Conjoined twins in West Africa

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SUMMARY 12 cases of conjoined twins from West Africa were reported between 1936 and 1978. Eight sets were liveborn and were surgically separated either in local hospitals or abroad. Four were stillborn. Two new cases of stillborn conjoined twins were recently delivered at this hospital. The most common type and the ones most likely to be born alive were the omphalopagi. Surgical separation was successful in 5 cases but the twins separated at Zaria died about a month later. Emergency operations were performed on the pygopagus and ischiopagus, and one member of the former but both of the latter died. The thoracopagus and dicephalus twins were stillborn. However, necropsy findings in one of the thoracopagi indicate that surgical separation would have been feasible had the twins been born alive. The internal mechanical factors causing cardiac defects in such twins may be relevant to the study of the pathogenesis of congenital cardiac malformations.

The incidence of conjoined twins in West Africa is not precisely known. Studies of such twins contribute to the knowledge of embryonic duplication, fetal development, and the mechanism of congenital malformations. Noonan¹ has stressed the relevance of thoracopagus conjoined twins in the study of the mechanism of cardiac malformations. We have reported on the 2 sets of conjoined twins separated at this hospital in Zaria.² This report is a collective review of all cases of conjoined twins in West Africa up to 1978, plus 2 new cases of stillborn conjoined

twins delivered recently in this hospital—namely, a thoracopagus and a dicephalus.

Material

All data relating to cases of conjoined twins reported from West Africa between 1936 and 1978 were collected. The clinical, epidemiological, surgical, and necropsy data were analysed. The system of classification recommended by Potter and Craig³ is used to describe such twins (Table 1).

Results

Stillbirths (Table 2). There were 6 stillborn sets of twins comprising 2 dicephalus twins, 2 thoracopagi, and 1 set each of diprosopus and omphalopagus. Although the dicephalus twins of Dakar were delivered vaginally,⁴ the recent dicephalus twins of Zaria were delivered by caesarean section because of shoulder dystocia and rupture of the uterus. The former had 3 upper limbs, dipus tribrachius, while the latter had only a pair, dipus dibrachius (Fig. 1).

Table 1 *Type and sex distribution of conjoined twins born in West Africa between 1936 and 1978*

Type	Liveborn (n=8)	Stillborn (n=6)	Male (n=2)	Female (n=12)
Omphalopagus (n=6)	5	1	1	5
Dicephalus (n=2)	0	2	1	1
Thoracopagus (n=2)	0	2	0	2
Diprosopus (n=1)	0	1	0	1
Ischiopagus (n=1)	1	0	0	1
Pygopagus (n=1)	1	0	0	1
Heteropagus (n=1)	1	0	0	1

Table 2 *Stillbirths*

Year of report	Author	Country	Age of mother (years)	Type	Sex	Birthweight (kg)	Shared organs
1940	Bowesman ²⁵	Gambia	?	Omphalopagus	F	4.5	No necropsy
1958	Stiggelbout ⁶	Nigeria	25	Thoracopagus	F	2.4	Pericardium
1971	Engman and Arkutu ⁵	Ghana	26	Diprosopus	F	1.1	Y-eosophagus, Y-trachea, heart
1975	Sylla <i>et al.</i> ⁴	Senegal	29	Dicephalus	F	?	Humerus, heart, liver, jejunum
1980	Mabogunje	Nigeria	21	Thoracopagus	F	2.3	No necropsy
1980	Mabogunje	Nigeria	25	Dicephalus	M	2.6	Heart, liver, jejunum

Fig. 1 Stillborn dicephalus twins *dipus dibrachius* delivered by caesarean section in Zaria, September 1978.

Most of the internal viscera were fairly similar with 2 brains and 2 spinal cords, 2 tracheas, 2 oesophagi, 2 stomachs, and 2 duodenums with a common jejunum and a pancreas. The Dakar twins had 2 hearts which shared a common right auricle. The cardiac anatomy of the Zaria twins is the subject of another report.

The monocephalus diprosopus of Accra⁵ were anencephalic with 2 faces and spina bifida, but they had no recognisable spinal cord in the duplicated cervico-thoracic and the fused lumbosacral canals. Similarly, the tracheas and oesophagi were Y-shaped. There were 4 brachiocephalic trunks but the 2 median ones continued as carotid arteries only.

The thoracopagus twins necropsied by Stiggelbout⁶ in Kaduna had 2 normal hearts within a single pericardium. There was a large, fused central liver mass with a much smaller lateral lobe for each twin.

However, the recent thoracopagus twins of Zaria were not necropsied (Fig. 2).

Livebirths (Table 3). Four of the omphalopagus twins were delivered vaginally. The omphalopagus of Ho, Ghana,⁷ were members of a triplet pregnancy. The first was a boy delivered vaginally, following which caesarean section was performed to deliver the conjoined girls. The omphalopagus of Azare,² had a prominent umbilical hernia (Fig. 3).

The surgical data are summarised in Table 4. Both members of 4 omphalopagi were successfully separated, but the Azare twins died 35 days and 37 days postoperatively from intractable nosocomial

Fig. 2 Stillborn thoracopagus twins delivered vaginally in Zaria, March 1978.

Table 3 Livebirths

Year of report	Author	Country	Age of mother (years)	Type	Sex
1936	McLaren ¹⁸	Nigeria	25	Omphalopagus	F
1954	Aird ¹¹	Nigeria	20	Omphalopagus	F
1956	Holgate and Ikpe ¹⁷	Nigeria	24	Omphalopagus	M
1966	Gupta ⁸	Nigeria	25	Pygopagus	F
1970	Takyi ⁷	Ghana	35	Omphalopagus triplets	F
1972	Bankole <i>et al.</i> ⁹	Nigeria	30	Ischiopagus	F
1978	Mabogunje <i>et al.</i> ²	Nigeria	30	Omphalopagus	F
1978	Mabogunje and Lawrie ¹⁰	Nigeria	20	Heteropagus	F

Table 4 Surgical data

Author	Type	Birthweight (kg)	Age	Indication	Shared organs	Result of separation		Cause of death
						Twin 1	Twin 2	
McLaren ¹⁸	Omphalopagus	?	6 months	Elective	Xyphoid	Survived	Survived	—
Aird ¹¹	Omphalopagus	3.5	4 months	Elective	Xyphoid	Survived	Died	Cardiovascular collapse (1 hour)
Holgate and Ikeme ¹⁷	Omphalopagus	3.6	7 hours	Elective	Xyphoid, liver	Survived	Survived	—
Gupta ⁸	Pygopagus	3.6	5 months	Emergency	Gluteus, sacrum, spinal cord, anus, rectum	Survived	Sacrificed	Hypoplastic lungs
Takyi ⁷	Omphalopagus	3.7	? days	Elective	Xyphoid, pericardium	Survived	Died	Sepsis (9 days)
Bankole <i>et al.</i> ⁹	Ischiopagus	5.2	60 hours	Emergency	Ileum, colon, bladder, cloaca	Died	Died	Circulatory failure (30 hours)
Mabogunje <i>et al.</i> ²	Omphalopagus	?	9 days	Elective	Xyphoid, liver	Died	Died	Enteric sepsis (35 and 37 days)
Mabogunje and Lawrie ¹⁰	Heteropagus	2.6	5 weeks	Elective	Liver	Survived	—	—

Fig. 3 Omphalopagus twins of Azare separated at the ABU Hospital, Zaria, April 1975.

Fig. 4 Heteropagus twins of Ndu Sule separated at the ABU Hospital, Zaria, June 1976.

enterocolitis. Also one member of the omphalopagus of Ho died of sepsis 9 days after separation (H K Takyi, 1979, personal communication). Emergency separation of the pygopagus was performed because of intestinal obstruction and recurrent spells of apnoea in one of the twins who died during the operation. Necropsy showed that her lungs were hypoplastic and unaerated. She had apparently been

kept alive by her conjoined sister and would have been incapable of an independent existence.⁸ The ischiopagus twins also had emergency separation because of intestinal and urinary obstruction, but both developed multiple cardiorespiratory arrests shortly after surgery terminating soon after in congestive heart failure with pulmonary oedema and peripheral cyanosis.⁹

After the separation of the heteropagus (Fig. 4), the autosite was followed up regularly for 10 months; during this time her development was normal.¹⁰ Later however, we received a report from her local dispensary that she had died at age 19 months during the cerebrospinal meningitis epidemic of 1977. The surviving member of the omphalopagus of Ho is now 9 years old and in good health (H K Takyi, 1979, personal communication), and the longest known survivor is Wariboko of the Kano omphalopagus, separated by Aird.¹¹ Now 26 years old, she is a trained nurse and midwife on the staff of this hospital. Recently, she delivered her second healthy baby boy.

Discussion

Conjoined twins are monozygotic twins, incompletely separated. The separation probably occurs at the first blastomeres but it may take place at the gastrula stage.¹² In humans, long or irregular menstrual circles appearing in the very young or in premenopausal women may be associated with delayed ovulation and over-ripening of the ovum which would then be more susceptible to blastomeric duplication.^{3 13} However, the oldest mother in this series was only 35 years old, and the others ranged from 20 to 30 years. This is similar to the findings of Milham,¹⁴ who studied 22 sets of conjoined twins from New York State, USA, and is difficult to reconcile with the concept of the ageing gamete. Milham also found among the conjoined twins, a pronounced tendency to prematurity, low combined birthweight, an excess of stillbirths, and a preponderance of females, features which are all present in this series.

The rate of monozygotic twinning appears to be similar for all races, while the dizygotic twinning rate differs considerably and is known to be high in some parts of West Africa.¹⁵⁻¹⁶ Working in Eastern Nigeria, Cox¹⁵ suggested that the effect of protein and riboflavin deficiencies on the ovary may cause multiple ovulation or polyovular follicles, or otherwise cause delayed implantation in the endometrium. Any of these effects may account for the high frequency of multiple births observed in that region. Although most of such births would be expected to be dizygotic it would be of great interest to examine the surviving obstetrical records of the

same region during the recent civil war (Biafran war), when malnutrition was widespread, in order to ascertain whether there were still more multiple births or whether more conjoined twins were delivered.

The most common conjoined twins are the omphalopagi, accounting for over 40% of the collected series of Bankole *et al.*⁹ and for 43% of the West African series. They are also the most often separated, sometimes with quite simple anaesthetic and surgical facilities.¹⁷⁻¹⁸ Next in frequency are the thoracopagi in which there is currently great interest, because of the new insight afforded by the study of their cardiac conjunction in relation to the pathogenesis of other cardiac malformations. The cardiac findings in these twins are of three types. There may be pericardial fusion only with 2 normal hearts, atrial connection but ventricular separations, or ventricular connections usually associated with multiple cardiac defects.¹⁹ The twins necropsied by Stiggelbout⁶ for instance were of the pericardial type and would have been eminently separable had they been born alive. The possibility of successful separation in the twins of other types would depend on the particular clinical and electrocardiographic findings. Furthermore cardiac catheterisation and angiocardiology may be indicated in order to assess the defects, as is the practice in other cases of congenital cardiac malformations. Indeed Edwards *et al.*²⁰ have presented a plan for the potential surgical salvage of at least a member of such conjoined twins with complex atrial and ventricular fusions.

The cardiac anomalies described in thoracopagus conjoined twins include anomalous pulmonary venous return, common atrium, single ventricle, cono-truncal abnormalities, and endocardial cushion defects. In addition to genetic control, it is possible that mechanical factors arising from thoracic conjunction are involved in the pathogenesis of these anomalies.¹ This possibility is further enhanced by the demonstration by Gessner²¹ of cardiac lesions produced in chick embryos by mechanical means. The problem then would be to identify the types of intrauterine factors which may constitute comparable mechanical forces leading to the development of cardiac defects even in the single fetus.

A prenatal diagnosis of conjoined twinning would obviously be of help in planning surgical separation, and would also prompt elective caesarean section to improve the chances of liveborn twins. Prenatal diagnosis is facilitated by using the radiographic criteria established by Gray *et al.*²² These include close approximation of the chest of the twins who face each other and have an exaggerated lordosis. Diagnostic ultrasound has also been recently used for this purpose.²³ It is remarkable that only 2 sets of

twins in this series were delivered by caesarean section and, in one instance, the twins had already died and the uterus ruptured after prolonged labour.

The reason for a greater number of females in most series of conjoined twins is unknown. Milham¹⁴ suggested that it might be due to the early loss of conjoined male embryos. In more recent reports from the USA and Sweden the male: female ratio seems to be approaching 1:1. Therefore, Källén and Rybo²⁴ have suggested that better prenatal care may be decreasing such male losses resulting in a more even sex distribution. If this explanation is correct, the male: female ratio of 1:6 in West Africa would thus reflect our continuing deficiencies in antenatal care.

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