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Case Report

A Rare Case of Conjoined Pygopagus Twins: Case Report

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ABSTRACT

Introduction and importance: *Conjoined twins are a complication of monozygotic pregnancies with an incidence of 1:50,000-200,000 life births. Pygopagus twins are joined back-to-back, facing each other, commonly sharing the gluteal region, terminal spine, and lower gastrointestinal, urological, and reproductive tracts. Pygopagus is found in around 17% of all types of conjoined twins and has a mortality rate of 23%. Proper diagnosis and management, in this case, are important.*
Presentation of case: *A 25-year-old primigravida woman presented at 22 weeks of gestation with a Pygopagus ultrasound result at 17 weeks of gestation. MRI was performed at 25 weeks of gestation, showed conjoined pygopagus twins with lumbosacral spina bifida, without a visualized bone union, separated bladder, rectum, and anus were difficult to evaluate. Caesarean section (CS) is planned at 36 weeks of gestation, and a postpartum MRI will be performed in preparation for the separation surgery. The patient was admitted to the hospital at 35 weeks of gestation in the active phase of the first stage of labor, and then an urgent caesarean section was performed. Both babies were born with an APGAR score of 2/4/7. The second baby died a few hours after delivery. The separation surgery was performed as an emergency, but the first baby died during the procedure.*
Conclusions: *Pygopagus requires holistic management, starting from diagnosis, preparation for delivery, to*

postpartum complications. Proper holistic management is expected to reduce infant and maternal morbidity and mortality.

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1. INTRODUCTION

Conjoined twins are a rare complication of monochorionic-monoamniotic (monozygotic) pregnancies.[1] Incidence 1: 50,000-200,000 life births, with a male to female ratio of 1:3.[2] Conjoined twins have high morbidity and mortality rates, 28% will die prior to delivery, 54% will die soon after birth, and 18% will survive.[3]

Conjoined twins are classified into 8 types according to the part of the body that undergoes fusion, such as Omphalopagus (umbilicus), Thoracopagus (chest), Cephalopagus (head), Ischiopagus (hips), Craniopagus (cranium), Rachipagus (spine), Pygopagus (rump), Parapagus (side).[4] The Pygopagus incidence is around 17% of all types of conjoined twins and has a mortality rate of 23%. Conjoined twins of the Pygopagus type undergo dorsal fusion, either in the gluteal region, terminal spine, lower gastrointestinal, urological, or reproductive tracts.[2,5]

2. CASE PRESENTATION

A 25-year-old primigravida woman was referred to Dr. Sardjito Hospital at 22 weeks of gestation because her ultrasound result at 17 weeks of gestation was conjoined twins with fused dorsal parts (Pygopagus), the sacrum looked separated with the impression that could be separated (Fig. 1). The first day of the patient's last menstrual period was November 4th, 2020. The patient had no family history of multiple pregnancies or genetic diseases. The ultrasound examination at Dr. Sardjito Hospital showed the same results as the previous ultrasound (Fig. 2a and 2b).

MRI examination performed at 25 weeks of gestation showing conjoined Pygopagus twins with lumbosacral spina bifida (fusion from subcutis to thecal sac at lumbosacral level with craniocaudal fusion diameter of about 3.68cm), without visualized bone union, separated bladder, rectum and anus were difficult to evaluate, the placenta was located in the left uterine fundus, two separated fetal heads were on the right side, and each fetus had complete of feet (Fig. 3). From the results of the above examination, the diagnosis of Conjoined Pygopagus twins was established. The patient's family was given an explanation regarding the examination results, diagnosis, prognosis, and complications that can occur. After listening to the explanation, the parents are given informed consent to continue the pregnancy by accepting all possible consequences.

At 26 weeks of gestation, a multidisciplinary team consisting of obstetric and gynecologist doctors, a pediatric surgeon, a neonatologist, and an anesthesiologist planned the delivery of the patient. From the discussion results, an elective caesarean section was planned at 36 weeks of gestation, with an anesthetic plan similar to caesarean section (CS) generally. The patient carries out routine antenatal care until 36 weeks of gestation. Another result of the discussion from the multidisciplinary team was the evaluation of the baby's condition after birth, and a postpartum MRI would be performed in preparation for the separation surgery.

This patient had several ultrasounds performed at 22, 25, 27, 30, 32, 33, 34, and 35 weeks of gestation with normal fetal development results, breech-breech presentation

since 32 weeks of gestation. At 35 weeks of gestation, the patient came to the Emergency Room (ER) with complaints of uterine contractions, bloody mucous, leaking amniotic fluid, and active fetal movements. The patient was declared to be in the active phase of the first stage of labor, with breech-breech presentation then an urgent CS was immediately performed with the outcome of Conjoined Pygopagus Twins, Neonatal Asphyxia, Low birth weight babies (Fig. 4). The patient gave birth to twin girls (conjoined in the gluteal, separated anus) with a total birth weight of 3900 grams. The appearance of the first baby is larger than the second one and the second baby looks paler, with an APGAR Score of both babies 2/4/7. Both babies had intercostal retraction and 94% oxygen saturation with the respiratory rate in the first baby was 68 x/minute and the second baby 70x/minute, resuscitation with intubation was done. Serum laboratory results revealed haemoglobin level of 19.4 g/dL in the first baby and 17.3 g/dL in the second baby. Blood gas analysis revealed respiratory alkalosis on the first baby and respiratory acidosis on the second baby. Double antibiotics were given to both babies. Echocardiography revealed normal on the first baby and the second baby had obstruction of the left pulmonary artery and ASD right to left shunt. Postpartum MRI could not be performed because the babies' condition was unstable. 22 hours after birth, the second baby was deteriorating, and she died with the diagnosis of severe asphyxia. After the second baby died, a multidisciplinary team consisting of a neurosurgeon, pediatric surgeon, orthopedic doctor, neonatologist, and anesthesiologist decided to perform the emergency separation surgery. During the surgical procedure, the first baby's condition continued to deteriorate, and she died due to bleeding when her sacral venous plexus was dissected on the second hour of the surgical procedure.

Fig. 1. Ultrasound at 17 weeks age of gestation showed fused dorsal parts, the sacrum looked separated

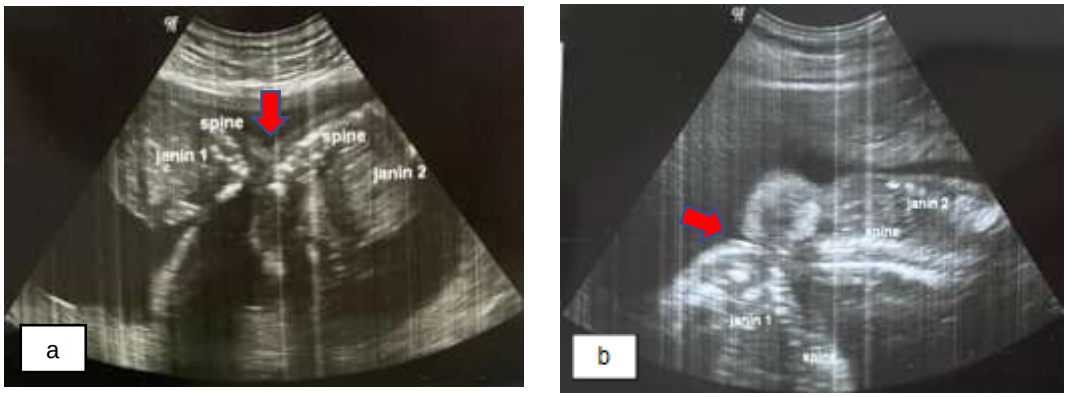

Fig. 2. Ultrasound at 22 weeks age of gestation showing fused dorsal parts (Pygopagus), there was a separatable sacrum. **a)** Baby I: transverse lie, head in left side of abdomen, fetal back is facing up. **b)** Baby II: transverse lie, head in left side of abdomen, fetal back is facing down.

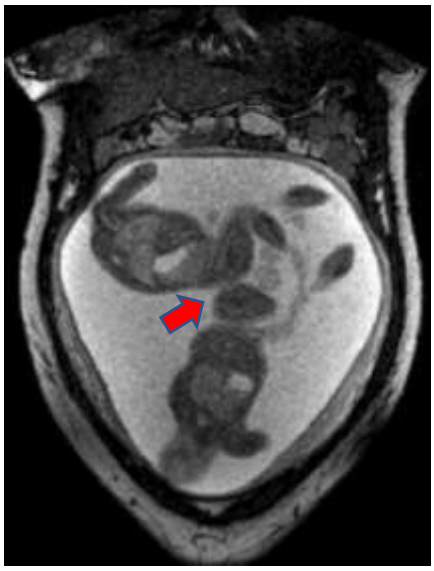

Fig. 3. MRI at 25 weeks age of gestation showing conjoined Pygopagus twins with lumbosacral spina bifida (fusion from subcutis to thecal sac at lumbosacral level with craniocaudal fusion diameter of about 3.68cm), without visualized bone union, separated bladder, rectum and anus were difficult to evaluate, the placenta was located in the left uterine fundus, two separated fetal heads were on the right side, and each fetus had complete of feet

Fig. 4. Clinical photograph showing female pygopagus twins joined back-to-back facing away from each other with attachments at the buttocks and separated anus

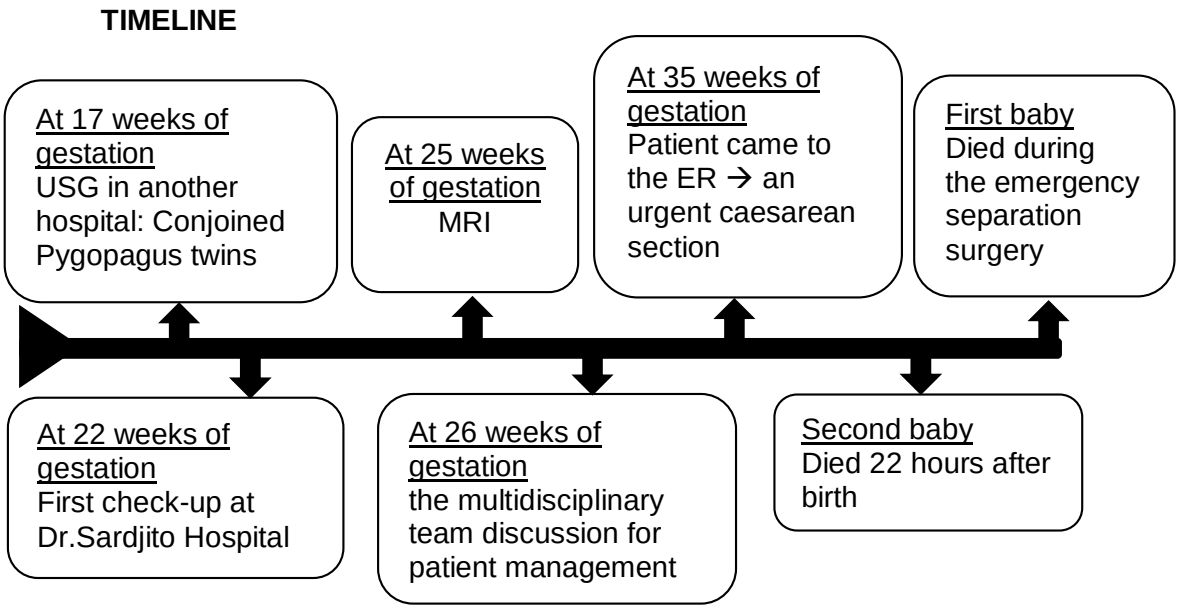

Fig. 5. Timeline of case report.

3. DISCUSSION

Conjoined twins are a rare complication of monochorionic–monoamniotic (monozygotic) pregnancies, but it has high morbidity and mortality rates. Conjoined twins are preceded by failure to separate from the embryonic disc at day 13 or later after fertilization, which is associated with monochorionic-monoamniotic. Another theory says this is also caused by a secondary fusion process that occurs between two different monovular embryonic discs (fusion theory).[6]

Accurate prenatal diagnosis is needed to see the anatomy and anomalies of the fetus using ultrasound or MRI. Ultrasound in the first trimester of pregnancy is the best

supporting examination for early detection of conjoined twins. The best time to diagnose conjoined twins is 11-14 weeks of gestation.[4] Specific sonographic features indicative of conjoined fetuses include: (a) bifid appearance of the fetal pole in the first trimester; (b) absence of separating membrane(s) between fetuses; (c) non-separation of fetal bodies and skin contours; this finding should persist and it should always be visualized at the same anatomic level; (d) detection of a variety of fetal anomalies; (e) finding more than three vessels in the umbilical cord and the identification of single placenta; (f) sonographic evidence of fetal heads and bodies lying in the same plane; (g) unusual extension of the fetal spines; (h) unusual proximity of the fetal extremities; (i) the fetuses do not change position to one another after movement or manipulation or as time passes by.[4,7] Ultrasound is the preferred investigation modality because it's non-invasive, non-ionizing, real-time imaging, low acquisition time, low costs, and broad availability which has sensitivity 86.2% and specificity 92.2%. The limitations of ultrasound are maternal obesity, severe oligohydramnion, limited field of view, and lower tissue resolution (Table 1.).[8] Prenatal fetal MRI help define the precise anomalies and connecting anatomies between the fetuses due to higher spatial resolution, excellent soft tissue contrast, unlimited tissue depth penetration, lack of ionizing radiation, and their non-invasive nature.[9] Postpartum MRI extensively to evaluate anatomical relationships, including the union of blood vessels that have not been/difficult to evaluate in prenatal fetal MRI examinations. Postpartum MRI will be performed in preparation for the separation surgery. Limitations of MRI are expensive, long acquisition time, limited sensitivity, and not as readily accessible. The accuracy of MRI is 97% compared to 90.4% of ultrasound (Table 2.).[8] In our case, conjoined pygopagus twins were first diagnosed at 17 weeks of gestation by ultrasound.

Table 1. Advantages and limitations of using USG as modality for diagnosis

Advantages of USG	Limitations of USG
Non-ionizing and Non-invasive	Maternal obesity
Broad availability	Severe oligohydramnion
Low costs	Limited field of view
Real-time imaging and Low acquisition time	Lower tissue resolution
Highly sensitive (Sensitivity 86.2 and Specificity 92.2)	
Accuracy 90.4%	

Table 2. Advantages and limitations of using MRI as modality for diagnosis

Advantages of MRI	Limitations of MRI
Define the precise anomalies and connecting anatomies between the fetuses	Expensive
Higher spatial resolution and excellent soft tissue contrast	Long acquisition time
Lack of ionizing radiation and Non-invasive	Limited sensitivity
Extensively to delineate anatomical details, especially of the cardiovascular system	Not as readily accessible

provides an overall assessment of the pregnancy during all 3 trimesters	
Unlimited tissue depth penetration	
Accuracy 97%	

The prognosis of conjoined pygopagus twins depends on the degree of vascular anastomosis, neurological connection, and spinal cord and vertebral column fusion. Vertebral fusion is more common than spinal cord fusion.[7] In our case, the MRI showed spina bifida at the lumbosacral level (fusion from the subcutis to the thecal sac) without visualized bone union. In the case of conjoined twins, intrauterine complications that may occur are umbilical cord entanglement, intrauterine growth restriction, preeclampsia, polyhydramnios, premature birth, and intrauterine fetal death. The diagnosis, prognosis, and complications that can occur must be informed in detail to the parents so they can decide to terminate or continue the pregnancy and postpartum plan for separation surgery with a multidisciplinary team.

The method of delivery in the case of conjoined twins is by performing an elective caesarean section. According to Greco et al., an elective caesarean section of conjoined twins was performed at 34-36 weeks of gestation, considering the possibility of stillbirth and the risk of premature birth. In conjoined twins, the recommended incision for caesarean section is a midline vertical skin incision, then a vertical uterine incision (Classical Caesarean Section).[10] According to O'Brien et al., an elective caesarean section of conjoined twins was performed at 35 weeks of gestation by considering preterm birth.[11] According to the Maternal Fetal Evidence Based Guidelines in 2017 and the Oxford Textbook of Obstetrics and Gynecology in 2020, the delivery time for monoamniotic-monochorionic twins is 32-33 6/7 weeks of gestation.[3,12] According to Van Mieghem et al., delivery time at 33 weeks of gestation had the best outcome for monoamniotic twins with corticosteroids have been administered.[13] In our case, the patient was planned to undergo an elective caesarean section at 36 weeks of gestation, but the patient had uterine contractions and leaked amniotic fluid at 35 weeks of gestation, so an urgent CS was performed.

Conjoined twins with planned separation surgery have an 80-90% survival rate compared to emergency separation surgery of only 30-50%. This is related to having more time to evaluate anatomical relationships and detect congenital abnormalities that have not been/difficult to evaluate in previous examinations.[4] In separation surgery, coordination, communication, and careful planning are required from a multidisciplinary team which involves a neurosurgeon, pediatric surgeon, orthopedic doctor, neonatologist, and anesthesiologist. The psychosocial needs of the parents should be provided.[7] In our case, the multidisciplinary team planned the separation surgery after the babies were born with postpartum MRI first performed, but in reality, an emergency separation surgery was performed because one of the babies died.

Respiratory distress syndrome is the most common cause of death in preterm with low birth weight conjoined twins. Cardiac congenital anomalies are the most commonly seen in conjoined twins. Complications such as infection, bleeding, organ failure, and intraoperative hypothermia can occur during the separation surgery.[14,15] In our case, both babies were born preterm with an APGAR score of 2/4/7, 94% oxygen saturation, and low birth weight, not long after that, the second baby died with the

diagnosis of severe asphyxia, and the first baby died during the emergency separation surgery due to bleeding.

4. CONCLUSION

Conjoined Pygopagus Twins requires holistic management starting from diagnosis, preparation for delivery, complications that can occur, and providing appropriate education to patients and families in dealing with all possibilities and consequences that occur before, during, and after delivery as well as after separation from the fetus. Proper holistic management is expected to reduce future infant and maternal morbidity and mortality.

ETHICAL APPROVAL

Written informed consent was obtained from the patient who participated in this study.

CONSENT FOR PUBLICATION

Attached

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Conflict of Interest Statement:

The author declares that the case report was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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