CHAPTER 124
CONJOINED AND PARASITIC TWINS
Alastair J.W. Millar

Introduction
The birth of conjoined twins has always fascinated mankind, with the public’s view of malformed children greatly influenced by the prevailing culture and religious beliefs. In prehistoric times, conjoined twins were depicted in cave drawings, on pottery, or as figurines. In folklore, they were often regarded as an omen of impending disaster, eliciting strong emotions ranging from wonder and admiration to rejection and hostility. Although malformed children were treated compassionately at times, historical records show that infanticide was frequently practiced and the mother was often held responsible for causing the malformation.

Demographics
Although the worldwide incidence of monozygotic twinning is the same in all ethnic groups, the incidence of conjoined twins appears to be higher in sub-Saharan Africa, ranging from 1 in 50,000 to 1 in 100,000 live births, or 1 in 400 monozygotic twin births. The natural history that follows a prenatal diagnosis of conjoined twins confirms that a large number of infants die either in utero (28%) or immediately after birth (54%); in fact, only around 20% survive.

Aetiology and Pathophysiology
Conjoined twins are monozygotic, monoamniotic, and monochorionic. They are always of the same gender, with a 3:1 female preponderance. Their formation results either from failure of separation of the embryonic plate between 15 and 17 days gestation, or from secondary union of two separate embryonic discs at the dorsal neural tube or ventral yolk sac areas at 3 to 4 weeks gestation.

Spencer’s extensive embryological studies appear to favour the latter theory, but this remains controversial. Although genetically identical, one of the conjoined twins is almost always weaker or smaller than the other and may have additional congenital defects. These twins also develop dissimilar personalities from an early age. Conjoined twins are individual and deformed but symmetrical and proportional.

Classification
Conjoined twins are always joined at homologous sites, and the clinical classification is based on the most prominent site of union, combined with the suffix “pagus” meaning “that which is fixed”. There are eight recognized configurations, as shown in Figure 124.1: thoracopagus (chest), omphalopagus (umbilicus), pygopagus (rump), ischiopagus (hip), craniopagus (cranium), paragagus (side), cephalopagus (head), and rachipagus (spine).

Conjoined twins can be further described as symmetrical or asymmetrical. Asymmetrical, or incomplete, conjoined twins result from the demise of one twin with remnant structures attached to the complete twin, with the junction remaining at or near one of the common sites of union (Figure 124.2). Fetus-in-feto refers to asymmetrical monozygotic diamniotic intraparastic twins. Conjoined triplets are exceptionally rare, and their pathogenesis remains even more obscure.

Management
This section includes descriptions of the lessons learned at one hospital over a period of 42 years, supplemented by a literature review. These descriptions encompass prenatal diagnosis, obstetric intervention, special investigations, postnatal management, anaesthetic considerations, and surgical strategies, with the major focus on thoracopagus conjunction.

Prenatal Diagnosis
In developed countries, antenatal diagnosis is usually made by ultrasound (US) scan. Once the diagnosis is suspected, echocardiography and ultrafast foetal magnetic resonance imaging (MRI) may be used to confirm the diagnosis. The mother is referred for advice either to plan the mode of delivery because of obstetric implications or for consideration of termination of the pregnancy with the attendant ethical and moral considerations.
Despite all the extensive preoperative investigations (Figure 124.3). Unexpected anatomic variations frequently identified during surgery. The anatomical configurations encountered are often complex, with management carried out by an experienced multidisciplinary team. Of the special investigations, and correct anaesthetic and surgical preparation. Improved survival rates for conjoined twins are due to advances in comprehensive and functional description of normal and fused anatomy. Are thriving and all investigations have been completed, providing a valuable assessment of the twins. Elective surgery is best scheduled for when the infants are 6 months old and there is the possibility of saving at least one twin. Most children born normally do not sustain any damage to the connecting sites (bridges), except where there is an omphalocoele associated with thoraco-omphalopagus conjunction. Rupture of the exomphalos and evisceration of liver and bowel may occur. Maternal mortality during labour has also been reported.

Ideally, the immediate perinatal management of the babies is also planned, and in one case in which a twin with a normal heart perfused the co-twin with a rudimentary heart, the ex utero intrapartum treatment (EXIT) procedure was utilised due to concern that the normal twin would suffer immediate cardiac decompensation at birth. This EXIT to separation strategy allowed prompt control of the airway and circulation before clamping the umbilical cord and optimised management of a potentially lethal situation with survival of the normal twin.

Once born, the twins should be referred for appropriate investigation and surgical management. The therapeutic options to be considered range from conservative, nonsurgical management to emergency or planned surgery (see next subsection).

**Special Investigations**

Investigations should be directed towards identifying the anatomy of conjunction and consequently the viability of separation. The areas of fusion largely determine the imaging modalities chosen. Skeletal surveys, echocardiography, US, computed tomography (CT), MRI, and angiography provide excellent anatomical detail, demonstrating organ position, shared viscera, and vascular anatomy. Contrast imaging evaluates the gastrointestinal and urinary systems, and endoscopy is of further help in the urogenital assessment. Radioisotope scanning can assess regional perfusion fields.

Twins with no reasonable chance of survival, largely due to cardiac anomalies incompatible with life, and those with irreversible postnatal diseases such as necrotising enterocolitis totalis, should receive only palliative treatment and are not considered for separation. Where separation is clearly not possible without the inevitable death or unacceptable mutilation of both twins, it is perfectly reasonable to accept this situation and to provide counselling and all possible support for their future growth and development.

Emergency surgery is indicated when there is damage to the connecting bridge or when correctable anomalies threaten the survival of one or both twins and there is the possibility of saving at least one of the twins. Elective surgery is best scheduled for when the infants are thriving and all investigations have been completed, providing a comprehensive and functional description of normal and fused anatomy. Improved survival rates for conjoined twins are due to advances in perinatal and postnatal diagnostic techniques, meticulous interpretation of the special investigations, and correct anaesthetic and surgical management carried out by an experienced multidisciplinary team. The anatomical configurations encountered are often complex, with unexpected anatomic variations frequently identified during surgery despite all the extensive preoperative investigations (Figure 124.3).

Many examples exist of conjoined twins living full and productive lives without separation. The case for this acceptance is most eloquently made in the book *One of Us* by Alice Dreger.

The investigations used to fully evaluate conjoined twins are listed in Table 124.1. Prior meticulous clinical examination is conducted with necessary examination under anaesthetic and placement of catheters for later contrast radiological studies.

**Postnatal Management**

Immediate postnatal management consists of resuscitation and stabilisation of the twins. This is followed by a thorough physical examination with special investigations to define the relevant anatomy. If emergency surgery is anticipated, all twins should undergo echocardiogra-
phy and plain roentgenography, which provide limited but essential information. The site of conjunction will determine the type and order of special investigations. The information obtained will determine the surgical approach, the timing of separation, the allocation of organs and structures, and the eventual prognosis regarding survival and functional outcome. Important structures to evaluate are the cardiac, hepatobiliary, intestinal, urogenital, and spinal systems. The use of diagrams, three-dimensional (3D) organ models, and surgical rehearsal of the procedure will ensure the best possible outcome. Despite all these investigations and careful analysis of findings, however, preoperative interpretation may still be difficult, with incorrect conclusions drawn.

Emergency separation has resulted in up to a 70% mortality rate compared to 20% for elective procedures, emphasizing the need to stabilise the infants initially and to postpone surgery until the basic investigations have been completed. In the author’s experience, emergency surgery was necessary to alleviate intestinal obstruction, to treat necrotising enterocolitis, to manage a ruptured exomphalos, and, for deteriorating cardiac-respiratory status threatening survival of one or both twins. Delaying separation into early childhood may result in increased postnatal deformities and psychological problems. If separation is possible and desirable, surgery should be performed within the first 6 to 9 months, before the twins develop an awareness of their condition. Motor skills, sensory integration, and personality need to develop in separated twins.

**Anaesthetic Considerations**

Anaesthesia for separation of conjoined twins is a complex, demanding procedure that is facilitated by having two colour-coded anaesthetic teams representing each child. The author’s experience has highlighted the following anaesthetic considerations. The infants are often premature with pre-existing cardiac and pulmonary dysfunction, and the induction of anaesthesia is often compromised by the abnormal positions and proximity of the twins. During surgery, difficulties with vascular access, haemodynamic stability, and temperature control can be considerable.

To maintain haemodynamic stability, blood volumes transfused may range from 10% to 450% of the estimated blood volume. Blood loss can be especially extensive in thoracopagus and ischiopagus separations, and relative changes in position of the two infants during surgery lead to significant shifts in blood volumes. Due to cross-circulation, pharmacokinetics and pharmacodynamics are inconsistent, especially in thoracopagus twins, and altered drug responses must be expected. Anticipated postseparation problems include respiratory insufficiency, haemodynamic instability, fluid balance, temperature control, sepsis, wound closure difficulties with staged closure if necessary, and residual organ dysfunction.

**Surgical Strategies**

The first successful surgical separation took place in 1689 and more than 1,200 cases had been reported in the literature by 2000. The surgical separation of conjoined twins presents a great challenge and undoubtfully requires a multidisciplinary team. An unequal external union, variations in internal anatomy, and discordant anomalies mandate thorough elucidation of the anatomy of conjunction before planning the surgical procedure required to separate and individualise the twins.

Many descriptions of surgical procedures to separate the various types of conjoined twins have been published. Technical details are determined by the anatomy of conjunction, the allocation of sharing of organs and structures, and the planned reconstruction. Standard approaches are normally utilized, but variations may demand a novel surgical approach or alternative techniques.

Major factors that will govern successful separation include the order of separation, the distribution of organs between the twins, meticulous aseptic surgical techniques, the reconstruction of divided organs and structures, and wound closure. It is also necessary to distinguish between structures that are shared by both twins and those belonging only to one individual. Allocation of shared organs usually involves the anus, rectum, genitourinary tract, lower spine, and spinal cord. Unexpected anatomical variations are often encountered, including previously unrecognised cardiac, gastrointestinal, hepatobiliary, spinal, and genitourinary anomalies. Operation time is prolonged with the separation of the more complex thoracopagus and ischiopagus twins, and is in the order of 7–13 hours and 13–19 hours, respectively.

**Skin closure**

Whenever there is extensive sharing of body surface areas (e.g., thoracopagus and ischiopagus), closure of the disconnected surfaces may pose major problems, especially when separation is undertaken as an emergency. Subcutaneous tissue expansion is used to provide tissue for reconstruction or closure where insufficient natural tissue exists (Figure 124.4). This allows for primary tension-free closure, thereby minimising respiratory and wound complications. Closure under tension is poorly tolerated, and it is preferred that the body cavities (both chest and abdomen, if necessary) are left open for later staged closure with plastic reconstruction using skin and muscle flaps or split skin grafting onto granulation tissue. Vacuum dressings may assist a more rapid healing, earlier grafting, and wound closure. Unfortunately, tissue expansion is not always possible and has a nearly 60% incidence of complications due to factors such as placement over bony areas with little subcutaneous tissue, wound sepsis, and skin necrosis. Skin expanders must be correctly sited; placements are best tolerated in older infants. It takes 6 to 8 weeks to gain maximum advantage.

**Cardiovascular system**

Experience with 22 thoracopagus sets of twins has shown that evaluation requires the use of every tool available—from clinical evaluation to angiography. The ECG is generally unreliable because two separate ECGs do not rule out significant sharing of cardiac structures. The mainstay of the evaluation is echocardiography, generally best accomplished by a pair of investigators who meticulously double-check each other’s findings. The investigators may be left with apical and suprasternal
views only. Tracheo-oesophageal echo is frequently not possible, given
the size of the infants. The newer multislice CT scanning and modern
MRI machines will clearly have roles to play in future evaluations,
despite the radiation exposure and the need for a general anaesthetic.

In the author’s experience, an MRI investigation resulted in
“overcalling” ventricular sharing. Angiography under general
anaesthetic may still be needed, but it remains a high-risk procedure
with an unpredictable response to anaesthesia. In a set of twins with
venous-pole sharing, induction of anaesthesia resulted in asystole
in the twin with a myopathic ventricle, resulting in brain death and
an emergency separation. Prior to separation, the surgeon may still
not know the exact nature of venous connections, coronary arterial
anatomy, the branching anatomy of the head and neck vessels, and the
ture size of the right ventricle.

No twins with ventricular conjunction have ever been successfully
separated with both twins surviving; however, there is a report of
successful separation of thoracopagus twins with two normal hearts
joined by a myocardial bridge. In a situation where one twin is acardiac
or where the twins share ventricles, successful separation is impossible
without sacrificing one infant. The chest wall and skin of the sacrificed
twin are used to obtain skin cover and to create a firm structure to
protect the protuberant fused single heart, as any dislocation of the
heart from its natural position is likely to cause disturbance of function.
All the main inflow and outflow vessels from the sacrificed twin have
to be disconnected from the heart, and the whole cardiac complex is
then assigned to the infant selected to survive. In two cases of infants
surviving a sacrifice procedure, one survived 30 days and died from
aspiration and the other is a long-term survivor of nearly 15 years.
Subsequent reconstruction of the deformed chest is possible at a later
stage. Thoracopagus conjoined twins may be classified according to the
degree of cardiac conjunction.

Hepatobiliary system
The liver is shared in almost all ventral forms of conjoined twins.
Ultrasound, CT scanning, and radio nucleotide scanning provide the
best overall picture of hepatic conjunction, the biliary drainage system
including the gallbladder, and configuration of the pancreas. For suc-
cessful hepatic division, each liver has to have an inferior vena cava
to its own heart. Hepatic conjunction is along an oblique plane, and
venous connections may consist of a labyrinth of small venous channels
that may bleed excessively during surgery. In the author’s experience,
hepatic division has always been possible. Cardiac disconnection must
be accomplished before hepatic division because a large volume of
blood can circulate through the liver, creating a false impression that
both hearts are able to sustain independent life. The anatomy of the
extra hepatic biliary system (EHBS) needs to be confirmed, which may
require intraoperative cholangiography. Two gallbladders do not always
equate with two EHBSs, especially if there is fusion of the proximal
duodenum, which may be demonstrated by upper contrast radiography.
However, two gallbladders and two duodenums usually indicate two
separate extrahepatic bile ducts. Bile drainage is imperative, and in
the presence of a single EHBS, one twin should be allocated the EHBS,
while every attempt should be made to establish bile drainage through
a Roux-en-Y hepaticojejunostomy in the other twin. Anatomically,
the pancreas belongs to the duodenum, and is best left with the EHBS.

Gastrointestinal system
The intraabdominal gastrointestinal tract is frequently shared in ventral
and caudal types of junction and usually follows one of two patterns. In
the first, duodenal junction is often encountered in thoracopagus twins.
The junction can extend distal to the duodenum and involve the upper
small bowel up to the level of Meckel’s point, where it divides again
into two separate distal ilea. The second type of GIT sharing (ileocolic
or, rarely, only colonic) is commonly encountered in ischiopagus twins.
The single ileocolon, resembling a conjoined organ, opens into a single
anus. A double blood supply may facilitate longitudinal division of the
colon, thus preserving an anatomically normal or foreshortened colon
for each child. Alternatively, one child can be allocated the ileocecal
valve and the other the anus, with both sharing the divided colon.
Pygopagus twins always have a common anal canal. It is the author’s
practice to reconstruct the anorectal region at the time of primary divi-
sion. A previously placed colostomy, however, demands a different type
of allocation and reconstruction.

Urogenital tract
Complex and variable urogenital abnormalities accompany pelvic
fusion and are restricted to symmetrical and asymmetrical ischiopagus
and pygopagus twins. These abnormalities are less commonly seen in
thoracopagus conjunction. The incidence of shared pelvic organs is on
the order of 15% for pygopagus to 50% or more for ischiopagus twins.
An unobstructed continent urinary system with a physically acceptable
and functional genital system is the primary goal. Essential in the work-
up of urogenital abnormalities are genitourinary US, isotope renogra-
phy, micturating cystourethrography, and endoscopy.

The kidneys may vary in number, size, ectopia, degree of fusion,
and the course of the ureters. Most ischiopagus twins have four kidneys
and two bladders, with one ureter crossing to the ipsilateral and one to
the contralateral bladder. One or two bladders may be present,
lying side by side or fused in the midline with one draining into the
other. In most cases, despite these variations, a functional bladder can
be reconstructed. The presence of spinal fusion in its various forms
complicates the situation by introducing a neuropathic element into the
behaviour of the bladder, which has a significant influence on future
management. Crucial decisions regarding assignment are therefore
required when shared organs that cannot be divided are present.

The genital pattern varies widely, and every effort should be made
to achieve functional reconstruction, which may require an individual
approach. In females, urogenital sinuses or even cloacal abnormalities
are often present, requiring careful consideration during division,
allocation of organs, and reconstruction. In males, the status of the
external genitalia, urethra, and testes are important. Twins with two
sets of external genitalia can undergo successful separation; secondary
reconstructive genitoplasty may be required if only one set of external
genitalia is present. Staged procedures may be required to achieve
optimum outcome.

Central nervous system
Neurosurgical interest in conjoined twins has tended to focus on cra-
niopagus twins, who comprise only 2–6% of all conjoined twins but
present some of the greatest challenges in separation. A recent review
proposes a practical four-category classification based on the angle
of union (vertical or angular) and the degree to which the dural venous
sinuses are shared. Conjoined cerebral tissue may present an important
technical challenge, but preservation of the venous drainage of the
brain has emerged as one of the most critical determinants of outcome
following separation. Various surgical approaches have been reported.
Bony abnormalities of the spine, such as haemivertebrae remote from
the area of conjunction, put twins at risk for progressive spinal
deformity and scoliosis after separation.

Musculoskeletal system
The orthopaedic surgeon is predominantly involved early in the separa-
tion of ischiopagus twins. Three-dimensional reconstruction CT of
the pelvis is most helpful in ascertaining the anatomical configuration
of the pelvic ring and the possible junction of the vertebral columns. In
ischioptagus twins, the conjunction is at the pelvis with the twins lying
on their backs. The legs of each of the twins are widely separate with
the hips at right angles to the median plane. Diastasis of the pelvis is due
to external rotation of the posterior segment. Posterior osteotomies of
the iliac bones allow for medial rotation of the acetabula and symphy-
sis pubes, which restores the whole pelvic ring into normal alignment.
and facilitates anterior abdominal wall closure and urogenital closure, rendering stability to the perineum. Although osteotomy rarely prevents rediastration of the symphysis pubis, it helps early reconstruction of the pelvic anatomy and corrects acetabular retroversion to anteverision. The commonly encountered postoperative flexion deformities of the hips of 30°–50° usually resolve within 6 months. Associated with ischiopagus twins are spinal and cord abnormalities and lower limb abnormalities in nearly two-thirds of cases. Correction of the pelvic abnormalities ensured that all six ischiopagus children operated on at the author’s institution became community walkers.

Separation of asymmetric heteropagus twins requires the same detailed investigative approach and may also be a considerable surgical endeavour, but only one patient is at risk and thus skin cover can be supplied from the parasite and any organ conjunction is divided in favour of the autosite.

As mentioned previously, children with hemivertebrae, asymmetrical or diminutive chest cavities, and even those with caudal junction are prone to develop scoliosis. Progressive scoliosis in nonparalytic patients will not affect the hips—it is more a cosmetic deformity or affects respiratory capacity. Long-term follow-up is mandatory because rotational abnormalities, contractures, and dislocation of the hips, together with progressive scoliosis, can occur.

**Postoperative Management**

Cardiovascular and respiratory failures remain the most frequent causes of death in the immediate postoperative period. Further operations may be required for secondary wound closure or dehiscences and skin grafting. There is also hidden long-term morbidity and mortality. A number of infants have died later from factors such as unresolved aspiration from gastro-oesophageal reflux, bronchopneumonia aggravated by poor diaphragmatic function (particularly with thoracopagus), cerebral anoxia, gastroenteritis, urinary tract infections, biliary sepsis, and even malaria.

**Prognosis and Outcomes**

Inevitably, the ultimate prognosis will depend on the state of the conjoined organs and the potential for successful separation. Tragically in some, separation will not be possible. Detailed preoperative assessment is essential to determine the best surgical approach, reconstruction methods, and ultimate outcome. Despite successful separation, some children are left crippled and disabled, requiring lifelong follow-up and care. The overall survival for symmetrical twins is 33.3%, but it is 64.7% for those who underwent surgery. Emergency surgery had a dismal outcome with only around 33% surviving. Asymmetrical separation had a 92% survival rate.

**Prevention**

Better preconceptual maternal nutrition with folic acid supplementation is likely to reduce the incidence as it has the incidence of twinning abnormalities and spina bifida. If the diagnosis is made antenatally, then the decision to terminate the pregnancy may be taken after detailed evaluation and counselling.

**Ethical Issues**

Ethical considerations, which need to reconcile the best options for the twins and their parents, are playing an increasing role in present-day decision making. The sacrifice of one twin due to the inability to sustain life alone is the controversy that evokes the most anguish. The decision on whether to operate is rendered more complex by those surviving conjoined twins who consciously elect not to be separated and report that they have lived socially acceptable lives. Equal controversy surrounds those few conjoined twins who have survived to adulthood and then decide that separation should be attempted despite the operative risks and the potential for significant long-term morbidity as separate individuals.

Being conjoined does not necessarily negate individual development. Religious views may support only minimal surgical interference, especially when one twin has a high risk of dying at surgery. “We cannot accept one baby must die so that the other one may live. It is not God’s will”, which differs from the legal opinion “Why I must order twin baby to die”—both quotations were recorded in the press in a case in the United Kingdom when parents did not give consent to separation but doctors asked for legal support for separation. In the words of Eliza Chulkhurst, one of the most famous conjoined twins of the premedical era, “As we came together, we will also go together”.

From a practical point of view, the Great Ormond Street Ethical Guidelines for Conjoined Twin Separation have been widely accepted:

- Where separation is feasible with a reasonable chance of success, it should be carried out.
- When surgery is not possible, custodial care should be offered and nature allowed to take its course.
- Where one twin is dead or has a lethal abnormality and cannot survive independently from its normal twin and if unoperated both twins could die, separation to save the healthy twin should be attempted.

I acknowledge huge contributions of the teams of surgeons, anaesthetists, intensivists, nurses, social workers, physiotherapists and occupational therapists who, over the last 50 years, have so successfully managed our series of patients at the Red Cross War Memorial Children’s Hospital in Cape Town. Separation surgery was pioneered at this hospital by SidneyCywes and Jannie Louw and continued by my colleague Heinz Rode.

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**Key Summary Points**

1. Conjoined twins are usually symmetrical, of the same gender, and, in addition to the areas of conjunction, have an increased incidence of other congenital malformations.
2. Separation should be delayed for several months after birth, if possible.
3. Separation is not always possible nor indeed mandatory.
4. Success is achieved with meticulous attention to detail with a multidisciplinary team approach.
5. Long-term follow-up is always required to manage, in particular, musculoskeletal deformity and urogenital anomalies.
6. Expertise around the world can and should be shared.
**Suggested Reading**


