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## Thoracopagus parasitic twin with omphalocele rupture: a case report



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### ABSTRACT

**Background:** Heteropagus or parasitic twin is a rare congenital anomaly that affects two connected children with imprecisely understood anatomy and physiology. This case report aims to describe a thoracopagus patient with severe complications.

**Case presentation:** One-day-old female baby was born with thoracopagus and omphalocele rupture. She was the third child, born aterm from a 36-year-old mother, with a birth weight of 3000 grams. Laboratory and radiography examinations showed leukocytosis,

metabolic acidosis, prolonged coagulation profile, acute kidney injury, hypoglycemia, transaminitis, and congenital heart defects. She was treated in the intensive care unit and managed conservatively. Unfortunately, she died on the sixth day of treatment due to severe complications.

**Conclusion:** Conjoined twins have a poor prognosis. If it is coupled with other congenital anomalies, it will worsen the outcome.

**Keywords:** parasitic twin, thoracopagus, omphalocele.

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### INTRODUCTION

Heteropagus, or parasitic twins, is an uncommon congenital condition in asymmetric conjoined twins in which the tissues of one severely damaged twin (the parasite) depend on the circulatory system of the other, a completely intact twin (the autosite), for survival.<sup>1</sup> The incidence of heteropagus twinning is thought to be less than 0.1 per 100,000 live births, making it an extremely rare condition.<sup>2,3</sup> The illness was possibly first accurately described by a 16th-century French surgeon named Ambroise Pare, who wrote that one of his patients had an attached acephalus body to the abdomen. The actual rate of heteropagus twin births was between 0.05 and 0.1 per 100,000, according to a US database of 7.9 million births gathered over eight years in the 1970s. A more recent European investigation encompassing nearly 5 million babies found an incidence of 0.02

per 100,000.<sup>2</sup> There were 45 parasitic twin cases reported in an Indonesian hospital's database between 1987 and 2022, and 18 of those patients underwent surgery to separate the twins.<sup>4</sup>

Usually, the parasite attaches to the autosite through a soft tissue pedicle with a big blood artery and fetal-looking components with a gross fetal-like appearance. Thoracopagus, omphalopagus, craniopagus, cephalopagus, parapagus, ischiopagus, pyopagus, or rachipagus are the eight attachment sites that could be used. There are two main explanations for heteropagus twinning, while the exact etiology is uncertain. The "fission" theory suggests a partial separation of the embryo, in contrast to the "fusion" theory, which suggests joining two initially separate components. According to the most recent notion, one twin dies and experiences partial resorption due to vascular damage while still in the uterus.<sup>3</sup>

Antepartum evaluation, labor, delivery,

and a preoperative and surgical strategy are all parts of managing parasitic twins.<sup>2</sup> Twin parasites have a 7.5% survival rate, which is not good. Only 60% of the surgically separated patients made it out alive. Prenatal imaging, perinatal follow-up, postnatal surgery, tissue expansion during surgery, and cadaveric transplant for the twins' shared essential organs would all improve the mother's and child's survival at birth.<sup>5</sup>

In this study, we aimed to report our experience managing a case of a thoracopagus parasitic twin newborn with severe complications.

### CASE PRESENTATION

A day-old female infant with pale skin and a sluggish state was referred from a local hospital. The infant was spontaneously born, and he or she did not cry right away. There were anomalies in the body components, including extra superior and

inferior limbs linked to the right side of the chest and extra, correctly functioning genitalia and buttocks. The area of the abdomen was open, and the covering membrane had been torn, exposing the hepatic, gastric, and ileal organs (**Figure 1**). The infant, the mother's third child, weighed 3000 grams at birth and was delivered preterm by a 36-year-old woman. Throughout the prenatal stage, the mother consistently managed her pregnancy with an obstetrician and midwife. Neither prenatal fetal abnormalities nor a history of repeated or unusual pregnancies in the preceding trimester existed. Mother had no prior history of serious illnesses.

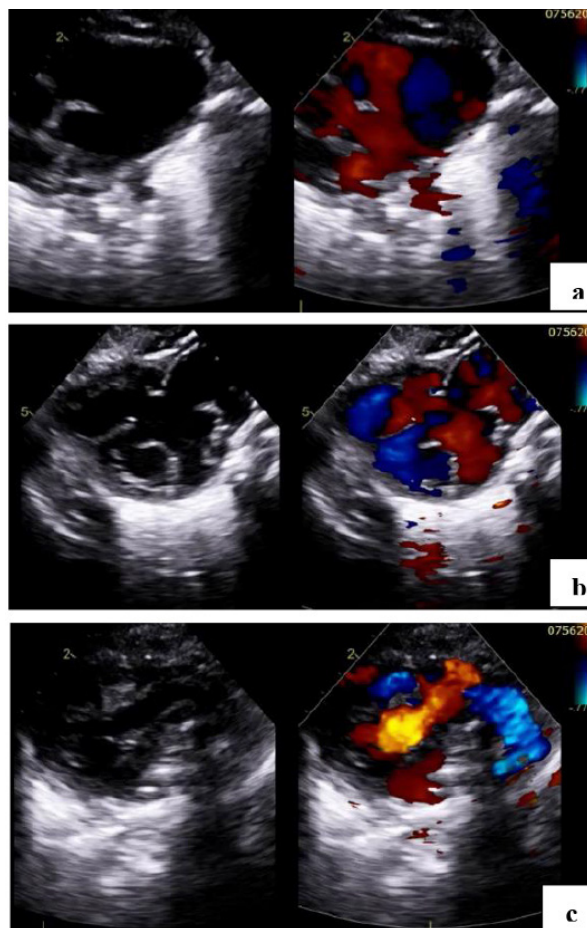
Laboratory examination showed WBC  $13.72 \times 10^3/\mu\text{L}$ , HB 16.66 g/dL, PLT  $88.79 \times 10^3/\mu\text{L}$ , metabolic acidosis with pH of 7.17,  $\text{pCO}_2$  of 35.1 mmHg, and bicarbonate levels of 12.4 mmol/L, prolonged coagulation profile (APTT 42.7 second, PPT 24.9 second, INR 1.81). Serum creatinine level was 2.08 mg/dL, BUN 32.60 mg/dL, which indicates neonatal acute kidney injury. Random blood sugar 55 mg/dL, sodium 142 mmol/L, potassium 5.68 mmol/L. The liver function test also increased (AST 150.5 U/L, ALT 61.6 U/L). We diagnosed the patient with term neonates, appropriate to gestational age, moderate asphyxia, parasitic twin (thoracopagus), omphalocele rupture (multiple congenital anomalies), and sepsis neonatal early onset. We gave empirical antibiotic ampicillin and amikacin. A blood culture was performed and found *Klebsiella pneumonia* on two-sided.

Radiological X-ray examination (**Figure 2**) found that the heart and lungs are challenging to evaluate because overlapping with the other organs. Echocardiography results show that the heart's position is horizontal, with the heart's apex on the right side. Congenital heart defects were a complete atrioventricular septal defect, single atrium, and large patent ductus arteriosus (**Figure 3**).

We consulted the patient with the pediatric surgeon's department. The ruptured omphalocele is an emergency condition and was managed with a silastic silo (schuster technique; **Figure 4**) that was modified with a urinary bag. After the procedure patient was intubated



**Figure 1.** Parasitic twin patient



**Figure 2.** Radiology examination

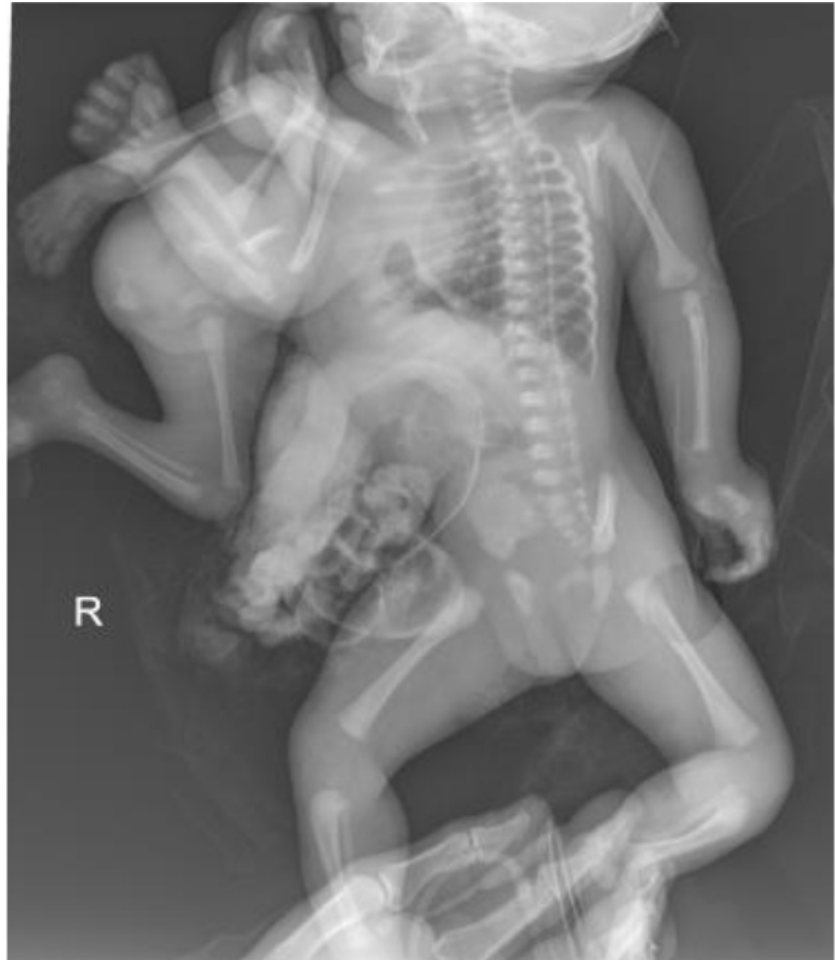
and treated in Neonatal Intensive Care Unit (NICU) with ventilator support. Intravenous antibiotics were continued. We gave total parenteral nutrition, fresh frozen plasma (FFP), cryoprecipitate, and packed red cells (PRC) were transfused simultaneously. The patient also received an injection of phytomenadione intramuscularly for three consecutive days. Unfortunately, the patient died on the sixth day of treatment due to sepsis and suggestive DIC.

**DISCUSSION**

Parasitic twin is a rare condition with an incidence of 1-2:1,000,000 birth in the USA and Europe. An estimate of the frequency of heteropagus pairs as a percentage of all conjoined twins is 4.5% to 15%.<sup>2</sup> Africa and Asia have greater parasitic twin incidence rates, which range from 1:14,000 to 1:25,000.<sup>4</sup> Epidemiologic information on heteropagus twins is scarce due to their rarity, the diverse terminology, and underreporting of anomalies.<sup>2</sup> Data from Cipto Mangunkusumo Hospital, the top referral hospital in Indonesia, also needs more data about this condition. The facility had split 18 conjoined twins from 45 cases that were registered between 1987 and 2022.<sup>4</sup>

The two leading theories for the embryologic origin of conjoined twins are fusion and fission. The former theory's proponents assert that between 13 and 15 days after conception, during the primitive streak stage, the blastocyst inner cell mass undergoes incomplete fission, resulting in two foci of axial growth that eventually maintain a link. On the other hand, "fusion" describes the secondary coalescence of two initially distinct inner cell masses.<sup>2,3</sup> One of the most widely proposed advanced ideas for the emergence of imbalance between autosite and parasite following these occurrences; the tissue of the parasitic twin becomes dependent on collaterals produced from the autosite due to vascular impairment. The parasite's bodily part that is being starved then experiences selective ischemic atrophy. Since then, this hypothesis has undergone several variations.<sup>2</sup>

Our patient is a thoracopagus-type parasitic twin. The place of fusion determines how this condition is described.



**Figure 3.** Echocardiography: (a) Single atrium; (b) Complete Atrioventricular Septal Defect; (c) Large patent ductus arteriosus



**Figure 4.** Schuster technique

Thoracopagus has an attached point in the chest. Some attachment sites are possible (thoracopagus, omphalopagus, craniopagus, cephalopagus, parapagus, ischiopagus, pyopagus, or rachipagus). The most frequent cases are thoracic-omphalopagus (joined at thorax and abdomen), 28%, and thoracopagus (joined at the thorax) in 18.5% of total cases of parasitic twins.<sup>6</sup> The most frequent type in Cipto Mangunkusumo Hospital Indonesia was thoracic-omphalopagus 26 cases (58%) from the registry 1987-2022.<sup>4</sup>

In our case, the mother is 36 years old, was in her third pregnancy, and was born aterm, weighing 3000 grams. Mother routinely controlled her pregnancy with a midwife and obstetrician. She has no history of multiple or unusual pregnancies or fetal abnormalities throughout the prenatal period. Mother had no prior history of serious illnesses. According to data from numerous studies, the median maternal age for mothers of heteropagus twins is 26 years old, with a range of 17 to 36 years, and the median parity is 0, with a range of 0 to 5. Studies found no difference in maternal age between symmetric and asymmetric conjoined twins. The congenital relationship between the mother and father of heteropagus twins has not been documented. In addition, asymmetric conjoined twinning has never been associated with maternal use of illegal drugs, alcohol, or tobacco, except for one mother who smoked a half-pack of cigarettes every day while pregnant.<sup>1</sup> But another study found that risk factors for developing this rare condition include a positive history of twin births, ovulation-inducing medications, infertility treatments, and radiation exposure.<sup>7</sup> But none of these risk factors were mentioned in our patient.

The antepartum diagnosis influences the management of parasitic twins. Results are often favorable. Hence, heteropagus twins should be distinguished from more congenital severe abnormalities to prevent needless termination. Furthermore, fetal positioning and autosite abnormalities can be used to direct future obstetric and surgical care. Depending on the morphology of the conjoined twins being studied, prenatal echocardiography may occasionally be more technically possible

than ex-utero examination. Typically, this disease is detected in utero between 9 and 28 weeks. Imaging diagnosis by ultrasonography can detect abnormalities intra-uterine. Examining conjoined twins by prenatal ultrasound in the first trimester is essential due to the increased risk of perinatal morbidity and mortality. In the second trimester, ultrasound evaluation and magnetic resonance imaging can improve and clarify the diagnosis by thoroughly describing the anatomy of the two fetuses and the shared portions.<sup>2,8,9</sup> An early diagnosis is not always attainable. For instance, if the mother is obese or has insufficient amniotic fluid, an ultrasound scan may not be sufficient, and support from pelvic magnetic resonance imaging may be needed. A prenatal diagnosis is crucial when determining whether to abort a pregnancy or continue it with surgical twin separation. Because early pregnancy termination indicates a smaller emotional toll on the couple, which could be made worse by the frequent interdisciplinary follow-ups required during prenatal and postnatal care, it may be deemed a safer decision.<sup>9</sup>

Only 34 studies published from 1984 to the present detail the mode of delivery of heteropagus twins. In contrast to 10 (29%) cesarean deliveries, 18 pairs of twins (53%) were born vaginally. The parents chose elective termination in 2 cases (6%), while spontaneous abortion happened in 4 cases (12%). The heteropagus twin's anatomical arrangement and location may be a more vital indicator of the delivery method than their overall weight. Complicated labor and mechanical dystocia were the two most common causes of cesarean delivery in heteropagus twins. Omphalopagus, rachipagus, ischiopagus, and pygopagus were among the parasitic twin configurations that required cesarean delivery.<sup>2</sup> In our case, the patient was delivered spontaneously because the mother did not know about abnormalities in her pregnancy. She could not choose whether to terminate or continue the pregnancy and the delivery method according to the abnormalities.

Imaging preoperative separation surgery includes computed tomography (CT), ultrasound, and magnetic resonance imaging (MRI). Echocardiography is

frequently added. Thoracopagus, by definition, suggests a heart problem, which an echocardiography might clarify. As previously stated, a cardiac abnormality is linked to 25% to 39% of omphalopagus autosites.<sup>2</sup>

Although there have been instances of heteropagus autosite twins carrying all or some parasites into preadolescence and late adolescence, surgical separation is typically done perinatally. In addition to physical and psychosocial considerations, separation is necessary because parasites frequently result in respiratory distress and may limit the growth of autosites. Not much information is available on surgical methods for separating heteropagus twins. These details might not be beneficial even when offered because autosites and parasites have different vascular, soft tissue, and bone connections.<sup>2</sup> Although it is technically possible to divide many cases of thoracopagus twins, the results of such efforts historically have been appalling. Since 90% of cases have shared pericardium, and 75% have conjoined hearts.<sup>10</sup> Our patient did not survive until the preoperative stage due to severe complications.

In our case, the patient was complicated with a ruptured omphalocele. Omphalocele is a defect in the abdominal wall. Ruptures or tears result in the exposure of the contained viscera (the intestine, stomach, liver, bladder, spleen, ovaries, and testes). To avoid the accompanying morbidity and mortality, ruptured omphaloceles must be diagnosed and treated immediately. They are also problematic surgical conditions. In order to treat a ruptured omphalocele, first resuscitation and surgical therapy are used. Preserving hydration and normothermia should be the main goals of resuscitation. These newborns lose much fluid through evaporation and the third space. Hypovolemia can cause hypotension. Hence immediate intravenous access is required. Broad-spectrum antibiotics should be started. One option for procedural intervention is a silastic silo known as the Schuster technique. The silo is fastened to the fascia or skin with a non-absorbable suture. The infant's abdomen is then covered by the silo, enabling gravity to help with reduction.<sup>11</sup>

A ruptured omphalocele in a baby may have consequences. The most frequent consequence is sepsis. The child's concomitant abnormalities ultimately influence the prognosis for omphalocele. Patients with huge omphaloceles and severe malformations require prolonged intubations, respiratory assistance, and prolonged hospital stays compared to those with isolated defects or mild anomalies.<sup>11</sup> Conjoined twins typically have a dismal prognosis. There is a 7.5% survival rate. Only 60% of cases with surgical separation make it.<sup>12</sup> Our case has a bad outcome due to multiple anomalies and complications.

The limitation of our case is that the parasitic twin condition did not diagnose prenatally, so the mother did not go to the tertiary hospital for further treatment and birth planning. It is made a delay in treatment and complication of severe systemic infection increased morbidity and mortality in our case.

## CONCLUSION

The one-day-old female baby, the third child, born at term from a 36-year-old mother, was diagnosed with thoracopagus and rupture of omphalocele. The condition was worsened with sepsis neonatal early onset. The supporting examination found an overlapping of the heart and lungs and multiple congenital heart defects. She was treated intensively and managed with a silastic silo. Unfortunately, she died due to severe complications.

## CONFLICT OF INTEREST

There is no competing interest regarding the manuscript.

## FUNDING

None.

## AUTHOR CONTRIBUTIONS

All authors equally contribute to the study by selecting cases, evaluating the outcome, and reporting the study results through publication.

## PATIENT'S INFORMED CONSENT

The patient's parents has signed a written informed consent and agreed to this study's publication.

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