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## Surgical management of pygopagus parasiticus in a developing country: Challenges and review of the literature

A.O. Ademuyiwa<sup>a,c,\*</sup>, E.O. Alabi<sup>b</sup>, H.O. Idioidi-Thomas<sup>c</sup>, O.B. Bankole<sup>d,e</sup>, O.A. Elebute<sup>c</sup>, F.M. Alakaloko<sup>c</sup>, I. Desalu<sup>f,g</sup>, C.O. Bode<sup>a,c</sup><sup>a</sup> Paediatric Surgery Unit, Department of Surgery, College of Medicine, University of Lagos, Lagos, Nigeria<sup>b</sup> Orthopaedic and Trauma Unit, Department of Surgery, Lagos University Teaching Hospital, Lagos, Nigeria<sup>c</sup> Paediatric Surgery Unit, Department of Surgery, Lagos University Teaching Hospital, Lagos, Nigeria<sup>d</sup> Neurosurgery Unit, Department of Surgery, Lagos University Teaching Hospital, Lagos, Nigeria<sup>e</sup> Neurosurgery Unit, Department of Surgery, College of Medicine, University of Lagos, Lagos, Nigeria<sup>f</sup> Department of Anaesthesia and Intensive Care, Lagos University Teaching Hospital, Lagos, Nigeria<sup>g</sup> Department of Anaesthesia and Intensive Care, College of Medicine, University of Lagos, Lagos, Nigeria

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## ABSTRACT

Conjoint twins are a rare aberration of monozygotic twinning which could be symmetric or asymmetric. Pygopagus parasiticus is a form of asymmetric twinning in which the parasitic or incomplete twin is attached to the autosite (dominant twin). We present our recent experience in the management of a case of pygopagus parasiticus highlighting the challenges peculiar to developing economies such as ours and how such challenges could be surmounted.

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Conjoined twins are a rare aberration of monozygotic twinning which has fascinated humanity and challenged the medical community for centuries. While symmetric conjoined twins comprise a set of equally developed twins with varying degrees of anatomic union, asymmetrical conjoined twins are characterized by a severely defective or incomplete twin (parasite) attached to and dependent on its largely intact co-twin (autosite). Asymmetric twins are also referred to as heteropagus or parasitic twins. The point of attachment or conjunction is the basis for the classification of the different anatomic types of conjoined twins [1].

A major challenge to the management of this anomaly lies in its relative rarity and thus low individual surgeon exposure. In fact, a lot of the published work to date is based on isolated case reports [2]. Many recent reports from developed countries do not take into account peculiar challenges in the low and middle income

countries (LMICs). Our recent experience in the successful management of this case aims to highlight such challenges and how they can be surmounted as well as review existing literature on the subject.

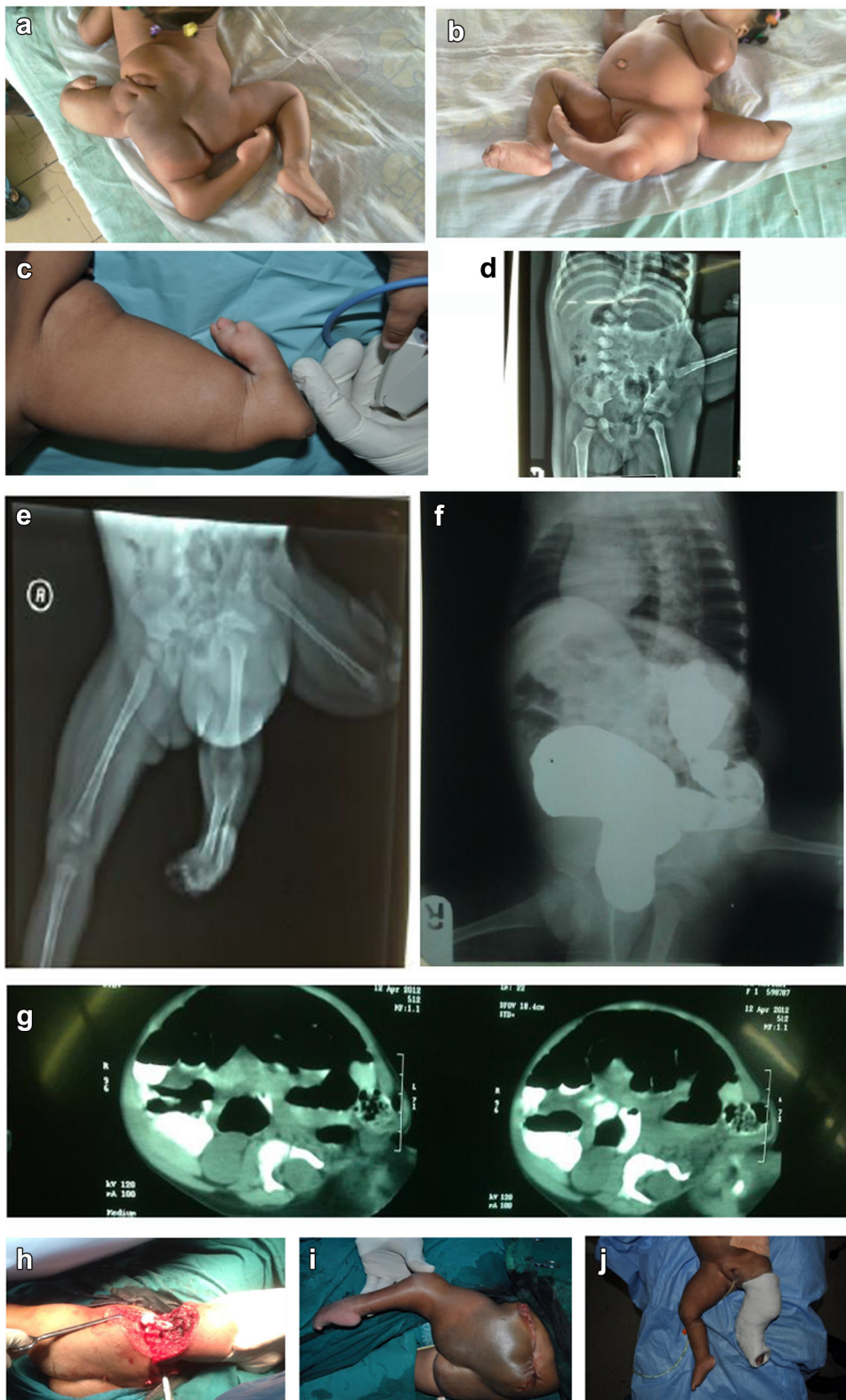
## 1. Case report

The patient is a 16 month old female child from a low socio-economic background with dysmorphic features since birth which included median cleft palate, mandibular mass with intra-oral extension, bifid tongue and accessory, malformed lower limbs. She had elective tracheostomy, excision of mandibular mass and repair of bifid tongue at the age of 3 months before being referred to our unit. Index presentation was to address the additional lower limb which had increased in size as she had grown with associated ambiguous movement. At 16 months, the patient could not sit without support, could not crawl nor walk. She was constantly being wrapped up by her mother to hide the three lower limbs.

In the antenatal period, patient's mother took herbal concoctions and never received orthodox medical care. Delivery was term,

\* Corresponding author. Paediatric Surgery Unit, Department of Surgery, College of Medicine, University of Lagos, Lagos, Nigeria. Tel.: +234 705 5787 110.

E-mail addresses: [adesojiademuyiwa@yahoo.co.uk](mailto:adesojiademuyiwa@yahoo.co.uk), [aoademuyiwa@cmul.edu.ng](mailto:aoademuyiwa@cmul.edu.ng) (A.O. Ademuyiwa).



**Fig. 1.** (a–c) pre-operative pictures of index patient demonstrating lower limbs including parasitic limb. (d) and (e): shows kyphoscoliosis with concavity to the left, midline spinal defect in T10–L5 vertebral bodies extending down to the sacrum but covered by soft tissue (spina bifida occulta); a left accessory pubic–ischial complex with a corresponding 3rd (appendage) femur attached which was not connected to a tibia or fibula. The left femur, tibia and fibula were shortened. The right femur, tibia and fibula were normal. (f) contrast enema outlining the large bowel with no communication with the external dimple. (g): abdominal computerized tomography scan showing normal gastrointestinal tract, spina bifida occulta and a left posterior abdominal wall weakness with herniation. (h–j) Intra-operative and immediate post operative pictures of index patient demonstrating disarticulation, increased range of knee movement following soft tissue release and left above knee POP cast.

at home; per vaginam with no assistance as had been the mother's practice in the delivery of her other children. All gross anomalies were noticed at birth. She is the 7th of 8 children in a monogamous setting. Her mother is a 40 year old petty trader and her father is a 47 year old Islamic cleric. The gross monthly income for the family is approximately 250 USD. They all reside in a 3 bedroom rented apartment. There was no family history of congenital anomalies or twinning until index case.

Physical examination showed a playful child with ocular hypertelorism, healed scars on the face and neck from previous surgeries, and normal chest, cardiovascular and abdominal examination findings. She had a normal right lower limb, a hypoplastic left lower limb with fixed flexure deformity at the knee and talipes equinovarus. She had an extra left lower limb stump consisting of a thigh and rudimentary foot (Fig. 1a–c).

The parasitic thigh demonstrated both active and passive movement as would be expected from a ball and socket proximal joint. At the proximal attachment of the accessory limb were 2 dimples suggestive of a rudimentary female perineum. Based on the posterolateral attachment of the accessory limb a diagnosis of *pygopagus parasiticus* was made.

Several multidisciplinary pre-operative conferences were held by the pediatric surgeons with the orthopedic, plastic and cardiovascular surgeons as well as anesthetists to plan removal of the parasitic limb. Investigative work up required to plan surgery for this patient was severely constrained due to finances. It took the concerted efforts of hospital management, social workers, charitable organizations and individuals to be able to bear the cost of pre-operative investigations and subsequent management. This translated to several months' delay in investigating and eventually operating her. There was also the ethical challenge of convincing the family and financial sponsors that she had a right to live a full, functionally independent life and not be abandoned to a dependent existence or worse due to her gross physical anomalies. The parents were already totally financially depleted after the first surgery at 3 months and several counseling sessions were held with the parents to get them not to give up on further care and support of this child. Some of the investigations performed included radiologic investigations for pre-operative planning:

- (i) X-rays of the lumbosacral spine, pelvis and lower limbs (Fig. 1d, e) showed: kyphoscoliosis with concavity to the left, midline spinal defect in T10-L5 vertebral bodies extending down to the sacrum but covered by soft tissue (*spina bifida occulta*); a left accessory pubic–ischial complex with a corresponding 3rd (appendage) femur attached which was not connected to a tibia or fibula. The left femur, tibia and fibula were shortened. The right femur, tibia and fibula were normal.
- (ii) Barium enema (Fig. 1f) was normal; and in particular there was no communication with the dimples in the rudimentary perineum proximal to the accessory limb.
- (iii) Triple contrast abdominal computerized tomography scan (Fig. 1g) confirmed a normal gastrointestinal tract, *spina bifida occulta* and a left posterior abdominal wall weakness with herniation of a low lying left kidney and some bowel loops. The right kidney was normal in location. Both kidneys showed normal excretory function.

Magnetic resonance imaging and arteriography were not done on account of cost which is a major limiting factor in LMICs such as ours although these investigations are superior to CT scan in outlining soft tissue outline and vascular anatomy. In the absence of a

pre-operative roadmap of the vascular anatomy of the accessory limb, blood products were cross matched ready for possible massive intra- and post-operative transfusion. Preoperative full blood count and serum electrolytes were normal.

Intravenous access was secured via an upper limb venous cut down the night before surgery and an internal jugular central line once the patient was under anesthesia. At surgery (by pediatric and orthopedic surgeons), the openings in the rudimentary perineum were confirmed to be blind ending with no communication with autosome hollow viscera. An elliptical incision was made to encompass the rudimentary accessory limb and blind ending stomata. The limb was excised by disarticulation at the ball and socket joint. Soft tissue release as well as manipulation and plaster of paris (POP) application for the residual left limb was then done (Fig. 1h–j). She did well post operatively and had serial manipulations and cast re-applications over a 4 month period before being lost to follow-up. She is scheduled for a limb lengthening procedure by the orthopedic surgeons and continuous physiotherapy. However, phone conversations and counseling sessions to bring her back for follow-up and further management have so far been fruitless.

In the absence of progressive neurologic deficit, no neurosurgical intervention was planned for the *spina bifida occulta* at the time she defaulted from her outpatient clinic visits.

## 2. Discussion

Conjoined twins have been and continue to be regarded as a medical curiosity. The exact etiology is unknown, the spectrum of presentation is wide and management of this anomaly still poses a challenge today. Ambroise Paré, a French surgeon, was among the first to describe different varieties of conjoined twins in the 16th century, but figurines and other prehistoric art forms with the appearance of conjoined twins date back as far as the 6th millennium BC [1,3].

In one of the largest recorded series, Edmonds and Layde [4], found a crude incidence of 10.25 conjoint twins per million live births in the United States, with parasitic twins making up just 10%. This is similar to the estimated incidence of parasitic twins being about 1 case per million live births quoted in more recent reports [1,2]. It is likely that the rarity of this condition has contributed to its fascination amongst different cadres of health workers. For pediatric surgeons who play a major role in separating these twins, the peculiar challenges faced with each anatomic type and the innovations developed in handling these difficulties are also of interest.

### 2.1. Challenge of limited surgical experience

Worldwide, there have been about 210 documented cases of *heteropagus* twins in the medical literature to date [1,2,5–16]. (157 cases reported in the literature up to 2003 [1,2], 39 more from 23 articles collated up to 2010 by Sharma and colleagues [1,2], and we found 14 more cases from 12 case reports in the literature between 2010 and 2013 [5–16]) Our index patient will add to these cases. This number however trickles down to usually less than 4 cases per individual surgeon (in a lifetime) in most centers with a few exceptions in large specialist centers. Dr. James O'Neil, emeritus professor of surgery, has participated in the surgical separation of 23 sets of conjoined twins (symmetric and asymmetric) and this is probably more than any other surgeon [17].

While it is desirable for the most experienced surgeons to handle these rare congenital anomalies [18], this may not always be feasible considering the specialist/patient ratio which for pediatric surgery in sub-Saharan Africa is about 1:5,000,000 [19]. The index

patient was the first asymmetric conjoined twin operated on by the authors (except COB).

A tested method that has improved outcome in the management of rare anomalies is to concentrate care into certain centers such that surgeon's expertise is improved with increased work load of these rare anomalies. This method has worked well in the management of biliary atresia in the UK [20]. A good approach by developing countries may be to copy this model for the management of conjoined twins in order to obviate the limited surgeon experience in the management of conjoined twins.

### 2.2. Challenge of poor health insurance coverage

In most developing countries including Nigeria, health services are accessed on fee-for-service basis. Although the Nigerian Health Insurance Service (NHIS) scheme has been operational since 2005, coverage is less than 4% which constitutes mainly civil servants and some of the organized private sector [21]. In addition, poverty is pervasive in many LMICs, and as a result, many patients cannot afford cost of health care.

This was a major challenge in the index patient as presented earlier. At a time, the hospital management had to underwrite the cost of the hospital bills completely as the patient was on admission for over 10 months with no means in sight of upsetting the increasing bills. It is vital that policy makers in health care delivery in developing countries increase the coverage of health insurance schemes to mitigate against this challenge. In the interim, significant subsidized rates should apply to cases of this nature and hospital managements should establish grants and specialized funds that can be accessed when cases like this arise.

### 2.3. Challenge of ethical considerations

Surgery for symmetric conjoined twins is aimed at saving two lives even though this may not always be possible. This brings in an ethical dilemma aptly discussed by Raffensperger [22] and Atkinson [23] in separate articles. In asymmetric conjoined twins, this ethical dilemma is removed as surgery is aimed at complete excision of the parasite to preserve the autosite [16]. As such; no medical sacrifices are made on the autosite while the aim is to completely remove the parasite. However, other ethical issues which we experience in the sub-region include stigmatization, cultural and religious taboos, abandonment of the patient as well as low socio economic status being a determinant of treatment as in the index patient.

It is important to have in-depth counseling sessions with the parents to enlist them as allies in the management of this medical curiosity. In our patient, several multidisciplinary based discussions were held with the parents to explain the epidemiology, possible etiology and likely outcome. Parents' mind were disabused from superstitious beliefs and metaphysical influences as a cause. Parents were encouraged to ask questions and some of these questions stemmed from what family members and acquaintances raised with them. The parents' cooperation was reassuring and this made it easy for the managing teams to proceed with the perceived best mode of care.

Surgical separation of the parasite from the autosite is technically less challenging than the separation of symmetric conjoined twins as they rarely share major organs and blood vessels, unlike the latter [1,2]. However, the distorted anatomy of the autosite-parasite union as well as associated congenital anomalies in the autosite are challenges worthy of adequate pre-operative investigations and multidisciplinary pre-operative planning. The need for detailed preoperative evaluation (including imaging studies in particular), multiple specialists' consultations and pre-operative conferences had already been emphasized as far back as

1988 by O'Neil and colleagues [18]. In our index patient, the confirmation of a normal gastrointestinal tract both on a barium enema as well as a triple contrast abdominal CT scan allowed us to excise the parasite rudimentary perineum without creating intestinal complications in the autosite. Also, the herniated ectopic left kidney could not have been detected without imaging studies which saved us from possible on-table catastrophe.

### 2.4. Challenge of multiple surgeries and follow up

Surgical separation of parasitic twins is often a single event. It is important to ensure the complete excision of the parasite in order to avoid need for further surgery to remove the remnant from the autosite. Occasionally, as in our patient, other associated congenital anomalies may be present in the autosite that require further surgeries. In our index case, prior surgery had been performed for median cleft palate, a bifid tongue and mandibular mass before being referred to us. It is also clear that further surgeries are needed to correct the shortening of the left lower limb.

From the foregoing, close follow up of these patients is important. However, clinical follow-up after initial intervention has been a major challenge in most developing countries [24–26]. In spite of the goodwill the patient enjoyed while on admission, it has been difficult to convince the parents to bring the patient for the necessary surgery to correct the limb deformity at no cost to them. It may be necessary to intensify efforts on health education and having a robust social work paradigm to overcome this scourge in LMICs.

## 3. Conclusion

The management of parasitic twins in LMICs poses several peculiar challenges. A multidisciplinary team approach with dedicated specialist hospitals to cater for rare conditions such as parasitic twins, better health insurance coverage and robust health education and social work programs may improve outcomes of patients in the face of such challenges.

### Conflict of interest

None declared.

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None.

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