears able to survive whilst the other appears weak and obviously incapable of survival, as in Spencer's (1956) case, or may be dead, as in the present case.

SUMMARY

A case is described of conjoined twins diagnosed during labour and delivered by caesarean section. One twin was stillborn and the other died 36 hours after surgical separation.

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Addendum

Since this paper was written a further case, of thoracopagus twins, has occurred in Southern Rhodesia. Delivery was spontaneous by the vaginal route and both infants died shortly after birth.

The African orderly who sent the case to Dr. Heber states: "We see quite a lot of these cases in the African dispensary!"