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Case Report

# Separation of ischiopagus tetrapus conjoined twins in Dr. Sardjito Hospital, Yogyakarta

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Conjoined twins are one of the greatest challenges to modern pediatric surgery. Despite extensive preoperative investigations, the precise description of the conjoined anatomy is often only possible during surgery.[1-3]

Social attraction, medical-ethical problems, and sociological impacts are three characteristics faced by conjoined twins or Siamese twins, so called due to the famous conjoined twins Chang and Eng Bunker, born in 1811 in Siam. They were united at the umbilicus by 17 cm wide bridging tissue; they had never undergone surgery and lived together for 63 years.[4-6] The existence of conjoined twins was mentioned in the old Egyptian records. The first successful separation was performed in Bazilea by Johannes Fotio. The twins were joined at the region between the xiphoid process and the umbilicus. Fotio succeeded in separating the umbilical vessels, as well as tying and transecting the bridging tissue. This case was presented in 1689 by Koenig.

This paper reports a case of conjoined twins who were successfully separated in Sardjito Hospital, Yogyakarta, Indonesia in 2006.

## Case report

The babies were born in a rural hospital to a healthy 28-year-old mother in January 2006, full-term, and delivered spontaneously with the combined birth weight of 5454 grams. The mother died after delivery due to uncontrolled bleeding. We labeled the first baby as Baby A and the second one as Baby Y. They were conjoined at the pelvic region with their lower limbs parallel on both sides, arising from lateral part of their bodies. The anterior abdominal wall ran from one to the other without clear demarcation. There was only one umbilical cord located exactly in the middle of the anterior abdominal wall, two anuses and two vulvae (**Figures 1** and **2**).

All organ systems were evaluated with x-rays, renogram, ureterocystogram, barium enema, and CT scan. The bony pelvis formed a ring and joined with the normal lower extremities. The upper and lower gastrointestinal studies demonstrated normal and separate stomachs, small intestines, and a single cecum, single ascending colon with single appendix. The two small intestines joined at the terminal ileum and entered the solitary cecum. Each baby had two

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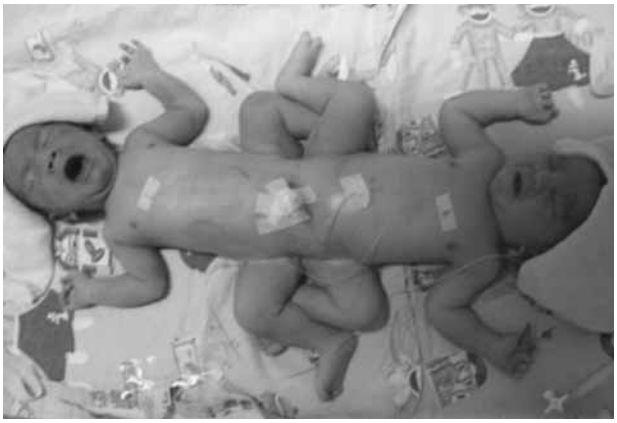

**Figure 1**. The anterior abdominal walls

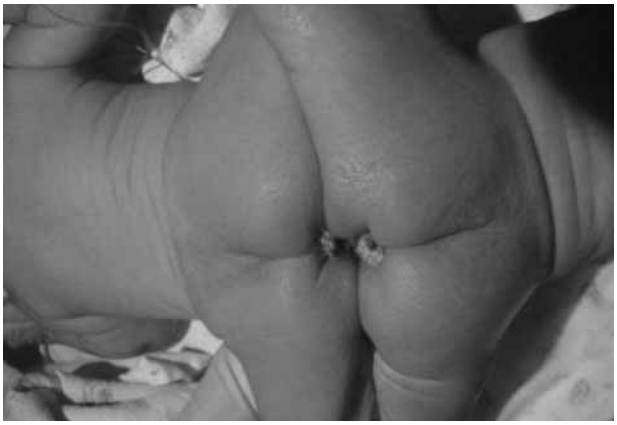

**Figure 2**. The posterior side with 2 anuses

kidneys in the shared pelvis with two ureters which emptied into two bladders that was attached one to another. The functions of micturition were normal except for Baby A who had hydroureter with reflux and mild hydronephrosis caused by occlusion of the urethral orifice. Each of them had one normal uterus. Operation was performed when they were seven months old to prevent further damage to Baby A's kidney. Their combined weight was 8000 grams.

Preparations for surgery were including forming a team consisted of 40 experts, who were anesthetists, surgeons, intensive care specialists, electrical engineers, blood bank and laboratory staffs. The major surgical considerations included draping procedure, operative approach, the sequence of organ separation, preservation of urinary tract, pelvic reconstruction and the closure of the huge abdominal wounds in both twins.

A large transverse incision nearly 40 cm in length was made across the anterior abdominal wall. Exploration confirmed the accuracy of the preoperative evaluation. Each infant had a normal stomach, duodenum, liver, gallbladder, pancreas, spleen and small intestines. Two small intestines were in normal length and joined for about 15 cm from the single cecum. Besides the single cecum with single appendix, there were also single ascending colon, single transverse colon, and two sigmoids with two rectums.

The two small intestines were transected at the junction where they joined the common terminal ileum. The transverse colon was then divided and an ileotransversostomy anastomosis was performed (**Figure 3**).

There were two separate kidneys lying in the pelvis, and the bladder was separated. A Gore–Tex patch was used to reconstruct the distal part of the anterior abdominal wall. A thick layer of subcutaneous fat was spread over the patch and the skin layer was finally reconstructed (**Figure 4**).

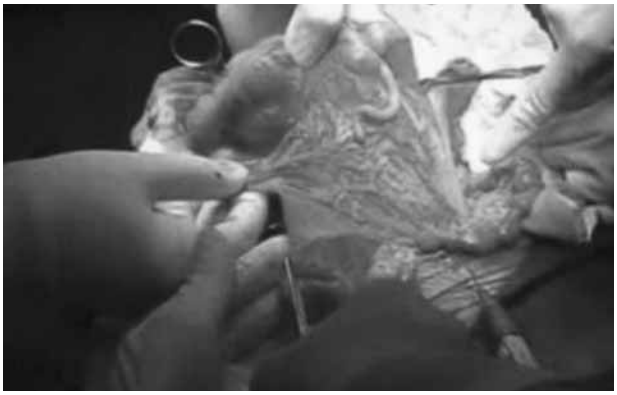

**Figure 3**. The transverse colon was divided and an ileotransversostomy anastomosis was performed

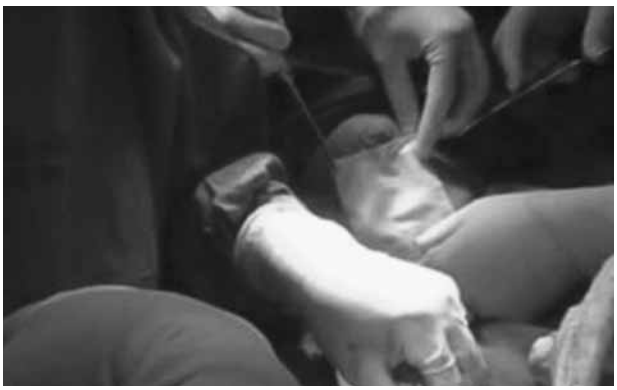

**Figure 4**. The reconstruction of the distal part of the anterior abdominal wall using Gore-Tax patch

The difficulties during operation were when the separation of the bridging vascular system in the pre-sacral region, separation of the fused bones, and reconstruction of the pelvis, the pelvic floor and the anterior abdominal wall were performed (**Figure 5**).

There was no hemodynamic instability during the operation. There was only minimal blood los. Ten minutes after the seven hours of surgery, both twins moved their limbs and opened their eyes. Three days later, Baby A developed peritonitis, caused by leakage, which was successfully treated by colostomy and re-anastomosis.

**Figure 5**. After separation

**Figure 6**. The twins at the age of two

There was no problem in nutrition and they began to walk at 13 months. Their mental development is excellent; there were no ambulatory problems and they speak fluently (**Figure 6**).

## Discussion

The exact incidence of Siamese twins varies from one per 14,000 live births, to one per 200,000 live births. Approximately two thirds are female, and all pairs are of the same sex. There are five main types of conjoined twins: thoracopagus (40%), omphalopagus (33%), pygopagus (19%), ischiopagus (6%) and craniopagus (2%).[7]

In thoracopagus twins, the joining includes the thorax and may extend from manubrium to umbilicus. Seventy–five percent of the twins have varying degrees of cardiac fusion. In omphalopagus, the joining is at the abdominal wall and fusion of the liver must be anticipated when the junction of the twins includes the umbilical area. In pygopagus, there is attachment at the buttock area and vertebral columns, usually in continuity but spinal cords are usually separated. In ischiopagus, the fusion extends from the umbilical area to include the lower trunk and pelvis with three or four limbs. Craniopagus twins might be partial or total conjoining forms, having a junction at brow, vertex, or parietal bone. [8-10]

Identical twins develop from the division of a single fertilized ovum. By the sixth day after fertilization, the human zygotes become blastocysts. At one pole of the blastocyst, some cells aggregate, known as the inner cell mass. From this cell mass, the embryo, amnion, and yolks sac develop. Inner cell mass is totipotent and may split to form two germinal discs which can develop into two identical individuals. Division of the zygote within the first seven days after fertilization yields monozygotic identical twins.[1,2,6]

The ultrasound examination in the 12$^{th}$ week of gestation is very useful to detect polyhydramnios with one placenta, one umbilical cord with more than three blood vessels. Prenatal CT examination or magnetic resonance imaging may detect the exact anatomy of the malformation and the presence of other associated anomalies.[1,4]

Preoperative examinations are performed to evaluate the internal anatomy and to determine

whether one or both twins can survive after separation. The anatomy, the function of all major organ systems, and the skeleton are needed to be evaluated. In thoracopagus conjoined twins, CT scan of chest and liver, isotopic biliary tract scan, contrast studies of the gastrointestinal tract and ECG are needed. Omphalopagus needs CT and isotopic scans of the liver and biliary tract. Head CT scan, arteriogram and EEG are important in craniopagus conjoined twins. CT scan of the abdomen and pelvis, contrast studies of gastrointestinal and urinary tract, abdominal arteriography and genitography are needed in ischiophagus conjoined twins. Pygopagus conjoined twins need CT scan of abdomen and pelvis.[1,3,7]

The separation of conjoined twins is a surgical challenge, especially that of ischiopagus type. The planning for eventual surgical separation should be unhurried. It needs multidisciplinary team. The main goal is to achieve life as normal as possible. A variety of diagnostic studies is necessary to create diagrams of organ sharing and demonstrate possible coexisting congenital anomalies. Separation is best performed on an elective basis when the babies are 9 to 12 months of age, but the timing depends on the abnormalities. On the other hand, urgent separation may be required, even in the newborn because of the emergency conditions such as stillborn one of the twins, intestinal obstruction, ruptured omphalocele, obstructive uropathy, heart or respiratory failure.

The difficulties in the present case were creating proper anastomosis due to the condition of the cecum, terminal ileum and transverse colon; separating the bridging vascular system in the pre-sacral region; separating the fused pelvic bones; and reconstructing of the pelvis, the pelvis floor and the anterior abdominal wall. Factors that make this separation successful were careful preoperative planning and detailed investigation, recognition of potential intraoperative and postoperative problems, intensive intraoperative monitoring and good management.

In conclusion, ischiopagus tetrapus is a very rare variation of conjoined twins. With a team approach and good preparation, they can be successfully separated.

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