Minimally conjoined omphalopagi: emphasis on embryogenesis and possibility of emergency separation

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Minimally conjoined omphalopagus twins (MCOTs) has been recognized in the last decade as a special subgroup in which omphalopagus twins have union of peritoneal cavities through anterior lower abdominal wall defect with union of distal small intestine and patent urachal structures and associating anorectal malformation.

A careful review of the current literature revealed that MCOTs have usually been separated in emergency situations within the first hours of life due to ruptured omphalocele, gastroschisis, stillbirth of one of the twins, intestinal obstruction, or requirement of enterostomy for cloacal anomaly.

Pediatric surgeons should be familiar with MCOTs and ready for emergency separation with thorough knowledge of the anatomical relationships of the connecting structures and the embryologic basis for this anomaly. We present a new set of MCOTs separated in emergency conditions with a review of the relevant English literature. We give special emphasis to the common surgical characteristics and a brief discussion on the embryogenesis of this rare condition.

Key words: conjoined twins, omphalopagus, anorectal malformation, imperforate anus, cloacal anomaly, minimally conjoined omphalopagus.

Conjoined twinning is one of the rarest congenital anomalies, with an incidence of one in 50,000-100,000 births, and is one of the greatest challenges in modern pediatric surgery. It is predominantly encountered in females in live births. Conjoined twins (CTs) have been reported to be recognized as monozygotic, monochorionic and monoamniotic twins of the same sex with identical chromosomal patterns. Although they are classified according to site of attachment as thoracopagus, omphalopagus (or in combination, thoracoomphalopagus), pygopagus, ischiopagus and craniopagus, every pair of conjoined twins represent a unique event.

Omphalopagus comprises up to 10% of all cases and is usually in the form of thoracoomphalopagus[1,2]. Minimally conjoined omphalopagus twins (MCOTs) presenting with union of the infraumbilical abdominal wall without union in the perineum or the bony pelvis has been described in the last decade[3,4]. Union of peritoneal cavities through abdominal wall defect, union of distal intestinal tract, union of patent urachi, and anorectal malformation were the main abnormalities in these cases. We present a new set of MCOTs and review the relevant medical literature to determine the common surgical characteristics, and to speculate briefly on the embryogenesis of this rare anomaly.

Case Report

Female omphalopagus CTs were admitted to the emergency unit following birth to a 27-year-old female (gravida 1, para 1). The delivery occurred following a 30-week gestation by vaginal route in a rural hospital. There was no positive history for drug exposure during pregnancy or congenital anomalies in the family. An obstetrical ultrasonography had revealed a twin pregnancy but not evidence of CT.

One of the twins was dead (Twin B) at admission and the other (Twin A) was pale, cyanotic and bradycardic as in a septic condition. The combined weight of the twins was 2550 g. Both had large defect in the abdominal wall and both were conjoined through a ruptured large edematous omphalocele sac (Fig. 1). Two separate umbilical cords, each containing three vessels, were attached to the sac. Both twins had imperforate anus and single perineal outlet without separate urethral and vaginal openings, which were consistent with cloacal anomaly. Emergency separation was decided because of the deteriorating condition of the living twin and the presence of a dead sibling and ruptured omphalocele.

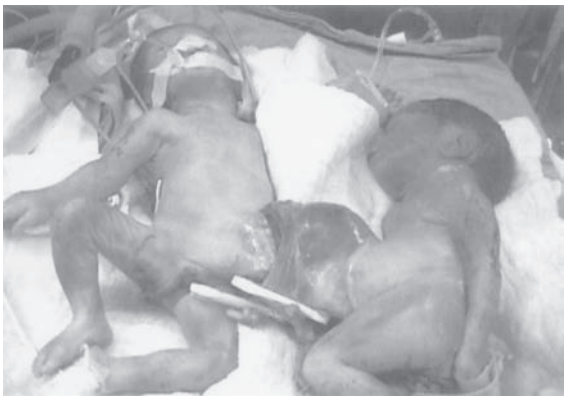

Fig. 1. Minimally conjoined omphalopagus twins (Right: twin A, Left: twin B).

The living twin was intubated. The ruptured area of the omphalocele sac was enlarged. There were several loops of small intestine and the liver of Twin B in the sac. Twin A had an apparently normal intestinal tract with small intestine and colon, which was opening into the cloacal cavity through the posterior wall (Fig. 2). Twin B had a normal length of small intestine and its terminal ileum was joined to the terminal ileum of Twin A at a point 10 cm proximal to the cecum. Twin B had no colon. There was a bridging tubular urachal structure in the anteroinferior part of the omphalocele sac, extending from the dome of one bladder to the dome of the other. Both twins had bicornuate uteri, two Fallopian tubes, two ovaries and a single vagina.

The bridging urachal structure was divided, dissected up to the bladder and excised. The bladder was closed. The terminal ileum

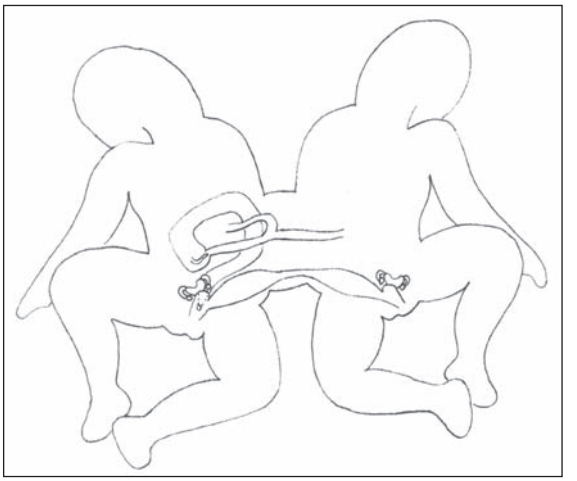

Fig. 2. Schematic drawing shows bridging structures: omphalocele sac, intestinal and urachal connections, and cloacal malformation (Right: twin A, Left: twin B).

of Twin B was separated at the attachment point to the ileum of Twin A. The hole in the ileum of Twin A was closed in two layers and an end-colostomy was created in the left lower quadrant. The liver, spleen and kidneys appeared normal. The abdominal wall was closed without tension. Twin A deteriorated and died postoperatively within three hours.

Discussion

Various names have been used to describe this association, such as minimally conjoined omphalopagus, minimally united ischiopagus, minimally conjoined twins and vitellopagus[3-6]. We identified 13 sets with MCOT through evaluation of only original articles in the literature[3,5-15]. We reviewed them systematically and included the present set (Table I).

Most of the cases were as one twin set except two sets reported by Poenaru et al.[3]. Female gender was encountered in 70% of twin sets, similar to the gender rate of the general CT population. The types of placentation in human monozygotic twinning are monochorionic-diamniotic (60-70%), dichorionic-diamniotic (30-40%) and rarely monochorionic-monoamniotic[1]. Although human CTs are generally believed to present with monochorionic-monoamniotic placentation (the rarest type), there are some examples of diamniotic placentation in MCOTs[5,6,13,14]. The umbilical cord may be single, double or forked and may contain various numbers of vessels. Since the data including the type of placentation and shape of cord is lacking in most

Table I. Demographic and Macroscopic Features and Indication for Separation of MCOT

Set no.	Year, author	Sex	Placenta	Umbilical cord	Vessels in the umbilical cord	Timing and indication for separation
1	1964, Riker[7]	M	–	Single	–	ES, intestinal obstruction
2	1964, Cywes[8]	F	MC, MA	Single	2A	ES, stillbirth and ruptured omphaloceles
3	1968, Gans[9]	F	–	–	–	ES, ruptured omphaloceles
4	1982, Votteler[10]	M	–	–	–	ES, severe malformation and ruptured omphaloceles
5	1990, Weston[5]	M	MC, DA	Forked	2/3	ES, hemorrhage and trauma to bridging ileum
6	1991, Walton[11]	F	Single	Double	2A+1V/2A+1V	ES, gastroschisis
7	1993, Zoppini[12]	F	–	Single	–	Medical abortion
8	1994, Poenaru[3]	F	–	Single	–	ES, ruptured omphalocele
9	1994, Poenaru[3]	F	–	–	–	ES, ruptured omphalocele
10	1994, Kapur[13]	M	MC, DA	Double	2A+1V/2A	ES, early postnatal death of one of twin pairs
11	1997,Gold/fischer[14]	F	DA	Double	3/3	Elective separation
12	1998, Koltuksuz[15]	F	–	Single	4A+2V	ES, intestinal obstruction
13	2005, Charles[6]	F	MC, DA	Single	1/1	Premature delivery (both stillborn)
14	2006, Karnak*	F	–	Double	2A+1V/2A+1V	ES, stillbirth and ruptured omphaloceles

Note: Dashes indicate that data were not available. *Present set.
MCOT: Minimally conjoined omphalopagus twins. M: Male. F: Female. ES: Emergency separation. MC: Monochorionic. MA: Monoamniotic. DA: Diamniotic. A: Artery. V: Vein.

reported cases, any comment may be misleading. Therefore, the anatomy of the placenta and membranes, cord and cord vessels should be carefully recorded during delivery of CTs.

A delayed separation is usually recommended in CTs. This approach provides time for detailed evaluation of the anatomy, decreases the risk associated with anesthesia and allows planning of surgical strategies. However, emergency separation may be indicated in twin sets with a stillborn or a just dead pair, ruptured omphalocele or gastroschisis, trauma to bridging structures, intestinal obstruction, obstructive uropathy, congestive heart failure, respiratory embarrassment and when compromising one twin versus the other. Interestingly, all but one case among MCOTs required emergency separation due to ruptured omphalocele, stillbirth or death of one of twins, hemorrhage and trauma to intestinal bridge and intestinal obstruction. We think frequent association of ruptured omphalocele was due to lack of awareness of the conjoined status of the twins. MCOTs may have a flexible connection and can move independently in some extent. This might result in difficulty in recognizing the conjoining structures at the prenatal ultrasonography. Pediatric surgeons should be familiar with the anatomical features of MCOTs and be prepared for emergency surgery. Furthermore, all MCOTs have an anorectal malformation and necessitate diversion of fecal stream within the first few days of life. Thus, early separation would be logical in MCOTs with creation of ileostomy or colostomy in each.

The detailed description of associating abnormalities, physical findings, surgical findings and surgical procedures are given in Tables II and III. Basically, an abdominal defect, urachal anomaly, connection of distal small intestine and anorectal malformation are all components. Abdominal wall defect was frequently an omphalocele with its usual characteristics. Gastroschisis, exstrophic cloacal structure and direct connection of peritoneal cavities with fusion of abdominal walls without interposing sac were encountered in a few sets of MCOTs. The closure of the abdominal wall was usually achieved without difficulty and no prosthetic material was required. Associated anomalies of other systems could have been evaluated by postmortem computerized tomography in the present case. However, the family did not give consent for postmortem investigations.

Urachal anomaly was usually a tubular bridging structure between the domes of bladders. Fusion of two patent urachi at their distal ends close to the umbilicus can be assumed. It is usually located at the lower part of the connecting sac. Urachal bridge can be divided, dissected up to bladder and excised without difficulty.

Table II. Details of Associated Anomalies in MCOT

Set no.	Sex	Abdominal wall defect	Urachal/allantoic anomaly	Intestinal connection*	Type of anorectal malformation	Appearance of perineum**	Pubic separation
1	M	Direct connection	Conjoined U	TI-TI	AA/AA+RV fistula	N male, bifid scrotum in one	NAD
2	F	Omphaloceles	Conjoined U	TI-TI	Both cloacal anomaly	Single opening in both	+/+
3	F	Omphaloceles	Conjoined U	SB-TI	Both cloacal anomaly	Single opening in both	NAD
4	M	Omphalocele	Elongated U	Ileum-ileum	AA/AA	N male in both	+/–
5	M	Omphalocele	One patent U	SB-TI	AA/AA+RV fistula	UDT and hypospadias in each	+/–
6	F	Gastroschisis	One patent U	TI-TI&	Cloacal anomaly/AA	Single opening/N female	+/–
7	F	Gastroschisis	NAD	SB-Colon	AA/AA+vaginal fistula	NAD	NAD
8	F	Omphalocele	Conjoined U	SB-TI	Both cloacal anomaly	Single opening in both	NAD
9	F	Omphalocele	Conjoined U	SB-TI&	Both cloacal anomaly	Single opening in both	+/+
10	M	Omphaloceles	Conjoined U	Ileum-ileum	AA/AA+RV fistula	N male in both	+/+
11	F	CCE	Cloacal bridge	SB-SB	Both cloacal anomaly	No perineal opening in both	+/+
12	F	Omphalocele	None	TI-TI	AA/cloacal anomaly	N female/single opening	NAD
13	F	Omphaloceles	NAD	SB-SB	Both cloacal anomaly	Single opening in both	NAD
14	F	Omphaloceles	Conjoined U	TI-TI	Both cloacal anomaly	Single opening in both	+/NAD

*A common colon was demonstrated distal to the conjunction point of small bowels in all twin sets except Set 13. **All twins had imperforate anus. &Associated with ileal atresia. MCOT: Minimally conjoined omphalopagus twins. M: Male. F: Female. CCE: Covered cloacal exstrophy. U: Urachus. TI: Terminal ileum. SB: Small bowel. AA: Anal atresia. RV: Rectovesical fistula. N: Normal. UDT: Undescended testicle. NAD: No available data. +: Pubic separation present. –: No pubic separation.

Table III. Details of Surgical Procedures, Findings and Outcome in MCOT

No, sex	Surgical procedure (Twin one/twin two)	Internal genitalia	Outcome
1, M	UE, TI, RAW/UE, ileal repair and LC, RAW	–	EX (4h)/EX (8d)
2, F	TI, urinary diversion with ileal conduit, repair of cloacal anomaly, RAW	HU, 2FT, 2O in both	One survived
3, F	UE, TI, RAW/UE, ileal repair and LC, RAW	HU, 2FT, 2O in both	One survived/EX (4d)
4, M	UE, ileostomy, RAW	–	One survived/EX
5, M	UE (?), colostomy, RAW/Ileostomy, RAW	–	Both survived
6, F	UE, TI-MF, RAW/TI, RAW	DU, 2FT, 2O in both	Both survived
7, F	No surgical intervention (medical abortion)	DU and double vagina in both	Medical abortion
8, F	UE, ileostomy, RAW/UE, colostomy, RAW	U, 2FT, 2O in both	One survived/EX (1h)
9, F	UE, ileostomy-MF, RAW/UE, colostomy, RAW	U, 2FT, 2O in both	Both survived
10, M	–	–	One survived/EX (4h)
11, F	UE, V, TI, RAW/UE, V, end-colostomy, RAW	HU, 2FT, 2O in both	Both survived
12, F	TI, RAW/ileal repair and colostomy, drainage of hydrometrocolpos, RAW	BU, HMC, 2FT, 2O in one	EX (2d)/EX (6m)
13, F	No surgical intervention (both stillborn)	2O in both	Both stillborn
14, F	UE, ileal repair and end-colostomy, RAW	BU, 2FT, 2O in both	EX (3h)/EX

Note: Dashes indicate that data were not available.
MCOT: Minimally conjoined omphalopagus twins. M: Male. F: Female. UE: Urachus excision. TI: Terminal ileostomy. LC: Loop colostomy. RAW: Repair of abdominal wall defect. MF: Mucous fistula. EX: Exitus. HU: Separate hemi-uteri. FT: Fallopian tube. O: Ovary. DU: Didelphys uterus. U: Uterus. BU: Bicornuate uterus. V: Vesicostomy. HMC: Hydrometrocolpos.

Intestinal connection was certainly at the level of the distal ileum at a point 10-15 cm proximal to the cecum. Ileal atresia close to the connection area was present in two cases[3,11]. The distal intestine was usually common and one of the twins had no colon. The ileum was usually separated at the level of connection and brought to the skin as an ileostomy. The small intestine may be short. A part of the colon can sometimes be suitable for interposition to the other twin to lengthen the intestine (one of the twins in Set 9)[3]. Therefore, a critical judgement may be required at this step before separation to avoid adverse effects of ileostomy such as fluid loss (one of the twins in Set 12)[15]. In usual cases, the hole in the ileum was closed and a colostomy was created in the other twin.

Anorectal malformation was another component. Imperforate anus was encountered in all twin sets. Males revealed anal atresia with or without fistula to bladder according to presence or absence of the colon. Males also revealed normal external male genitalia in most cases. Cloacal anomaly was encountered in most of the female twins and there was only one perineal opening in all but one set of them (Set 11)[14]. Separate vaginal and urinary openings were encountered in only two of the twins (one in Set 6 and one in Set 12) in females with single perineal opening[11,15]. Internal genitalia were hemi, duplicated or bicornuate uteri with normal Fallopian tubes and ovaries, which can be encountered in cases with cloacal anomaly. Pubic separation was encountered in one or both twins in most sets. We think it has been usually overlooked or unreported in previous cases. It is seen in exstrophic anomalies such as exstrophia vesicae or cloacal exstrophy. Therefore, observation of pubic separation should place MCOTs between cloacal anomaly and cloacal exstrophy complex.

It has been previously suggested that 40% of CTs are stillborn and 60% live-born. Only 15% of all CTs lived long enough to be candidate for separation[16]. The mortality rate of urgent or emergent operations in CTs was noted as 40-80%[17]. The rate of stillbirth was 15% and 54% of twins (excluding twin Set 7) survived after separation in MCOTs. This survival rate may emphasize a better prognosis for MCOTs.

Minimally conjoined omphalopagus twins present invariable abnormalities: infraumbilical abdominal wall defect, conjoined ilea and abnormalities of derivatives of the cloaca and the cloacal membrane itself (urachal anomaly, anorectal/cloacal malformation). The observation of this spectrum indicates that union of both embryos should be in the area around the allantois and caudal portion of yolk sacs[4,18]. Since the normal development of the cloacal membrane requires a delicate environment and sensitive cellular interactions, fusion of the allantois and/or occurrence of lower abdominal abnormality may prevent normal cloacal development. The mechanism may be due to the simple suspended position of the allantois resulting in both prevention of midline closure of the abdominal wall and disturbance of the cloacal membrane. The union of the intestinal tract may also produce some augmentation on this anatomical barrier.

Minimally conjoined omphalopagus twins present with union of peritoneal cavities through abdominal wall defect, conjoined urachal anomaly, union of ilea and anorectal malformation, namely, anal atresia with or without fistula, or cloacal anomaly. In contrast to the general belief about CTs, diamniotic placentation has been recognized in some sets of MCOTs. Separation would be performed in emergency situations in presence of definite indications, or within the first days of life because of the need for diversion of fecal stream for accompanying anorectal malformation. The intestinal tract should be separated carefully with creation of ileostomy and colostomy, one in each twin. Division of urachal bridges and closure of abdominal wall defects do not present difficulty. Pubic separation may be encountered. The prognosis is better than that of the other CT types. The embryological basis is still obscure. However, urachal and intestinal unions may cause mechanical prevention of midline closure of the abdominal walls or they may disturb normal cloacal development by creating upward traction on the cloacal membrane.

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