Systematic Review

Conjoined Thoracopagus Twins: A Systematic Review of the Anomalies and Outcome of Surgical Separation

Rahul Saxena, Arvind Sinha, Manish Pathak, Kirtikumar J Rathod

Department of Pediatric Surgery, All India Institute of Medical Sciences, Jodhpur, Rajasthan, India

Abstract

Introduction: Conjoined twin is an extremely rare condition and requires a thorough knowledge of anatomy, and a multidisciplinary approach is essential to successfully separate the twins. Thoracopagus twins lie face to face and are attached from chest to upper abdomen. They are the most common among all the varieties but have a poor survival rate. **Materials and Methods:** This study is a review of literature from 2019 to the oldest via PubMed and Google Scholar using keywords: Conjoined twins, Thoracopagus twins, Thoracoomphalopagus and Thoraco-omphalopagus twins. The articles were reviewed for the description of the anatomy of shared organs, management and outcome of these twins. **Results:** One hundred and fifty-eight sets of thoracopagus and thoraco-omphalopagus twins including our twins were included in this study. Out of 158 reported thoracopagus twin sets in literature, with M: F ratio of 1:2.3, 71 sets were found to be non-operable and all of them subsequently expired; 82 sets were operated upon, out of which 83 babies survived, suggesting an overall surgical success rate of about 50%. **Conclusion:** Thoracopagus twins have a dismal prognosis. The most important decisive parameter for successful separation is the extent of sharing of organs between twins. The role of a motivated multidisciplinary team is also indispensable and cannot be overemphasised.

Keywords: Conjoined twins, the survival of thoracopagus twins, thoraco-omphalopagus twins, thoracopagus twins

Introduction

Conjoined twins are one of the rarest congenital anomalies and have always been an interesting topic for clinicians. The occurrence of conjoined twins has always been highlighted in the media and has always caught the attention of society in general. The reported incidence of conjoined twins is 1:50,000, but due to the high rate of stillbirth, its true incidence remains at 1:200,000 live births. Females are three times more common than males.[1-4] Incidence is more common in sub-Saharan Africa reaching up to 1 in 400 monozygotic twins[2] and also in South American countries.[4]

Thoracopagus twins commonly share hearts and have complex cardiac anomalies which make separation difficult and worsen the outcome of surgery. A detailed pre-operative evaluation of the anatomy of shared organs and pre-operative preparation may help in the smooth conduction of the surgery. In this study, we have reviewed the literature about the imaging, pre-operative preparation, the complexity of shared organs and the outcome of thoracopagus twins.

Received: 27-06-2022 **Accepted:** 12-08-2022 **Available Online:** 19-01-2023

Access this article online

Quick Response Code:

Website: https://journals.lww.com/AJPS

DOI: 10.4103/ajps.ajps_77_22

Materials and Methods

The literature review was performed from the oldest available report in English to analyse all cases of thoraco-omphalopagus twins by searching online databases (PubMed/Medline, Google Scholar and Scopus) for the following keywords – 'conjoined twins', 'thoracopagus twins', thoracoomphalopagus and 'thoraco-omphalopagus'. We have included both thoracopagus and thoraco-omphalopagus twins because they were used to describe similar anatomy in previous literature. The study aimed to evaluate the survival rate after surgical separation of thoracopagus twins. The primary endpoints were age at the separation and survival after surgery. We also reviewed the variables such as birth history, born alive or stillborn, operable or non-operable, sex, shared organs and incidence of complex intracardiac anomalies. The categorical data were expressed

Address for correspondence: Dr. Rahul Saxena, Department of Pediatric Surgery, All India Institute of Medical Sciences, Jodhpur - 342 005, Rajasthan, India.
E-mail: drrahulsaxena@gmail.com

How to cite this article: Saxena R, Sinha A, Pathak M, Rathod KJ. Conjoined thoracopagus twins: A systematic review of the anomalies and outcome of surgical separation. Afr J Paediatr Surg 2023;20:157-65.

in percentages and continuous data were expressed in median and interquartile ranges (IQR).

Results

Including our patients, we found data of 158 thoracopagus and thoraco-omphalopagus sets in 7 case series,[1-7] 2 review articles,[8,9] and 36 case reports.[10-46] The age at surgery was mentioned for 63 sets ranging from 11 h–144 months, with a median age of 2.75 months (IQR = 5.5). Sex was documented in 77 sets with 23 males and 54 females (M: F = 1:2.3). Out of 158 sets, 71 (45%) sets were not operated as 20 sets were stillborn and 51 sets were found to be non-operable. All those who were non-operable ultimately died. The complex cardiac anomalies were present in 84 out of 158 sets (53%). The shared organ were liver in 14 (9%), hepatopancreatobiliiary and small intestine in 7 (4.4%) while the sharing of pericardium along with other organs was present in 27 (17%) patients.

Eighty-two sets (164 babies) were operated, and the outcome of 5 sets was not documented. Out of 164 babies, 83 babies survived after surgery, 5 were sacrificed during surgery and 76 patients were lost after surgery suggesting an overall surgical success rate of about 50.6% [Table 1].

The 83 survivors have resulted from the separation of 82 twin sets with a survival rate of 50.6%. The heart was shared with the presence of complex cardiac anomalies in 7 (8.4%) patients, while 40 patients (48%) had only common pericardium. The liver was shared in 70%, the small bowel in 14% and the diaphragm in 10% of patients [Table 2]. The complex cardiac anomalies were present in 45 (55.5%) out of 81 patients who were lost, which was significantly higher than that in survivors (Chi-square 42.03, $P < 0.0000$), while the hearts were shared with pericardium in 8 (10%) patients [Table 3].

Discussion

Conjoined twins always belong to the same sex as they are monozygotic, monochorionic and monoamniotic.[2] The defect in embryogenesis is either a failure of separation of embryonic discs at 15–17 days[47] or the fusion of two embryonic discs.[2,47] Moreover, there are no definite genetic or environmental predisposing factors associated with them.[48]

They are classified according to the fused body part and added with the suffix 'pagus' which is a Greek term meaning fixed.[49] Thoracopagus are those who have fused chest and upper abdomen walls, and usually, they have some degree of the sharing of hearts. Omphalopagus twins are fused in the area of the umbilicus and often include the lower chest but never share the heart.[48,49] Thoracopagus variants account for almost 40% of all varieties, and omphalopagus accounts for 32% of cases.[1] According to some authors, the thoracopagus twins always share a part of the abdomen along the thorax, they may have an exomphalos and share organs such as the heart (75%), pericardium (90%), liver (100%), bile ducts (25%) and upper small intestine (50%) and there is no need of defining combined types like thoraco-omphalopagus.[1,47-49] However, many authors used the term thoraco-omphalopagus to describe the twins which are fused from the thorax to the upper abdomen.[10-13,21-23,25-27] The thoraco-omphalopagus twins account for 70% of conjoined twins and are associated with the highest mortality rates of 51% due to complex cardiac anomalies.[5,10,13]

The separation of conjoined twins is a challenging process and requires a multidisciplinary team approach. The overall success rate of separation is around 50.6%.[1-4] The first case of antenatal ultrasound (US) detection of conjoined twins was reported in 1977,[3] and the earliest antenatal diagnosis has been reported at 12 weeks of age. However, the details are usually well confirmed around 20 weeks of gestation.[2,5] The diagnosis of conjoined twins is suspected in cases of twin pregnancy with polyhydramnios with a single placenta and an absence of separate amniotic membranes.[5] The criteria for making a diagnosis of conjoined twins on ultrasonography include various signs such as inseparable foetal bodies and skin contours heads at the same level as the fixed position of foetuses, an abnormal number of cord vessels (>3), an unusual extension of the spine, bi-breech or bi-cephalic presentation and the presence of a single heart with complex anomalies.[2,5,50] An echocardiogram is required to diagnose the associated cardiac anomalies and compatibility to life. The termination of pregnancy is advisable when the investigations demonstrate a shared heart or complex anomalies, which preclude a safe separation. This should be accompanied by proper and fully informed parental counselling.[2,5] All attempts are directed towards the termination of pregnancy in the second trimester, especially in cases that demonstrate anomalies incompatible with life. This allows for a termination of pregnancy by the vaginal route, avoids caesarean section and minimises maternal morbidity. The findings of ultrasonography may be complemented with foetal magnetic resonance imaging, which helps in assessing the feasibility of separation, especially in cases with fused hearts, and is extremely helpful in deciding to continue the pregnancy. Whenever the decision to continue a pregnancy is taken, the delivery should be performed by caesarean section at 36–38 weeks to avoid stillbirth, dystocia and vaginal injuries.[1,2,5,6]

Post-natal investigations and imaging are required to evaluate the degree of fusion and associated anomalies to assess the feasibility of separation and compatibility of life. Thorough pre-operative planning should include imaging to evaluate the cardiac, hepatopancreatobiliary anatomy and gastrointestinal anatomy [Table 4].

Providing safe anesthesia is a major challenge in separation surgery, and two separate colour-coded teams for each twin should be designated. The whole team should be sensitised to the anticipated problems likely to be encountered during the separation, and the course of possible corrective actions should be discussed among the team members. The doses of drugs are calculated for the total weight and half is instituted

Table 1: Outcome of thoracopagus twins

Study	Number of thoracopagus sets (*n*=158)	Non-operable (*n*=71)	Outcome of non-operated cases (*n*=71)	Operated (*n*=82 sets; 164 babies); 5 - outcome not reported	Survival after surgery (*n*=83)	Mortality after surgery (*n*=76 and 5 sacrificed)
Spitz and Kiely[1,5]	10	6	Death - 6 sets	4 sets (3 emergencies, 1 elective)	3	5
Rode *et al*.[2]	22	8 stillborn, 5 born alive but died within 2 months=13 sets	Death - 13 sets	9 sets (1 - emergency, 8 - elective)	10	8
O'Neill *et al*.[3]	5	2	Death - 2 sets	3 sets (2 - emergencies, 1 - elective)	0	6; two late death
Tannuri *et al*.[4]	7	6	Death - 6 sets	1-emergency (one twin died and a separated twin died at 11 months)	0	2; one late death
Al Rabeeah[6]	10	10 sets - 1 stillborn; 9 alive	Death - 10 sets within 3 weeks	none	-	-
Saguil *et al*.[7]	12	5 sets	Death - 5 sets	7 sets (2 emergency and 5 elective=7 sets)	5	9
McMahon and Spencer[8]	33	10 stillborn; 6 alive Non-operable=16 sets	Death- 16 sets within 1 day	12 operated; outcome not reported for 5 sets	4	20
Mulcare *et al*.[9]	14	-	-	14 sets	13	13; 2 sacrificed
Thompson *et al*.[10]	1			Elective	2	0
Boles and Vassy[11]	1			Elective	2	0
Singh *et al*.[12]	1			Elective	2	0
Lalwani *et al*., 2011[13]	1			Elective	2	0
Saranrittichai *et al*., 2007[14]	1			Elective	2	0
Elizondo *et al*., 2017[15]	1			Elective	2	0
Hedrick, 2003[16]	1			EXIT	1	1; sacrificed
Tug *et al*., 2009[17]	3	3	Death - 3 sets	-	-	-
Chen *et al*., 2012[18]	1	-	-	Emergency	2	0
Ambar *et al*., 2010[19]	1	1	Death at 18 h	-	-	-
Asaranti *et al*., 2012[20]	1	1	Stillborn	-	-	-
Ekenze *et al*., 2009[21]	1	-	-	Elective	2	0
Bahador *et al*. 2020[22]	2	-	-	2; elective	4	0
Rossetti *et al*., 2020[23]	1	-	-	Elective	2	0
Wu *et al*., 2018[24]	2	-	-	2 sets (1 elective and 1 emergency)	2; elective	2; emergency
Freitas *et al*., 2019[25]	1	-	-	emergency	1	1 sacrificed
Richtsfeld *et al*., 2018[26]	1	-	-	elective	2	
Abdullah *et al*., 2017[27]	1	1	Death on 2nd day of life			
Wood *et al*., 2017[28]	3	-	-	3 sets (2 - elective 1 - emergency)	4-elective ones	2-emergency
Park *et al*., 2016[29]	1	-	-	emergency	0	2
Aneja *et al*., 2013[30]	1	1	Death immediately after birth			
Tekgündüz *et al*., 2013[31]	1	1	Death immediately after birth			
Wen *et al*., 2013[32]	1	-	-	Elective	2	0
Chelliah *et al*., 2012[33]	1	1	Death at 7th day			
Hamdan *et al*., 2010[34]	1	1	Death at 50th day			
Piaseczna-Piotrowska *et al*., 2009[35]	1	-	-	Elective	2	0

Contd...

Table 1: Contd...

Study	Number of thoracopagus sets (*n*=158)	Non-operable (*n*=71)	Outcome of non-operated cases (*n*=71)	Operated (*n*=82 sets; 164 babies); 5 - outcome not reported	Survival after surgery (*n*=83)	Mortality after surgery (*n*=76 and 5 sacrificed)
Karpelowsky and Millar, 2010[36]	1	-	--	Elective	2	0
Wataganara *et al*., 2008[37]	1	-	-	Elective	2	0
Mair and Mair, 2006[38]	1	1	Death at 6 months		-	-
Ray *et al*., 2004[39]	1	-	-	Elective	2	0
el Gohary, 1998[40]	3	1	Death at 7th day	2 sets - elective	0	4
Meyers and Matlak, 2002[41]	1	-	-	Emergency	2	0
Jaffray *et al*., 1999[42]	1	-	-	Emergency	0	2
Jung *et al*., 1997[43]	1	-	-	Elective	2	0
Simpson, 1969[44]	1	1	Death immediately after birth	-	-	-
Micheli *et al*., 1978[45]	1	-	-	Emergency	0	2; one sacrificed and the other died after 11 h
Sinha *et al*.,[46]	1	-	-	Elective	2	0
Total	158	71 sets - 20 stillborn; 51 born alive		82 sets operated; 5 not reported (*n*=82×2=164 babies)	*n*=83	76 deaths; 5 sacrificed

EXIT: *Ex utero* intrapartum treatment

for each baby. Intubation may be difficult due to the positioning and prematurity of babies and may need several attempts. Nasal intubation may provide better stability while shifting. Full arterial and central venous access should be taken for proper monitoring and massive transfusions. All lines and monitoring cables should be colour coded, to avoid confusion while separation and transportation.[2,5] The intraoperative blood loss is significant in separation and its replacement is always challenging, as it is difficult to attribute the exact volume lost by a baby. To expedite the surgery and avoid hypovolaemic shock, two separate colour-coded surgical teams should be formed. This allows each team to perform the reconstruction simultaneously saving time and preventing confusion among the teams.[3]

The post-natal course of conjoined twins may fall into non-operative management, emergency separation and elective separation.[1-4] The non-operative management is followed, where complex cardiac anomalies preclude a separation and the possibility of reconstruction of even a single working heart. The emergency separation is reserved for those sets of conjoined twins who develop complications such as rupture of exomphalos, cardiac instability, liver injury causing blood loss, volvulus and necrosis of the intestine and death of one twin.[1] Elective separation is performed when the infants are in an optimal physiological state.[2] The optimal time for surgical separation is 3 months as in our case, however, it may be extended up to 3 years depending upon individual cases.[1-5] This pre-operative interval allows for the completion of the exhaustive work-up, which is required to assess the unique anatomy of conjoined twins and for accurate pre-operative planning and rehearsal. This also allows babies to attain pulmonary and immunologic maturity to tolerate surgical stress. The waiting period can be utilised, to settle the moral and ethical considerations when the survival of one or both babies is at risk. The parents should also be active participants in this ethical decision-making, and all attempts should be made to inform the parents of the risks and to take their input and consent for the many difficult decisions that are required to be taken. The waiting period is also used for the preparations for the proper set-up of surgical separation and for skin growth by tissue expanders to allow for closure.[1-4] CT scan with 3D reconstruction allows 3D printing to create models, which can help in deciding the site of injection and the amount of tissue expanders. These models can be used to measure the dimensions of anticipated soft tissue defects and to design the flaps to cover the defect.[28]

The reported survival rate in previous literature for emergency separation of conjoined twins is around 30% while that of elective separation reaches up to 80%.[1-4] The cumulative survival rate after surgical separation in our series is 50.6% and it includes both emergency and elective separations.

Other factors responsible for the outcome are the distribution of organs between twins, surgical technique used in separation, reconstruction method followed, wound closure technique and the provision of dedicated post-operative care. McMahon and Spencer reported a 100% incidence of congenital heart defects in 314 thoracopagus twins.[8] Thoracopagus twins may have a spectrum of cardiac anomalies and are classified as group A: separate hearts, separate pericardium; group B: separate hearts, common pericardium; group C: fused atria, separate ventricles and group D: atrial and ventricular fusion.[8,50] The majority of thoraco-omphalopagus have group A and most thoracopagus have group C and D cardiac anomalies.[51]

Table 2: The shared organs in the sets where at least one of the twins survived after the surgery

Studies	Operated	Survival	Mortality after surgery	Shared organs	Associated anomalies
Spitz and Keily[1,5]	1; emergency	1	1	Pericardium. Diaphragm, liver, CBD, small bowel	PA
	1;elective	2	0	Pericardium, liver, CBD, small bowel	Exomphalos, EHBA
Rhode *et al*.[2]	9; (1-emergency, 8- elective)	10	8	not mentioned	
Saguil E *et al*.,[7]	1; elective	1	1	Liver, pericardium	
	1; elective	2	0	Liver, pericardium	
	1; elective	2	0	Liver, pericardium	
	1	1	1	LH - normal; RH - common atrium, SV (IM), PTA	
	1	2	0	normal hearts	
	1	1	1	LH - ASD, VSD; RH - Dextrocardia, hypo RV, PAT, TAT, LVSC-CS	
Mulcare *et al*., [7]	1	1	1	Liver	None
	1	2	0	Liver	None
	1	2	0	Liver	Exocardia
	1	1	1	Liver, small bowel	Collapsed ileum and colon of one twin
	1	2	0	Liver	None
	1	2	0	Liver	None
	1	2	0	Liver	None
	1	1	1	Liver	None
Thompson JL *et al*.,[10]	1; elective	2	0	Pericardium, liver, CBD, duodenum	Ectopia cordis, large bridging portal vein extending from one twin to another twin portal system
Boles *et al*.,[11]	1; elective	2	0	Pericardium, liver	None
Singh M *et al*.,[12]	1; elective	2	0	Pericardium, liver	
Lalwani J *et al*.,[13]	1; elective	2	0	Liver	None
Saranrittichai S *et al*.,[14]	1; elective	2	0	Liver, bile duct, small bowel from the duodenum to the terminal ileum	Three GB, single bile duct
Elizondo *et al*.,[15]	1; elective	2	0	Diaphragm, liver and portions, of the lung, were shared, and bowel loops were comingled, shared bladder and crossing ureters with HDN	
Holly L. Hedrick[16]	1; EXIT	1	1 sacrificed	Common liver and a portion of the umbilical vein, rudimentary heart in complete heart block	
Chen G *et al*.,[18]	1; emergency	2	0	Liver and heart Twin A PDA; Twin B d-TGA and PDA	
Ezenke SO *et al*.,[21]	1; elective	2	0		Ruptured omphalocele
Bahador A *et al*.[22]	1; elective	2	0	Pericardium and left lobe of liver	
	1; elective	2	0	Pericardium and left lobe of liver	
Rossetti *et al*.[23]	1; elective	2	0	Pericardium	
Wu S *et al*.[24]	1; elective	2	0	Pericardium	
Freitas MH *et al*.,[25]	1; emergency	1	1 sacrificed	liver	
Richtsfeld M *et al*.,[26]	1; elective	2	0	Shared liver, a shared pericardial sac and a venous connection between the right atria of both twins	Twin A normal heart Twin B - TA, ASD, d-TGA and VSD
Wood BC *et al*.,[28]	1; elective	2	0	Pericardium and liver	Twin A ASD and Twin B malpositioned ventricle and VSD
	1; elective	2	0	Pericardium and liver	Twin A - Tetralogy of Fallot
wen X *et al*.,[32]	1	2	0	Pericardium, liver, diaphragm	Major vascular connection between hearts and liver of twins
Piaseczna-Piotrowska A *et al*.,[35]	1 elective	2	0	Pericardium and liver	Small VSD and PDA in both twins
Karpelowsky JS *et al*.,[36]	1; elective	2	0	Pericardium and liver	

Contd...

Table 2: Contd...

Studies	Operated	Survival	Mortality after surgery	Shared organs	Associated anomalies
Wataganara T *et al*.,[37]	1; elective	2	0	Liver	
Ray AK *et al*.,[39]	1; elective	2	0	Pericardium, liver, diaphragm and pleura	
Meyer RL *et al*.,[41]	1; emergency	2	0	Liver with shared circulation, common giant cystic duodenum/jejunum	Twin A biliary atresia, twin B large VSD/double outlet in right ventricle
Jung PM[43]	1; elective	2	0	Pericardium, liver, diaphragm	
Sinha A *et al*.[46]	1; elective	2	0	Pericardium and liver	
		83	81		

ASD: Atrial septal defect, CBD: Common bile duct, EHBA: Extrahepatic biliary atresia, EXIT: Ex utero intrapartum treatment, d-TGA-Dextro-transposition of the great arteries, GB: Gallbladder, HDN: Hydronephrosis, IM: Indeterminate Morphology, LH: Left heart, LVSC-CS: Persistent left superior vena cava with intact coronary sinus, PTA: Patent truncus arteriosus, PA: Pulmonary atresia, PDA: Patent ductus arteriosus, RV: Right ventricle, RH: Right heart, SV: Single ventricle, TAT: Tricuspid Atresia, TA: Tricuspid atresia, VSD: Ventricular septal defect,

In thoracopagus twins with shared hearts, the ventricles may lie ventral and caudal to the atria, but rarely, the atria may remain caudal to the ventricles.[8] These may be associated with congenital cardiac anomalies such as atrioventricular septal defects, aortic stenosis, tetralogy of Fallot, anomalous pulmonary veins, pulmonary hypoplasia and atresia and abnormal vena cava and origin of major arterial vessels from the aortic arch.[2-6] The most common atrial malformation is a common atrium with a large atrial septal defect, and the most common ventricular malformation is a single ventricle with an infundibular outlet chamber and a large ventricular septal defect.[33] The high incidence of complex cardiac anomalies is responsible for the dismal prognosis of thoracopagus twins. When pre-operative investigations reveal that the two hearts are not surgically separable, a decision can be made to sacrifice one of the twins. The separation is then accomplished by ligating the aorta and vena cava of the sacrificed twin, and the entire heart and vascular tree are placed in surviving twin. Most of the children separated for ventricular fusion do not survive for any extended period and even after the sacrifice of one twin at separation and the other twin demised within 2–6 weeks of such procedure.[1-3,5] Spitz[5] and Al Rabeeah[6] reported only 3 out of 13 and 1 out of 7 thoracopagus sets, in whom the sharing of hearts was limited to pericardium only.

The liver is shared in almost all the sets of thoracopagus twins. The conjoined liver is oriented in an oblique plane so that the liver of one twin is anterior and another twin is posterior with superimposition of pancreaticobiliary trees.[3,4] The separation is feasible when each liver has separate hepatic veins draining into its vena cava. When there is a single shared hepatic vein, the hepatic veins of one twin may traverse the liver to enter the heart of another twin and the twin without hepatic veins should be sacrificed whereas the heart is autotransplanted to the twin with hepatic veins.[3] Moreover, it is not always possible to define biliary anatomy preoperatively and is best identified during surgery. The thoracopagus twins are frequently associated with anomalies of the biliary tree. Pre-operative hepatobiliary scintigraphy is recommended, to delineate the biliary anatomy, but it may not be helpful when the anatomy of biliary systems is truly complex and obstructed.[41] The two separate gallbladders should be identified, but there may be a single extrahepatic biliary system (EHBS) when there is a fusion of the proximal duodenum. These cases may require hepaticoduodenostomy, Roux-en-Y choledochojejunostomy or hepaticojejunostomy, external drainage and allocating duodenum and EHBS along with pancreas to one twin and performing hepaticojejunostomy in another one.[2,3,14,41] Rarely, the fusion of the duodenum may extend up to the level of Meckel's point, separating into two ilea. The proximal duodenum of two twins may be opening into common cystic duodenum/jejunum, which may complicate by perforation.[41,42] The closure of the defect is always a concern in thoracopagus twins due to its large size. The tissue expanders help in closure by increasing tissue surface for reconstruction, but most of the authors advise against its use due to frequent complications such as wound infection and skin necrosis in up to 60% of cases and most of the authors prefer closure with the help of polypropylene mesh or skin grafts.[2-4,6] A plastic liner should be placed beneath the mesh to avoid adhesion of viscera to mesh and fluid losses. The mesh can be plicated on alternate days to achieve the contraction of wound. The final closure is usually possible within two weeks and at that time the plastic liner is removed.[1] The prosthetic patches are made up of polytetrafluoroethylene like GORE-TEX® soft tissue patch (W. L. Gore and Associates, Flagstaff, Arizona) and Permacol® (Tissue Science Laboratories PLC, Aldershot, UK); the acellular sheet of porcine dermal collagen can be used to close pericardium and abdominal defects. The chest wall defects can be reconstructed by absorbable platings.[28,36]

The prognosis of thoracopagus twins is very dismal. Out of 158 reported thoracopagus twin sets, 71 sets were found to be non-operable and all subsequently expired; 82 sets (164 babies) were operated, out of which 83 babies survived, suggesting an overall surgical success rate of about 50% [Table 1].

Table 3: The shared organs in the sets where at least one of the twins died after the surgery

Study	Operated	Survival	Mortality after surgery	Shared organs	Associated anomalies
Spitz and Keily[1,5]	1; emergency	1	1	Pericardium. diaphragm, liver, CBD, small bowel	PA
	1; emergency	0	2; one early other at 6 weeks	Cardiac atrial, liver	Unilocular heart
	1; emergency	0	2	Heart, diaphragm, liver	Diaphragmatic hernia, unilocular heart
Rhode *et al*.[2]	9; emergency-1, planned 8	10	8	Not mentioned	
O'Niell *et al*.,[3]	1	0	2 - one sacrificed and one late death at 4 months	Heart - 4 chambers in one; 3 chambers in the other twin communicating at the ventricular level	
	1	0	One died at 4 weeks and the other died at 6 months	Liver, biliary system, duodenum and jejunum	
	1	0	2	Heart	No hepatic veins in one twin and single left ventricle, severe PS and left superior vena cava
	1	0	1, one late death	Pericardium, liver and duodenum	
Saguil E *et al*.,[7]	1; planned	1	1	Liver, pericardium	
	1; emergency	0	2	Liver pericardium	Intracardiac defects in one twin; one died of cardiac failure and the other of NEC
	1; planned	0	2	Liver pericardium	
	1; planned	0	2	Liver, xiphoid	
	1; planned	0	2	Liver, xiphoid	
Mc Mahon CJ and Spencer R.,[9]	1; emergency	0	2; one immediately other after 2 months	Heart - 2 atria, 2 ventricles ASD, VSD, right TGA, left PS	
	1	0	2	Heart - Common right atrium, shared LV, VSD, another twin: hypo RV, PS, shared coronary artery	
	1	0	2	Common right atrium, shared LV, VSD, another twin: Hypo RV, PS, shared coronary artery	
	1	0	2; within 11 h	Single QRS, one twin single ventricle, another twin; VSD, TGA, shared coronary	
	1	0	2 at 36 h	Single QRS, fused RV	
	1	0	2	LH-Dextrocardia pulmonary atresia, VSD, ASD, LSVC-CS, RH - tricuspid atresia, ASD, VSD	
	1	0	2	LH - d-TGA, IAA, PDA; RH - ASD/ VSD, interrupted IVC, connection right liver to left mesenteric artery	
	1	1	1	LH - normal; RH - common atrium, SV (IM), PAT	
	1	0	2	LH - coarctation, VSD; RH- Common atrium, Hypo RV, D-TGA, PS	
	1	0	2 - both died by 8 h	LH - LVSC-CS; RH- normal	
	1	1	1	LH - ASD, VSD; RH - Dextrocardia, hypo RV, PAT, TAT, LVSC-CS	
Mulcare RJ *et al*.,[9]	1	1	1	Liver	None
	1	0	2	Liver	Exocardia
	1	0	2	Liver, heart	Cardiovascular, multiple
	1	0	2=1 at 6 h, sacrificed-1	Liver, heart	Cardiovascular, multiple
	1	0	2	Liver	None
	1	0	2	Liver	Cardiovascular, multiple

Contd...

Table 3: Contd...

Study	Operated	Survival	Mortality after surgery	Shared organs	Associated anomalies
Mulcare RJ *et al.,*[9]	1	1	1	Liver, small bowel	Collapsed ileum and colon of one twin
	1	0	2=1 at 4 h, sacrificed-1	Liver, heart	None
	1	1	1	Liver	None
Holly L. Hedrick[16]	1; EXIT	1	1 sacrificed	Common liver and a portion of the umbilical vein, rudimentary heart in complete heart block	
Wu S *et al*.[24]	1; emergency	0	2	Ventricles and liver	
Freitas MH *et al*.,[25]	1;emergency	1	1 sacrificed	Liver	
	1;emergency	-	1 died at POD 7 other on POD 94	Pericardium and liver	Twin A - Tetralogy of Fallot
Park *et al*.[29]	1; emergency	0	1 died after 5 h and other on 100th day	Pericardium, joined atria and liver	Twin A - TAPVR, d-TGA, VSD, Twin B - TAPVR
elGohary[40]	1; planned	0	Both died of all after 3 months		
	1; planned	0	Both died; first after 1 month and second after 2 months later	Common liver, duodenum and part of the jejunum	Communication between atria of two twins
Jaffray *et al*.[42]	1 emergency	0	2	Liver and dilated common jejunal loop	
Micheli *et al*.[45]	1; emergency	0	1 sacrificed and the other died 11 h after surgery	Heart with shared atria and ventricular wall, liver, duodenum and jejunum	TGA

ASD: Atrial septal defect, CBD: Common bile duct, d-TGA: Dextro-transposition of the great arteries, EXIT: Ex utero intrapartum treatment, IAA: Interrupted aortic arch, IM: Indeterminate morphology, IVC: Inferior vena cava, LH: Left heart, LV: Left ventricle, LVSC-CS: Persistent left superior vena cava with intact coronary sinus, NEC: Necrotising enterocolitis, PA: Pulmonary atresia, PAT: Pulmoary atresia PDA: Patent ductus arteriosus, POD: Post-operative day, PS: Pulmonary stenosis, RH: Right heart, RV: Right ventricle, SV: Single ventricle, TAPVR: Total anomalous pulmonary venous return, TAT: tricuspid atresia, VSD: Ventricular septal defect

Table 4: List of investigations recommended for thoracopagus twins

Investigations	Indications
Plain X-ray	Bony anatomy and fusion
Ultrasound abdomen and Doppler	To assess fusion of liver and presence of separate GBs, pancreas, spleen, kidneys, great vessels of abdomen and hepatic veins
Electrocardiogram	Single QRS complex indicated poor chances of separation although two separate ECG does not rule out significant fusion of hearts[2]
Echocardiogram	Anatomy of heart, detection of associated congenital cardiac anomalies and understand cardiac connections[19]
GI contrast studies	Identification of separate GI systems
Computed tomography	Assess liver and vascular connections, functioning of kidneys, ureteric and bladder anatomy
Hepatobiliary scintigraphy	Details of biliary anatomy like whether GBs and extrahepatic biliary tract are separate
MRI	3D reconstruction of cardiac, vascular and hepatobiliary anatomy for accurate pre-operative planning of separation

GI: Gastrointestinal, MRI: Magnetic resonance imaging, 3D: Three-dimensional, GBs: Gallbladders

Conclusion

Conjoined twins bear a poor survival rate and very few successful separations have been reported until now in literature. The success of separation surgeries in thoracopagus twins is dependent mainly on the extent of shared organs between twins. A shared heart and a single set of hepatic veins preclude the survival of both twins. A small proportion of such twins who have separate hearts and common pericardium have better survival rates and with exhaustive pre-operative investigations and rehearsals, successful separation can be achieved.

Ethical approval

This article does not contain any studies with human participants or animals performed by any of the authors.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Spitz L, Kiely EM. Experience in the management of conjoined twins. Br J Surg 2002;89:1188-92.
2. Rode H, Fieggen AG, Brown RA, Cywes S, Davies MR, Hewitson JP, *et al*. Four decades of conjoined twins at red cross children's hospital – lessons learned. S Afr Med J 2006;96:931-40.
3. O'Neill JA Jr., Holcomb GW 3rd, Schnaufer L, Templeton JM Jr., Bishop HC, Ross AJ 3rd, *et al*. Surgical experience with thirteen conjoined twins. Ann Surg 1988;208:299-312.
4. Tannuri AC, Batatinha JA, Velhote MC, Tannuri U. Conjoined twins: Twenty years' experience at a reference center in Brazil. Clinics (Sao Paulo) 2013;68:371-7.

5. Spitz L. Conjoined twins. Prenat Diagn 2005;25:814-9.
6. Al Rabeeah A. Conjoined twins-past, present, and future. J Pediatr Surg 2006;41:1000-4.
7. Saguil E, Almonte J, Baltazar W, Acosta A, Caballes A, Catangui A, *et al*. Conjoined twins in the Philippines: Experience of a single institution. Pediatr Surg Int 2009;25:775-80.
8. McMahon CJ, Spencer R. Congenital heart defects in conjoined twins: Outcome after surgical separation of thoracopagus. Pediatr Cardiol 2006;27:1-12.
9. Mulcare RJ, Bhokakul P, Potitung P, Wheeler B. The surgical separation of the thoracopagus conjoined twins of Korat, Thailand. Ann Surg 1970;172:91-7.
10. Thompson JL, Zarroug AE, Matsumoto JM, Moir CR. Anatomy of successfully separated thoracopagus-omphalopagus conjoined twins. Clin Anat 2007;20:814-8.
11. Boles ET Jr., Vassy LE. Thoraco-omphalopagus conjoined twins: Successful surgical separation. Surgery 1979;86:485-92.
12. Singh M, Jacob R, Naik V, Baines D. Separation of thoraco-omphalopagus twins in a rural secondary hospital: Perioperative management. Indian J Anaesth 2012;56:442-7.
13. Lalwani J, Dubey K, Shah P. Anaesthesia for the separation of conjoined twins. Indian J Anaesth 2011;55:177-80.
14. Saranrittichai S, Niramis R, Siripornadulsilp S, Thianjaruwatthana W, Hompleum T, Tipsunthonsak N, *et al*. Successful separation of thoracopagus conjoined twins with a single extra-hepatic biliary system. J Med Assoc Thai 2007;90:994-1000.
15. Elizondo RA, Au JK, Roth DR, Olutoye OO, Cass DL, Koh CJ. Urologic considerations in the separation of thoracoomphalopagus conjoined twins. Urology 2017;99:231-3.
16. Hedrick HL. Ex utero intrapartum therapy. Semin Pediatr Surg 2003;10:190-5.
17. Tug N, Sahinoglu Z, Cam C, Eren S. Route of delivery of thoracoomphalopagus twins: Analysis of three cases. Arch Gynecol Obstet 2009;280:815-8.
18. Chen G, Jia B, Zhang WB. Successful surgical repair of d-transposition of the great arteries in a separated conjoined twin. World J Pediatr 2012;8:371-3.
19. Ambar SS, Halkati PC, Patted SV, Yavagal S. Twin heart with a fused atria and separate ventricles in conjoined twins. Ann Pediatr Cardiol 2010;3:196-8.
20. Asaranti K, Pranati M, Tushar K, Jagadish B, Susmita B, Amarendra N. Autopsy findings in conjoined twin with single heart and single liver. Case Rep Pathol 2012;2012:129323.
21. Ekenze SO, Ibeziako SN, Adimora GN, Anyanwu PA, Ekwunife H, Ajuzieogu VO. Ruptured omphalocele in thoracoomphalopagus conjoined twins. Int Surg 2009;94:221-3.
22. Bahador A, Nikeghbalain S, Foroughi M, Beizavi Z, Sahmodini MA, Eghbal MH, *et al*. Successful surgical separation of thoraco-omphalopagus symmetrical conjoined twins in Iran: Two case reports. Iran J Med Sci 2020;45:143-7.
23. Rossetti E, Giudici DL, Bianchi R, Chiusolo F, Grimaldi C, Cetrano E, *et al*. Effective neurally-adjusted ventilator assist weaning off mechanical ventilation in separated conjoined thoraco-omphalopagus twins with sternal MeDPor implant patch. Minerva Anestesiol 2020;86:220-1.
24. Wu S, Guo K, Xiao P, Yang J, Sun J. Body wall reconstruction for conjoined twins: Our experience and lessons learned. Ann Plast Surg 2018;81:S66-70.
25. Freitas MH, Lima LC, Couceiro TC, Costa MC, Freitas MH. Anesthesia and perioperative challenges for surgical separation of thoraco-omphalopagus twins: Case report. Braz J Anesthesiol 2019;69:214-7.
26. Richtsfeld M, Konia M, Hiremath G, Kloesel B. Anesthetic management during atrial septostomy in a conjoined thoraco-omphalopagus twin with tricuspid atresia and d-transposition of the great arteries before separation: A case report. A A Pract 2018;10:298-301.
27. Abdullah H, Abdul Wahab N, Abu Bakar K. Fetal MRI of thoraco-omphalopagus conjoined twins. BMJ Case Rep. 2017 Jun 13;2017:bcr2017219793. doi: 10.1136/bcr-2017-219793.
28. Wood BC, Sher SR, Mitchell BJ, Oh AK, Rogers GF, Boyajian MJ. Conjoined twin separation: Integration of three-dimensional modeling for optimization of surgical planning. J Craniofac Surg 2017;28:4-10.
29. Park JS, Park JJ, Kim DY, Eom JS. Emergency transplantation of free flap between separated thoraco-omphalopagus conjoined twins. Arch Plast Surg 2016;43:97-9.
30. Aneja A, Rajanna DK, Reddy VN, Mayilvavaganan KR, Pujar P. Conjoined twins: A rare case of thoraco-omphalopagus. J Clin Diagn Res 2013;7:1471-2.
31. Tekgündüz KŞ, Olgun H, Caner I, Ceviz N. Thoraco-omphalopagus conjoined twins [corrected]. Anadolu Kardiyol Derg 2013;13:284-5.
32. Wen X, Parajuly SS, Lu Q, Xiang B, Jiang X, Tang H, *et al*. Thoraco-omphalopagus conjoined twins: Impact of ultrasound assessment on successful surgical separation. Clin Imaging 2013;37:138-42.
33. Chelliah KK, Faizah MZ, Dayang AA, Bilkis AA, Shareena I, Mazli M. Multimodality imaging in the assessment of thoraco-omphalopagus conjoined twin: Lessons to learn. Case Rep Radiol 2012;2012:564036.
34. Hamdan MA, Begam M, Mirghani HM. Inseparable thoraco-omphalopagus twins. Pediatr Cardiol 2010;31:1252-3.
35. Piaseczna-Piotrowska A, Chilarski A, Krajewski W, Godlewska-Tarka J. Thoraco-omphalopagus twins: Different perinatal circumstances, different outcome. Arch Med Sci 2010;6:461-3.
36. Karpelowsky JS, Millar AJ. Porcine dermal collagen (Permacol) for chest and abdominal wall reconstruction in thoraco-omphalopagus conjoined twin separation. Pediatr Surg Int 2010;26:315-8.
37. Wataganara T, Sutanthaviboon A, Ngerncham S, Vantanasiri C. Three-dimensional power Doppler in the diagnosis and surgical management of thoraco-omphalopagus conjoined twins. Ultrasound Obstet Gynecol 2008;32:236-7.
38. Mair AA, Mair EA. Two babies, one heart, and no airway. Int J Pediatr Otorhinolaryngol 2006;70:1489-93.
39. Ray AK, Mukherjee NN, Mukherjee G, Patra R, Ghosh DK. Separation of thoraco-omphalopagus Siamese twin. Indian J Pediatr 2004;71:755-7.
40. el-Gohary MA. Siamese twins in the United Arab Emirates. Pediatr Surg Int 1998;13:154-7.
41. Meyers RL, Matlak ME. Biliary tract anomalies in thoraco-omphalopagus conjoined twins. J Pediatr Surg 2002;37:1716-9.
42. Jaffray B, Russell SA, Bianchi A, Dickson AP. Necrotizing enterocolitis in omphalopagus conjoined twins. J Pediatr Surg 1999;34:1304-6.
43. Jung PM. Successful separation of thoraco-omphalopagus and ischiopagus tetrapus twins in Korea. Pediatr Surg Int 1997;12:607-10.
44. Simpson JS. Separation of conjoined thoracopagus twins, with the report of an additional case. Can J Surg 1969;12:89-96.
45. Micheli JL, Sadeghi H, Freeman J, Bozic C, Queloz J, Genton N. An attempt to separate xiphopagus twins sharing a common heart, liver, and duodenum. J Pediatr Surg 1978;13:139-42.
46. Sinha A, Saxena R, Pathak M, Rodha MS. Conjoined thoracopagus twins-Our experience of successful separation. J Indian Assoc Pediatr Surg 2021;26:354-7.
47. Spencer R. Theoretical and analytical embryology of conjoined twins: Part 1: embryogenesis. Clin Anat 2000;13:36-53.
48. Mutchinick OM, Luna-Mun˜oz L, Amar E, Bakker MK, Clementi M, Cocchi G, *et al*. Conjoined twins: A worldwide collaborative epidemiological study of the International Clearinghouse for Birth Defects Surveillance and Research. Am J Med Genet C Semin Med Genet 2011;157:274-87.
49. Spencer R. Anatomic description of conjoined twins: A plea for standardized terminology. J Pediatr Surg 1996;31:941-4.
50. Sabih D, Ahmad E, Sabih A, Sabih Q. Ultrasound diagnosis of cephalopagus conjoined twin pregnancy at 29 weeks. Biomed Imaging Interv J 2010;6:e38.
51. Andrews RE, McMahon CJ, Yates RW. Echocardiographic assessment of conjoined twins. Heart 2006;92:382-7.