# BLADDER RECONSTRUCTION DURING SEPARATION OF A CONJOINT TWIN ISCHIOPAGUS TRIPUS

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***ABSTRACT***

***Objective:*** *Conjoined twins represent one of the rarest congenital anomalies occurring with a varying incidence of about approximately 1 : 50.000 - 1 : 200.000 births. Ischiopagus conjoined twinning (Fuse at the hip region) is even rare representing only 6% of all conjoined twins. Here we report our experience in lower urinary tract reconstruction during separation of a conjoined twin ischiopagus tripus in Hasan Sadikin Hospital, Bandung.* ***Case(s) Presentation:*** *Two years old female conjoined twins, weighing in total of 8 kilograms, were brought to our hospital. The twins were fused at the lower halves of their bodies with two separate lower limbs at the right side and a fused left lower limb. External genitalia were absent. From CT Angiography we found four kidneys, two in each child with two separate bladders, with right ureter of baby A and left ureter of baby B enter the right bladder, while left ureter of baby A and right ureter of baby B enter the left bladder. There was one cloaca that drains urine and feces. During the separation surgery, we performed ureteroneocystostomy and temporary ureteral stenting, followed by cystostomy. The pediatric surgeon performed bowel separation by stapling technique, pelvic wall separation, ileostomy, and pelvic floor reconstruction. The orthopedic surgeon performed femur separation, pelvic floor reconstruction, reconstruction of the pelvic ring, and femur reconstruction. The patients underwent defect closure, which performed by plastic surgeon. The operation was successful, without any complication to the kidney and the urinary tract of both babies.* ***Discussion:*** *Ischiopagus conjoined twins are the most complex conjoined twins and occur in only 6% of all conjoined twin cases. Ischiopagus is usually located along the axis with the head located on the opposite side. This case usually has one umbilicus and the two bodies are fused below this level, the union occurs in the lower abdomen and the pelvis. The internal organs that usually coalesce in cases of ischiopagus conjoined twins are liver, lower gastrointestinal, and genitourinary. The fused gastrointestinal part is the ileo-colic part of terminal ileum (Meckel's diverticulum).* ***Conclusion:*** *The separation of conjoined twin is a unique challenge due to its complex anatomy and physiology. Although advancement in imaging and monitoring has improved the survival rate separation can be successfully achieved only with meticulous planning and team work.*

***Key words:*** *Bladder reconstruction, conjoint twin, ureteroneocystostomy.*

***ABSTRAK***

***Tujuan:*** *Kembar siam mewakili salah satu anomali kongenital terlangka yang terjadi dengan insiden bervariasi sekitar 1 : 50.000 - 1 : 200.000 kelahiran. Kembar siam ischiopagus (bersatu di daerah pinggul) bahkan hanya mewakili 6% dari seluruh kasus kembar siam. Berikut kami laporkan pengalaman kami dalam rekonstruksi pemisahan saluran kemih bagian bawah kembar siam ischiopagus tripus di Rumah Sakit Hasan Sadikin, Bandung.* ***Presentasi kasus:*** *Kembar siam perempuan berusia dua tahun, dengan berat total 8 kilogram, dibawa ke rumah sakit kami. Kembar ini menyatu di bagian bawah tubuh mereka dengan dua tungkai bawah terpisah di sisi kanan dan tungkai bawah kiri menyatu. Alat kelamin eksternal tidak ada. Dari CT Angiography, ditemukan empat ginjal, dua di setiap anak dengan dua kandung kemih terpisah, dengan ureter kanan bayi A dan ureter kiri bayi B memasuki kandung kemih kanan, sedangkan ureter kiri bayi A dan ureter kanan bayi B memasuki kandung kemih kiri. Terdapat satu kloaka yang mengeluarkan air seni dan tinja. Selama operasi pemisahan, kami melakukan ureteroneocystostomy dan stenting ureter sementara, diikuti oleh cystostomy. Dokter bedah anak melakukan pemisahan usus dengan teknik stapel, pemisahan dinding panggul, ileostomi, dan rekonstruksi dasar panggul. Ahli bedah ortopedi melakukan pemisahan tulang femur, rekonstruksi dasar panggul, rekonstruksi cincin panggul, dan rekonstruksi tulang femur. Untuk mengurangi defek pemisahan dan rekonstruksi ini dilakukan oleh ahli bedah plastik. Operasi itu berhasil, tanpa ada komplikasi pada ginjal dan saluran kemih kedua bayi.* ***Diskusi:*** *Kembar siam ischiopagus adalah kembar siam yang paling kompleks dan hanya terjadi pada 6% dari semua kasus kembar siam. Kasus ini biasanya terletak di sepanjang sumbu yang berlawanan dengan kepala. Kasus ini biasanya memiliki satu umbilikus dan dua tubuh menyatu di bagian perut bawah dan panggul. Organ-organ internal yang biasanya menyatu dalam kasus kembar siam ischiopagus adalah hati, pencernaan bagian bawah, dan genitourinari. Bagian gastrointestinal yang menyatu adalah bagian ileo-kolik dari terminal ileum (divertikulum Meckel).* ***Simpulan:*** *Pemisahan kembar siam merupakan tantangan*

*unik karena anatomi dan fisiologinya yang kompleks. Meskipun kemajuan dalam pencitraan dan pemantauan telah meningkatkan kelangsungan hidup pasien, hal ini dapat berhasil hanya dengan perencanaan yang cermat dan kerja sama tim multidisiplin yang baik.*

***Kata kunci:*** *Rekonstruksi kandung kemih, kembar siam, ureteroneosistostomi.*

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## INTRODUCTION

Conjoined twins is challenging case for the medical world. Conjoined twin cases are rare, with an estimated incidence of 1 in 50,000 births.[1] with most cases occurred in women.[1,2] However, in most cases, the pregnancy leads to abortion and stillbirth; only 18% of all conjoined twins survive, and about 35% of live births die within the first 24 hours, and only 18% of all conjoined twins last more than 24 hours.[1]

Conjoined twins are monozygotic, monochorionic twins, in which there is an incomplete separation of the inner cell mass at day 13 to 15 of pregnancy. The classification of conjoined twins depends on their attachment sites, the ventral union, the dorsal union, and the lateral union. The ventral union group consists of the cephalophagus (head), thoracophagus (chest), omphalophagus (umbilicus), and ischiopagus (pelvis). The dorsal union group consists of pyophagus (sacrum), rachiphagus (spine), and craniophagus (cranium). The last group consists of the paraphagus (unification on one side). In addition, conjoined twins can be classified according to the number of existing lower limbs, such as tetrapus (four), tripus (three), and bipus (two).[3]

The surgical separation of conjoined twins is now a challenge for all surgeons because of the rare incidence and complexity of anatomy, although the surgical correction of the persistent cloaca is complex, the upper and lower urinary tract outcome can be favourable.[4] In this study, we describe the reconstruction of the lower urinary tract on the separation of conjoined twin ischiopagus tripus.

## CASE(S) PRESENTATION

A pair of conjoined twins, 2 years old, weighing 8 kilograms in total, admitted to our hospital. These conjoined twin case have unified on the lower part of their bodies with 2 separated lower limbs on the right side and a fused lower limb on the left side.

The external genitalia cannot be identified/was absent. Vaginal atresia occurs with a fistula (The channel in which both ileum empties into a terminal ileum into a caecum, colon, rectum (sharing ileum, caecum, colon/rectum). Rectum empties down to 1 common channel which is also the estuary of 2 pieces of urethra.

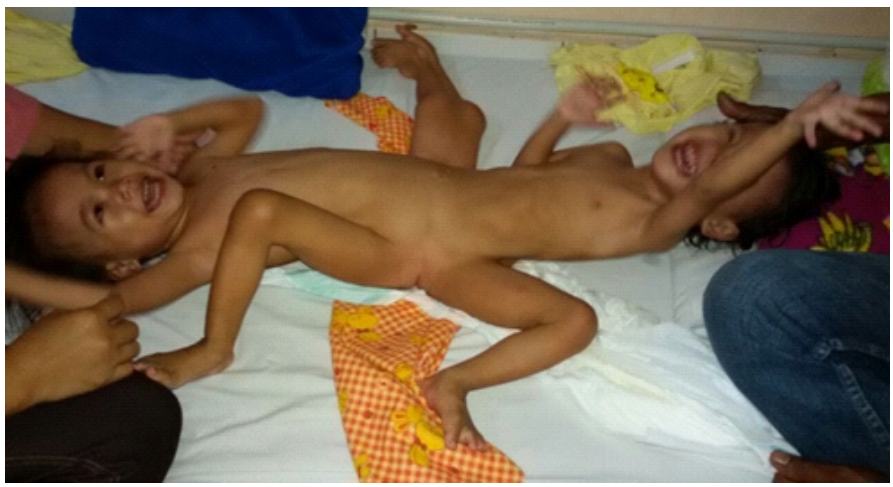

**Figure 1.** Clinical presentation of ischiopagus tripus.

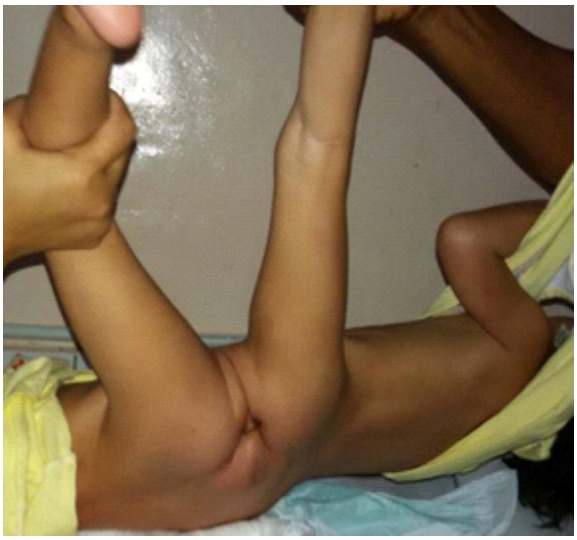

**Figure 2.** Vaginal atresia.

Angiographic CT was performed. There were 4 kidneys, 2 in each child, and two separate bladders, with the right ureter of child A and the left ureter of child B entered the right bladder, while the left ureter of child A and the right ureter of child B entered the left bladder. There was 1 cloaca that drains urine and stool.

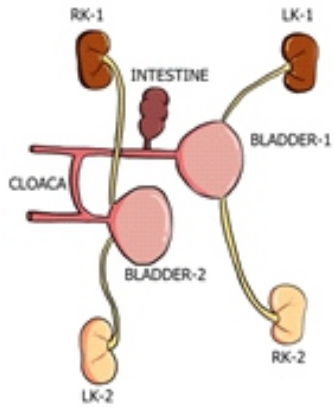


**Figure 3.** Anatomy before separation.

During the separation surgery, we performed bladder reconstruction, ureteroneo-cystostomy and temporary ureteral stenting, followed by cystostomy. The duration of surgery was 210 minutes in total, the blood loss was 1600 cc. The surgical results were successful, with no complication in the kidneys and urinary tracts in both children.

The importance of intestinal reconstruction is to make the intestinal system able to absorb nutrients, water, and various substances that are useful to the body.

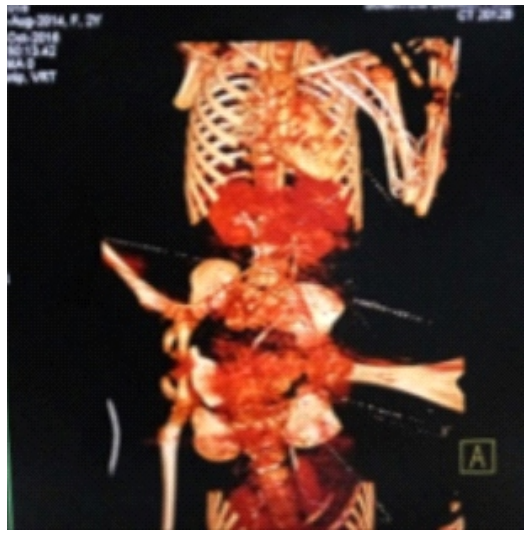

**Figure 4.** CT Angiography of abdomen and pelvis.

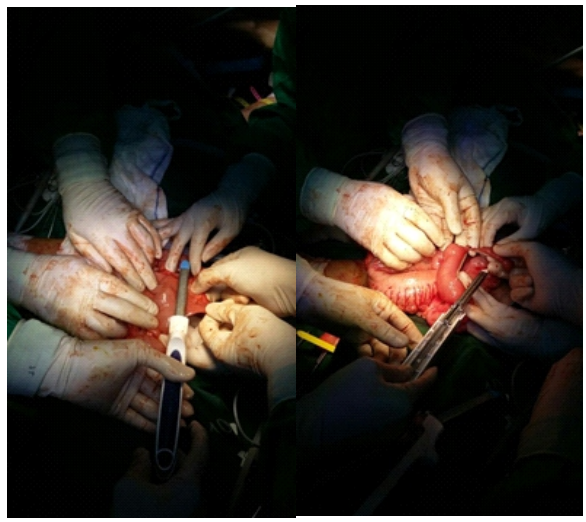

**Figure 5.** Ileal dissection using stapler

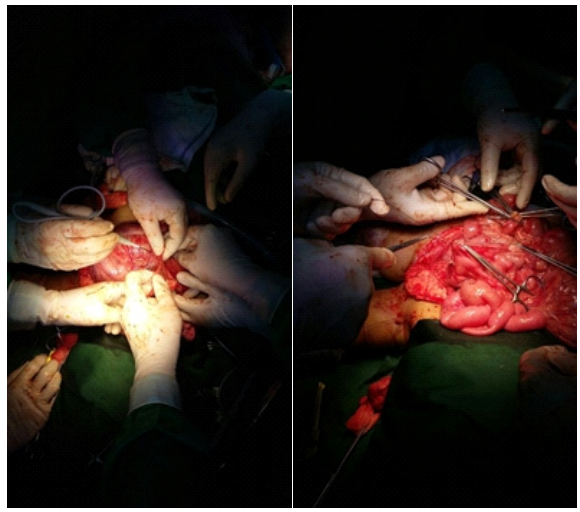

**Figure 6.** Mesenterial dissection.

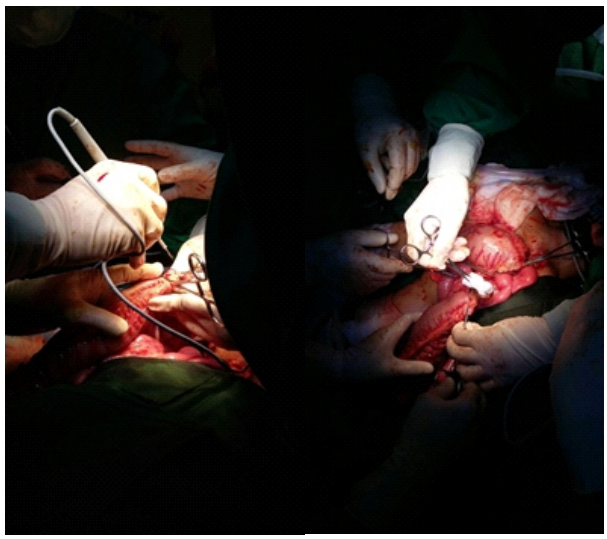

**Figure 7.** Colon dissection.

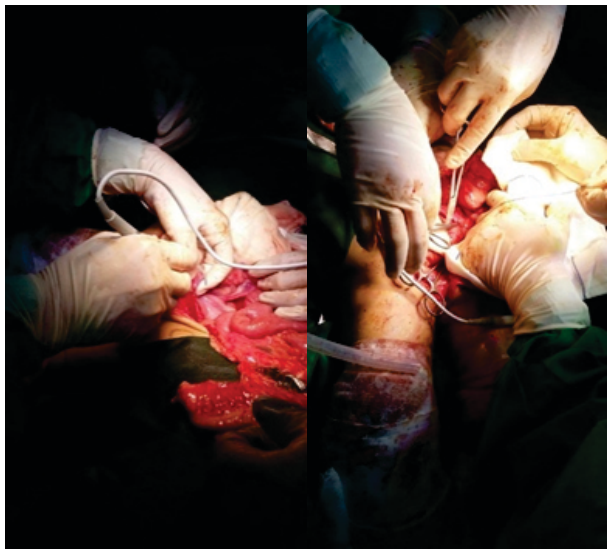

**Figure 8.** Mesenterial dissection and ovarial separation.

The skeleton of child A child developed thoracolumbar vertebral scoliosis with a thoracic apex as high as the Th-8 vertebra, apex of the lumbar vertebra as high as the L-2 vertebra. Child B had thoracic vertebral scoliosis, no hemivertebra or spina bifida. The left femur of child A fused with the right femur of child B, and the ischium also fused with pubic symphisis. The ilium-sacrum parts were normal.

In the vascular section, child A had iliac artery branch as high as the L-3 vertebra with a left-sided common iliac artery smaller than the right side, the right renal artery out of the aorta as high as L-1 vertebrae and the left renal artery as high as L-2 vertebra. The femoral artery to the left femur had a small diameter. In child B, the right and left iliac artery branches were as high as L-4 vertebrae with the same relative diameter respectively, the right femoral artery had a relatively normal diameter.

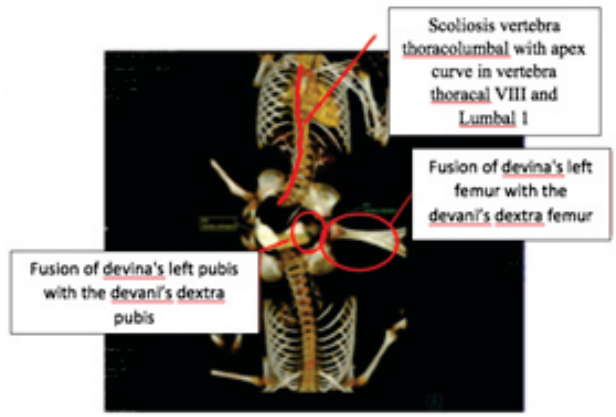


**Figure 9.** CT Angiography of proximal lower extremities.

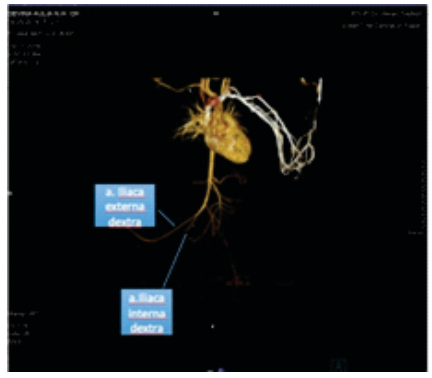


**Figure 10.** CT angiography.

In the genitourinary tract, the left ureter entered into the left urinary vesica, the right ureter entered into the right urinary vesica.

In the gastrointestinal tract section, child A experienced gastric passage disorders with gastroesophageal reflux, each child had a jejunum and an ileum. The rectums unifies with a possible shared colon between child A and B

Surgical separation of conjoined twins is a risky operation, which can be avoided by minimizing surgical time and anesthesia. In addition, the need for good technical abilities and experiences are critical to successful surgery.[5,6] Treatment planning was done well, several meetings involving all specialist doctors (anesthesia, pediatric surgery, urology, orthopedics and plastic surgery), nurses,

therapists, and administrative teams had been undertaken to ensure the surgery proceeds well.

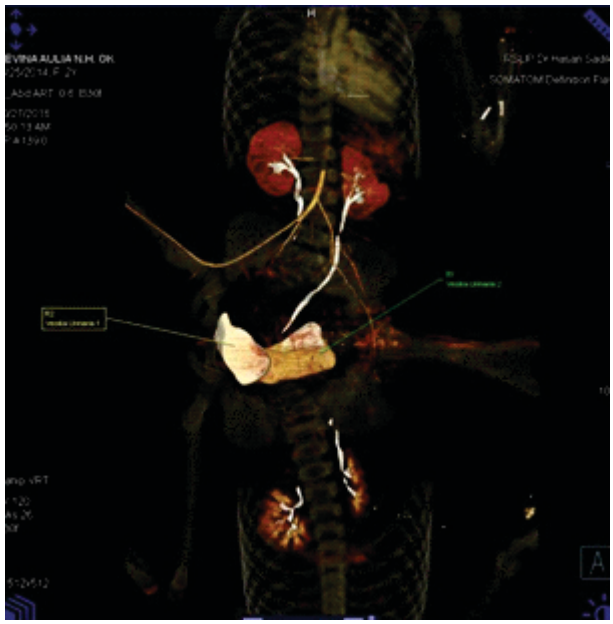

**Figure 11.** CT Angiography of genitourinary tracts.

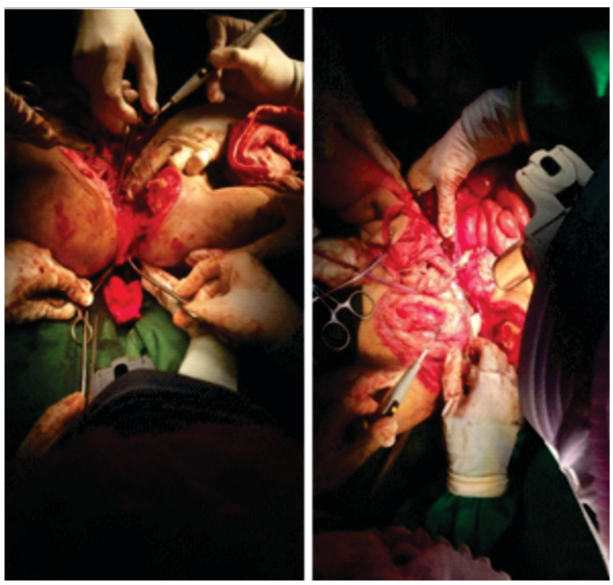

**Figure 12.** Separation process.

In this case, conjoined twins had 4 kidneys and 2 bladders, with 1 ureter emptying on the ipsilateral side and the other leading to the contralateral side. Cystoscopy and catheter or ureter stent insertion were conducted, then the separation of ureters (with temporary stenting on 2 ureters) and the separation of the bladders were performed. When the 2 children had been separated, ureteral reimplantations were performed on each child. The surgery completed successfully, with no complications in the kidneys and urinary tracts in both children.

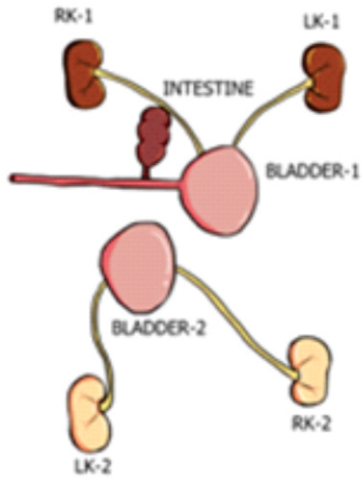


**Figure 13.** Anatomy after reparation.

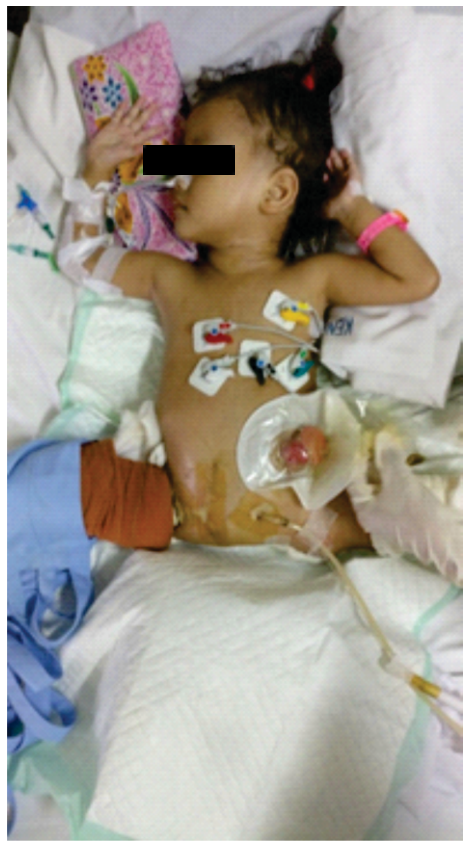

**Figure 14.** Patient A after separation.

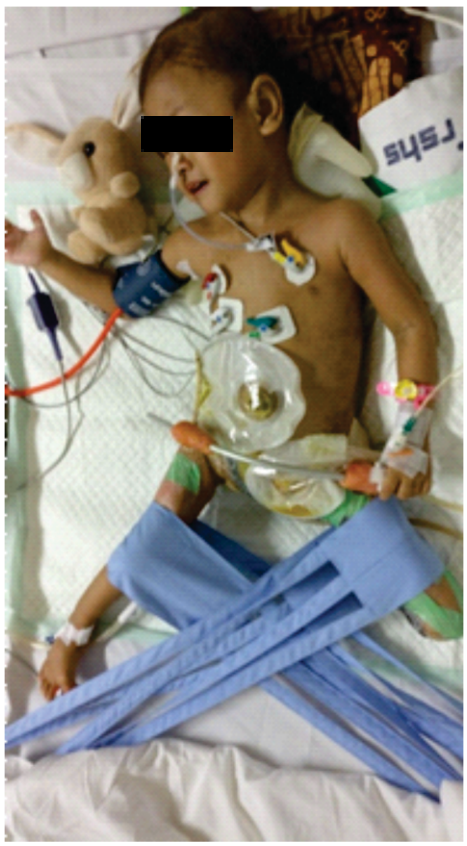

**Figure 15.** Patient B after separation.

## DISCUSSION

Conjoined twin cases are rare. The rapidly developing prenatal ultrasound ability to detect congenital anomalies causes the incidence of conjoined twins to decline. The etiologies of conjoined twins are still a controversy.[1] Some experts described that as a result of the incomplete diffusion of the embryonic disc, another theory suggested that due to reunion of the formerly separated monozygous embryonic disc.[1] The separation process of conjoined twins is a complex surgery and must be well-planned. The ideal time for separation is during the first year of life.[5] Extensive investigations and separation efforts of conjoined twin cases require a multidisiplint approach, involving various specialist doctors. In our case, the separation surgery was done at the age of 2 years, so that there had been postural deformity and psychological problems.

Ischiopagus conjoined twins are the most complex conjoined twins and occur in only 6% of all conjoined twin cases. Ischiopagus is usually located along the axis with the head located on the opposite side. This case usually has one umbilicus and the two bodies are fused below this level, the union occurs in the lower abdomen and the pelvis. The internal organs that usually coalesce in cases of ischiopagus conjoined twins are liver, lower gastrointestinal, and genitourinary. The fused gastrointestinal part is the ileo-colic part of terminal ileum (Meckel's diverticulum). Approximately 51% of ischiopagus twins have pelvic organ fusion.[6] In such case, diagnostic examinations are performed, including skeletal examination, ultrasonography, imaging with contrast of gastrointestinal and urinary system, and angiography to assess abnormalities in the skeletal, vascular, genitourinary tract, and gastrointestinal tract.

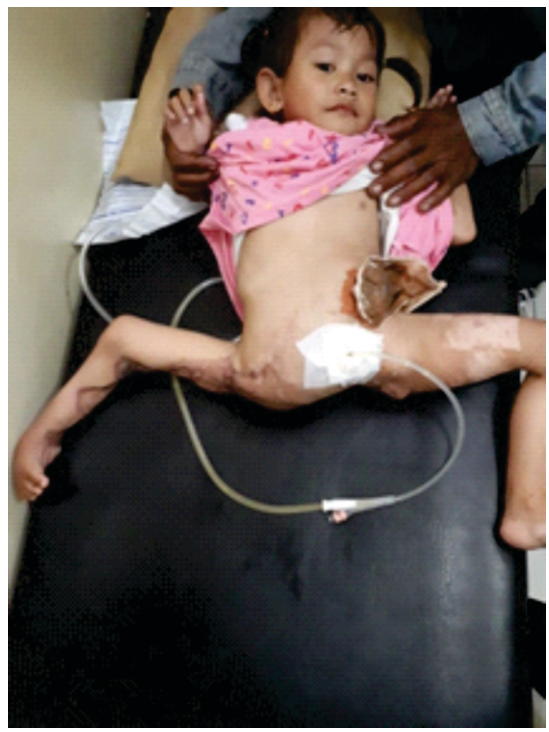

**Figure 16.** Patient A 7 months post operation.

A detailed anatomic study of the twins and surgical planning must precede separation. A well-prepared pediatric surgery team is sufficient to surgically manage conjoined twins.[7] Complex urological problems occur in ischiopagus and pygopagus conjoined twins. Beyond the challenge of surgical separation, they continue to provide management challenges requiring secondary reconstructive surgery and close urological monitoring.[8]

**Figure 17.** Patient B 7 months post operation.

Facing conjoined twin cases, surgical separation is the only way to provide normal conditions in patients. With a comprehensive and multi-disciplinary planning process, more conjoined twin surgeries are successful. The successful separation of ischiopagus twins requires multiple reconstruction and long-term follow-up to prevent complications. Strict observation and adequate interventions are necessary to maintain renal function, bladder continence, and maintain fertility function in order for patients to have a better quality of life.

## CONCLUSION

The separation of conjoined twin is a unique challenge due to its complex anatomy and physiology. Although advancement in imaging and monitoring has improved the survival rate separation can be successfully achieved only with meticulous planning and team work.

## REFERENCES


1. Rode H, Fieggen A, Brown R, Cywes S, Davies M, Hewitson J, et al. Four decades of conjoined twins at red cross children's hospital - lessons learned. South African Medical Journal. 2006; 96(9): 931-40.
2. Spitz L. Conjoined twins. Current Paediatrics. 2001; 11: 386-9.
3. Spencer R. Anatomic description of conjoined twins: A plea for standardized terminology. Journal of Pediatric Surgery. 1996; 31(7): 941-4.
4. Rink RC, Herndon CDA, Cain MP, Kaefer M, Dussinger AM, King SJ, et al. Upper and lower urinary tract outcome after surgical repair of cloacal malformations: A three-decade experience. BJU International. 2005; 96: 131-4.
5. Wilcox DT, Quinn FM, Spitz L, Kiely EM, Ransley PG. Urological problem in conjoined twins. British Journal of Urology. 1998; 81: 905-10.
6. Khan YA. Ischiopagus tripus conjoined twins. APSP J Case Rep. 2011; 2: 5.
7. Tannuri AC, Batatinha JA, Velhote MC, Tannuri U. Conjoined twins-twenty years experience center in Brazil. Clinics. 2013; 68(3): 371-7.
8. Lazarus J, Raad J, Rode H, Millar A. Long term urological outcomes in six sets of conjoined twins. Journal of Pediatric Urology. 2011; 7: 520-5.