Conjoined (Siamese) Twins in Zambia

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The university Teaching hospital is the main referral centre for Zambia a country that has a population of over 11 million people. Fourteen sets of conjoined twins have been seen between 1970 and 1999. Of these fourteen sets of twins, three cases were stillborn (21.43%). The remaining 11 (78.57%) sets were brought in alive. Five (35.71%) of these eleven sets had multiple congenital abnormalities; and were not subjected to operation. All five died within a period of two weeks. Six (42.86%) sets were successfully separated. An attempt has been made to establish the incidence of conjoined twins in Zambia. Basic principles of management have been suggested and constraints likely to be encountered in a third world country like Zambia have been discussed.

Introduction

A study has been undertaken to find out the incidence of the conjoined twins in Zambia.
Because of the rarity and variance of conjoined twins, no textbook has a comprehensive chapter on this subject. There is also paucity and a scarcity in the case reports. In this paper an attempt has been made to comment on a few important issues on its management in view of constraints encountered in a developing world.
Another purpose of this paper is to present an analysis of fourteen sets of conjoined twins which may form the basis of future reference on this subject here in Zambia.

Material and Methods

This retrospective study of conjoined twins consisted of case reports, referral cases, and clinical records from Gynaecology, Obstetrics, and Neonatal Paediatric wards in order to complete the data. The diagnosis of this condition was made on simple clinical examinations, inspection, Table II, auscultation and physical examination, Table II. Classifications of conjoined twins and their associated abnormalities were over 70% made using these methods. Electrocardiography, Radiology (simple and special x-rays) ultrasound, and blood studies, were used in confirming the clinical diagnosis. CAT-scan MR1, and any other sophiscated investigation were not available due to financial constraint. However, CAT scan has just been available at the time when this data was collected.

Results

Fourteen sets of conjoined twins were seen in period of 29 years (1970 to 1999). Eight sets (57.14%) were male while six (42.86%) were female, giving male to female ratio of 1.33 to 1. The twins were seen at the ages ranged from 2 hours to 78 hours after birth giving an average of 6 hours. Three sets (21.43%) of stillborn were seen at University Teaching Hospital from 1970 to 1977 a period of 8 years.
Eleven cases of conjoined twins (78.57%) were seen in 21 years (1978 to 1999). These were borne alive. Five cases (35.71%) had multiple congenital abnormalities, which were incompatible to life. These died within two weeks. Six sets (42.86%) were successfully separated. Table I.

Case reports

Case 1

An 18-year-old primigravida gave birth to hetropagus (fig. 1) female conjoined twins on 5th January 1983. It was a prolonged vaginal delivery in Mtendere Mission Hospital in the South West part of Zambia. Birth weight was 3.420 kg. The baby and the mother were referred to University Teaching Hospital for operation, which was done 30 hours after birth. The finding were noted:

A common pelvic floor; a loop of small intestine of the of the living baby was housed in the pelvis of the non-living baby; blind pouches from rectum and vagina were attached to a cystic mass of the monster baby; bladder, uterus, urethra and the ovaries of the viable baby were normal and properly placed but the vulva was both shared.

The surgical procedure included freeing of the rectum,

and vagina with meticulous preservation of sphincters. The pelvic floor was reconstructed by using rotation advancement of gluteus maximal myocutaneous flap from the deformed baby. The small intestine was replaced in the abdomen. The vulva was divided and reconstructed. A large flap from the residual appendage was used to cover defect over the hip and sacrum. The child recovered well with normal urinary and bowels function and lost follow up after 5 years.

Case 2

A 30 years old multiparous mother gave birth to a hetropagus female conjoined twins xiphomphalopagus born at University Teaching Hospital, on 12th April 1983. Birth weight was 3.110kg. The viable child had large exomphalus. Radiology showed bony connection at xiphisternal area. Separation was done after 18 hours. On exploration upper abdominal cavities of the twins were communicating. The liver of the living child was joined to small hypoplastic liver of the deformed baby; otherwise there were no other shared organs. The liver was separated using diathermy with minimal blood loss. Two large flaps were raised from the buttocks of the deformed child to cover the thoraco – abdominal defect and omphalocele of the living baby. The child made a quick recovery and remained well with herniation. Scoliosis and pectus carinatum became apparent at the age of six years and has been referred abroad for management.

Case 3

A 28-year-old multiparous mother gave birth to a set of male diplopagus omphalopagus on 15th October 1984 in University Teaching Hospital. Birth weight was 4.260kg. After initial stabilisation and routine investigation, separative operation was done after 40 hours. On operation a tubular connection between the two large independent bowels was found and was separated easily. Difficulty was encountered during the covering of the abdominal wall defects due to lack of synthetic materials. However, the cover was achieved with the rotation advancement flap of the residual abdominal tissue. Postoperative recovery was uneventful. Follow up was lost after four years.

Case 4

A 36-year-old multiparous mother gave birth to a set of male twins diplopagus omphalopagus on 12th January 1990.in the University Teaching Hospital. Birth weight was 4.580 kg. The set of twins were nursed in a special Neonatal wing for a month. During this time, investigations were done and revealed to normal babies connected to the two urinary bladders by a huge tube. On operation this was confirmed and operation was easily done, the tube was removed. Abdominal muscle walls were normal; the small area where the twins were connected was repaired by approximating the muscle walls while the small skin defects left on both twins were covered by mobilizing the skin. The twins were lost from clinic follow up at the age of 8 years. However, one of the twins reappeared after two years at the age of 10 years.

Case No.5

A 16 year old school going child gave birth to a set of female twins diplopagus omphalopagus in University Teaching Hospital on 21st August 1993.Birth weight was 4.100 kg. Both twins were found to be normal. Investigations revealed complete individual twins connected by skin. There was no muscle defect.

The set was operated after 74 hours. The skin defect was easily covered after mobilizing of the skin. The twins had been attending clinic up to the age of five years when they were lost for follow up.

Case 6. Craniopagus

A 36-year Zambian multiparous mother gave birth to male conjoined twins (craniopagus) on 27th January 1997 at University Teaching Hospital. The weight at birth was 4.800 kg. Investigations showed two separate babies sharing one cranio-cavity with two normal brains separated by fibrous meninges. The babies were nursed in a special ward up to the of 9 months when they were sent to South Africa where a 40 hour operation was carried out by joint team consisting American, South African and Zambian surgeons. The patients were later sent back to Zambia where they are being nursed. The babies are waiting for plastic surgery to provide a protective shield to their heads.

Case 7. Thoracipagus

A 40-year-old Zambian multiparous mother gave birth to a set of twins with two heads four arms joined to the chest, one abdomen with a normal pelvis and a pair of legs. The weight at birth was 4.1 kg and was delivered by caesarean section at University Teaching Hospital. Plain chest x-ray and ultrasound revealed that the patient had one heart and one liver. The babies were fed through nasal gastric tube. They were kept in intensive care unit but died after two weeks due to chest infection (pneumonia).

A total number of three sets of thorapagus conjoined twins were seen in this institution, two male and one female. All died after resuscitation and hospitalisation in Neonatal Surgical Ward the average length of stay was ten days.

TABLE 1. Conjoined twins (Siamese twins) seen in Zambia in 29 years

	Male	Female	Total (%)
Still birth	1	2	3 (21.4)
			5 (35.7)
Alive	Inoperable 3	2	6 (42.9)
	Operable 4	2	
Total	8	6	14 (100)

Table 2. Anatomical site where twins were joined

Anatomic site		Number of cases	Percentage
Thoracopagus	(Chest)	3	21.4
Omphalopagus	(Abdomen)	6	43.0
Pygopagus	(Buttocks)	1	7.1
Ischiopagus	(Pelvis)	3	21.4
Craniopagus	(Head)	1	7.1
Total		14	100

Table 3. Findings on physical, ultrasonographic and radiographic examination

Shared organs	Number of cases	Percentage
Heart	4	28.6
Large intestines	3	21.4
Liver	2	14.3
Bladder	2	14.3
Small intestines	2	14.3
Skin	1	7.1
Total	14	100

Table 4. Successfully separated conjoined twins

Type of twins	Number	Percentage
Omphalopagus	4	28.6
Ischiopagus	1	7.1
Craniopagus	1	7.1
Total	6	42.8

Discussion

AETIOLOGY AND INCIDENCE

The aetiology of conjoined twins remains obscure. There is either an incomplete division of the inner cell mass at the blastula stage of morphogenesis resulting in incomplete separation of monozygomatic, monoamniotic twins, or there is initiation of two areas of axial growth, resulting in abnormal budding from embryonic plate[3].

World literature suggests that the incidence of conjoined twins is between 1:20,000 and 1: 200,000 births. The cause of this vast discrepancy in figures has not been fully analysed. In our scenario, most of the population is found in the rural areas with strict belief and cultural background. Many conjoined twins either abort or are

stillborn which go unreported and may be one of the causes of maternal death during labour. Even live birth with appendages are uninteresting and unwanted, therefore, they are destroyed and never reported. As an illustration of this point, the mother of Case No. 4 had signed a paper in the mission hospital where she delivered that she would not want the child even after a successful operation.

The controversy regarding possibility of 'epidemic' of conjoined twins in Southern Africa still exists[4]. This apparent increase of conjoined twins in our community may be due to better reporting awareness, improvement in health care delivery system and identification of aborted stillborn conjoined twins. Until 1986, when facilities for delivery were extended to various urban clinics, UTH was the only maternity hospital for Lusaka. Between 1970 and 1977, there were 126,952 deliveries in UTH over that 8 year period[1] and in the next years, between 1978 and 1985, the deliveries numbered 156,273.This gave an incidence of the conjoined twins of 1:63,000 and 1:52,000.

CLASSIFICATION

Classification of conjoined twins is well compiled and described by Guttmacher[5] and Potter and Craig[6]. Conjoint twins are rare and diverse. Each twin may be a potentially viable individual or often one is deformed and remains as an extraneous appendage on the other twin. Well-formed twins are known as *diplopagus* (symmetric or mirror image) but one twin attached with an incomplete foetus is known as *hetropagus* (asymmetrical). Thoracopagus twins (joined at the chest) account for 40% of cases, omphalopagus, joined at the abdomen, in 34%, those joined at the buttocks (pygopagus) in 18%, ischiopagus (joined at the pelvis) in 6% and the craniopagus (joined at the heads) in 2%. Thoracoxiphomphalopagus comprise almost three quarters (75%) of all conjoined twins. These are relatively easy to separate depending upon the type and extent of visceral involvement. Sharing of organs varies from completely shared to a thin band separating the two organs. Joining of the liver has been reported in 100%, pericardium in 90%, heart in 75% and gastrointestinal tract in 50%[7]. Conjoined twins attached to the thorax and abdomen constituted 61% in our series, which was close to the 57% reported from West Africa[8]. Due to the scarcity of the forensic pathologists in UTH, no autopsy reports were available[8].

MANAGEMENT

Management of the conjoined twins requires a multidisciplinary approach. In the developing world, obstetric and gynaecologic services are not developed in the rural population where the majority of deliveries are conducted without expert supervision. Tan estimated that 1 in 546 twins are conjoined twins[9].

Twins associated with hydramnios and prolonged labour should be suspected to be conjoined twins. Its live birth per vagina is not rare and is dependent on its rigidity \t the site of union. Conjoined twins delivered spontaneously or by Caesarean section have a better chance of survival than the set, which underwent prolonged labour.

Gray[10] suggested that conjoined twins should be suspected if:

(i) Heads are at the same level and plane,
(ii) Unusual extension and proximity of spines,
(iii) Relative position of foetus remains unchanged even after manipulation and movement on series x-rays[10].

Ultrasound shows that echoes arising from the body surface of each foetus will converge into a single echogenic band with no clear separation of the foetus. Shared organs may be delineated. Failing to establish diagnosis or with equivocal results, amnioscopy or even amniography could be done using double contrast media of oil and water. CT scan will be an additional help in the antenatal diagnosis. Means and methods of prenatal diagnosis and safe delivery are not well organized in the developing world and have to be upgraded for overall successful surgical separation.

Following a referral of the conjoined twins, there should be an emergency meeting of the interested team namely the surgeons, anaesthesiologists, radiologists, neurologists and the staffs from the operation theatre. They should draw up a plan of action for future activities including timing for next meetings. Involved team must undertake meticulous clinical evaluation by sophisticated equipments to diagnose between inoperable and operable lesions. Lack of these might be one of the causes of death in our two cases (Cases No. 9 and 11). The team is also responsible in procuring materials like Teflon and Vicryl mesh and other gadgets needed for their efficient management. In addition to routine, the following investigations may be needed to elucidate more about them – blood chemistry, haematocrit, x-rays I.V.P, cholecystography, E.C.G., Barium swallow, cross-circulation test by methylene blue via Nasal gastric tube and glucose tolerance, TC. 99m liver scanning, ultrasound, CT scanning, N.R.M. imaging, Umbilical vein cardiac catheterisation, CT angiocardiography and use Doppler. Every effort must be made to know the precise problems before operation. In view of existing complexity, surgery to separate can be categorised in three headings.

A. Urgent Emergency Operation

Where life of one or both twins is threatened by intestinal obstruction, obstructive uropathy or impending rupture of omphalocoele or tracheo-oesophageal fistula etc; a minimal surgery of temporary or permanent nature is advisable to overcome the emergency.

B. Early Elective Operation

When a gross extrapendage is found in an otherwise living healthy baby, early separation is indicated because the unwanted part is sharing oxygen and nutrition, thereby putting stress and stain on the cardio-respiratory system of the living one.

C. Delayed elective Operation

Where the diplopagus conjoined twins are healthy and there is intention of saving both, the plan for operation has been outlined by Kling[11] such as special investigation, rehearsal, preparation for special equipments and organizing competent monitoring system services. During the waiting period, intensive care including hyper alimentation is needed to improve the general health; constant monitoring is also required to detect any bizarre clinical feature. Some of the minor procedures may also be performed to ease the final operation such as tissue expansion such as tissue expansion[12] or screw clamping of superior sagittal sinus[13].

Comments

It has been evident from our report that in our environment, success in the separation of conjoined twins is dependent on various factors.

Firstly, dissemination of the fact that the conjoined twins are just one of the congenital deformities such as cleft lip and cleft palate which are amenable to surgery.

Secondly. Meticulous assessment and management of the foetus in the pre-, peri- and post-natal period. This demands improvement in obstetric and gynaecological services by providing specialized manpower and equipments. Practitioners in the field should take urgent measures to transfer cases to the referral hospital. They should also take interest in reporting stillborn conjoint twins. Thirdly, to achieve success in the surgical separation, it requires modern sophisticated material and equipments to be used in investigations, assessment, monitoring in pre-, peri- and postoperative period. From our discussion, it has been apparent that none of the above three exists in our environment and needs to be analysed and provided. These are not only for the conjoined twins but also for overall management of the surgical cases.

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