Hunterian Lecture

Surgery for conjoined twins

Lewis Spitz

Department of Paediatric Surgery, Institute of Child Health, and Great Ormond Street Children's Hospital, London, UK

Conjoined twins have fascinated both the medical profession and the general public since classical times. Interest has ranged from fear of their birth being a sign of impending disaster through exhibitionism and curiosity to intense media attention. The incidence of conjoined twins is 1:50,000–100,000 pregnancies but as 60% are still-born or die shortly after birth, the true incidence is 1:200,000 live births. Females predominate in the ratio of 3:1.

There are two theories regarding the pathogenesis of conjoined twins. The most widely accepted theory is failure of complete separation of the embryonic disc at the 15–17 day stage of a uni-ovular gestation (fission theory). The alternative theory is that of fusion of adjacent parts (fusion theory) of two originally separate monovular embryonic discs.¹

Conjoined twins are classified according to the most prominent site of union together with the suffix 'pagus' (Greek term for fixed). Thoracopagus accounts for 40% of cases, followed by omphalopagus (33%), pygopagus (19%), ischiopagus (6%) and craniopagus (2%). Recently, the term 'parapagus' has been introduced to indicate extensive side-to-side fusion.

Historical highlights

The earliest example of conjoined twins is a 17.2 cm marble statuette portraying parapagus twins, called the 'double goddess' dating from the sixth millennium BC. The statue of the sisters of Catathoyuk is housed in the Anatolian Civilisation Museum in Ankara, Turkey.² Another early example is a stone carving of pygopagus twins dated to 80 BC found in Fiesole and housed in the Museo San Marco, Florence (Fig. 1).³

Figure 1 Stone carving of pygopagus twins dated to ~80 BC, Fiesole, Museo San Marco, Florence, Italy.

The first reasonably well-documented case of conjoined twins is of male ischiopagus twins born in around 940 AD in Kappadokia, Armenia, who lived together for 30 years. When one twin died aged 30 years, surgeons in Constantinople tried to save the surviving twin by separating him from his dead brother, but he too died 3 days after the operation. This represents the first recorded attempt at surgical separation of conjoined twins.²

In 1100, Mary and Eliza Chulkhurst, joined at the 'hips and shoulders', were born in Biddenden, Kent. They lived together for 34 years. When Mary fell ill and died, Eliza was advised to be separated but absolutely refused saying 'as we came together, we will also go together' and she died 6 h later. They bequeathed to the parish 20 acres

Correspondence to: Prof. Lewis Spitz, Nuffield Professor of Paediatric Surgery, Department of Paediatric Surgery, Institute of Child Health (University College London), and Great Ormond Street Children's Hospital, 30 Guilford Street, London WC1N 1EH, UK
Tel: +44 20 7829 8691; E-mail: l.spitz@ich.ucl.ac.uk

of land with the instructions that the proceeds were to be spent on the distribution of cakes, bearing an impression of their images, to be given away on each Easter Sunday to all strangers in Biddenden – a practice which endures to the present day.⁴

The Scottish brothers born in 1490 were two complete individuals above the waist but had only one set of lower extremities. They were brought before the Court of King James IV of Scotland who ordered that they be carefully brought up and educated. They were talented musicians and proficient linguists and, by the age of 20 years, could speak eight languages. They died in 1518, aged 28 years.⁵

In 1495, Geoffroy-St Hilaire reported on craniopagus females. They were joined at the forehead, causing them to

stand face-to-face; their noses almost touched. When one twin died, aged 10 years, separation was attempted but failed.⁶

William Durston documented the first example of oesophageal atresia in one of female thoracopagus twins in Plymouth in 1670.⁷

The first successful separation of conjoined twins was carried out by Fatio in 1689. Elizabeth and Catherine were xypho-omphalopagus twins. In the presence of the lord mayors of Basel and a galaxy of medical men, he separated the twins by 'tracing the umbilical vessels to the navel where he tied them separately. He then transfixied and tied the bridge between the two infants with a silken cord and cut the isthmus'. The ligature fell off on the ninth postoperative day and both children survived. Koenig, an observer at the operation, published the case as his own and to him is attributed the first success.⁸

John Hunter was also interested in abnormal births and there are six specimens of conjoined twins (Fig. 2) in the Hunterian Museum at The Royal College of Surgeons of England.

The most celebrated pair of conjoined twins was Chang and Eng born on a river boat on the Mekong river in Siam in 1811. They were joined at the xiphisternum by a short band (xiphopagus) which, over time, stretched so that they were

Figure 2 Specimen of thoracopagus conjoined twins in the Hunterian Museum, The Royal College of Surgeons of England.

Figure 3 Photograph of Chang and Eng, xiphopagus twins. From an oil painting by Irvine in The Royal College of Surgeons of England.

Table 1 Clinical details of conjoined twins where no separation was attempted

Case	Year	Type	Sex	Shared organs	Associated anomalies	Outcome
3	1986	Thoracopagus	F	Complex cardiac fusion, pericardium, liver, common bile duct, small intestine	Pulmonary atresia, oesophageal atresia and tracheo-oesophageal fistula	Both died – 3 days
5	1989	Thoracopagus	F	Complex cardiac fusion, small intestine, liver	Exomphalos, VSD, coarctation	Both died – 4 weeks
9	1994	Thoracopagus	F	Complex cardiac fusion	No necropsy	Both died – 3 days
12	1996	Parapagus	F	Thoraco-abdomino-ischio-pagus	(Normal triplet). No necropsy	Both died – 5 days
17	1999	Thoracopagus	F	Complex cardiac fusion	Hydrops. No necropsy	Both died – 1 day
20	2002	Thoracopagus	F	Complex cardiac fusion, duodenum	Truncus arteriosus, exomphalos	Both died – 19 days

VSD, ventricular septal defect.

able to stand side-by-side (Fig. 3). They were 'discovered' by a travelling Scottish merchant, Robert Hunter, and taken to the US where they were displayed by the famous showman, Phineas Barnum. In 1829, they were examined by the 'most eminent professors of surgery and medicine' in the Egyptian Hall in London, amongst whom was Leigh Thomas, then

Figure 4 Thoracopagus twins with complex cardiac fusion.

President of The Royal College of Surgeons of England. With reference to the practicability of separation, all concurred that it would involve 'an operation of such risk, as not to be warranted unless in the event of the death of one of them'. They married sisters, lived in separate houses in North Carolina and had 22 children between them. During the last year of their life, Chang, who three years previously had suffered a stroke, expressed concern that they could not survive together much longer. Their surgeon agreed that, in the event of one of them dying, he would operate to separate them immediately. Chang died during the night of the 17 January 1874. Due to the icy conditions, the 3-mile journey to the twins' house took the surgeon an hour, during which Eng died. They had lived together for 63 years.^{9,10}

The diagnosis of conjoined twins can be made by the 12th week of gestation on prenatal ultrasonography. By 20 weeks of gestation, precise anatomical detail of shared organs can be determined on ultrasound scan. It is important to carry out chromosomal analysis and echocardiography in order to counsel the parents. Prenatally, there is the opportunity for recommending elective termination particularly in the presence of a chromosomal abnormality, or cardiac fusion in thoracopagus twins or cerebral fusion in craniopagus twins. The parents may elect for termination if the predicted extent of deformity would be so extensive as to be unacceptable to them. If pregnancy is allowed to proceed, elective Caesarean section should be planned at 38 weeks' gestation. The birth should take place at, or close to, the surgical centre where further management will be carried out.

Clinical experience

Over the past 34 years (1968–2002), I have personally been involved in the management of 20 sets of symmetrical conjoined twins. Their management can be divided into three categories.^{11,12}

Non-operative management

A total of 6 cases summarised in Table 1. No attempt at separation was made in the presence of complex cardiac fusion (5 cases; Fig. 4) or where the extent of deformity was unacceptable to the family (1 case). All these infants were given custodial care and all died within a few days of birth.

Emergency separation

A total of 7 cases summarised in Table 2. Separation was performed as an emergency where one twin was dead (2 cases) or dying (3 cases) and threatening the survival of the other twin, where there was a correctable congenital anomaly present which if unoperated would lead to the death of both twins (1 case), or where there was serious damage to the connecting bridge (1 case). Only 4 of the potential 14 infants survived emergency separation (28.6%), but if two infants already dead and three with lethal abnormalities are excluded the survival rate increases to 44%.

Planned separation

A total of 7 cases summarised in Table 3. Where the infants are stable, it is prudent to allow them to thrive which permits time for detailed investigations to be carried out, such as CT, MRI, ECHO cardiography,

angiography, urography, to plan the operative procedure. It also allows time to apply methods, such as tissue expansion, keratocyte culture, to achieve primary wound closure following separation. In our experience with 7 sets undergoing planned separation, 11 of a possible 14 infants survived (79%) and two of the three deaths occurred 6 months after the separation from ‘cot death’.

Illustrative cases

Two illustrative cases that have not been previously published are described below.

Case 15

Xipho-omphalopagus twins were diagnosed prenatally at 17 weeks’ gestation and were delivered by elective Caesarean section for early onset of labour. Their condition at birth was satisfactory with a combined birth weight of 5500 g. Arrangements were made to transfer the infants the following day but, due to the deterioration of one twin, they were retrieved as an emergency by air. On admission, the exomphalos between the two infants was ‘black’ indicating an intra-abdominal catastrophe (Fig. 5). An echocardiogram was normal in both infants. They were resuscitated and taken immediately to theatre where a necrotic midgut volvulus was found. All necrotic intestine was resected. A duodenal atresia in one twin was repaired and end-to-end small intestinal continuity restored. Both

Table 2 Clinical details of conjoined twins undergoing emergency separation

Case	Year	Type	Sex	Age at operation	Shared organs	Associated anomalies	Outcome
2	1985	Thoracopagus	F	3 days	Pericardium, diaphragm, liver, common bile duct, proximal small intestine	Pulmonary atresia	1 A&W, 1 died at 6 weeks
7	1993	Omphalopagus	F	1 day	Liver, distal small intestine, anorectal anomaly (cloaca)	Exomphalos ruptured at birth and liver split. One dead	Both died – 1 day
11	1995	Thoracopagus	M	8 days	Cardiac (atrial communication), pericardium, liver	1 unilocular heart, 1 absent hepatic vein	1 immediate death and 1 died at 6 weeks
14	1997	Thoracopagus	F	1 day	Cardiac fusion, pericardium, liver	Diaphragmatic hernia, unilocular heart	1 immediate death and 1 died at 1 day
15	1998	Omphalopagus	F	2 days	Liver, upper small intestine	Exomphalos, volvulus of small intestine, proximal and distal intestinal atresia	Both A&W
16	1999	Omphalopagus	M	3 h	Small intestine, bladder	Hypoplastic lungs and horseshoe kidney	1 died pre-operatively, 1 A&W
18	2000	Parapagus	F	1 day	Lung, diaphragm, aorta, liver, intestine	AV canal. Persistent pulmonary hypertension	1 died intra-operatively, 1 died post-operatively

A&W, alive and well.

Table 3 Clinical details of planned separation

Case	Year	Type	Sex	Age at operation	Shared organs	Associated anomalies	Outcome
1	1968	Craniopagus ¹³	F	2 months	Sagittal sinus	–	1 A&W, 1 died at 6 months
4	1987	Ischiopagus ¹⁴	M	8 months	Liver, distal ileum and colon, crossed ureters, genitalia	Imperforate anus	Both A&W
6	1992	Parapagus ¹⁵	F	3 years	Pericardium, diaphragm, liver, distal ileum and colon, bladder and vagina	Single kidneys	1 A&W, 1 died at 3 days
8	1993	Parapagus ¹⁶	M	10 months	Pericardium, liver, distal small intestine and colon, crossed ureters, genitalia	Anorectal anomaly	1 A&W, 1 died at 6 months
10	1994	Thoracopagus ¹⁷	F	3 months	Pericardium, diaphragm, liver, bile ducts, small intestine	Exomphalos, extrahepatic biliary atresia	Both A&W
13	1997	Omphalopagus	F	3 months	Liver	–	Both A&W
19	2001	Pygopagus	F	2 months	Spinal cord, vertebral column	Anorectal anomaly	Both A&W

A&W, alive and well.

had severely shortened small bowel (28 cm, 30 cm). The liver was divided and the abdomens closed with prolene mesh due to the massive defect. The meshes were removed and the abdomens repaired 1 week later. The postoperative courses were complicated by an intestinal perforation in one twin, but intestinal adaptation occurred rapidly and the infants were on full enteral nutrition at the time of discharge at the age of 6 months.

Figure 5 Xipho-omphalopagus twins (case 15) showing dark colour of exomphalos between the twins.

Case 16

Omphalopagus conjoined twins were referred at 17 weeks' gestation for prenatal evaluation. Ultrasound scan showed bladder and intestine within the exomphalos between the twins (Fig. 6). There appeared to be two amniotic sacs with a 'membrane' covering the face of one twin. Echocardiography was normal. The twins were delivered by elective Caesarean section at 37 weeks' gestation and, immediately after delivery, one twin required mechanical ventilation for respiratory distress. Immediate transfer was effected, but the twin experiencing respiratory distress developed bilateral pneumothoraces, which required tube thoracostomies, and he died *en route* to our hospital. The bridge of tissue connecting the twins was digitally compressed to prevent the live twin exsanguinating into his dead brother. On arrival, they were taken directly to the operating theatre where the dead twin was separated. The conjoined bladder was repaired and the junction between the infants' intestine was divided in the distal ileum and a colostomy performed for an anorectal agenesis. The infant made an uneventful recovery and subsequently underwent a posterior sagittal anorectoplasty. Necropsy on the deceased twin revealed severe pulmonary hypoplasia.

Conclusions

Success in the management of conjoined twins is dependent on an experienced team and the resources available only in a tertiary referral centre where the full range of medical, nursing and surgical specialties are present. Advances in diagnostic techniques – particularly imaging modalities including prenatal ultrasonography and ultrafast magnetic resonance imaging (MRI)^{18,19} and post-natal three-dimensional

Figure 6 Antenatal ultrasound scan of omphalopagus twins showing the bladder and intestine within the exomphalos between the twins.

computerised tomography and MRI – are of value in planning the separation procedure.²⁰ The operation requires two sets of experienced anaesthetists with individual monitoring of each twin separately as they may require different volume replacement and drug dosages.

Prior surgical experience is invaluable as the operative approach is from an unusual angle, the anatomical configuration is usually complex and variants in the anatomy may dramatically alter the operative plan. The separation may involve a variety of specialists, such as cardiac surgeons, urologists, orthopaedic surgeons, neurosurgeons and plastic surgeons, depending on the area of union.

Postoperatively, the infants require meticulous monitoring in the intensive care unit with careful regulation of their fluid and electrolyte requirements, mechanical ventilation and prevention of infection to large areas of open wounds covered only by mesh in some instances.

Separation is recommended in all cases where this is feasible with the anticipated survival of both infants. Where there is complex cardiac or cerebral fusion, survival is unlikely and custodial care only is offered. Where one twin is dead or has a lethal abnormality and cannot survive independently from its normal twin and if unoperated both twins would die, separation to save the healthy twin should be attempted.²¹

Acknowledgements

The surgery of conjoined twins is a supreme example of team work. I wish to express my sincere gratitude to my

colleague, Mr E Kiely, with whom I have been involved in 18 of the 20 cases. Thanks also to all other surgical specialists, anaesthetists, intensivists, and nursing staff as well as radiologists for their invaluable contribution in the management of these complex cases.

This Hunterian Lecture of The Royal College of Surgeons of England was delivered at the British Association of Paediatric Surgeons' Annual Conference in Cambridge, July 2002.

References

1. Spencer R. Theoretical and analytical embryology of conjoined twins. Part I: embryogenesis. *Clin Anat* 2000; **13**: 36–53.
2. Geroulanos S, Jaggi F, Wydler J, Lachat M, Cakmakci M. Thoracopagus symmetricus. *Gesnerus* 1993; **50**: 179–200.
3. Courtesy of The Museum of San Marco, Florence, Italy.
4. Bondeson J. The Biddenden maids: a curious chapter in the history of conjoined twins. *J R Soc Med* 1992; **85**: 217–21.
5. Bondeson J. *The Two-headed Boy and Other Medical Marvels*. London: Cornell University Press, 2000; 164.
6. Geoffroy St Hilaire I. *Histoire général et particulière des anomalies de l'organisation chez l'homme et chez les animaux*. Paris: Baillière, 1832–1836.
7. Durston W. A narrative of a monstrous birth in Plymouth October 22, 1670; together with the anatomic observations taken thereupon by William Durston, Doctor of Physick, and communication to Dr. Tim Clerk. *Philos Trans R Soc* 1670; **V**: 2096.
8. Rickham PP. The dawn of paediatric surgery: Johannes Fatio (1649–1691) – his life, his work and his horrible end. *Prog Pediatr Surg* 1986; **20**: 94–105.
9. Hunter K. *Duet for a Lifetime*. London: Michael Joseph, 1964.
10. *An Historical and Descriptive Account of the Siamese Twin Brothers from Actual Observations* (Entered at Stationers Hall). London: W. Turner, 1829.
11. Spitz L, Kiely EM. The success rate for surgery of conjoined twins [letter]. *Lancet* 2000; **356**: 1765.
12. Spitz L, Kiely EM. Great Ormond Street experience with conjoined twins. *Br J Surg* 2002; **89**: 1188–92.
13. Wolfowitz J, Kerr EM, Levin SE, Walker DH, Vetten KB. Separation of craniopagus twins. *S Afr Med J* 1968; **42**: 412–24.
14. Spitz L, Capps SNJ, Kiely EM. Xipho-omphalo-ischiopagus tripus conjoined twins: successful separation following abdominal wall expansion. *J Pediatr Surg* 1991; **26**: 26–9.
15. Spitz L, Stringer MD, Kiley EM, Ransley PG, Smith P. Separation of branchio-thoraco-omphalo-ischiopagus bipus conjoined twins. *J Pediatr Surg* 1994; **29**: 477–81.
16. Wilcox DT, Quinn SM, Spitz L, Kiely EM, Ransley PG. Urological problems in conjoined twins. *Br J Urol* 1998; **81**: 905–10.
17. Spitz L, Crabbe DCG, Kiely EM. Separation of thoraco-omphalopagus conjoined twins with complex hepato-biliary anatomy. *J Pediatr Surg* 1997; **32**: 787–9.
18. Kingston C, McHugh K, Kumar Devan J, Spitz L, Kiely EM. The role of radiology in the assessment of conjoined twins. Paper presented at *The Radiological Society of North America*, Chicago, 1999.
19. Casele HL, Meyer JR. Ultrafast magnetic resonance imaging of cephalopagus conjoined twins. *Obstet Gynecol* 2000; **95**: 1015–7.
20. MacKenzie TC, Crombleholme TM, Johnson MP, Schnauer L, Flake AW, Henrick HL *et al*. The natural history of prenatally diagnosed conjoined twins. *J Paed Surg* 2002; **37**: 303–9.
21. Ward Lord Justice, In the Supreme Court of Judicature, Court of Appeal (Civil Division), On Appeal from the Family Division. Case No: B1/2000/2969. Royal Courts of Justice, Strand, London, 22nd September 2000.