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## Management of pyopagus conjoined twins

Received: 16 March 2004  
Published online: 15 July 2004  
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**Abstract** *Introduction:* Approximately 18% of conjoined twins (1 in 200,000 live births) are joined at the sacrum ("pyopagus"). As the joined structures are not life-threatening, there is generally a good prognosis, with time for investigation and planning of operative separation. *Case report:* This paper reports on the management of pyopagus twin girls, the first in the UK delivered at 36 weeks by Caesarean section. The diagnosis had been made by ultrasound at 12 weeks' and confirmed by MRI scan at 26 weeks' gestation. Each twin had wasting and weakness below one knee, but no deficit in the other leg. The perineum had two urethras but only one anus. One infant had a colostomy in the immediate neonatal period. With appropriate further imaging, surgery was planned in two stages. First, a balloon expander was inserted in the bridging area. At 3 months of age a team comprising Plastic, Paediatric and Neurosurgeons undertook the definitive separation. *Discussion:* Details of the multidisciplinary planning, operative separation and follow-up of these two children will be discussed.

**Keywords** Conjoined twins · Pyopagus

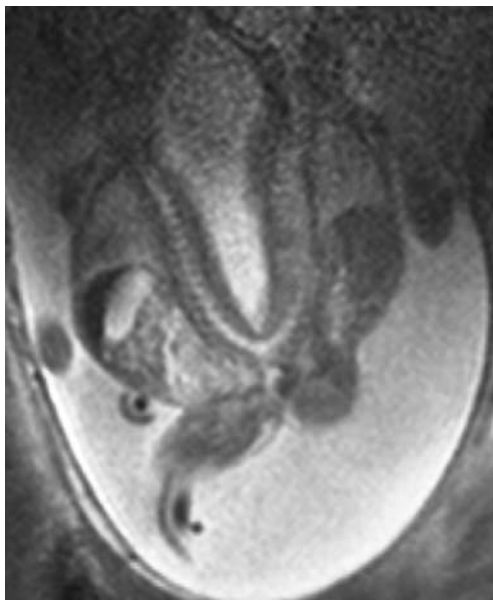
## Introduction

The incidence of conjoined twins is estimated to be 1 in 50,000, but, as around 60% are stillborn, the true incidence is 1 in 200,000 live births [7]. Girls predominate with a 3:1 ratio. Twins are classified according to the most predominant site of connection, most frequently front to front, with thoracopagus and omphalopagus accounting for approximately 75% of cases. The term “pyopagus” (in earlier literature and strictly speaking more correctly “pygopagus”) is given to twins joined at the sacrum and accounts for approximately 18% of cases.

There have been several successful surgical separations of pyopagus twins reported [1, 2, 4, 5], but only a limited number having conjoined spinal cords (J. Peter, personal communication). Anaesthesia for separation of conjoined twins has also been previously well described [3, 6]. The following case report represents the first experience of such a case in the UK. Pyopagus twins generally have a good prognosis as the joined structures are not life-threatening. There is time for the children to thrive, have appropriate investigation and plan operative separation.

## Case report

Conjoined twins joined at the sacrum (pyopagus) were diagnosed at 12 weeks' gestation by ultrasound and confirmed at 26 weeks by MRI scan, which demonstrated fusion of the lower spinal cords (Fig. 1). The parents were informed and, in addition to attending the antenatal clinic, were also seen for consultation in the Paediatric and Neurosurgery clinics. After discussion by all concerned it was planned that the twins would be delivered by Caesarean section at



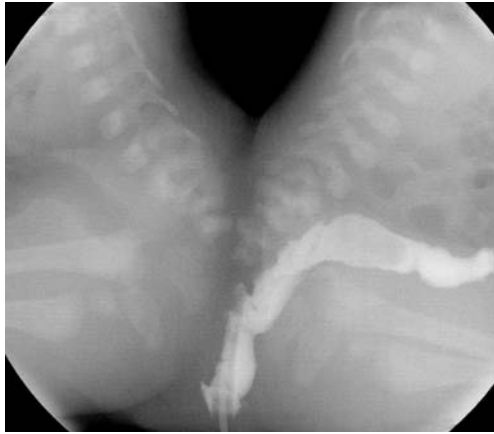
**Fig. 1** Antenatal MRI scan at 26 weeks



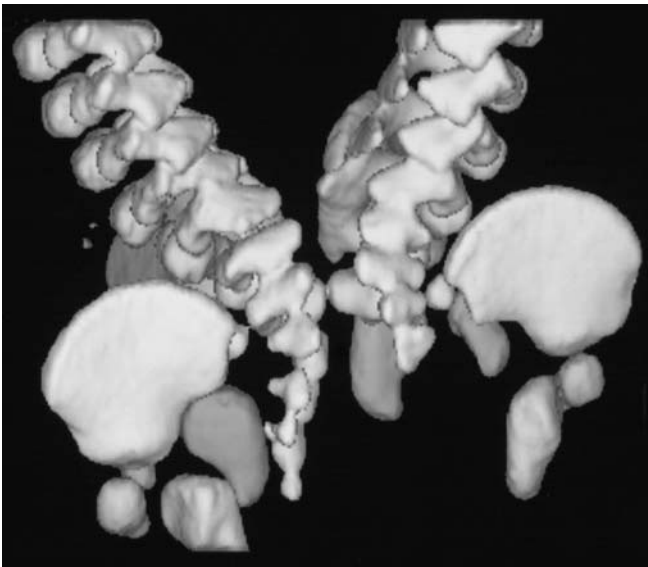
**Fig. 2** Conjoined twin girls at birth

or after 36 weeks and that surgical separation would be the aim of management. The twins were in fact delivered at 36 weeks on account of maternal toxæmia. The twin girls were lively at birth (Fig. 2), weighting 4.5 kg in total. Initial examination confirmed the join at the lumbosacral level with abnormal perineal anatomy. Essentially, the two vaginas lay back to back, and each infant had an urethra. One twin had no anus, and there was meconium visible from the anal orifice of the other twin. Each baby had a normal leg neurologically with full movements and no wasting. The right leg in one baby and the left in the other did show neurological deficit below the knee, which included wasting in the calf and reduced movement in the feet with some deformity.

Once delivered and stabilised, the conjoined twins were transferred to the Children's Hospital. The first priority was to identify the anatomy of the lower intestinal tracts, so that both infants could receive adequate nutritional intake. A retrograde contrast study identified which twin's intestine was connected to the single anus (Fig. 3) and on the 2nd day of life the other baby had a transverse colostomy. Both twins needed to be anaesthetised for this procedure and this was a useful rehearsal for the same anaesthetic team who were to be involved in the definitive separation. Under a further general anaesthetic, appropriate CT and MRI scans were undertaken (Figs. 4, 5). A detailed first multidisciplinary planning meeting then followed, with representation from all specialities that had been and were continuing to be involved in the care of these children. It was decided to plan separation at or after 3 months if all was well. In order to facilitate skin closure at the time of separation,



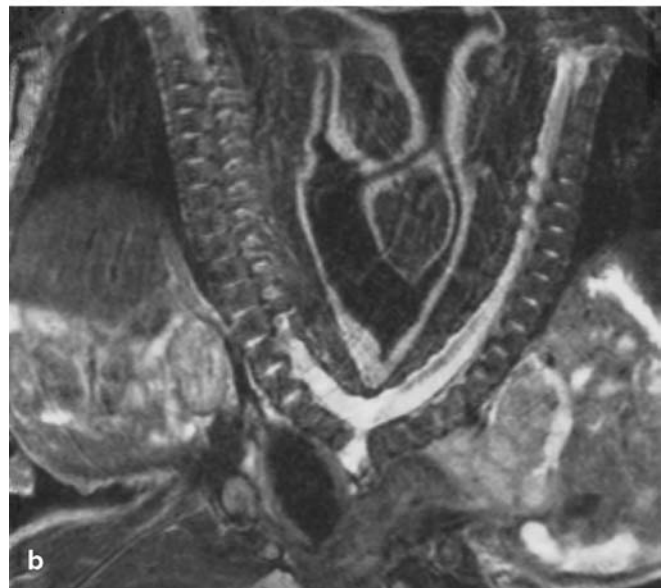
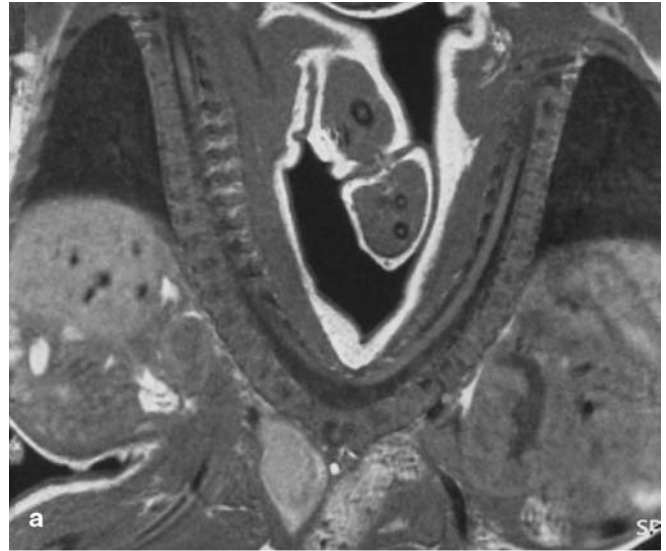
**Fig. 3** Retrograde water-soluble contrast study to demonstrate one infant with continuity of large bowel to the anus



**Fig. 4** CT scan to demonstrate relationship of lower bony spinal canals

it was decided as a preliminary to insert a tissue expanding balloon beneath the joining skin bridge.

Accordingly, when aged 1 month, this procedure was undertaken and repeated saline inflation of the balloon by medical staff and then parents at home produced a significant additional area of skin. The infants thrived at home. A second planning meeting was held with the agreed surgical plan to deal with the spinal cord separation first, including their dural tube closure, before correction of the perineal anatomy, leading to separation with individual repair and closure of skin defects. The infants would be placed on the operating table in the lateral position, back to back. A preliminary incision on the side opposite the balloon with insertion of a gauze swab in the subcutaneous space was planned, which would be helpful in identifying the surgical "midline" with certainty during the deep part of separation from the other side. This would involve turning the twins over; once finally separated one twin would be taken to the adjacent operating room and both would then be



**Fig. 5 a, b** MRI scans to demonstrate fusion of lower spinal cords

repositioned prone for completion of their operative repair. There were two anaesthetic, neurosurgical, plastic surgery and paediatric surgery teams, one of each designated to accompany each infant. A list was prepared of essential personnel who would only have access to the operating rooms. Closed circuit TV was available for surgical colleagues and trainees.

With regard to anaesthesia, there were three main concerns:

1. The practical approach to the twins in terms of anaesthetic and surgical access to the babies
2. The unpredictable physiological and pharmacological response to each twin under anaesthesia
3. The massive blood loss anticipated in an infant weighing approximately 2.25 kg (with an estimated circulating blood volume of 180 ml) and subsequent coagulopathy

The twins had been anaesthetised on two occasions prior to the surgical separation by the same two anaesthetists. The anaesthetic

access to the twins was therefore familiar to the anaesthetists. The position of the airway and access to the neck veins and upper body arterial venous vessels were all reasonably accessible. One major undertaking was to rotate the twins 180° in order to make the second stage of surgery possible. All the infusion lines and breathing circuits were colour coded in order to avoid confusion during and after this positional change.

There was insufficient time to perform angiography to delineate cross-circulation between the twins prior to the first anaesthetic due to the urgency of surgery. The twins were therefore induced by gaseous induction simultaneously and paralysis of one twin did not lead to paralysis in the second twin. In addition, one anaesthetist as part of the anaesthetic only used nitrous oxide. Nitrous oxide was not detected by end-tidal gaseous analysis in the second twin. As a result of this, it was concluded that no significant cross-circulation exists. This was later confirmed at the surgical separation when the twins behaved as two separate physiological entities.

As anticipated, both twins required massive blood transfusion in order to maintain haemodynamic stability and oxygen-carrying capacity. One twin received 100 ml/kg of packed red cells and the other 80 ml/kg. No other blood products and platelets were clinically indicated.

As planned, the infants were first placed in the lateral position, with the balloon on the underside. A vertical incision was made beneath the skin bridge down to palpable bone and a betadine-soaked ribbon gauze pack inserted. The skin was closed with a continuous nylon suture.

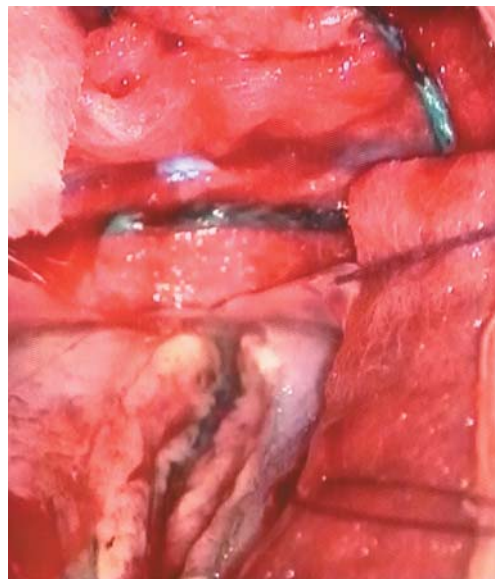
The twins were turned into a reverse position so that the balloon was uppermost. Each infant was now catheterised and electrode needles were inserted into all four limbs and buttocks for the purpose of spinal cord/root monitoring. The operative site was prepared and a vertical incision over the balloon allowed its removal, leaving a well-defined capsule. On deepening the incision the spinous processes and shared posterior sacral bony elements of each infant were exposed. In several areas the bony covering was deficient, allowing incision of the underlying ligamentum flavum. With punch forceps a generous exposure of a "Y"-shaped dura was achieved. With the operating microscope, the dural sac was opened in the midline (surgeon's midline) and extended on each side at the top end. As the MRI had shown, there was a corresponding "Y"-shaped connection of the two lower spinal cords. Nerve roots were seen passing from each side (Fig. 6). A dorsal midline incision was made from the junction point at the top and then down the stem of the "Y", assuming this would be between the posterior columns of each cord. As the incisions deepened it was possible to separate the two spinal cords and expose the arachnoid and dura on the far side (Fig. 7). A traversing bar of bone in the lower part of the exposure, crossing from one infant to the other, with its own dural envelope, had to be excised as it was tethering one of the spinal cords. Essentially, this was a very low "diastematomyelia". Caudal to this bone the filum from each spinal cord was identified and divided.

With complete separation of the cords, there was some fine traversing rootlets at the upper part of the exposure, stimulation of which produced a positive response in one of the twins' right L5 and S1 recording electrodes. They had to be divided in order to fabricate two dural tubes. The dura of the far side was incised vertically and a fascial graft on each side was needed to achieve closure of the separate dural tubes without tension.

The General Surgeons now carried out the perineal separation, essentially separating the rectum of each twin and fashioning a separate vagina for each. In the infant without an anus a narrow recto-perineal fistula was identified. After completion of the perineal separation, it was now possible to divide the bony and ligamentous connecting structures on the far underside of the cords. This was facilitated by visualising the initial ribbon gauze, which was exposed, and effectively the children were now separated, just needing removal of the skin suture from the under surface.



**Fig. 6** Appearance of conjoined lower spinal cord at operation



**Fig. 7** Lower "cords" separated

Both infants were now placed prone in adjoining theatres. In each infant the posterior repair was completed and some of the balloon capsule tissue was incised and used to enhance the two dural closures. Primary skin closure was possible in each infant. The anus and rectum were now appropriately sited in each twin. The whole operative procedure had been of 15 h duration and each baby had effectively had a blood volume replacement. It was elected to continue each infant on assisted ventilation overnight. The following day both babies were well and there was no demonstrable additional motor deficit in either infant. Perhaps the



**Fig. 8 a,b** The twins following their surgical separation

divided nerve rootlets had been sensory with retrograde stimulation, or if motor did not represent the total L5/S1 outflow.

Following surgical separation, the twins (Fig. 8) needed continued multidisciplinary follow-up, with initially at this stage the most involved specialities being orthopaedics and urology. Problems in these areas need careful surveillance to maximise limb and renal function. There are issues for the twin with a colostomy in whom the question may later arise whether to perform a corrective pull-through procedure.

The twin girl who has the colostomy continues to have regular dilatation of her recto-perineal fistula. Currently, this is considered too small to become an anus. She only has some muscle function below her left knee and in the ankle, but wears a special boot to improve foot posture. Now aged 14 months she is crawling, but not yet walking independently. Both her hip joints look normal. She does have crossed ectopia of her kidneys, but without dilatation of their ureters.

The other twin has a right-sided hip dislocation, shortening of her right leg, particularly below the knee, with wasting of calf muscles and a small stiff right foot. She shows very little movement in the ankle or foot. She has a partially fused ectopic right kidney, with a duplex draining system including a dilated ureter. There have been problems with urinary infection. She is sitting independently and has just had an open reduction of her right hip. From a neurosurgical point of view, clinical and imaging follow-up are necessary to assess vertebral growth and spinal cord anatomy.

## Discussion

This paper describes our first experience in managing pygopus conjoined twins. Because the joined structures were not life threatening there was generally a good prognosis with time for investigation and planning operative separation. With reliable antenatal ultrasound and MR imaging, planning for the care of our two patients started before their birth. Good multidisciplinary co-operation was absolutely essential and in the early phase this mainly involved discussion between the surgical team, obstetrician and neonatal paediatrician. Our obstetrical colleague planned the extent and technique for Caesarean delivery using a model of two dolls stuck together. A full paediatric resuscitation team was available when the twins were delivered in the obstetrical unit. Once stabilised later that day the Neurosurgeon and Paediatric Surgeon examined the children with the Paediatrician, and the events outlined above followed.

Because of its rarity, and the special facilities required it was important that these infants were treated in an appropriate specialised paediatric centre. The Birmingham team were very fortunate to be joined by Professor Lewis Spitz with his special experience in the field of conjoined twins. From a neurosurgical point of view, we also very much appreciated the advice given by Professor Jonathan Peter in Cape Town, who had treated an almost identical case shortly beforehand and whose operative video was so helpful.

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