Case Report

Successful Separation of Thoracopagus Conjoined Twins with a Single Extra-hepatic Biliary System

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A pair of thoracopagus conjoined twins were separated at the age of 3 months at Khon Kaen Regional Hospital, Thailand on November 19, 2004. Pre-operative investigations showed separate hearts, joined duodenum, and fusion of the livers. Separation of the extra-hepatic biliary systems was suspected. Operative findings revealed fusion of the intestines from the second part of the duodenum to the terminal ileum with two normal colons. An intussusception was found at the terminal ileum. Fusion of the livers with only one extra-hepatic biliary system was noted. In one of the twins, the gastrointestinal tract was anastomosed with Roux-en- Y enteric loop to one area of good bile drainage at the cut surface of liver. Post operative course was hectic but both twins recovered satisfactorily. Both are doing well at present, two years after the separation.

This was the first reported case of thoracopagus conjoined twins with complex biliary tract anomalies in Thailand. From the literature, pre-operative investigations in most cases of these conjoined twins failed to define the precise anatomy of the biliary system and may be misleading as in the presented case. The mortality rate remains high. Meticulous pre-operative planning, decision-making in the operative field and postoperative management as well as a multidisciplinary team are very important for a successful separation.

Keywords: *Conjoined twins, Siamese twins, Thoracopagus, Biliary system, Intussusception, Separation*

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Separation of conjoined twins remains one of the greatest challenges to surgeons and multidisciplinary teams. The authors herein report a successful separation of thoracopagus conjoined twins with a single extra-hepatic biliary system at Khon Kaen Regional Hospital. This was the first reported case of thoracopagus conjoined twins with biliary tract anomalies in Thailand. The authors approach to the problem was described and the literature was reviewed for other's experiences.

Case Report

A 29-year-old woman, gravida 2, para 1, was referred from a district hospital with polyhydramnios and twin pregnancy at 38-week gestation. On August 11, 2004, male thoracopagus conjoined twins were delivered by emergency caesarean section with a total weight of 5,590 gm. They were joined from the chest wall down to a common 6 x 8 cm omphalocele with an umbilical cord containing one vein and four arteries (Fig. 1). Pre-operative investigations showed that the sternum of each twin was joined in the lower part. Upper gastrointestinal study showed separate stomachs. There was a connection between the second part of the duodenum of each twin. Barium enema showed that they had their own colons. The study was not able to demonstrate the joining part of the lower gastrointestinal tract. Echocardiography and computed tomographic (CT) angiography showed separate hearts, fusion of the livers and good demarcation of the vascular

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supply to the livers. Each baby had two well functioning kidneys and one urinary bladder. Color Doppler ultrasonography (US), magnetic resonance imaging (MRI) and magnetic resonance cholangio-pancreatography (MRCP) of the abdomen revealed two hepatic veins, two portal veins, two common bile ducts and one gall bladder. The pancreas could not be identified. From the investigations, separation of extra-hepatic biliary systems was assumed. Since this was the first pair of conjoined twins at the provincial tertiary care hospital, the authors set up a conference and invited experts who had experience in conjoined twins to participate in discussions of the case. It was suggested that the twins should be separated at the age over 3 months so they would be strong enough to tolerate the major operation. The authors arranged a special room and nursing staff to care for the twins and their parents in Khon Kaen Hospital. Before surgery, multidisciplinary teams detailed conferences, which were held to plan the pre-operative, perioperative and post operative care.

Fig. 1 Three- day-old thoracopagus conjoined twins

The Operation

Separation of the twins was performed at the age of 3 months. The combined weight of the twins was 10.7 kg. Operative findings revealed joined pleura and diaphragm, fusion of the intestines from the second part of the duodenum to the terminal ileum with two normal colons (Fig. 2). Dual vascular supply of the common small bowel was observed. Surprisingly, a tight intussusception was found at the terminal ileum of twin B and invaginated retrogradedly into the joined jejunum. Manual reduction was performed successfully with some difficulties. Meckel's diverticulum was located at the distal portion of common small bowel and subsequently found to contain heterotropic pancreatic tissue. Each twin had his own pancreas. Fusion of the livers had an ill-defined line of demarcation. The gall bladder of twin B was not present, but there were three gall bladders in twin A. Only one extra-hepatic biliary system was noted. Intra-operative cholangiogram through one of the three gall bladders visualized only single extra-hepatic biliary anatomy in twin A and a long intra-hepatic duct of twin A joining the intra-hepatic duct of twin B who had no extra-hepatic bile duct (Fig. 3). The gastrointestinal tract and the liver were separated. The authors chose to give the entire common small bowel (75 cm) to twin B as there was sufficient bowel length to construct a Roux-en-Y enteric loop for biliary drainage of twin B. So twin A's duodenum was anastomosed to the distal small bowel, which was in continuity with his colon. Twin B's duodenum

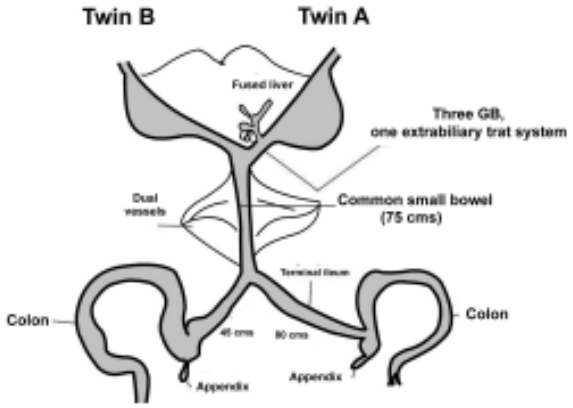

Fig. 2 Diagram of operative findings

was in continuity with the upper small bowel to his terminal ileum and colon. One area of good bile drainage at the cut surface of twin B liver was noted. It was anastomosed with a Roux-en-Y enteric loop. The diagram of operative anatomy of both twins after gastrointestinal tract and liver separation is shown in Fig. 4. Primary closure of twin B's abdominal wall was performed successfully. However, the abdomen of twin A was closed with Gore-Tex dual synthetic fascial

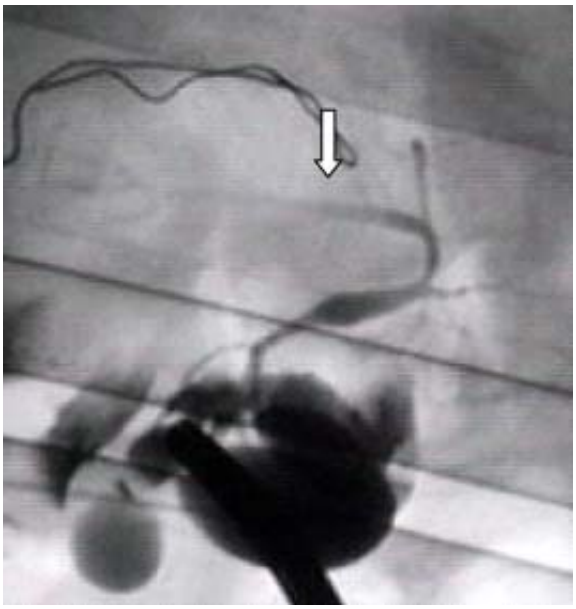

Fig. 3 Intraoperative cholangiogram through one gall bladder, showed a long intrahepatic duct of twin A to join the intrahepatic duct of twin B (Arrow)

substitute mesh and bilateral musculocutaneous skin flaps. Total operative times were 16 and 19 hours for twin A and B respectively. The operation teams consisted of 8 surgeons (4 pediatric surgeons, 2 plastic surgeons, and 2 general surgeons), 4 anesthesiologists, 10 anesthetic nurses, 30 scrub nurses, and other paramedical personnel.

Progress in the postoperative period and follow up

The post operative course was hectic. Immediate post operative complication of twin A was severe upper gastrointestinal bleeding. Coagulation time was prolonged. It was corrected by administration of fresh frozen plasma and cryoprecipitate. He also developed convulsion, hypokalemia, hypoalbuminemia, and septicemia. Hemoculture revealed MRSA (Methicillin-resistant *Staphylococcus aureus*). Abscess of abdominal wall and partial skin flap necrosis occurred 7 days after the separation. Pus culture from the wound revealed *E. coli* and *Klebsiella pneumoniae*. The dual mesh was removed. The wound was locally treated and eventually required a skin grafting.

Two days after the operation, twin B also developed convulsion. *Klebsiella* septicemia, hypomagnesemia, hypokalemia were found. Anterior abdominal wall wound infection was noted and accordingly drained.

Both twins recovered satisfactorily within one month. Twin B had an episode of infection possibly from ascending cholangitis. The development of twin A is normal but twin B has a slight delay. The twins were discharged from the hospital at 6 months of age (Fig. 5). Both twins are now two years after separation. They are doing well.

Discussion

Conjoined twins or "Siamese Twins" are rare congenital anomalies. "Siam" is the ancient name of "Thailand". The most well known Siamese twins were

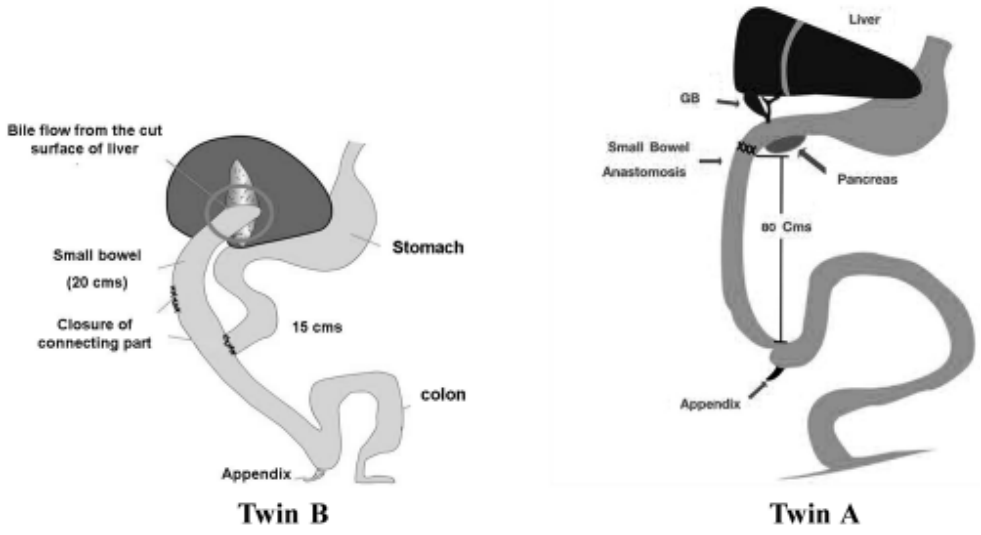

Fig. 4 Diagram of operative anatomy of twin A and B after separation

Fig. 5 Both of twins were doing well 3-months after separation

Eng and Chang Bunker, born in Thailand in 1881. The incidence of conjoined twins has generally been reported to be approximately one in 50,000 births. However, a study in Thailand reported an incidence of one in 34,000 births[1]. There have been more than 22 reported cases of successful separation of conjoined twins in Thailand so far[2-8].

Thoracopagus twins are united face to face from the upper thorax to the umbilicus with a common sternum, diaphragm, and upper abdominal wall[9-11]. In 1967, Nichols et al reviewed 42 case series of thoracopagus twins. There were a bridge of liver (100%), conjoined hearts (75%), joined intestinal tract (50%), joined biliary tree (25%)[12,13]. Hoyle[14] reported 48 sets of thoracopagus twins. The liver was conjoined in 40 (83%) cases. The small intestine was conjoined in three cases and the biliary tree in just one case. Meyers[15] reported his cases and eight additional cases of complex or conjoined biliary anatomy in conjoined twins. All of these twins had coexistent conjoining of the duodenum. It was shown that if there were no cardiac anomalies, the mortality rate was 40%. If there were cardiac anomalies, mortality rate was up to 87.5% (Table 1).

The suitable time for separation of conjoined twins are related to the condition of those twins and to the type of twins[24]. If potentially correctable life-threatening anomaly is present and the critical condition of one twin threatens the life of the other, immediate or emergency surgical correction is indicated. If the children are doing well, it is general considered to wait before carrying out the separation. On the other hand, if separation is delayed much beyond 1 year, there is some evidence to suggest that the twins may have difficulty in developing a separate identity[19].The optimal age for separation is 3-12 months[3], allowing for growth and physiological stabilization. A delayed separation was clearly beneficial. The risk of surgery and anesthesia are lower[13]. It also provides time for pre operative assessment and multidisciplinary teams' preparation.

Pre-operative investigation for thoracopagus twins, the severity of the cardiac abnormality determines the prognosis, survival, and feasibility of separation and requires accurate assessment. Cardiopulmonary assessment is best accomplished with echocardiography and it usually should be done early as the initial investigation to establish the degree of cardiac conjunction and associated structural heart abnormalities[3,11]. Pre-operative evaluation of the liver and pancreatico-biliary system was crucial. It can be performed with US, contrast-enhanced CT, MRI, and

Table 1. Reported cases of complex biliary anatomy in conjoined twins

Authors	Biliary Tract Anatomy	GB	Twin A	Twin B
Simpson (1969) [13]	Single bile duct to twin A GB, atresia of other bile duct	2	D*	D*
Micheli (1978) [14]	Single CBD	2	D*	D*
Saing (1987) [15]	Single extrahepatic biliary system	2	S	D
O'Neill (1988) [16]	Single biliary tree	2	D	D*
Lobe (1989) [17]	Single CBD	2	D	S
Cywes (1997) [18]	Single CBD	2	S	D*
Spitz (1997) [19]	Single bile duct to twin A GB, atresia of other bile duct	2	S	S
Jaffray (1999) [20]	Separate CBD in to joined duodenum	3	D	D
Meyers (2002) [12]	Twin A, proximal biliary atresia Twin B, biliary hypoplasia	1	S	S
Present report	Single extrahepatic biliary system	3	S	S

S = Survival , D = Death , D* = Death, associated with cardiovascular anomalies
CBD = Common bile duct, GB = Gallbladder

MRCP. These investigations will give information regarding hepatic vessels, the intra- and extra-hepatic biliary tree, the number of gallbladders, and whether the pancreases are conjoined or separate. Demonstration of separate hepatic venous drainage into the inferior vena cava and right atrium of each twin is especially important, as absent or anomalous hepatic venous drainage in one twin is incompatible with survival after surgery[19]. The pre-operative investigation in the present case failed to define the precise anatomy of the biliary system. Although preoperative Technetium 99m Hydroxyiminodiacetic acid (^{99m}Tc-HIDA) scan has been recommended to clarify the biliary anatomy in twins with conjoined liver, it is not always helpful[15,20-22]. This emphasizes the need for careful dissection and surgical ingenuity at the time of separation[22].

The methods of operation for the cases that have complex biliary anatomy in conjoined twins remain a challenging exercise for any surgeon. The biliary treatment strategies include control of external biliary drainage from the cut surface of the liver, hepatico-duodenostomy, Roux-en-Y choledocho jejunostomy, hepaticojejunostomy or an appendix portoenterostomy[15-23]. The authors chose a reconstruction with anastomosis of a Roux-en-Y enteric loop to one area of good bile drainage at the cut surface of one twin's liver because there was sufficient bowel length to construct a Roux-en-Y. Bile from the liver can drain directly into the proximal small bowel.

Abdominal wall closure is usually difficult in thoracopagus twins. The use of tissue expanders has some advantages that there would be adequate abdominal wall coverage but it needs an additional operation. It also has the risk of infection and ischemic compromise of the skin[13,24-26]. Tissue expanders were not used in the presented case because preoperative assessment of the amount of shared surface area suggested that a primary abdominal closure could be successfully accomplished. The authors planned to use synthetic mesh facial substitutes if the abdominal wall was closed with tension.

The post operative period was very important. The twins were cared for in the intensive care unit, each with one nurse and one pediatrician. The total cost of this surgical separation was 1.7 million Baht (about 40,000 USD).

When the twins were discharged from the hospital, the authors contacted the district hospital, local administration, and private organizations to support their family not only with health considerations but also with the economic problems.

The experience in the presented case suggested that conjoined twins could be successfully separated at a provincial tertiary care hospital in Thailand. Meticulous pre-operative planning, appropriate decision-making in the operative field, and post operative management as well as a multidisciplinary team are very important for a successful separation of conjoined twins.

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การผ่าตัดฝาแฝดสยามชนิดที่ตัวติดกันบริเวณหน้าอกส่วนล่างและหน้าท้องส่วนบนและมีทางเดินน้ำดีภายนอกตับชุดเดียว

สุรชัย สราญฤทธิชัย, รังสรรค์ นิรามิษ, สุรชัย ศิริพรอดุลศิลป์, วิเชียร เทียนจารุวัฒนา, ธานินทร์ หอมปลื้ม, นคร ทิพย์สุนทรศักดิ์, อักษร พูลนิติพร, ศิริจิตต์ วาสนะวัฒน, วิทยา จารุพูนผล

ฝาแฝดสยามชนิดที่ตัวติดกันบริเวณหน้าอกอายุ 3 เดือนได้รับการผ่าตัดแยกร่างที่โรงพยาบาลขอนแก่นเมื่อวันที่ 19 พฤศจิกายน พ.ศ. 2547 จากการตรวจโดยเครื่องมือต่าง ๆ ก่อนการผ่าตัด พบว่า มีหัวใจแยกกัน มีลำไส้เล็กส่วนต้นร่วมกัน มีตับติดกัน มีทางเดินน้ำดีแยกกัน แต่จากการผ่าตัดพบมีการเชื่อมต่อลำไส้เล็กส่วนต้นถึงลำไส้เล็กส่วนปลายลำไส้ใหญ่แยกกัน พบลำไส้เล็กกลืนกัน ตับติดกัน และมีทางเดินน้ำดีภายนอกตับเพียงชุดเดียว ได้รับการผ่าตัดแบ่งลำไส้และตับ โดยทางเดินน้ำดีได้แยกให้แฝดคนหนึ่ง ส่วนแฝดอีกคนได้ผ่าตัดโดยใช้ลำไส้เล็กส่วนหนึ่งครอบบริเวณตับที่ผ่าแยกกัน และเป็นตำแหน่งที่มีน้ำดีซึมออกมา ขณะนี้หลังผ่าตัด2 ปีแฝดทั้งสองคนสมบูรณ์ดี

ฝาแฝดสยามคู่นี้เป็นคู่แรกของประเทศไทยที่มีทางเดินน้ำดีภายนอกตับชุดเดียว จากรายงานของต่างประเทศการตรวจวินิจฉัยก่อนผ่าตัดในฝาแฝดสยามชนิดนี้ จะไม่สามารถบอกลักษณะทางกายภาพของทางเดินน้ำดีได้ชัดเจนและมีโอกาสวินิจฉัยผิดพลาดได้ อีกทั้งการผ่าตัดยังมีอัตราการเสียชีวิตที่สูง ดังนั้นการวางแผนก่อนการผ่าตัดอย่างละเอียดถี่ถ้วน การตัดสินใจขณะผ่าตัด การดูแลหลังผ่าตัดรวมทั้งการประสานงานและความร่วมมือกับผู้เกี่ยวข้องและการทำงานเป็นทีม มีส่วนสำคัญอย่างมากต่อความสำเร็จในการผ่าตัดแยกร่างฝาแฝดสยามคู่นี้