CASE REPORT

Ischiopagus Tetrapus Conjoined Twins

Nartono Kadri,[1] Harapan Parlindungan Ringoringo,[1] Darmawan Kartono[2]

(Departments of Child Health[1] and Pediatric Surgery[2] Medical School, University of Indonesia - Cipto Mangunkusumo Hospital, Jakarta)

ABSTRACT We report a case of ischiopagus tetrapus conjoined twins with one passageway (cloaca type) and born at term by spontaneously vaginal delivery, with a combined birth weight of 5000 grams. There was no history of twin in the family. During hospitalization, the conjoined twins suffered from sepsis due to ascending infection from cloacal portion. At the age of 17 days, an exploratory laparatomy and divided colostomy was performed. The postoperative complications were bleeding and sepsis. The bleeding could be properly handled only in 4 days after the treatment, whilst sepsis could not be overcome. Even in the 16th day after the operation, a peritonitis occurred followed by intestines prolapse from the edge of operative incision (on the 19th postoperative day) resulting in the death of the conjoined twins. **[Paediatr Indones 1996; 36:169-176]**

Conjoined twins are rarely found congenital anomaly, and always attract atention from both medical personnel and laymen. Medically, the etiology and pathogenesis of this anomaly are as yet unclear. The structure of these conjoined twins might appear to be weird and scary. The most famous conjoined twins are Chang and Eng, they were born in Siam (currently, Thailand) in 1811.[1] The twins Chang and Eng were connected at the front portion or xipho-ompahalo-pagus. At the age of 31 years,[2] they were married to two sibling women. Chang had 10 children, and Eng had 9 children. Both Chang and Eng died at the age of 63 years.

The conjoined locations varied considerably, and the conjoined body organs are in variation as well. These factors affected the prognosis of these conjoined twins.[3] Spencer (1956) is the first surgeon to successfully carry out the separation operation of ischiopagus.[4] The aim of this paper is to demonstrate that in the management of

Accepted for publication: July 23, 1996. Author's address: Dr. Nartono Kadri, Department of Child Health, Medical School, University of Indonesia, Jalan Salemba 6, Jakarta 10430, Indonesia. Phone 62-21-3907742, Fax 390-7743.

ischiopagus conjoined twins, a reasonably coordinated cooperation among various disciplines of study is essential, and the complexity of both preoperative and postoperative problems, as well as the duration of operative procedure should be taken into consideration.

Report of the Case

Both conjoined twins were hospitalized at the Department of Child Health, Faculty of Medicine, University of Indonesia - Cipto Mangunkusumo Hospital from February 21, 1995 to March 27, 1995. These conjoined twins were born on February 20, 1995 at home, by a full term spontaneously vaginal delivery, and assisted by a midwife. The combined birth weight of both conjoined twins was 5000 g. The first twin was delivered by cephalic presentation, and cried immediately; the second twin was born by breech presentation, and also cried immediately. The passageway was only one. Both conjoined twins were then referred to Dr Cipto Mangunkusumo Hospital.

The patients were the seventh and eighth children of the overall 8 brothers and sisters. The age of the patients' mother was 30 years, and the father was 40. There was no history of twin in the family. The mother had antenatal care as many as three times to a midwife. Before and during pregnancy, the patients' mother had not had any complaints of any disease or felt any abnormality. During the pregnancy, the mother consumed sufficient food, did not smoke, and frequently took in herbal concoction.

On physical examination, the first and the second twins appeared to be connected at the umbilical portion in a longitudinal position with opposite cephalic positions, and at dorsal portion, urethral orifice and anus were connected (see Figures 1 and 2). The general condition as well as vital signs of both twins was good, having neither dyspnea nor cyanosis. Each of the twins had 2 hands and 2 limbs respectively. Both feet of the first twin were clubbed, and such was the case of the right foot of the second twin. Other physical examination did not reveal any abnormality. At that time the working diagnosis was ischiopagus tetrapus conjoined twins.

The patients were treated at perinatal ward. At the age of third day, both conjoined twins appeared to be yellow-colored with indirect bilirubin levels of 7.57 mg/dl and 7.58 mg/dl, direct bilirubin levels of 0.56 mg/dl and 0.86 mg/dl for the the babies respectively. At the age of the fourth day, the second baby suffered from convulsions, and at the age of the fifth day the first baby appeared to have cyanosis. At the age of the eighth day, both babies had sepsis with indirect bilirubin levels of 10.98 mg/dl and 11.90 mg/dl, direct bilirubin of 7.59 mg/dl and 6.98 mg/dl, and the thrombocyte value of 2000/µl and 2000/µl for the respective babies. Cefotaxime and amikacin were administered to both babies. As the clinical conditions of both babies were getting worse, at the age of the tenth day an exchanged transfusion was done.

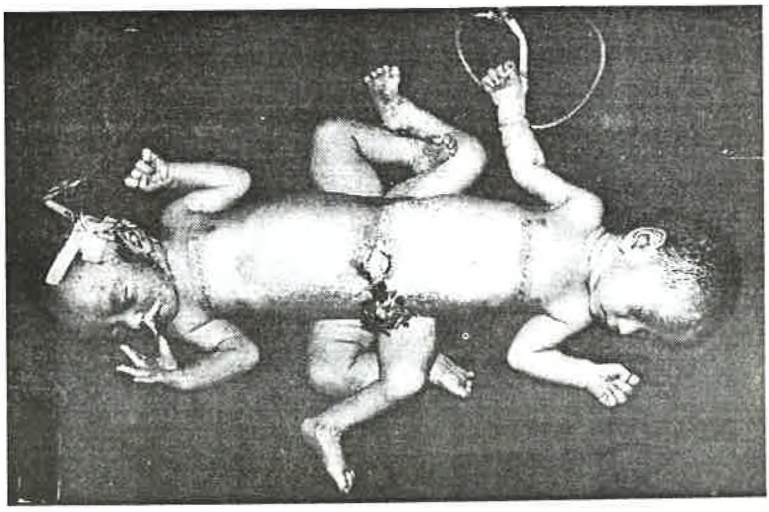


Figure 1. Anterior view of ischiopagus tetrapus conjoined twins.

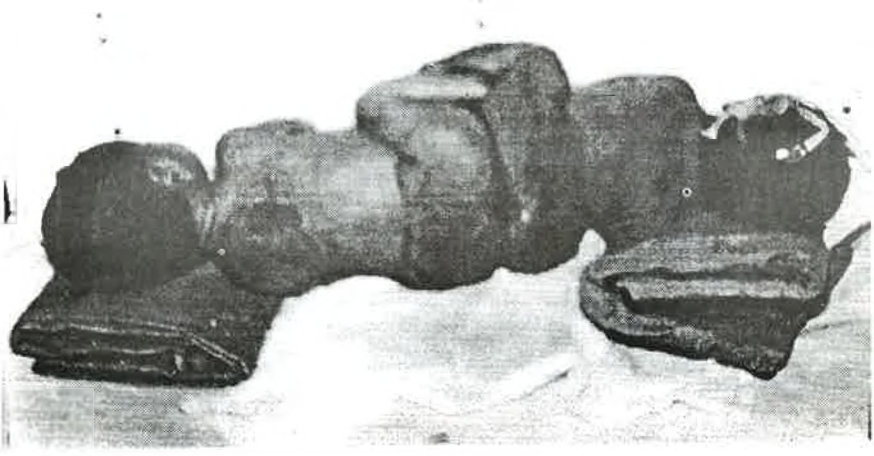
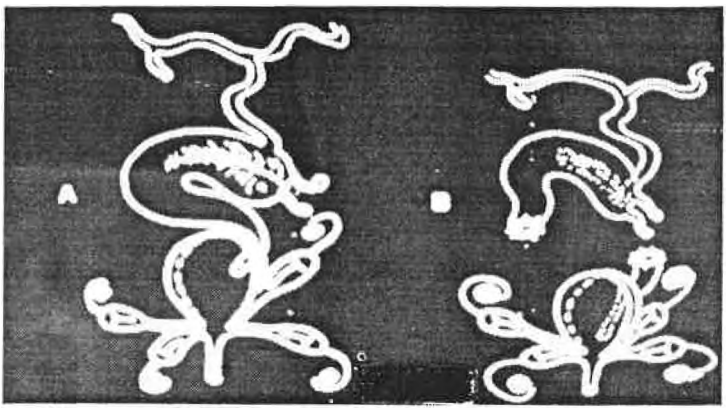


Figure 2. Posterior view of the twins.

Ischiopagus Tetrapus Conjoined Twins



A. Operative Findings.
Duplication of caecum, appendix, uterus, vagina, and bladder

B. Emergency operation : devided colostomy

Figure 3. Operative findings and divided colostomy of the conjoined twins.

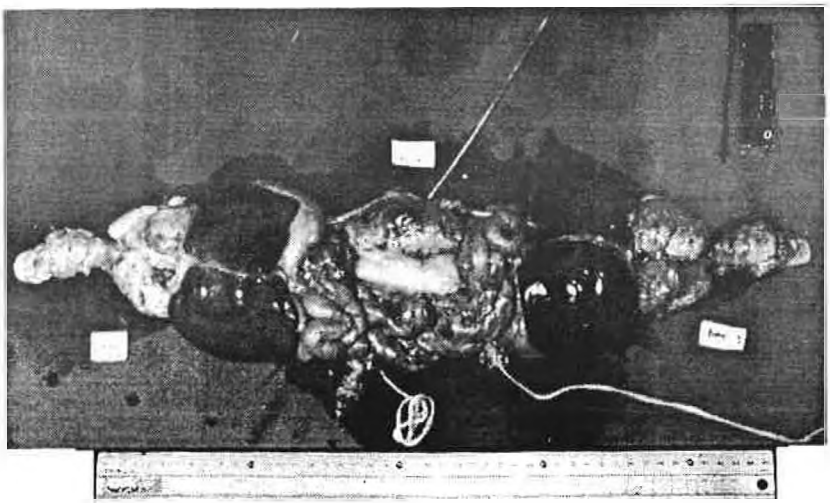


Figure 4. A part of autopsy findings of ischiopagus tetrapus conjoined twins

The clinical conditions of the patients improved after the exchanged transfusion has been performed. However, at the age of the 15th day their clinical conditions got worse and a melena was present, so that the exchanged tranfusion was repeated. After being carefully evaluated, it was suspected that the sepsis occurred due to ascending infection from cloaca. A clinical conference was then arranged among Departments of Child Health, Anesthesiology, and Pediatric Surgery. The meeting came up with a decision that before an exploratory laparotomy and colostomy were carried out, an exchanged transfusion should be done again to ensure an optimum condition of the patients.

The supporting examination showed that chest X-ray, echocardiography, cephalic and liver ultrasonography were normal. Abdominal plain photo revealed the unification of the digestive tract. Kidney ultrasonography revealed the presence of both kidney in each of the babies. During the exchanged transfusion, it appeared that there was a connection of blood vessel system in both babies. The blood type of the two babies was B with Rhesus (+), whilst chromosome analysis evidenced that both babies were of female sex (46,XX).

The operation was at last done on March 9, 1995, the patients' ages at that time were 17 days. During the operation, a union at ileum terminalis was found and terminated in one colon; there were unified vesicas, two ovaria and two Fallopian tubes in each of the babies. The first and second uterus attached to the edge of vesica. A divided colostomy was then attempted: an orifice colostomy was done in the first baby for urine discharge, and another orifice colostomy in the second baby for defecation (see Figure-3). During the operation, each of the babies received platelet transfusion of 30 ml and fresh frozen plasma of 50 ml. The patients were afterward monitored in the pediatric intensive care unit.

The postoperative problems were bleeding through the edge of operative wound and sepsis. The bleeding was properly managed in four days, while sepsis could not be handled until the death of the patients. On the 14th day of the care, the patients had acute diarrhea (feces culture revealed an infection of *Proteus vulgaris*). On the 19th day of the care, an intestinal prolapse occurred, and first baby finally died at 23:00 local time and the second baby at 23:30. The direct cause of the death was sepsis.

The next day an autopsy was done. The autopsy results revealed the presence of a union at terminal ileum terminating in one colon; there were two appendices situated in each side, while the livers were separated. The right kidney and right ureter of the first baby was smaller; there were two vesicas situated in the lateral portion and connected to each other through a tiny space in the middle part; the left ureter terminated in the gallbladder at the left edge, while the right ureter ended up in the gallbladder at the right edge. In the first baby there were 2 ovaria, 2 Fallopian tubes, 2 uterus, 2 vagina terminating in cloaca; whilst in the second baby there were 2 ovaria, 2 Fallopian tubes, 2 uterus, and 1 vagina. A part of autopsy findings can be seen in the Figure 4. There was a connection between the exit of abdominal aorta of the first and the exit of abdominal aorta of the second baby. The cause of their death was sepsis.

Discussion

Conjoined twins belong to the group of monozygotic twins and are rarely diagnosed at early pregnancy. Although the male monozygotic twins are more numerous than the female monozygotic twins, the life-born CT are generally female ones (with a ratio of 3:1).[3-7] The conjoined twins' data collected from 14 monitoring centers of congenital anomaly in the world showed that there were 312 cases out of >28 million births.[5] The incidence is 1.3:100,000 births,[6] and the highest incidence is in African-Asia race.[8] Of 117 cases of conjoined twins, the incidence of thoracopagus was 73.4% (pure thoracopagus 40% and omphalopagus 33.4%), pygopagus 18.8%, ischiopagus 5.5.%, and craniopagus 4.7%.[4] The medical records at Department of Child Health, Faculty of Medicine, University of Indonesia-Dr Ciptomangunkusumo Hospital revealed 9 cases of cojoined twins, consisting of 4 cases of ischiopagus, 2 cases of thoracoabdominopagus, 1 case of pygopagus, 1 cases of omphalopagus, and 1 case of caraniopagus.

Up to now, the etiology of conjoined twins is not clearly known. It is believed that all the factors contributing to the occurrence of monozygotic twins will likewise contribute to the conjoined twins. Among these are hypoxia, hypethermia,[2] protein and riboflavin deficiency,[9] the use of infertility drugs, and thyroid disease.[7] In this case, the causes remain unknown. Whether the habitual consumption of herbal concoction before or during pregnancy has been the cause is yet to clarified.

Pathogenesis of conjoined twins is not clear yet. Whether they result from a fusion process or the failure of fission process is a question to be answered. In this case, both the babies appeared to be identical with identical sex, one umbilicus; both of them had blood type B. This shows that these conjoined twins came from mono-ovular monochorion. This fact is in accord with the imperfect inner cell mass division before organogenesis.[3,8,9]

The skill of the midwife in assisting the delivery of the conjoined twins deserved to be complimented. This may be attributed to the relatively broad pelvis of the mother, and the fact that this delivery constituted the seventh one. Nevertheless, if we encounter a prenatal case of conjoined twins, it is advisable that the delivery be done by cesarean section in view of the high incidence of dystocia and stillbirth.[4,7,10]

If the sepsis due to ascending infection from cloaca had been suspected earlier, the operation would have been performed at an early stage, and the results would probably be better. Albert et al suggested that, if possible, the separation operation be postponed up to an average of 14 months (with a range of 4-27 months).[11] However, an emergency operation (absolute indications) must be done if:[3,4,8]

1. One of the twins died, is dying, or the conditions of one of the twins are critical and jeopardize the life of the other twin.
2. A condition making one of, or, the two babies unlikely to survive, such as: obstruction of digestive tract, obstruction of urinary tract, atresia ani, intraperitoneal bleeding, or omphalocele rupture.

The separation operation of ischiopagus conjoined twins should be done in two stages. The first stage of the operation is to remove the obstruction of digestive tract, whilst the second stage, i.e. a definitive operation should be conducted after three months. This gradual operation will permit the definitive operation to be carried out in a better clinical condition, and to cut short the duration of separation operation.[8,12,13]

In retrospective, based on autopsy findings, if the colostomy had been successful, it would make the definitive operation difficult to perform. The urinary tract which was similar to mirror reflection, the unified vesica, the complicated reproductive system, presence of unified blood vessels and the musculoskeletal problems, as well as definitive postoperative pelvic reconstruction problems are likely to cause an unfavorable result. Nevertheless, Shah and Chazotte reported a case of delivery by an 18-years old mother who had undergone a separation operation of ischiopagus beforehand. This suggests that although an extensive operation has been done involving abdominal portion, genitourinary system and extensive pelvic reconstruction, the reproductive functions of the mother can still be maintained to be able to deliver a healthy baby.[7]

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