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CASE REPORT

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Conjoined twins in Edo state of Nigeria; a report of the first surviving set

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Abstract: The term conjoined twins refers to babies who are physically joined at some point. It is a rare condition with an estimated incidence of 1 per 200,000 live births. We report our experience with conjoined twins over a twelve year period in tertiary hospital in Nigeria and a case of the first set of conjoined twin survivors in Benin City, Nigeria. Over the last twelve years (1999-2011), three cases of conjoined twin have been recorded in our teaching hospital. A set of **t**horacoomphalopagus twins (females) were delivered in 1999 and they survived for only 36hrs. Another set of female omphalopagus twins were delivered in 2009 and survived a separation surgery. A third set of female **t**horacoomphalopagus was delivered in another institution same year and referred to our unit but they only survived for 48 hours.
The first surviving twins were omphalopagus, sharing a single liver, and common bile duct emptying into a common duodenum. The stomach, as well as the jejunum was normal and unshared. Surgical separation of the liver was done and biliary reconstruction procedure performed for twin II. A three -year follow up showed good outcome.

Introduction

The care of conjoined twins continues to pose a daunting medical challenge that includes adequate care of pregnancy, well planned delivery, critical care in early neonatal life, advance surgical intervention and last but not the least is the ethical issues[1,2]. We report our experience with conjoined twins over a twelve year period in a tertiary hospital in Nigeria and a case of the first set of conjoined twin survivors in Benin City, Nigeria. The incidence of conjoined twins in Nigeria is unknown; however an article published in 2001 suggests that over the preceding 60 years there were 12 published cases nationwide,[3] excluding our own cases. The cases seen in our center have so far not been reported perhaps because this center had not recorded any survival of conjoined twins since its existence three and half decades ago. Therefore under reporting may be due to the poor prognosis and stigmatization associated with this condition.

Conjoined twining is one of the most fascinating human malformations but it is not exclusive to our specie as it has also been reported in other mammals, reptiles, birds and fishes.[4] The term conjoined twins refers to babies who are physically joined at some point. It results from incomplete splitting of monozygotic (identical) twins after 12 days of embryogenesis. Some authors recently had postulated that it actually results from "fusion" of stem cells of already separated embryo[4]. Conjoined twinning occurs sporadically with no risk in future pregnancy. Overall the condition is rare with an estimated incidence of 1 per 200,000 live births[4]. Though there are more live born females conjoined twin with a female to male ration of 3:1, however this condition occur more in male foetuses as evidenced by its higher rates in male stillborn. Male conjoined twins are also more likely to die shortly after birth, implying that female conjoined foetuses have better chance at survival than their male counterparts.
The site of union forms the basis of the terms used for classifying conjoined twins: Thoracoomphalopagus (joined at the chest, abdomen or both) – 74%
Thoracopagus or xiphopagus (joined at the chest) - 40%
Omphalopagus (joined at the abdomen) - 34%
Pygopagus (joined at the buttocks) – 18%
Ischiopagus (joined at the ischium) – 6%
Craniopagus (joined at the head) – 2%
A rare type occurs when one incompletely formed (parasitic) twin is dependent on the well-formed one. This is known as heteropagus twinning. The term "pagus" is a Greek word which means "that which is fixed"
The term Siamese twins comes from Eng and Chang Bunker (1811-1874), the famous conjoined twins from Thailand (previously known as Siam). They were thoracopagus twins and were exhibited in circus shows around the world before settling in the United States, where they married two sisters and had nearly two dozen children. They were successful businessmen and lived up to 63 years. There have been several other reports of

conjoined twins in different parts of the globe. In Nigeria the earliest report of conjoined twins were born in Sokoto on 20th December 1935 to a 25 year old para 3 woman at home.[5] They shared only abdominal wall and skin, but no shared internal organs. They were readily separated at the General Hospital in Sokoto by a British missionary doctor.[5] Twenty years later the Kano omphalopagus twins Tamonotanye and Waiboko, were separated in London by Ian Ard.[6] Since then there have been sporadic, howbeit scanty, reports of conjoined twin from the country, but a three year survival follow up of the survivors is generally scarce. Since the first documentation in 1935 till date, 18 cases have so far been reported as summarized in table 1. There are, nonetheless, some cases of conjoined twins that were found in the Nigerian news reports that never made the medical literature, Table 2

Table 1: Summary of conjoined twin reported from Nigeria

Place of delivery	Author(s)	Type of Conjoin Twins	No of pairs	Place of surgery	Outcome	
		Twin I Twin II				
Sokoto	McLaren, 1936	Omphalopagus	1	Sokoto General Hospital	S	S
Kano	Aird, 1945	Omphalopagus	1	Hammersmith, UK	S	D
Port Harcourt	Holgate and Ikpeme, 1956	Omphalopagus	1	Enugu General Hospital	S	S
Kaduna	Stigglebout, 1958	Thoracopagus	1	-	Still Born	Still Born
Ibadan	Gupta, 1966	Pygopagus	1	Hammersmith, UK	S	D
	Omokhodion et al, 2001	Thoracopagus	1	UCH, Ibadan	D	D
Warri	Bankole et al, 1972	Ischiopagus	1	UCH, Ibadan	D	D
Zaria	Mabogunje, 1978	Omphalopagus	1	ABUTH, Zaria	D	D
	1980	Thoracopagus	1		Still Born	Still Born
	1980	Dicephalus	1		Still Born	Still Born
	Sathiakumar et al, 1990	Pygopagus	1		S	D
NDU Sule	Mabogunje and Lawrie, 1978	Heteropagus	1		S	-
Anambra	Iroku and Anah, 1990	Pygopagus	1	UNTH, Enugu	D	D
Lagos	*2003	Thoraco-abdomino pagus	1	John Hopkins, Baltimor	S	S
Ile-Ife	Adejuyigbe et al, 2005	Ischiopagus	2	OAUTHC, Ile-Ife	S	S
Enugu	Ekenze et al, 2009	Omphalopagus	1	Germany	S	S
Maiduguri	Auwal et al, 2011	Ischiopagus	1	UMTH, Maiduguri	S	S
*Total			18			

*Total excluded the conjoined twins seen at the UBTH between 1999 and 2011

Table 2: News reports of Nigerian conjoined twins

Gender	Date	Birth place	Extent of joining	Place of surgery	Website
??	2005	Owerri	Chest Abdomen Pelvic girdle Genitalia		www.allafrica.com/stories/200503040379.html
F	2013	Oturkpo	Abdomen		www.newsinnigeria.org/2013/18
F	2012	Kano	Pyopagus Heart Chest to abdomen	India	www.punchng.com
F	2013	Nasarawa	Upper intestines		www.news2.onlinenigeria.com
M	2013	Enugu	Thoracoomphalopagus One heart Joined genitals		www.nigerianuniversitynews.com/2013/06
F	2011	Jos	Parasite twin – no head Chest down 3 legs One liver	Ibadan	www.enownow.com www.dailytrust.info/index.php/city-news/2010
M	2013	Abuja	One intestine		-abuja www.nigerianmonitor.com
F	2004	Abakaliki	??		www.business,highbeam.com/3548/article-IGI -121544947

Surgical separation of conjoined twins that results in the death of one, or both, of the twins raises complex moral, ethical and legal issues. Where organs such as brain or heart are shared there is a great risk of one or both twins dying if attempt is made at separation. Indeed, any shared organ is often not shared equally and the question often arises as to who should be left with what. Of particular concern is the potential for homicide charges against doctors.[2] The parents of the Manchester twins, Mary and Jodie born in 2000, refused to grant permission for surgery, despite the judges' ruling in favour of surgery. A circumstance, where Mary was sacrificed at surgery, was argued by some as "a murder Mary to save Jodie".[7]

Over the last twelve years (1999-2011), three sets of live conjoined twins were documented in our teaching hospital. The hospital has an average annual delivery rate of 1,600 and serves as a major referral center for a population of approximately six million people. There were two thoracoomphalopagus and one omphalopagus twins. One set of the thoracoomphalopagus twins was delivered elsewhere. Both sets of thoracoomphalopagus twins died within 48 hours following birth.

Case

The surviving twins were delivered on the 9th of September 2009 at 02:48hrs to a young couple at the University of Benin Teaching Hospital, in Benin City, Edo State which is in the South-South part of Nigeria. Their mother, a 29 year old lady registered this first pregnancy at our health facility at the 19th week of gestation. Obstetric ultrasound scan done at her antenatal booking revealed that she had a set of omphalopagus twins. The antenatal period remained otherwise unremarkable until the 34th week of gestation when she went into spontaneous preterm labour. She was delivered of live female omphalopagus twins by an emergency Caesarian Section (Fig 1). The twins were both small-for-dates. They had a combined birth weight of 3.4kg but the Apgar Scores were good. They were joined at the level of the xiphisternum to a point just above the umbilicus. There was a small exomphalos with separate umbilical cords. They both had mild respiratory distress syndrome which resolved within 72hrs following delivery. On the third day of life they developed jaundice requiring phototherapy: the highest bilirubin levels were 13.2 for twin 1 and 13.5mg/dl or twin 2. They were treated for Escherichia coli sepsis with ciprofloxacin and gentamycin guided by the antibacterial sensitivity. By the third week of life they had shown evidence of full recovery and had regained their combined birth weight. From the fourth week they were on full milk feeds and had satisfactory growth.

Thoraco-abdominal CT scan revealed that they both shared a single liver and proximal part of the gut. Twin I had dextrocardia without any functional abnormality. Extensive evaluation of the other systems was normal.

They remained in our newborn unit, until the age of nine months. Their combined weight was 9.8 kg, and a separation surgery was then performed at the Narayana hospital in Bangalore, India. Findings at surgery included a single liver that was "fused" in the midline with separate blood supplies. There was a common gallbladder and bile duct that emptied into a common duodenum which extended up to about 20cm in length. Each twin had her own stomach and jejunum. The liver was divided and biliary reconstruction procedure done for twin II. The duodenum was shared between the two by resection and re-anastomosis. The twins required initial mechanical ventilation and were weaned off by the 4th day. Post surgery they remained stable and were transferred back to the UBTH (fig 2). Physical therapy was instituted to enable them "catch-up" with their motor development that was hitherto made difficult whilst conjoined.

Twins I and II weighed 4.7kg and 4.8kg respectively after surgery. At 2 years postnatal age they weighed 8.5 and 8.6kg respectively while at 3 years they weighed 13.4kg and 13.6kg. Their psychomotor development was compatible with their age at 18 months using the Bailey Developmental Scale. Thorough clinical and laboratory re-evaluation at age three, paying particular attention to the cardiovascular, digestive and renal systems of the twins yielded normal findings.

Fig 1: The conjoined twins at two weeks of life

Fig 2: After surgery

Twin I

Twin II

Discussion

This case is a report of the first surviving conjoined twins in a decade of conjoined twins history in this center. Overall reporting of conjoined twins is low in the country. Review of available literature showed that 18 cases have been reported across the country in the last 76 years from 1935 to 2012. Five cases, 28%, were reported from a single center in the North, Zaria,[8] while the other cases were reported from Ibadan,[3] Ife,[9] Enugu[10] and few tertiary health centers in other parts of the country. The higher report from Zaria may be due to the heightened interest of the workers. None so far has been reported from Benin and some other tertiary centers across the country to enable a more countrywide data review. In contrast, 22 cases were reported from a single institution in Philippines, over a 30 year period (1974-2006).[11] An institution in Sao Paula, Brazil reported 14 cases over a 25 year period further reflecting

possible underreporting in Nigeria.[12] It is hoped that this report will be an important contribution to the few existing publications in the country. Although a small number of centers in the country have reported survival of conjoined twins, of note is that information on follow up morbidity and mortality were generally lacking. Our surviving conjoined twins were followed up for catch-up growth, psychomotor development, presence of organ dysfunction and possible late complications of the surgery. All these parameters turned out to be normal at the age of three years. Due to the complexity of the surgical separation, a follow up to evaluate long term survival and quality of life is useful reviewing surgical intervention in the future. In the recent case of separation of heteropagus twins in Maiduguri,[13] the twins had major reconstructive surgery, consequently, long term follow up of these twins will be complementary to our knowledge.

All three sets of conjoined twins seen at our center were females, which is in keeping with the female preponderance noted in the literature. Four out of the six babies suffered early neonatal deaths while 2 (index twins) survived at 3-year follow up. Due to paucity of data it is difficult to say, with any degree of accuracy, what the still birth rate or neonatal death rate for conjoined twins in the country is. Three of the reported 18 pairs where still born, all the reported live born had surgery, 11 in Nigeria and 4 abroad (Table I). Six out of the eight babies (75%) operated abroad survived (one baby was sacrificed to save the other twin). Twelve out of 22 babies (54%) operated in Nigeria survived. Success rate was fairer with ischio/pygopagus twins (58.3%) and poor with thoraco/omphalopagus twins, especially when internal organs are shared.

A careful review of the current case with surgeons in our institution and consult with other surgeons within and outside the country informed the choice of having the surgery done abroad to improved the chances of survival of both twins considering the shared organs. This reflects the need to build on the already existing capacity to handle such cases.

While some have questioned the decision to separate conjoined twins "when two are born as one",[2] having the twins separated may seem justified if it is adjudged that one or both twins would die without separation. This is the case in some heteropagus twin situations in which the parasite twin may die and/or cause the host twin serious physiologic embarrassment due to vascular, biliary or enteric anastomosis. Even though our omphalopagus twins are likely to survive into adulthood, the decision to separate was preferential because of the expected favourable outcome following surgery. In contrast to reports from other parts of the world, the author is unaware of any report of adult conjoined twins in the country. This situation may not be surprising as these babies might have been deprived of care and left to die largely because of stigma, poverty and ignorance. The current case required a lot of psycho-social support for the young parents who had initially abandoned these babies. On the contrary the Biddenden Maids, born in England in 1100, were famous and lived for 34 years. The Siamese twins were also wealthy and famous in the United States. We suggested that providing national awareness, special government support and opening national conjoined twins' registry will go a long way in improving the outcome of these babies, especially when surgery pose a survival risk to one or both twins.

Conclusion

There is under reporting of conjoined twins in Nigeria compared to other parts of the world. Experience from available literature showed that these can be largely prevented by demystifying the condition, providing more awareness and support for the families. These measures will go a long way to improving reporting as well as enhancing the survival of these babies. Secondly paediatricians and surgeons in Nigeria might want to review their decisions to separate when the risk to one or both twins is greater than the risk without the procedure.

Authors' contribution

Amuabunos AE, Eregie CO, Omoigberale AI Effiong V: All managed the patient and reviewed the manuscript

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