

Case reports

Surgical experience with conjoined twins

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Introduction

One of the most fascinating human malformations is conjoined twins, a rare complex anomaly of the newborn. Commonly known as Siamese twins, this phenomenon is shrouded in mystery and considered a curiosity by the general public. Frequently the twins are born dead but there are a few cases in which the twins survive. They require a highly experienced team and a centre equipped to deal with such challenging anatomy. Three sets of complete (thoraco-omphalopagus) and asymmetric, parasitic (heteropagus) conjoined twins were managed by our team over a three year period from 2006 to 2008.

Case report 1

A 30 year old mother who went into preterm labour delivered conjoined twins following an emergency caesarean section, at Base Hospital Polonnaruwa. She had no family history of congenital anomalies and had taken no medication during her pregnancy.

The twins were transferred to the Lady Ridgeway Hospital (LRH) the following day after initial resuscitation. One of the twins had sustained a fracture of left femur at the time of delivery. Although the twins appeared well during the first 12 hours after birth, they developed respiratory distress during the transfer and had to be intubated and ventilated soon after arrival at the surgical intensive care unit (ICU) at LRH.

On clinical examination it was noted that the twins were attached from the level of xiphisternum downwards. The twins, both females, were joined from the thorax and abdomen (thoraco-omphalopagus) with one common umbilical cord (Figure 1).

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Figure 1: *First conjoined twin*

They weighed 2.3 kg together and the smaller baby had poor saturation in the lower extremities on admission. The echocardiogram revealed a complex congenital heart disease in one of the twins, which was not compatible with life (single ventricle, overriding aorta and pulmonary hypertension). Two separate hearts were identified in a single pericardium sharing a single thoracic cavity. The liver was fused together in the midline with a common portal venous drainage.

Both children were put on tube feeding and passed urine and stools normally from separate external openings. After 5 days, the condition of the smaller twin started deteriorating, showing signs of sepsis and poor perfusion. By the 10th day this twin with the complex heart disease started deteriorating and a team of surgeons decided to sacrifice this twin to save the other twin. The team agreed to proceed with a semi-urgent separation.

After 12 days, clinical condition of the smaller twin deteriorated rapidly and had to be sacrificed to save the other twin by a team of paediatric surgeons and a cardiothoracic surgeon. After an 8 hour complex surgery the liver, hepatobiliary and gastrointestinal systems were successfully separated. As both hearts were covered with a single pericardium, the anterior

chest wall was only partially closed. As expected, the twin with the complex heart disease did not survive after separation. After surgery the other twin was managed in the surgical ICU. Initially, the child tolerated surgery very well but on the following day showed signs of cardiac failure due to the poor contractibility of heart due to its position and died.

Case report 2

Conjoined parasitic male twins (one well developed and the other underdeveloped) were delivered normally per-vagina to an 18 year old mother at Base Hospital, Akkaraipattu. Examination revealed a parasitic type of conjoined twins with a union of the lower trunk from the umbilicus downwards (Figure 2). One twin was small, less formed and dependent on the other. The parasitic twin was half formed with a rudimentary head, small thorax, abdomen and pelvis with intestines outside.



Figure 2: *Second conjoined twin*

Detailed ultrasound scanning of the twins for anomalies, revealed normal findings except the parasitic twin at the sacral region. The parasitic twin contained irregular lower limbs which moved freely. Preliminary imaging investigations were performed to identify the lower gastrointestinal and urinary system which appeared to be separate.

The parents were counseled and the baby underwent a successful separation on day four of life and made an unremarkable recovery. He was transferred back to the local hospital during the following week after removal of the urinary catheter and postnatal follow-up was normal at three months.

Case report 3

Another case of partially conjoined parasitic twins born to a 28 year old mother from Avissawella was

encountered with an extra portion of a pelvis and lower extremity attached to the back of the upper chest (Figure 3). The parasitic lower limb was totally excised. Post-operative period was uneventful and the newborn was discharged as healthy. The nine month postnatal follow-up was normal.



Figure 3: *Third conjoined twin*

Discussion

Conjoined twinning is a fascinating congenital abnormality with an incidence of one in 50,000 to 1 in 100,000 births. As many as 70% are girls. Neither incidence nor prevalence in Sri Lanka is known. The overall survival rate is around 25% as about half of conjoined babies are stillborn due to their deformities being incompatible with life¹.

Unfortunately, in all the cases reported here, abnormalities were not detected during the antenatal period. Early prenatal diagnosis and precise characterization of conjoined twins are essential for optimal obstetric, interventional and postnatal management as well as to reduce psychological trauma to the parents². Early prenatal diagnosis may change the concept of management of conjoined twins. As was seen in our first set of twins, birth trauma sustained could have been avoided. Planned delivery by elective caesarean section around 36 weeks is currently recommended if the fetuses are mature³. Although the latter two cases in this report were delivered vaginally, caesarean section is recommended in most third-trimester deliveries because of the high incidence of uterine dystocia and resultant fetal damage³.

The classification of conjoined twins is based on the site of union. The suffix “pagus” is commonly used to mean “fastened”. Thoracopagus has a shared thorax and 90% have a shared heart. Omphalopagus has a shared abdomen. Thoraco-omphalopagus, has a

shared thorax and abdomen and is one of the most common types. Ileopagus is connected at the iliac bone. Asymmetric and parasitic conjoined twins are rarer anomalies of monochorionic monoamniotic twins, consisting of an incomplete twin attached to the fully developed body of the co-twin.

Separation of conjoined twins is a complicated procedure. The importance of a multidisciplinary team with rehearsal of all aspects (surgical, anaesthetic and nursing) of the operative procedure cannot be overemphasized⁴. Although the outcome is influenced by careful planning and organization from all participants, the prognosis is often predetermined by the underlying anatomy which may preclude successful separation⁵. Delivery at a tertiary centre is recommended for optimal neonatal intensive care and paediatric surgical intervention.

References

1. Spitz L, Kiely EM. Experience in the management of conjoined twins. *British Journal of Surgery* 2002; **89**:1188-92.
2. Abdullah Al Rabeeah. Conjoined twins – past, present, and future. *Journal of Pediatric Surgery* 2006; **41**:1000-4.
3. Bondeson J. Dicephalus conjoined twins: a historical review with emphasis on viability. *Journal of Pediatric Surgery* 2001; **36**:1435-44.
4. Golladay ES, Williams GD, Seibert JJ, Dungan WT, Shenefelt R. Dicephalus Dipus Conjoined Twins: A surgical separation and review of previously reported cases. *Journal of Pediatric Surgery* 1982; **17** (3): 259-64.
5. Miller D, Colobani P, Buck JR, Dudgeon DL, Haller JA. New techniques in the diagnosis and operative management of Siamese twins. *Journal of Pediatric Surgery* 1983; **18** (4): 373- 6.