Characteristics and management of conjoined twins: A single-centre retrospective descriptive study

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ABSTRACT

Introduction: Conjoined twins (CT) is a rare congenital disorder characterised by the presence of malformations associated with secondary abnormal conjoined organ changes and abnormal hemodynamic superimposed effects about 1 in every 200 identical twin pregnancies, between 1 in 50,000 to 1 in 100,000 live births. The aim of this study is to describe the characteristics of conjoined twins.

Materials and methods: This was a retrospective descriptive study. All medical records of conjoined twins who were admitted to Hasan Sadikin Bandung General Hospital from January 1st, 2015, to June 30th, 2023, were reviewed for gender, conjoined type, birth order, risk factor and treatment.

Results: Of the 28 conjoined twins, 21 were girls (75%), and 7 were boys (25%); 19 (67,85%) were of the thoracoomphalopagus type; 11 (39,28%) were born as first children; 18 (64,28%) were born at 37 weeks of gestational age; and 22 twins' (78,57%) parents were aged between 21 and 35 years. None of the mothers had used medication, 13 (46,42%) took folic acid on occasion, five (17,85%) used traditional herbs, nine (32,14%) smoked and none drank alcohol. Parents who live in industrial areas were 18 (64.28%). There was no history of conjoined twins in previous pregnancies or deliveries or in the parent's family. Liver separation had been done in four (14.28%). Emergency separation in one twin. Nine (21.42%) patients died before surgery due to a worsening condition.

Conclusion: The conjoined twins were more common in girls, predominantly of the thoracoomphalopagus type. Risk factors that were commonly found were the first child, a gestational age of less than 37 weeks, and living in an industrial area.

KEYWORDS:
Conjoined twins, gestational age, medication, industrial area

INTRODUCTION

Conjoined twins (CT) are one of the rarest types of monochorionic monoamniotic twins. The incidence ranges from one out of every 200 identical twin pregnancies to almost always being identical, between one in 50,000 to one in 100,000 live births.[1,2] Some studies report prevalence as high as one in 2,800 live births in India,[3] and as low as one in 200,000 live births in the USA.[4] However, prevalence increases of 3.27: 100,000 live births[5] and 2.85: 100,000 live births[6] were reported in two studies in the Chinese population of surveillance programs at different times. A higher incidence is found in Africa, about 1: 14,000, and in Asia, 1: 25,000.[7]

CT are malformations associated with secondary changes in abnormal conjoined organs and abnormal hemodynamically superimposed effects.[8] The mechanism of the defect is due to changes in normal development processes, in which a pair of monozygotic twins (MZ) do not separate completely from one another and continue normal embryological development. MZ twins start with the separation and division of a single early embryo.[9-11] The mechanism of CT is still unclear. There are conflicting theories explaining the sequence of events in CT. CT are derived from a single fertilised egg. There are two theories to explain this phenomenon: 1) Fusion theory (more accepted): when a single fertilised egg is divided into two embryos. The phenomenon occurs between 13 and 15 days after fertilisation, resulting in failure to complete division. 2) The fission theory: when there is union of two embryos originally separated about 12 days after fertilisation. The theory supporting the 'fusion' process cannot explain the parapagus type, but it can explain the other type by the fusion of two separate embryos.[12-16] The formation of the parapagus can be explained by a single notochord bifurcation.[17] No theory of embryonic vertebral fission at any stage of development, from various planes and from various directions, can explain the selection of fusion sites, the details of fusion, or the delimitation of the specific area where twins are found joined.[13] On the other hand, proponents of fission theory claim that CT is the result of imperfect separation of the embryonic axis.[8,9,18-22] Except for parasitic twins, all CTs are symmetrical, and 'equal parts always join equal parts.'[9] The same authors state, 'If fusion, rather than fission, occurs in all cases of conjoined twins, the incidence of similar appearance is similar in all monoamniotic twins, whether conjoined or not,' and 'if the incidence of similar appearance is higher in CT than in separated twins, the fusion hypothesis is incorrect.' There are no records in the literature regarding the familial aggregation of CT or its preferential association with other unrelated anomalies.[23] Recently, it was found that low-dose radiation exposure triggers the incidence of twinning and the prevalence of CT.[24]

About 75% of conjoined twin pairs are female, with a female-to-male ratio of 3:1. Of the total, about 40% are stillborn, and 60% are live births; only about 25% live long enough to be

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candidates for surgery.[7] CT are classified based on the most common location of connection: thorax (thoracopagus), abdomen (omphalopagus), sacrum (pygopagus), pelvis (ischiopagus), skull (cephalopagus) and back (rachipagus). Based on the aspect of the embryonic disc, the most common type is the thoracopagus (19%).[25] Omphalopagus is the least common, with an incidence of 0.5%.[26] The overall survival rate of CT is around 25%.[27]

In recent studies, the diagnosis can be made in the first trimester, and because the family has already opted for the termination of pregnancy, further diagnostic intervention is not necessary. However, in developing countries, the lack of adequate maternal care facilities causes late diagnosis. Because this situation poses a great risk, early diagnosis and treatment during delivery are very important. Surgical separation of CT can vary from minimal risk to very complex, depending on the point of attachment and the internal parts that are shared.[1,2] This study wants to describe the characteristics of conjoined twins.

MATERIALS AND METHODS

This was a retrospective descriptive study at Hasan Sadikin Bandung General Hospital. From January 1st, 2015, to June 30th, 2023, 28 patients' data were collected on gender, conjoined type, birth order, gestational age, parent age, history of medication, traditional herbal, cigarette and alcohol consumption, residence and treatment.

RESULTS

There were few case series as example to compare characteristic in conjoined twins found at Hasan Sadikin Bandung General Hospital.

The patient was consulted from the paediatric department with the following diagnosis: conjoined twin parapagus + dd/Ebstein anomaly + pulmonary stenosis + single ventricle + hypoplasia distal phalanx digiti III-V manus dextra + CTEV (congenital talipes equino varus) + suspect CCAM (congenital cystic adenomatoid malformations). Attached body parts were the abdomen with two limbs, one umbilicus, one external genitalia, and one anus. Babies are born at a gestational age of 38 to 39 weeks.

There were one penis and one scrotum, one anus, two inferior extremities and the right lower extremity looked pale. On radiography, two vertebrae and one pelvis were found; on ultrasound, two livers and two separate bladders were found; on echocardiography, baby A with PDA and baby B with DORV (double outlet right ventricle), inlet VSD, CA-VSD, mild coarctation of the aorta, valvular PS, PDA: left to shunt right, and PLSVC (persistent left superior vena cava). Intraoperatively, each baby was found with 1 kidney facing into the pelvic area, with each ureter connected to one bladder, and two testicles in the pelvic area with normal shape, half the normal size, looked vital. The sacrococcygeal fusion was connected by fibrous tissue. Liver fusion (2.5 cm long, 1.5 cm thick) was separated with a harmonic scalper; ileal fusion 20 cm proximal to the Bauhini valve, sutured with 3.0 silk, with a single terminal ileum, caecum, ascending colon, and transverse colon. Duplicated colon in infant A, from splenic flexure to rectum, with fenestrated shared walls Meckel's diverticulum was found in baby A, 12 cm proximal to the Bauhini valve. Ileal fusion separated in infant B, proximal to ileal fusion. Left congenital diaphragmatic hernia was found in baby B (the organs that entered were the stomach, small intestine, left lobe of the liver and spleen)—a defect of about half the diaphragm. The intestines were separated. With an anterior incision approach, the pubic symphysis was separated, and the sacrum was separated into the right superior and inferior pubic rami. Postoperative findings were infant A with a complete gastrointestinal tract, from stomach to rectoanal, with duplication of descending colon to rectum, liver and spleen. Baby A was found with one left kidney, one left ureter connected to the bladder and urethra, one left testicle, one pelvis and one left extremity. The abdomen was closed in the same way as the first, and the part that had not been closed was given a prosthetic mesh to close the defect. Cutis and subcutis dissected the stretched part. The patient died 2 days postoperatively due to septic shock, hypoalbuminemia and suspected acute left lower extremity thrombosis.

One case of female thoracoomphalopagus was performed for emergency separation on the 4th day of life because baby 1 was declared dead. The patient was born as a preterm infant (31 to 32 weeks) and was not crying immediately. A liver fusion was found, each of which has a biliary vascular tree. In the intestines, no fusion was found, each having its own gastrointestinal system and its own urogenital system. A cross-vessel liver was found in the upper 1/3 of the fusion, with a diameter of 0.5 cm. Fusion was found in the proximal arch of the xiphoid process of the ribs. There was pericardial fusion at the apex ± 0.5 cm and intravesical pressure of 12.6 mmHg.

There were 3-year-old male conjoined twin ischiopagus with three inferior extremities, one bladder, and a shared intestine with one rectum, suggesting left kidney hypoplasia in conjoined twin 1 and left kidney agenesis in conjoined twin 2. The patient was born at term (40 weeks) and was crying immediately. In conjoined twin 1, forming the hepatogastric trunk (coeliac trunk variant type III, based on Uflacker's classification). Normal structures of the splenic parenchyma and splenic arteries are not visualised. The left renal artery appears smaller than the right renal artery. Patients underwent disarticulation of the supernumerary limbs, pelvic wedge osteotomy, ORIF P/S, tension band wiring and defect closure with a local advancement flap. Incisions in the proximal supernumerary limb were performed layer by layer; fat exposure, fascia and limb disarticulation were done. Wedge osteotomy at the pelvic bone, then pelvic reduction and approximation with SS wire. Drilling and setting recon plate surrel locking 6 mm with four cancellous screws, two screws installed, and SS wire tension band wiring There was a peritoneal defect measuring 6 cm in diameter with a small intestine exposed. Peritoneal defect closure was performed with chromic 4-0. At the regiolateral abdomen, there was an open wound post-disarticulation by an orthopaedic surgeon, 12 × 5 × 8 cm in size, with a muscle and fascia base.

Table I: Characteristics of patients with conjoined twins

Demography characteristic	Frequency (n = 28)
Gender	
• Girl	21 (75%)
• Boy	7 (25%)
Conjoined twins type	
• Thoracoomphalopagus	19 (67,85%)
• Abdominopygopagus	2 (7,14%)
• Ischiopagus	2 (7,14%)
• Cephalopagus	4 (14,28%)
• Parapagus	1 (3,57%)
• Rachipagus	0 (0%)
Birth order	
• First	11 (39,28%)
• Second	8 (28,57%)
• Third	9 (32,14%)
Gestational age	
• < 37 weeks	18 (64,28%)
• ≥ 37 weeks	10 (35,71%)
Parent age	
• < 21 years	6 (21,42%)
• 21-35 years	21 (78,57%)
• >35 years	0 (0%)
History of medication consumption	
• Analgesic and stomach ulcers medicine	4 (14,28%)
• None	24 (85,71%)
History of folic acid consumption	
• Always	8 (28,57%)
• Sometimes	13 (46,42%)
• Never	7 (25%)
History of traditional herbal consumption	
• Ever	5 (17,85%)
• Never	23 (82,14%)
Habitual smoking	
• Mother	0 (0%)
• Father	9 (32,14%)
Alcohol consumption	
• Positive	0 (0%)
• Negative	28 (100%)
Living in the industrial area	
• Yes	18 (64,28%)
• No	10 (35,71%)
History of childbirth with previous conjoined twins	
• Positive	0 (0%)
• Negative	28 (100%)
History of conjoined twins occurrence in family	
• Mother	0 (0%)
• Father	0 (0%)
Treatment	
• Rudimentary limb ablation	2 (7,14%)
• Bladder reconstruction	1 (3,57%)
• Liver separation	4 (14,28%)
• Abdominal wall reconstruction	4 (14,28%)
• Sternal and pericardial separation	3 (10,71%)
• Chest wall reconstruction	3 (10,71%)
• Disarticulation of the supernumerary limbs	1 (3,57%)
• None	20 (71,42%)
Inoperable	6 (21,42%)
Plan for elective surgery	5 (17,85%)
Death	9 (31,14%)

Data are presented as n (%)

Fig. 1: (Left) Two-months-old twin with thoracoomphalopagus, fusion of the chest and upper part of the abdomen with suspect congenital heart disease (right) Two-days-old cephalopagus (temporal bone fusion)

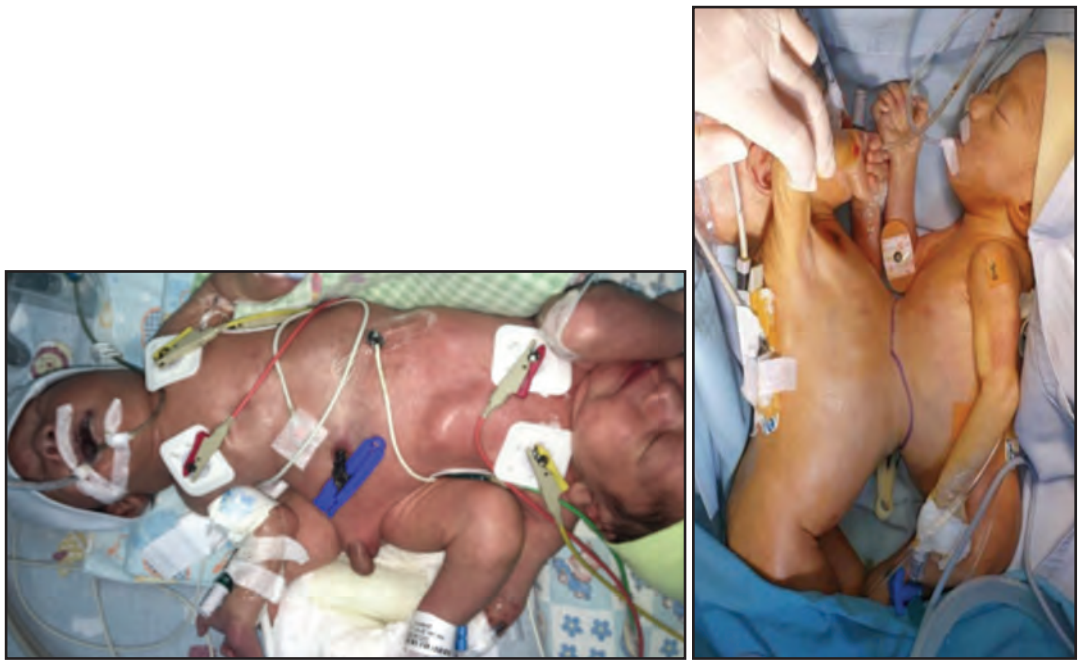

Fig. 2: (Left) One-day-old conjoined twin parapagus that the abdomen was attached with two limbs, one umbilicus, one external genitalia and one anus (right) Four-days-old thoracoomphalopagus with liver, proximal arch of the xiphoid process of the ribs, and pericardial fusion. Baby 1 died

There were six inoperable CT because of shared internal organ such as intestine and liver, shared external organ such as leg, penile, anus or inseparable ventricle and atrium of heart. Meanwhile nine twins were death before surgery due to worsening condition of asphyxia, pneumonia, sepsis, congenital heart disease such as tetralogy of Fallot, tricuspid atresia, ventricular septal defect etc.

DISCUSSION

The occurrence of CT is influenced by many factors that are not clearly known. The risk factors observed in this study were gender, birth order, gestational age, parent age, consumption of medication, folic acid, cigarettes and alcohol, living in an industrial area, history of CT in previous pregnancies or deliveries, and history of CT occurrence in family. Of 28 conjoined twins, 21 girls (75%) and seven boys (25%), according to one study, showed a 1.5–2.5 female sex predominance over male sex.[2]

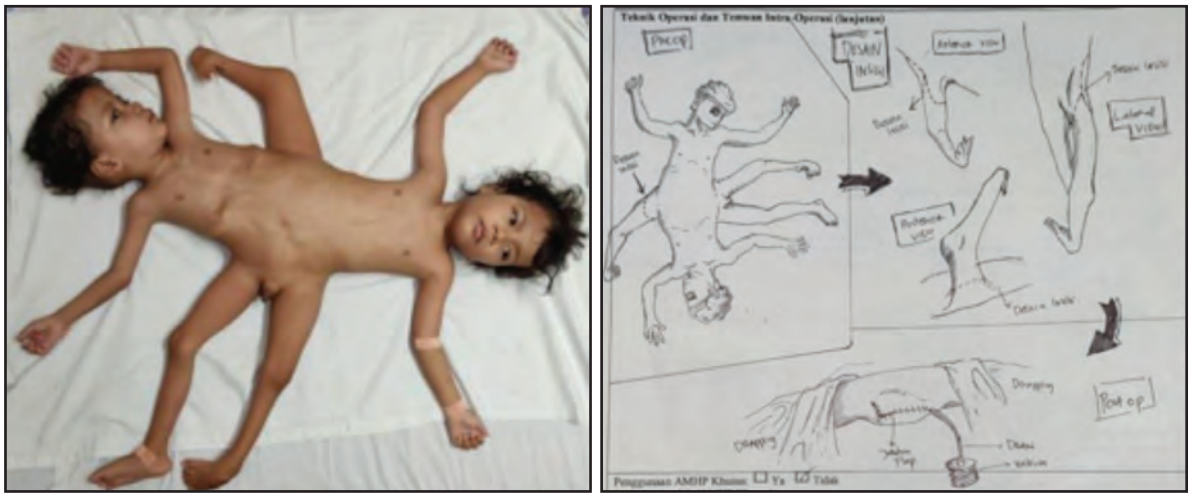

Fig. 3: (Left) 3-year- old ischiopagus with three inferior extremities, one bladder, shared intestine, and one rectum (right) incision design of surgery

A total of 19 twins (67,85%) were thoracoomphalopagus types, but based on the aspect of the embryonic disc, the most common type is thoracopagus (19%).[25] Omphalopagus is the least common, with an incidence of 0.5%.[26] 11 (39,28%) were born as first children; 18 (64.28%) were born at 37 weeks of gestational age; and 22 twins' (78,57%) parents aged between 21 and 35 years, the same as demographic characteristics such as mother's age, reproductive data, including gestational age, birth weight, and environmental factors such as exposure to and disease in the mother during pregnancy.[28]

In our series, risk factors such as consumption of medication, folic acid, traditional herbal and cigarettes were found in a small number (medication 0%, folic acid 13 (46,42%), traditional herbal 5 (17,85%) and cigarette 9 (32,14%)). No alcohol consumption was found. 18 (64.28%) CT were living in industrial areas but still required further study considering the limited number of samples for statistical tests. There was no history of CT in previous pregnancies or deliveries or in the parent's family. Generally, multiple factors, such as alcohol consumption, medication use, radiation or chemical exposure, and lack of folic acid supplementation during pregnancy, as well as family history, are reported risk factors for the development of congenital anomalies. Specific to conjoined twins, the exact aetiology has not been discovered yet.[29]

CONCLUSION

The incidence of CT was more common in girls, predominantly the thoracoomphalopagus type, with gestational age in first-born children at less than 37 weeks' gestation who were living in the industrial area. There was no history of consumption of medication, especially teratogenic, in this study. In the future, hopefully further research can be carried out regarding the incidence of CT being influenced by other risk factors and the management of conjoined twins.

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