



## Separation of conjoined twins in a resource constraint setting – Lessons learned and implications for global surgery initiatives



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### ARTICLE INFO

#### Keywords:

Conjoined twins  
Pediatric surgery  
Liver surgery  
Quality  
Global surgery

### ABSTRACT

Conjoined Twins are rare however, such cases are often highlighted in medical literature, social and electronic media. Separation of conjoined twins is no longer considered an exceptional event in the developed world but it remains a unique challenge to pediatric surgeons working in middle and low-income countries with limited well rounded health care facilities to deal with such cases. We report successful separation of a set of Conjoined Twins- Thoraco-Omphalophagus in a resource constraint setting to highlight lessons learned and its possible implications to global surgery initiatives.

### 1. Introduction

Birth of conjoined twins (CTs) is a rare (incidence: 1 in 50,000 births) event globally and these are well documented in English literature since early human history [1,2]. As 60% of the cases are still-born and many dies within few hours of birth, stated incidence is approximately 1 in 200,000 births. There is predilection for females (male: female ratio 3:1). CTs are reported more often in Central African, South-East Asian and Latin American population [1–3], and are classified according to site of fusion e.g. Thoraco-Omphalophagus, joined on front from lower chest to umbilicus, liver and variable portion of gastrointestinal tract is shared. The pericardium may be common, but heart is rarely shared [1].

Separation of CTs is no longer considered an exceptional event in the developed world [1,2,4,5]. However, it remains an ultimate challenge to pediatric surgeons and anesthesiologists working in the developing world; where access to care at a well-rounded health facility is sporadic. We report first successful separation and medium range outcome of a set of CTs-Thoraco-Omphalophagus at our university hospital (second in Pakistan) [6], to highlight challenges, lessons learned and its implications to global surgery initiatives.

### 2. Case

#### 2.1. Clinical parameters

A female set of CTs-Thoraco-Omphalophagus, was born to a

primigravida after a full-term pregnancy in rural Pakistan. Antenatal imaging was not performed due unavailability. Combined birth weight was 3.1 KG. The babies were managed at home after the delivery and self-referred to the Aga Khan university hospital, Karachi (AKUH.K), Pakistan, at the age of 4-months. Both (Twin A and Twin B) were facing each other and joined from lower chest to umbilicus weighed 9.1 Kg and their feeding was appropriate for age (Fig. 1). They were active, playful and would pull at each other during activity, and as a result developed pectus carinatum. Twin A had a systolic murrer on auscultation. Rest of examination including extremities, spine, genitalia and anal region was unremarkable.

#### 2.2. Laboratory and imaging

The twins were admitted for radiological studies and assessment to ascertain if optimization was required. Routine hematological and laboratory investigations were normal. ECG showed independent hearts with variable rates. Conduction times and rhythms were normal. An Echocardiogram revealed separate hearts, fused pericardium and Tetralogy of Fallot (TOF)-patent foramen of ovale (PFO), 6 mm Perimembranous VSD and overriding Aorta (pink TOF) in Twin A and PFO in Twin B.

US abdomen showed fused livers, normal gall bladders, common bile ducts, spleens, kidneys and urinary bladders and on contrast study gastrointestinal tract were separate. A contrast enhanced CT scan of abdomen under general anaesthesia (management in radiology suit is reported in literature by anaesthesia team) [7], showed fusion from

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Fig. 1. Photograph and CT Scan of Twin A and B, showing the extent of fusion.

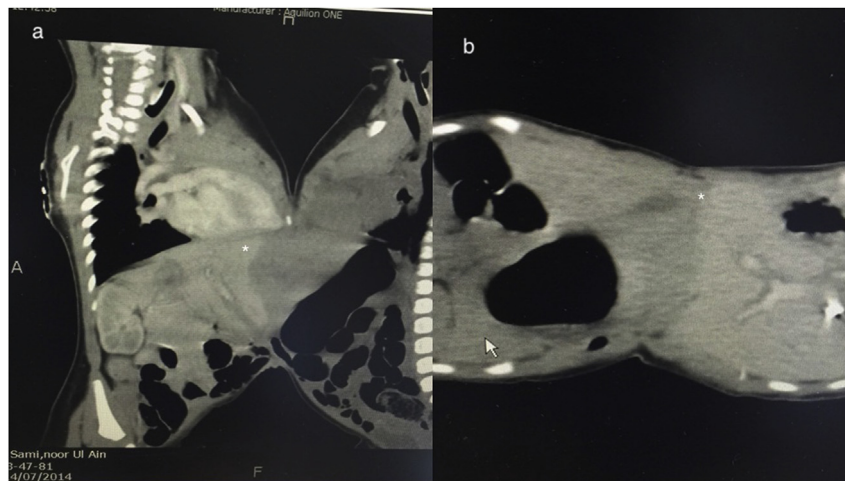


Fig. 2. Contrast CT Scan showing fusion of livers and cleavage line.

lower chest to umbilicus and no flow of contrast across the midline. Abdominal organs appear to be separate except fusion of livers (Fig. 2).

### 2.3. Preoperative preparation

Two teams including of pediatric surgeons, anesthesiologists, nurses, OR personals, Pediatric intensivists and a coordinator were constituted for the surgical separation. Regular meeting of the teams was held to discuss the clinical condition and anatomy as assessed on imaging and candid input was sought from the members to plan surgical procedure, anaesthesia and postoperative care. A day before the procedure, the plan was piloted using neonatal manikins. A total of 45 health care workers participated in the drill. An informed consent was administered to the parents after explaining the details of procedure, morbidity, mortality and success rates. Meanwhile, social and financial support was extended from the respective hospital departments and the public affair department handled the media to ensure privacy of twins and parents.

The twins were electively admitted a day before the procedure and evaluated by surgery, anaesthesia, pediatric intensive and cardiology teams. The twins were separated in a specifically organized operating room equipped with two set of operating tables, Anaesthesia machines, surgical instruments and monitoring equipment.

### 2.4. Surgical procedure

The twin was administered endotracheal anaesthesia. Anaesthesia managment is already reported in literature [7,8]. Foley's Catheter, Arterial and central venous lines were inserted and color coded to allowed independent monitoring of vital signs and fluids and broad-spectrum antibiotics (Amikacin and Augmentin) were administered. Incisions were marked to distribute sufficient abdominal wall to each twin to facilitate primary closure. We started posteriorly and deepened the incision to peritoneum. We stopped at this point and covered the wound with sterile dressing and repositioned the twins to approach anterior abdomen and chest. Incision was given from lower chest to umbilicus to enter common peritoneal cavity. Right and left lobes of livers were fused but there were two independent set of biliary, gastrointestinal and genitourinary tracts. Liver fusion was 5 cm long and 4.5 cm thick. With the help of preoperative contrast enhanced CT scan and intraoperative findings, the transection line was marked between the two gallbladders and bile ducts and the livers were encircled with two Penrose drains<sup>R</sup> (Bard Medical Division, Covington30014, GA) approximately 2 cm lateral to the transection line as described by Yang et al. [9] We preferred rubber drain over cotton straps as these are stretchable, easy to secure and provide batter hemostasis. The liver was transected with bipolar diathermy (Velleylab, Boulder, CO, US). Large blood vessels and bile ducts were suture ligated with 5/0 vicryl<sup>R</sup>

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