Anaesthetic management of a patient with thoracopagus

[a] **Kiran S**, MD, DNB
[a] **Kaur KP**, DA, DNB
[b] **Rattan KNR**, MS, MCh
[b] **Rattan SK**, MS, MCh
[a] Department of Anaesthesiology and Critical Care, Pt BD Sharma PGIMS, Rohtak, India
[b] Department of Paediatric Surgery, Pt BD Sharma PGIMS, Rohtak, India
Correspondence to: Dr Kiranpreet Kaur, e-mail: kiranpreet72@rediffmail.com
Keywords: thoracopagus; anaesthetic management

Abstract

We report the case of a 10-day-old male child weighing 3.5 kg, with mass over the sternal region, a set of four limbs and an omphalocoele that had undergone surgical separation. An exoparasitic twin had fully developed hind limbs, well-developed genitalia, one fully developed upper limb and another, underdeveloped upper limb. Echocardiography and a computed tomographic scan revealed no gross cardiac anomaly and the sharing of any other major organ was absent. The limbs of the parasite were lying in front of the neck and interfered with holding the mask in position. One anaesthetist held these limbs apart. We avoided the use of muscle relaxant out of fear that the large mass could hamper ventilation. The neonate was intubated successfully under deep inhalation anaesthesia. He had an uneventful recovery.

Ⓟ Peer reviewed. (Submitted: 2009-07-19, Accepted: 2010-06-04) ***S Afr J Anaesthesiol Analg 2010;16(4):33-36***

Introduction

Conjoined twinning is one of the rarest congenital anomalies. Because of its rarity, the terminology describing the types of malformation is difficult for most physicians to remember. The most famous conjoined twins were Eng and Chang Bunker, who were born in Thailand in 1811. They were called "Siamese twins". Since then, this term has been the best known for all kinds of conjoined twins.[1] Unequal and asymmetrically joined twins are termed as heteropagus. Heteropagus twins can be endoparasitic and exoparasitic. "Endoparasitic" is the type in which the abnormally developing embryo parasites the normal co-twin (autosite) by attaching to it internally. In the case of an exoparasite twin, the parasite is attached to the visible surface of the autosite.[2] Although the incidence of incomplete or heteropagus twinning is very rare, estimated to be 1 per 2 million live births, endoparasite twins are more common than exoparasite twins. It has been suggested that both environmental and maternal factors may play an important role in this regard.[3] There is a higher incidence of conjoined twins in Southeast Asia and in certain parts of Africa.[4] Intrauterine hypoxia and malnutrition have also been suggested as aetiological factors. An incomplete and asymmetric fission of inner cell mass and, more recently, "fusion theory" have been put forward.[3,5]

Several classifications of conjoined twins have been suggested. The classification by Potter is the most widely used (see Table I).[1]

Thoracopagus is the most common type of conjoined twins and occurs when twins are joined at the level of the thorax. Successful separation of thoracopagus twins, without sacrificing either of them, is dependent on the severity of cardiac involvement.[2,6,7] Conjoined twins are united at the level of the upper abdomen (xiphipagus), at the back (pyopagus), side to side at the level of the pelvis (ischiopagus), and head to head (craniopagus).[1,3,4] The first successful separation of conjoined twins was reported by Konig in 1689.[4] Surgical and anaesthetic management is often challenging in such patients, especially in patients having thoracopagus, because they may have shared cardiac chambers. Here we report on the anaesthetic management of a thoracopagus parasitic twin posted for surgical separation.

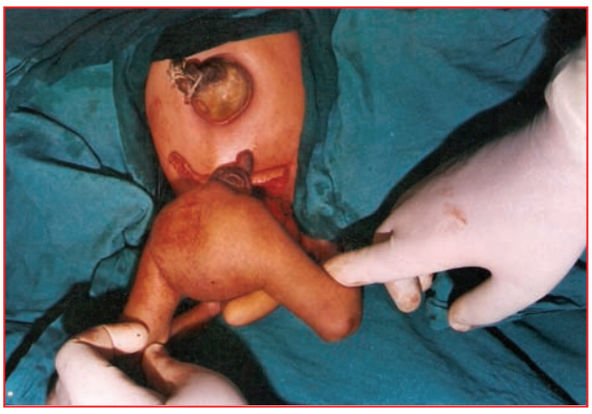
Table I: Classification of conjoined twins (modified from Potter)[1]

FORMS OF TWINS
I. Diplopagus (both twins are equal and symmetrical) A. Each twin is complete or nearly complete 1. Thoracopagus 2. Xiphipagus 3. Ischiopagus 4. Pyopagus 5. Craniopagus B. Each twin is not nearly complete 1. Duplication originating in the cranial region 1.1 monocephalus 1.2 dicephalus 2. Duplication originating in the caudal region 2.1 monocephalus (tripus dibrachius, tetrapus dibrachius) 2.2 cephalothoracopagus 3. Duplication of both cranial and caudal regions (dicephalus dipygus)
II. Heteropagus (unequal and asymmetrically conjoined twins)

Case report

A full-term 22-year-old primigravida with an unsupervised pregnancy delivered a 3.5 kg male baby at a nursing home. The 10-day-old baby, who was otherwise healthy, was subsequently referred to our centre for definitive treatment. Our institution is a tertiary care and referral centre catering to the needs of the people of Northern India. The baby had a grotesque appearance due to the presence of four limbs, a mass over the sternal region and an omphalocoele of 5 x 4 cm in size. On examination it was found that the mass attached to the baby's sternum was an exoparasitic twin. The exoparasitic twin had fully developed hind limbs, well-developed genitalia, one fully developed upper limb (with radial aplasia) and another underdeveloped upper limb (Figure 1). The parasitic twin was able to move its limbs in response to stimulus and occasionally passed urine.

Figure 1: The exoparasitic twin



At the preoperative assessment, the neonate was awake and alert, with no evidence of cyanosis. The cardiac status of the patient was normal, with no evident murmur. Preoperative investigations revealed haemoglobin to be 14 gm%, blood urea to be 34 mg%, blood sugar 55 mg%, serum sodium 148 mg%, and serum potassium 5.5 mg%. The autosite's transthoracic echocardiography and ultrasonography were found to be normal, whereas the exocytic parasite was found to be acardiac. A few loops of bowel were found inside the body cavity of the parasite, and a tubular urinary bladder-like structure was identified. No definite vascular connection with the autosite was identified. Spiral contrast tomography confirmed the clinical and sonographic findings.

Preoperative preparation

Preoperative preparation included consultation with the senior anaesthetist, the paediatric physician, the surgeon, radiologist, intensive care specialist, the blood bank and the biochemistry laboratory to arrange staff and to provide proper coverage during the perioperative period. Before taking up the patient for surgery, thorough preoperative preparation was done, including the arrangement of difficult airway equipment, including a stylet, different sizes of face mask, different sizes of endotracheal tube, a paediatric Miller and Macintosh laryngosco pe blade, a laryngeal mask airway (LMA), a Proseal laryngeal mask airway (PLMA), a fibre-optic bronchoscope and a tracheostomy set. The arrangement of an adequate amount of blood and blood products, proper nursing care and intensive care facilities, and continuous laboratory support, was done.

Induction of anaesthesia

The neonate was not premedicated. An intravenous line was maintained with 22 gauze cannula. On the patient's arrival in the operating room, all the monitoring devices were placed, including continuous electrocardiography, O_2 saturation of arterial blood, non-invasive blood pressure cuff, and a precordial stethoscope. Baseline arterial pressure, heart rate, respiratory rate and room air O_2 saturation were 90/60 mmHg, 156 beats/min, 48/min and 94%, respectively. After preoxygenation, general anaesthesia was induced with sevoflurane in O_2 by face mask. The limbs of the parasite were lying in front of the neck and interfered the holding the mask in place. One anaesthetist held these limbs apart. We avoided using muscle relaxant for fear that the large mass could hamper ventilation. The neonate was intubated successfully under deep inhalation anaesthesia. The child had a

markedly anterior larynx. The trachea was intubated using 3.5 mm Portex uncuffed tube with stylet. An injection of atracurium 1.5 mg was used to provide muscle relaxation. After induction, an oesophageal temperature probe and end-tidal CO_2 monitors were attached, the radial artery was cannulated and the arterial blood pressure was monitored continuously. Another cannula of 22 gauge was placed on the dorsum of the hand. The child's urinary bladder was also catheterised after the induction of anaesthesia for measuring the urine output during the perioperative period. Anaesthesia was further maintained using 33% O_2 in 66% N_2O (N_2O was used due to the non-availability of compressed air), sevoflurane and incremental dosages of atracurium. A total of fentanyl 7 µg was used to provide intraoperative analgesia. Radiant heat warmer, warm fluid and warm drapes were used to maintain the temperature of the neonate. Intraoperative fluid management consisted of replacing the preoperative fluid deficit (fasting deficit of four hours), intraoperative maintenance (4 ml/kg), and the estimated third space loss (8 ml/kg). The estimated blood loss was around 75 to 100 ml, and this was replaced.

Surgery lasted for one and a half hours. The surgical procedure included an elliptical incision, which was made around the omphalocele and the abdominal cavity of the autosite. During the surgical dissection, a prominent vascular band was found between the autosite and the parasite. About 15 cm of intestine was found entering inside the parasite, as well as in the abdominal cavity of the autosite through the omphalocele. The parasite's intestine was found communicating with the intestine of the host at the Meckel's diverticulum. The parasite's sigmoid colon was communicating with the urinary bladder. Using all possible precautions, the parasite was successfully detached from the autosite without any untoward event. The defect was subsequently covered with a skin flap. At the end of anaesthesia, the child was extubated using neostigmine and atropine, and shifted to intensive care for further monitoring. Analgesia was maintained, with local infiltration of the surgical wound with 6 ml of 0.125% levobupivacaine, and paracetamol suppositories in the postoperative period. The child was stable in the postoperative period and was moved to a ward after being monitored for 24 hours in ICU. His further stay in the hospital (10 days) was uneventful, except for a minor sternal wound infection. The child was discharged home after the stitches had been removed.

Discussion

Conjoined twinning is a rare phenomenon; conjoined twinning with parasite attachment is even more rare.[3] Conjoined twins with shared cardiac structures represent very difficult obstretical, paediatric and surgical problems, which are not often resolved with the survival of the infants.[8] Such types of complex surgery pose major problems for the anaesthestist,[4] requiring a highly experienced team and a centre equipped to deal with such challenging anatomy.[7] We deal with two to three cases of this type of congenital anomaly every year. For the appropriate management of these patients, we consulted paediatric medicine and radiology consultants about the exact planning and the surgical care.

Conjoined twinning of the thoracopagus variety carries an increased risk of experiencing problems. This variety usually shares complex cardiac anatomy with various cardiac anomalies.[7] The liver and the diaphragm are the other organs that may become involved. The most common congenital heart diseases, such as ventricular septal defect (VSD) or tetralogy of Fallot, may be present in 75% of thoracopagus twins, and some degree of pericardial sac in 90% of patients.[9,10] Our patient had no such anomaly. Accurate prenatal diagnosis of cardiac anomalies would help in the management of such cases. Foetal echocardiography may potentially supply the required diagnostic information. Data have revealed that, in twins with shared ventricular myocardium, there is virtually no chance for survival. If this could be determined prenatally, futile treatment may be avoided.[8]

Prenatal ultrasound and magnetic resonance imaging should be employed to provide information about the shared viscera.[7] The diagnosis of conjoined twins at an early stage of pregnancy gives the parents the option of termination.[10] Our patient was born to a 22-year-old primigravida after an unsupervised pregnancy with no prenatal ultrasound records. Besides the problem of shared viscera, twins can develop a reversed-arterial perfusion sequence. This is a condition that results from one or more vascular communications between twin foetuses, such that deoxygenated blood from the pump twin is diverted directly to the perfused twin before gas is exchanged in the placenta. The pump twin usually develops normally, but there is the possibility that it may develop intrauterine congestive heart failure. The perfused twin usually has aplasia or hypoplasia of the heart, head and arms, and therefore dies immediately after birth. Mortality among pump twins ranges from 50% to 75%. In view of this poor prognosis, it is often recommended that the umbilical circulation between the twins be interrupted prenatally,

thus sacrificing the acardiac twin before heart failure develops in the pump twin.[9]

Induction of anaesthesia in such patients requires an experienced anaesthetist. As far as conjoined twins are concerned, the recommended intravenous doses of anaesthetic agents for the combined body weight of the twins are usually halved and then divided into two equal doses, with one being administered to each twin. Reduced incremental doses are titrated against response and help to minimise the dangers of compounding the drug effect in one hour.[6]

An intraoperative concern in such patients is the maintenance of the core temperature (> 34°C), for which it is necessary to use heating blankets, blood warmers and heated humidifiers. A large volume of fluids and massive blood transfusions are often required in this type of surgery. Hypocalcaemia and dilution thrombocytopaenia may complicate a massive transfusion. Serum calcium should be determined at regular intervals during the massive transfusion.[4] In anticipation of this problem, a constant reserve of packed cell, fresh frozen plasma and platelet concentrate must be available throughout the surgery. In our patient, about 75 ml blood was lost, which was adequately replaced by packed cells. Intraoperative haematocrit and serum calcium were not measured in our patient, since blood loss was not massive in our case and the surgery lasted for only one and a half hours.

Arterial Pco_2, Po_2 and pH should also be monitored during surgery. While operating on thoracopagus, there is the possibility of pneumothorax or injury to the diaphragm. In our patient the blood gas analysis remained normal intraoperatively and there was no evidence of any pneumothorax. In relation to operating on xiphipagus-conjoined twins (attachment at the level of the upper abdominal wall), a few authors have recommended the use of corticosteroids at the time of induction and prior to the surgical separation of the xiphipagus-conjoined twins due to possible adrenal insufficiency in one of the twins.[4]

This surgical procedure is commonly undertaken after birth, but is usually delayed for a few weeks or months. Over time, the neonates become larger and anatomical relations between them can be delineated better, other congenital anomalies can be identified and the risks associated with anaesthesia can be minimised, which means that the separation procedure could be planned carefully.[9] In our patient the shared viscera was separated successfully without much difficulty. The separation of the parasite is easy in asymmetric twinning, as the parasite is usually non-viable. The prognosis for the autosite is generally good if no life-threatening anomalies are present.[3] Surgery and anaesthesia in our patient were totally uneventful, with no postoperative complications.

In conclusion, we recommend meticulous preoperative evaluation of patients of this nature prior to surgery. Proper intraoperative planning, team work and an integrated, multidisciplinary approach is necessary for a positive outcome to manage such complex cases. However, the prognosis for parasitic twinning is good as far as the autosite is concerned, unless life-threatening anomalies are present.

References

1. Watanatittan S, Suwatanaviroj A, Niramis R, Havanonda S. Conjoined twins: surgical separation in 11 cases. J Med Assoc Thai 2003;86(suppl 3):S633–43.
2. Rivera JRC, Leon JA, Gomez EV, et al. Unusual presentation of heteropagus attached to thorax. J Pediatr Surg 1997;32(10):1492–4.
3. Rattan SK, Rattan KN, Magu S, Gupta S, Narang R, Arora B. Thoracopagus parasites in two sets of twins: evidence for the 'fusion theory'. Pediatr Surg Int 2008;24:1255–9.
4. James PD, McLeod ME, Relton JES, Creighton RE. Anaesthetic consideration for separation of omphalo-ischiopagus tripus twins. Can Anaesth Soc J 1985;32: 402–11.
5. 5. Biswas SK, Gangopadhyay AN, Bhatia BD. An unusual case of heteropagus twinning. J Pediatr Surg 1992;27(1):96–7.
6. Szmukl P, Rabbl MF, Curry B, Smith KJ. Anaesthetic management of thoracopagus twins with cyanotic heart disease for cardiac assessment: special consideration related to ventilation and cross-circulation. British Journal of Anaesthesia 2006;96(3):341–5.
7. Shank E, Manohar N, Schmidt U. Anaesthetic management of thoracopagus twins with complex cyanotic heart disease in MRI suite. Anesth Analg 2005;100:361–4.
8. Danford DA, McManus BM, Nielsen SA, Levine MG, Needelman HW. Definition of inseparably fused ventricuar myocardium in thoracopagus: fetal echocardiographic utility and pathologic refinement. Pediatr Cardiol 1993;14:242–6.
9. Norwitz ER, Hoyte LPJ, Jenkins KJ, et al. Separation of conjoined twins with reversed-arteria-perfusion sequence after prenatal planning with 3-dimensional modeling. N Engl J Med. 2000 Aug 10;343(6):399–402.
10. Tansel T, Yazicioglu F. Cardiac and other malformation in parapagus twins. Arch Gynecol Obstet 2004;269:211–3