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The Ibadan conjoined twins: a report of omphalopagus twins and a review of cases reported in Nigeria over 60 years

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Summary An omphalopagus set of female conjoined twins, undiagnosed prenatally, who presented as obstructed labour needing operative delivery is reported. Their anatomic characteristics and clinical features, including overwhelming sepsis in twin II which forced early separation, and those which led to their demise are described. Twelve other cases reported in Nigeria over the past 60 years are reviewed with reference to the aetiology and epidemiology of conjoined twinning and the determinants of successful surgical separation.

Introduction

Reports from around the world estimate that one in every 50,000–75,000 births will be conjoined twins.[1–3] The incidence of conjoined twins in Nigerian is not known. To the best of our knowledge, 12 cases have been reported from Nigeria during the last 60 years.[4–14] This probably does not reflect the true incidence in Nigeria as most cases are not reported on account of the associated stigma. Cases of conjoined twins seen in hospital are rare and pose a management challenge when they do present because of a lack of experience in managing them. A set of omphalopagus twins recently encountered in Ibadan is therefore reported with a review of the 12 cases previously reported from Nigeria with a view to evaluating management options in the West African sub-region.

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Case report

A set of female conjoined twins were delivered at 34 weeks gestation by emergency lower segment caesarean section in a private medical facility in Ibadan to a 21-year-old unbooked primigravida. The indication for the section was prolonged, obstructed labour and prolonged rupture of membranes (PROM). The mother and her twins were referred to University College Hospital (UCH) 24 hours after delivery and the unhygienic transportation to UCH was an added risk factor for sepsis.

Initial clinical evaluation showed two preterm babies, joined from the xiphisternum to the lower limit of the abdomen where a small exomphalos was evident. There was a single umbilicus and a single cord. Their combined weight was 3.15 kg. They had similar core temperatures, 35.8°C in twin I and 35.6°C in twin II. Twin II had a grade 2/6 systolic murmur along the left sternal border. The heart sounds in twin I were normal and she had normal peripheral pulses. They were

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acyanotic, anicteric and not pale. The problems identified then included (i) abdominopagus twins, (ii) low birthweight (LBW), small-for-gestational-age (SGA), (iii) hypothermia, (iv) congenital heart disease in twin II, and (v) suspected sepsis on account of PROM and transportation to UCH. They were commenced on intravenous fluid therapy using 10% dextrose in 1/5 normal saline at 80 ml/kg and were given nil by mouth. They had full sepsis screening and were then given intravenous cloxacillin, 100 mg/kg bodyweight in four divided doses 6-hourly, and intramuscular gentamicin, 5 mg/kg bodyweight in two divided *doses 12-hourly*, pending results of the sepsis screening.

Laboratory profiles. The blood group in both twins was O Rhesus negative. Both had metabolic acidosis with serum bicarbonate levels of 15 and 10 mmol/l in twins I and II, respectively. *Klebsiella* species sensitive to ceftazidime were isolated from their blood cultures but both had normal cerebrospinal fluid.

Ultrasonography. Abdominal sonography in the left lateral decubitus position on both sides revealed a merging liver architecture of the same echogenic pattern at the midline. There was no demonstrable axis of separation of the liver tissue or capsule. The kidneys were of normal size, shape and position. The spleen was identified in both twins, and they were normal.

Plain radiographs. Lateral projections revealed two babies joined at the anterior abdominal wall from the level of D8 to the lumbosacral junction. The thoracic cage of twin I showed normal cardiac configuration and location with normal aeration of the lungs, while that of twin II showed an enlarged cardiac silhouette with pulmonary plethora. There were no bony abnormalities. Two stomach gas shadows were observed, that of twin I was J-shaped while that of twin II was distended with air with the suggestion of intrathoracic extension (herniation). The duodenal cap region and small bowels appeared in the midline, surmounted by a mushroom-shaped liver shadow that appeared to be common to both. The rectal gas shadows were separate.

Echocardiography. Twin I had a normal heart. Twin II, however, had an ostium primum atrial septal defect and a peri-membranous ventricular septal defect (a partial atrio-ventricular septal defect). Atrial situs was solitus and atrio-ventricular and ventriculo-arterial connections were concordant. The atrio-ventricular and arterial valves were normal. The ductus arteriosus was patent.

Barium studies. The barium was offered through a feeding bottle to twin I who sucked vigorously. The oesophagus and a J-shaped stomach were outlined with the duodenal cap at the midline beneath the inferior border of the conjoined liver shadow. No evidence of reflux or hiatus hernia was demonstrated. The duodenal loop appeared reversed. Some sharing of the barium at an early phase was noted in the small bowel of both babies. At 30 hours the barium was seen entering the terminal portion of the sigmoid colon of twin II and at 48 hours it was seen in the rectum of both babies. (Fig. 1).

Intravenous urogram showed normal excretion of contrast outlining normal separate pelvicalyseal systems, ureters and bladders in both babies (Fig. 2).

Medical management. They were nursed in an incubator. Following isolation of *Klebsiella* species from the blood, antibiotic therapy was changed on the 3rd day of life to intravenous ceftazidime, 100 mg/kg given in three divided doses 8-hourly to each baby. Metabolic acidosis was corrected using sodium bicarbonate. Thermal instability persisted in both babies and by the 7th day of life both had developed abdominal distension. Copious bile-stained fluid drained from a nasogastric tube in twin I while twin II showed signs of overwhelming septicaemia, including sclerematous skin changes and disseminanted intravascular coagulopathy. On suspicion of necrotizing enterocolitis, metronidazole was

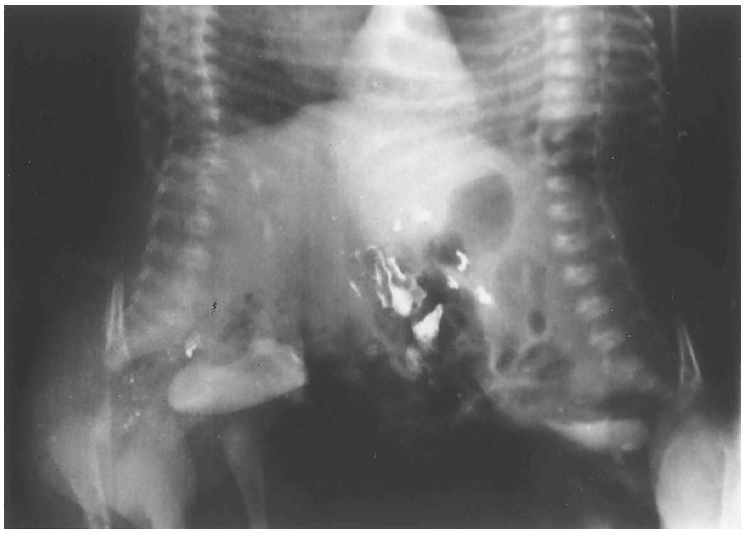

FIG. 1. Barium given to twin I showing in the colo-rectal regions of both babies.

added to the treatment regime, 10 mg/kg 8-hourly.

Surgical treatment. Initially, the plan was to attempt to separate the babies after about 3 months. However, by the 7th day of life, twin II, who had a grade 2/6 systolic murmur and was the smaller of the two, developed sepsis and necrotizing enterocolitis which did not respond to medical treatment, and it was decided on day 8 to separate the twins in order to give twin I a chance of survival.

At surgery, the twins were found to share a common liver with two separate bile ducts (Fig. 3). They also shared the same small bowel with the 'C' loop of twin II being reversed and on the left side of the abdomen. They had separate left-sided colons down to the rectum (Fig. 3). With these findings and the poor clinical condition, twin II was sacrificed in favour of twin I. The conjoined liver and the biliary systems were 'served' to twin I after careful separation from twin II. A thin portex sheet was sewn to the fascial edges of the abdominal wall of twin I without tension. Total operation time was 5 hours. Blood loss was minimal and the surgical procedure was considered satisfactory.

Twin I was transferred on assisted mechanical ventilation to the intensive care unit. She was on a combination of intravenous ceftazidime and intramuscular gentamicin over the period of surgical separation. Fifteen hours after surgery her vital signs were considered stable and weaning from mechanical ventilation was commenced. At 26 hours, extubation was attempted. However, she developed severe laryngeal spasm and re-intubation via the oro-pharyngeal route was extremely difficult. A tracheostomy was performed with intubation, but she had lapsed into cardio-respiratory arrest and failed to respond to resuscitation. At autopsy, the cause of death was established to be asphyxiation. She had also developed septicaemia with disseminated intravascular coagulopathy. Necrotizing enterocolitis was excluded in both twins.

Discussion

The aetiology of conjoined twins still generates debate. The age-old question of fusion or fission remains. It is contended by some that there is no evidence for an embryological process by which conjoined twins can be formed by fission and that evidence abounds to support fusion in all cases.[15–18] Intact ectoderm will not fuse to intact ectoderm, and seven

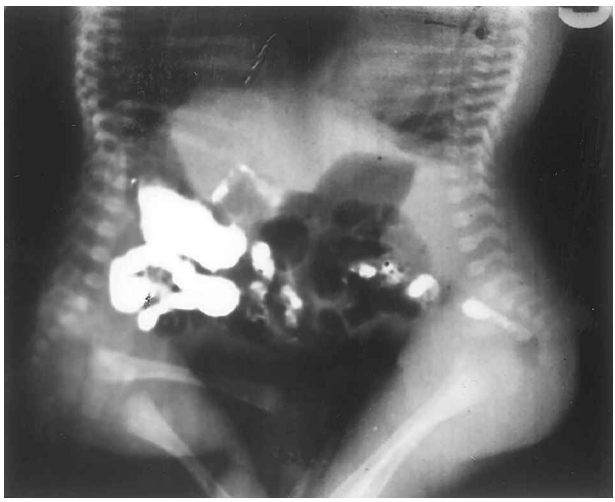

FIG. 2. Intravenous urogram ashowing separate bladders and the ureter in twin I on the left.

possible sites of union in the early embryo explain all seven types of conjoined twins. Spencer has enumerated these sites as: anterolaterally united parapagus twins resulting from two nearly parallel notochords in close proximity; craniopagi and pygopagi from fusion at the cranial and caudal neuropores, respectively; cephalopagi and ischiopagi from union at the pharyngeal and cloacal membranes, respectively; thoracopagi from merging of the cardiac anlage; and omphalopagi from fusion of the umbilicus or the edges of two embryonic discs in any area not including the above sites.[15] Parasitic twins result from the embryonic death of one twin, leaving various portions of the body vascularized by the surviving autosite.

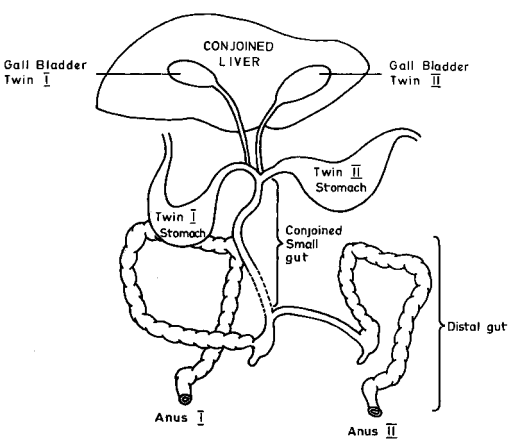

FIG. 3. Organs shared by twins I and II.

Since the twins are usually of the same sex and union is at similar anatomical parts and they usually have the same blood group, it has been suggested that the majority must be uniovular.[5,8] It is further argued that if separate twins became joined by fusion in the early stage of their development, it would require the disappearance of the intervening amnion. However, conjoined twins usually differ in size and appearance and are never as identical as separate mono-ovular twins.

Logrono *et al.*, reporting on heteropagus conjoined twins, showed by fluorescent *in situ* hybridisation studies that the parasitic twin was male and demonstrated dizygosity by DNA-typing studies.[16] The report demonstrates fusion, not fission, as the mode of formation.

Of the 13 reported conjoined twins born in Nigeria (including this case), three were stillbirths. The stillbirth rate in Nigeria is not known, but available data suggest a higher than national average stillbirth rate among mothers of conjoined twins.[12] Spencer reported a high stillbirth rate in his series and suggested

that a maternal factor might be of aetiological importance in conjoined twinning.[15]

This case was not detected antenatally, and the diagnosis was made only at emergency operative delivery. In good hands, early ultrasonic prenatal diagnosis of conjoined twins is easy.[19–21] High-resolution vaginal sonography makes diagnosis possible as early as the 8th week of gestation.[22]

The success of surgical separation is usually determined by, among others things, the complexity of the union and timing would also appear to be a key determinant of success. The major organs shared in this case included the anterior abdominal wall, hepatobiliary system (except the gall bladder, cystic duct and common bile duct), greater and lesser omenta and their vascular supply, as well as the duodenal bulb to about 'one foot' from the ileocaecal junction of twin II. The operative findings and the overwhelming septicaemia in twin II compelled the surgical team to secure the shared organs for twin I, in effect sacrificing twin II in the hope of saving twin I. A similar decision was made in the case reported by Gupta[8] and is in keeping with a recent landmark judgment in Manchester, UK in which it was ruled that the life of one twin could be sacrificed to save the other, even against the parents' desire to let nature take its course.[23,24]

The presence of other malformed organs can also be a confounding factor. Twin II had congenital heart disease, a partial atrio-ventricular septal defect with persistent ductus arteriosus, which led to cardiac decompensation which further compromised her survival. Twin I had a normal heart which gave her a better chance of survival, but there was extensive sharing of other organs that rendered twin I just as susceptible. Danford *et al.*[25] and Gelis *et al.*[26] have detailed various congenital heart defects encountered in their series of conjoined twins and the findings in our case are not unusual.

Surgical separation of conjoined twins is not a procedure for the occasional surgeon and having a specialised national team to operate on such infants is desirable. However, the cost-effectiveness of such a team must be viewed in the context of the meagre resources available in developing countries where there are greater and more pressing health problems. Though limited, there is some experience of managing conjoined twins in our centre,[8,9] and the decisions made in this case were in line with fairly standard management guidelines.[27,28] In retrospect, however, extubation of twin I post-operatively should perhaps have been delayed.

This and other previously reported cases in Nigeria suggest a uniform spread of cases across the country. The apparent preponderance of cases in Zaria probably reflects the presence of an interested team there. An apparent lack of cases from the Savannah belt does not necessarily indicate non-occurrence of conjoined twins, and it is more likely that cases have simply not been recorded.

Anatomically, seven of 13 cases were joined ventrally (thoracopagus and omphalopagus), there were two pygopagus unions, two heteropagi, one case was joined dorsally (ischiopagus), and there was a case of the rare dicephalus type.

There is a consensus that females predominate and some workers have reported a F:M ratio of 2.3:1. In Nigeria, females predominate in an even higher ratio, 5.5:1, the highest reported in sub-Saharan Africa. The highest F:M ratio, 20:1, was reported by Milham.[29] It is not known why females predominate but it has been postulated that either the early female zygote is more likely than the male to undergo this type of change, or that male conjoined twins, once formed, are not as viable and are lost early in pregnancy before ascertainment is possible.[25,29]

The ages of the eight mothers on whom information was available ranged from 20 to 30 years, similar to Milham's findings,[29] and it is difficult to reconcile this with the concept of the ageing gamete.

Perinatal mortality was high in the Nigerian series: only eight of 21 first or second twins survived and six of the deceased were stillborn. There appear to be no obvious differences in mortality rates between gender, type of conjoining and delivery by elective or emergency surgery. The commonest prob-

TABLE I. Cases reported previously from Nigeria

Author(s)	Year reported	Type of "-pagus"	Place of birth	Gender	Age at surgery	Indication surgery	Outcome	
							Twin I	Twin II
McLaren	1936	Omphalo-	Sokoto	F	6 mths	Elective	S	S
Aird	1954	Omphalo-	Kano	F	4 mths	Elective	S	D
Holgate & Ikpeme	1956	Omphalo-	Port Harcourt	M	7 hrs	Elective	S	S
Stigglebout	1958	Thoraco-	Kaduna	F	Stillborn	—	D	D
Gupta	1966	Pygo-	Ibadan*	F	5 mths	Emergency	S	Sac
Bankole *et al.*	1972	Ischio-	Warri	F	60 hrs	Emergency	D	D
Mabogunje *et al.*	1978	Omphalo-	Zaria	F	9 days	Elective	D	D
Mabogunje & Lawrie	1978	Hetero-	NDU Sule	F	5 wks	Elective	S	—
Mabogunje	1980	Thoraco-	Zaria	F	Stillborn	—	D	D
Mabogunje	1980	Dicephalus	Zaria	M	Stillborn	—	D	D
Sathiakumar *et al.*	1988	Hetero	Zaria	F	—	Elective	S	D
Iroku & Anah	1990	Pygo-	Anambra	F	—	Elective	D	D

S, survived; D, died; Sac, sacrificed; * assumed by deduction from the text.

TABLE II. Further details of cases reported previously from Nigeria

Author(s)	Age of mother (yrs)	Gravida	Para	DOB	Birth-weight (kg)	Place of birth/ separation	Shared organs	Cause of death
McLaren	25	—	2	20/12/35	—	Sokoto General Hospital	Skin and xiphoid	—
Aird		—	—	25/7/53	3.35	Hammersmith Hospital, London	Skin, liver, xiphoid	CVS collapse 1 hr post-op.
Holgate & Ikpeme	24	—	1	?	3.6	Enugu General Hospital	Skin, liver, xiphoid	
Stiggelbout	25	—	3	23/5/58	2.4	Stillbirth, Kaduna General Hospital	Pericardial cavity	IUD
Gupta	25	—	5	24/12/61	3.6	Hammersmith Hospital, London	Gluteus, sacrum, spinal cord, anus, rectum	Shared organ to twin I, hypo-plastic lungs
Bankole *et al.*	30	8	7	—	5.2	UCH, Ibadan	Ileum, colon, bladder, cloaca	CVS collapse 30 hrs post-op.
Mabogunje *et al.*	30	5	4	13/4/75	—	ABUTH, Zaria	Xiphoid, liver	Enteric sepsis 34 & 37 days post-op.
Mabogunje & Lawrie	20	3	2	24/5/76	2.26	ABUTH, Zaria	Liver	—
Mabogunje	21	—	—	—	2.3	Stillbirth	—	—
Mabogunje	25	—	—	—	2.6	Stillbirth	Heart, liver, jejunum	—
Sathiakumar *et al.*	—	—	—	—	2.5	ABUTH, Zaria	Skin parasite	—
Iroku & Anah	—	—	—	—	3.8	UNTH, Anambra	Skin, anus	—

IUD, intrauterine death; CVS, cardiovascular; UCH, University College Hospital; ABUTH, Ahmadu Bello University Teaching Hospital; UNTH, University of Nigeria Teaching Hospital.

lem appears to be a lack of prenatal diagnosis, as in our case; babies were therefore born in centres not equipped to cater for their special needs and had to be transported over long distances without transport incubators to better equipped centres. Such babies can be successfully transported across great distances if it is done under appropriate conditions, as happened several decades ago.[5,8] Additional epidemiological details on the cases reported from Nigeria are given in Table II.

Conjoined twinning might not be as uncommon as currently thought in Nigeria. Educating the public to change their perception of conjoined twins as monsters originating from witchcraft and therefore deserving destruction, combined with qualified antenatal supervision of pregnancies, would go a long way to ensuring early prenatal diagnosis. This, in turn, will ensure early referral of such cases to adequately equipped centres where elective operative delivery would improve survival. We hope that our report will help achieve this goal.

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