THORACOPAGUS

by

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Of all congenital malformations, conjoined twins are without doubt the most spectacular. Ballantyne (1902) wrote "few medical men are called upon to conduct a case of labour in which the product consists of double monster and even obstetricians of considerable experience will see no more than two or three confinements so complicated in a life time. Nevertheless, a practitioner may find himself any day face to face with such an emergency." As surgical advances have made separation of some of these twins possible, this subject has assumed even greater importance.

Conjoined twins must have occurred throughout history, but apparently the first case was not recorded until 945 A.D. when an omphalopagus from Armenia was exhibited in Constantinople. The famous twins, Eng and Chang, who gace rise to the name 'Siamese Twins,' were born in 1811 in Smut Song Gram, west of Bankok, Siam (Thailand). When they were 18, they travelled to America and later went to Europe and many other countries and appeared before royalty and at public exhibitions. They lived normal lives and married North Carolina Quaker sisters who bore their children—Eng 11, and Chang, 10. Illness did not affect them at the same time, and temperamentally they were different. Chang was the more aggressive but physically the weaker; he drank. Eng, more stolid and not given to drink, had to support part of his brother's weight during the drinking bouts and also for the last five years of their lives after Chang developed a right-sided hemiplegia due to a cerebrovascular accident. In 1874 they died. Chang died first from an embolus when Eng was asleep and it is believed that Eng's death was due to sheer fright when he awoke to find his brother dead beside him (Callahan, 1966).

Conjoined twins have been reported from time to time in the Indian literature. Some of these reports have been summarised in the table. In all these instances both the babies were females and had been stillborn. Except for one instance of omphalopagus all were cases of thoracopagus.

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Received for publication on 28-10-1968.

Case Report

Phoolmati, 30 years, was admitted to the U.I.S.E. Maternity Hospital, Kanpur, on January 22, 1968, at 1.30 P.M. as an un-

TABLE I

Author (Year)	Type	Stillborn or liveborn	Weight	Method of delivery
Bhargava et al (1960-61)	Thoracopagus	Stillborn	6 lb. 14 oz.	Division of bridge & decapitation.
Shah (1960-61)	,,	,,	10 lb. 6 oz.	Embyotomy.
Soonawalla & Patel (1960-61)	,,	,,	7 lb. 12 oz.	Decapitation
		,,	8 lb. 2 oz.	Decapitation
Chaphekar & Chapekar (1963)	,,	,,	11 lb.	Assisted delivery p.v
Varghese (1968)	Omphalopagus	,,	11½ lb.	Assisted breech p. v
Gupta & Wakhaloo (1968)	Thoracopagus	,,	1700 G.	Embryotomy.

booked case of full term pregnancy with labour pains. There was no history of exanthematous fever or of drug intake during pregnancy. She had delivered twin male babies 3 years before and both are doing well.

The general and systemic examination did not reveal any abnormality. Her blood pressure was 126/78 mm. of mercury and pulse was 86 per minute. The abdominal examination revealed it to be unduly enlarged. It was tense on palpation and multiple foetal parts were felt. Foetal heart was audible. Initial vaginal examination revealed the os to be two-fifth dilated and cervix to be half taken up. The presenting part was foot and the bag of membranes was tense. The pelvis was adequate. Hind water rupture was done and about 200 c.c. of liquor amnii was drained off. Vaginal examination was repeated after 4 hours and the os was now found to be fully dilated and cervix fully taken up. The patient was having strong pains. The feet were lying in the vagina Breech extraction was tried but there was no descent of the presenting part. The patient was then anaesthesized and one hand was put inside the uterine cavity. Another pair of feet were found and these were taken out. Traction was applied to all four feet but again there was no descent of the presenting part. Further examination revealed a conjoined twin. At this stage the patient was taken to the operation theatre and a lower segment caesarean section was done and a thoracopagus was delivered. Attempts at resuscitation were ineffective. Both the babies were females and together weighed 4.7 kg.

Autopsy findings:

Externally, the union was found to extend from the level of the third costal cartilage to just above the umbilicus. (Fig. 1) Apart from the following abnormalities the viscera of these babies were separate and normal.

(a) Heart: A single heart was shared by these babies. There were two ventricular chambers, but there was only one chamber which represented the atria. The superior and inferior vena cavae, aorta and pulmonary veins were separate for both the foetuses.

(b) The diaphragm of these babies was fused near the central tendon.

(c) A single liver with one gall bladder was shared by both the foetuses.

Discussion

The incidence of conjoined twinning is difficult to ascertain, as this is a very infrequent anomaly. The following is the estimate of incidence given by various authors.

Authors	Incidence
Mortimer and Kirshbaum (1942)	1:283,000
Potter (1961)	1:50,000
Freedman et al (1962)	1:80,000
Lu and Lee (1967)	1:25,367

It has been observed that females outnumber males by 3:1 in these cases of conjoined twinning.

Embryology

Aird (1959) suggested that, as both twins are of the same sex and the

union is at similar parts of their external surface, the majority are uniovular. So far, all conjoined twins have been found to be of the same blood group. It is believed that the anomaly is initiated before the end of the second week of gestation, after the formation of the embryonic disc. If, at this time the two centres of growth are formed too close together, incomplete separation of the embryos might result. Such joined twins may be symmetrical and each apparently a complete individual; on the other hand, although symmetrical, each of the joined twins may be less than a complete individual. Joined twins may be asymmetrical, have a mass of tissues in which parts of another individual may be discerned (a parasitic twin), is attached to the surface of, or lies within (included twin) the normal child (Farris and Bishop, 1950).

Alternatively, it has been suggested that an initially single embryo may undergo incomplete fission. This might be the more probable explanation in the cases of twinning in which each of the pair is considerably less than a complete individual, the fore —or the hind ends of a single body being duplicated (Willis, 1962).

It seems unlikely that a secondary fusion of the initially separated embryos can explain the phenomenon of symmetrical congenital twinning (MacGregor, 1960).

This is not a genetic or familial condition and its recurrence in a family or in the children born to separated twins has not been reported. Chang and Eng's descendents probably now number a thousand and no recurrence of their handicap has been recorded (Callahan, 1966).

Terminology

The following varieties of conjoined twins have been reported.

Craniopagus	joined by the head
Thoracopagus	„ „ chest
Xiphipagus	„ „ lower sternum
Omphalopagus	„ at the naval
	„ posteriorly in the sacral area
Ischiopagus	„ at the pelvic outlet
Dicephalous	these have two heads, two necks, two or four upper limbs and a body showing some signs of duplication.
Syncephalous:	there is fusion in the region of the heads and upper part of the trunk, but separation in the lower part of the trunk and lower limbs.

Robertson (1953), from an analysis of 117 cases collected from the literature by Taruffi (1891-4), found that 73% were thoracopagus, 19% were pygopagus, 6% were ischiopagus and 2% were craniopagus.

Ante-natal diagnosis and management

It is often difficult to establish the diagnosis of conjoined twins before delivery. In cases of twins where the delivery is difficult, exploration of the uterus with the entire hand may aid in the diagnosis. The abnormality may be suspected in cases of twin pregnancy when both babies present as breech or vertex and the straight film of the abdomen reveals:

(a) twins facing each other,
(b) both foetal heads at the same level,
(c) the thoracic cages of the foetuses are together,

(d) unusual backward flexion of the cervical spine (Gray *et al* 1950).

Gray *et al* (1950) and Mathew (1956) have reported correct antenatal diagnosis of congenital twins. Franklin (1964) states that the diagnosis is possible and desirable because it may influence the choice of delivery. However, Kreutner *et al* (1963) feel that antenatal diagnosis is difficult and Freedman *et al* (1962) suggest that the frequent presence of hydramnios makes the radiographic findings inconclusive.

The size of the conjoined twins render their passage through the pelvis very difficult and in certain circumstances even impossible. If diagnosis is not made, rupture of the uterus is likely as a result of obstructed labour or it may take place during internal manipulations. This is to a certain extent prevented as the foetuses are small or premature or macerated and mostly present by pelvic presentation.

Vaginal delivery without destructive operations have been reported by Lu and Lee (1967)), Roddie (1957) and Stiggelbout (1958). With small infants and roomy pelvis it may be possible to deliver a thoracopagus twin as parallel breech (Foster, 1948). Vaginal delivery may be achieved following a destructive operation such as division of the junction between the thoracopagus twins (Gibberd, 1925; Dwyer and Ripman, 1959) or decapitation (Jones, 1962) or cleidotomy (Mortimer and Kirschbaum, 1962) for dicephalic twins. Such destructive operations are recommended in exceptional circumstances or when the foetuses are dead. Caesarean section is the safest mode of delivery if the foetuses are of term size (Ligat, 1912; Badaway and Shehata, 1961).

Summary

A thoracopagus twin, delivered by caesarean section, has been reported The relevant literature has been reviewed.

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Fig. on Art Paper IV