

Minimally Conjoined Omphalopagus Twins with a Body Stalk Anomaly

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Am J Perinatol Rep 2015;5:e124–e128.

Abstract

Keywords

- body stalk anomaly
- intestinal conjunction
- minimally conjoined omphalopagus twins

Introduction This report will discuss a case of minimally conjoined omphalopagus twins (MCOTs) with a body stalk anomaly (BSA).

Case Report We experienced monochorionic diamniotic (MD) twins born at 31 weeks. One infant was suspicious of BSA before birth, and another infant was normal. But normal infant had anal atresia with small intestine which was inserted behind the umbilicus. Twins had very short common umbilicus and infant with BSA had intestinal conjunction, two appendixes at the site of the colon, and a blind-ending colon. We diagnosed MCOTs.

Discussion On the basis of the Spencer hypothesis, the etiology of MCOTs was that MD twins shared a yolk sac. However, this could not explain the presence of a BSA. It is necessary to consider the possible reasons for a singleton BSA. In addition, intestinal fusion occurred unequally in this case, although two appendixes were found in the same place, which might have occurred because of the balanced fusion.

The incidence rate of conjoined twins is estimated to be one in 50,000 to 100,000 births.¹ Among conjoined twins, 10% are omphalopagus, with some being minimally conjoined omphalopagus twins (MCOTs), in which the twins share a common umbilicus without thoracopagus.²

A body stalk anomaly (BSA), characterized by the presence of a major abdominal wall defect, severe kyphoscoliosis, and a rudimentary umbilical cord, is reported in about 1 per 14,000 births.³ This report details the case of MCOTs with a BSA, with the written informed consent of the twins' parents.

Case Report

A 32-year-old mother (gravida 1, para 1; no consanguinity) became pregnant naturally with monochorionic diamniotic

(MD) twins. At 12 weeks of gestational age, ectopia cordis and nuchal translucency were observed in one of the twins. The other twin was found to be "normal," but a large cyst was observed between the two. At 13 weeks, scoliosis and a lower abdominal mass were observed in the abnormal twin. At 19 weeks, the umbilical cords from each twin were found to be joined at the cyst and inserted into the placenta together. The cyst later resolved spontaneously. At 29 weeks, the increased uterine contractions led to the mother's admission to the hospital. At 31 weeks and 2 days, bradycardia of the "normal" twin was observed, and an emergency cesarean delivery was performed.

Twin 1 was male, with a birth weight of 1,154 g and 1- and 5-minute Apgar scores of 1 and 6, respectively. He was found

received

February 3, 2015

accepted after revision

March 5, 2015

published online

May 8, 2015

DOI <http://dx.doi.org/>

10.1055/s-0035-1549300.

ISSN 2157-7005.

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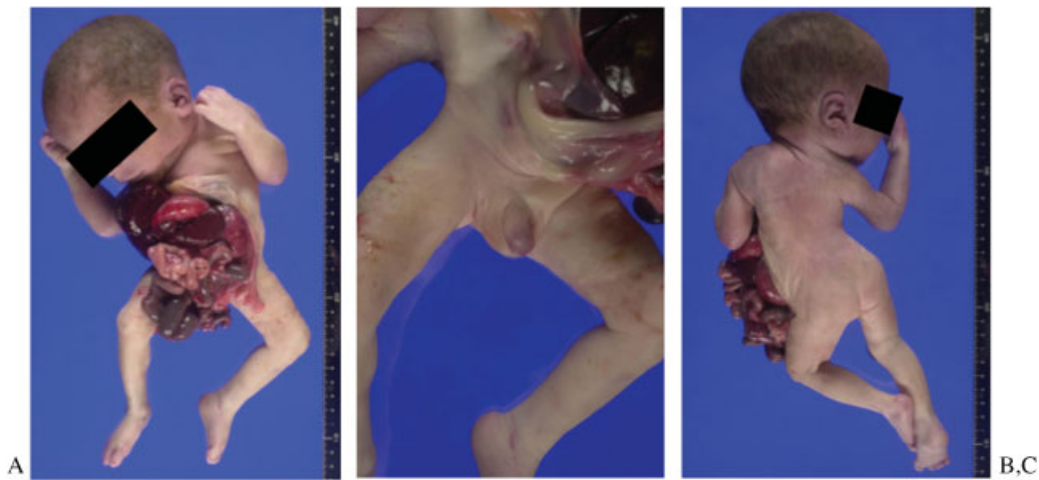


Fig. 1 Overview of the infant with body stalk anomaly. (A) Gastroschisis was found. (B) Normal male genitalia. (C) Scoliosis and anal atresia were found.

to have anal atresia, with no other abnormal findings. Surgical repair of twin 1's anal atresia was performed on day 1, by inserting small intestine behind the umbilicus, at 60 cm from the ligament of Treitz. Intestinal atresia and the absence of a colon were observed, and a jejunostomy (or ileostomy) was performed.

Twin 2 was male, with a birth weight of 1,211 g, and 1- and 5-minute Apgar scores of 1 and 1, respectively. He died 7 minutes after birth (►Fig. 1). Macroscopically, ectopia cordis, gastroschisis, diaphragmatic defect, scoliosis, adhesion of the amniotic membrane to the abdominal wall, single umbilical artery, short common umbilical cord, anal atresia, intestinal conjunction, two appendixes at the site of the colon, and

blind-ending colon were found. Twin 2 was diagnosed with BSA.

Pathological findings indicated a lung weight that was 0.4% of birth weight, indicating lung hypoplasia. There was no Meckel diverticulum present. No abnormal findings were observed in the bladder, ureter, or pubis. For intestinal conjunction, twin 1-derived colon was fused with twin 2-derived small intestine (►Fig. 2). Histologically, twin 1-derived intestine showed the characteristics of small intestine and colon. This tissue was fused with twin 2-derived small intestine. From the point of the fusion to the appendix, histology showed a mixture of small intestine and colon tissues (►Fig. 3).

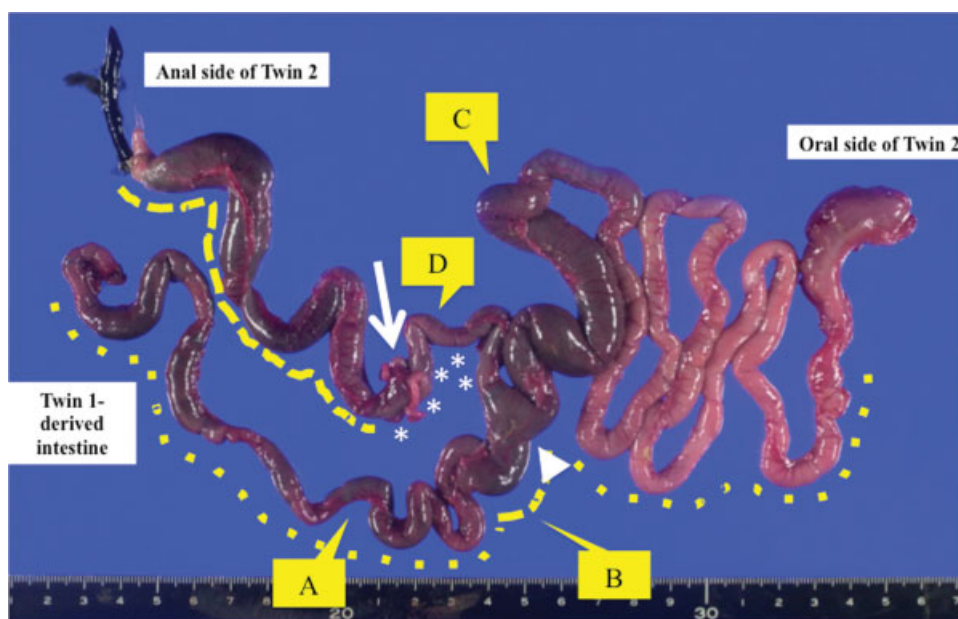


Fig. 2 Overview of the intestine. Twin 2's stomach was placed at the right side. Twin 2's intestinal tract was fused at the small bowel with Twin 1's colon (arrowhead). After that, there were two appendixes (arrow), followed by a blind-ending colon. Histological findings: small intestine, dotted line; colon, dashed line; mixture of small intestine and colon, asterisk(*). Marks A through D are the same as in ►Fig 3.

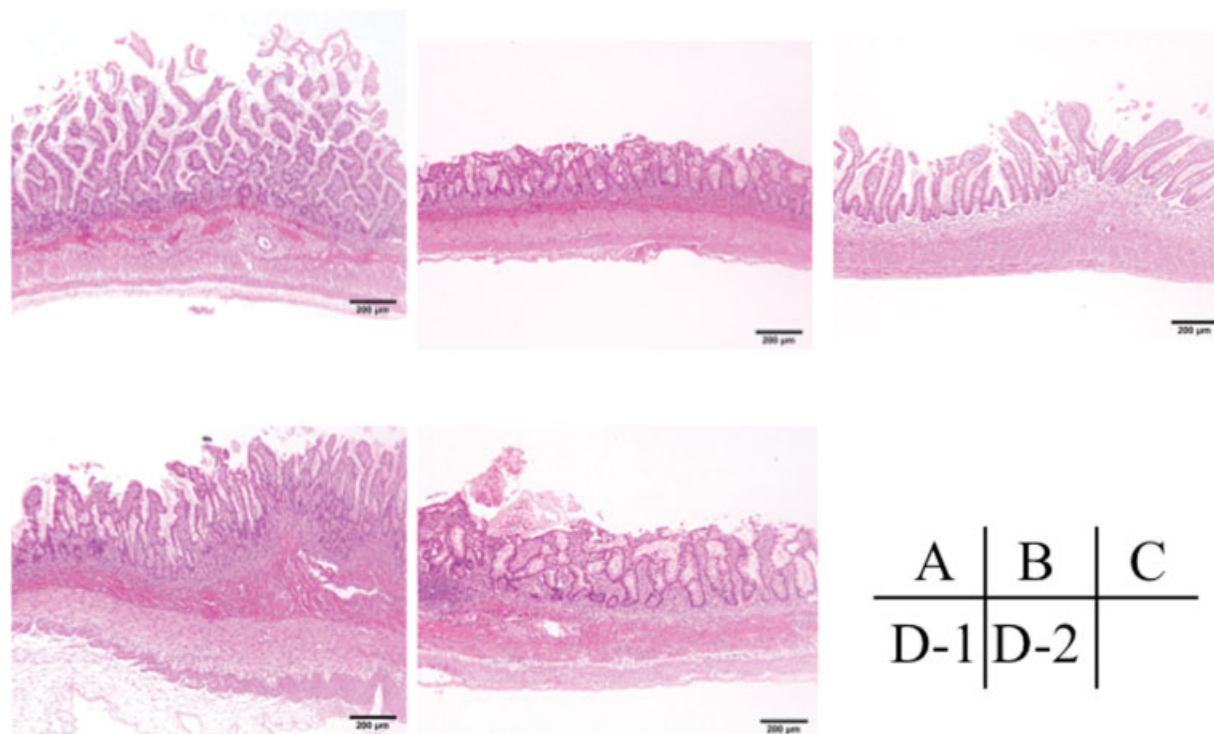


Fig. 3 Histology (hematoxylin-eosin staining). (A) Twin 1–derived intestine, with the characteristics of small intestine. (B) Twin 1–derived intestine, colon. (C) Twin 2–derived intestine, small intestine. (D) Intestine after conjunction, mixture of small intestine and colon. (D-1) small intestine. (D-2) colon.

Both umbilical cords were fused, with twin 1 having two arteries and one vein, and twin 2 having one artery and one vein. Between them was an urachal remnant, and placental findings indicated MD twins.

Discussion

This case was diagnosed as MCOTs because of the umbilical and intestinal conjunction. Some MCOTs have common urogenital sinuses and the connection of the colon to the cloaca,^{1,4–8} neither of which was found in the current case. The diagnosis of BSA was not difficult because of many characteristic features. MCOTs with a BSA would be rare. Collins documented a similar case, in which the smaller twin was observed to have severe kyphoscoliosis and a diaphragmatic defect.⁹

The hypothesis of Spencer¹⁰ posited that the etiology of MCOTs included MD twins that shared the same yolk sac. The common yolk sac would induce adjacent intestinal conjunction. The cyst observed at 12 weeks of gestational age would be a yolk sac. In the case of Walton et al,¹¹ a cystic structure was also found in the anterior place of placenta at 34 weeks of gestation. There were other hypotheses about MCOTs. Kapur et al⁴ hypothesized that the embryo consisted of a flattened disc in which two primitive streaks were established at one pole. But it could not explain MD twins. Shih et al¹² suggested incomplete separation of the monozygote and lack of a common yolk sac, but it could not explain intestinal conjunction. Of those mentioned, the hypothesis of Spencer is dominant, although it does not explain the presence of a BSA. In discussing the reasons behind the observed singleton BSA, the following three major hypotheses were raised: amnion rup-

ture,¹³ germ disc theory,¹⁴ and early generalized compromise of embryonic blood flow.¹⁵ It is also possible that the short umbilicus caused BSA.¹⁶ We could not clarify the cause of BSA.

On the basis of the twin 1's operational findings and twin 2's pathological findings, twin 1's intestine was suspected to leave the body through the umbilicus. Walton et al showed the similar omphalopagus conjoined twins.¹¹ They shared the conjoined bowel. One twin had gastroschisis and another twin's small bowel connected to the common cecum. Intestinal conjunction is almost always seen in MCOTs.^{1,4–8} In this case, a mixture of small intestine and colon histologically were observed. This could mean that both of the intestines fused at a certain length, rather than via end-to-side anastomosis. Some patterns of conjunction were reported, such as triplication,⁶ and side-by-side anastomosis.⁸ This case had two appendixes, which also has been reported previously.^{6,7} Intestinal fusion occurred unequally in this case, although two appendixes were found in the same place, which might have occurred because of balanced fusion. The mechanism of intestinal conjunction is still unclear.

Regarding the loss of twin 1's colon, hypothesis of Weston et al⁸ stated that: "This original pattern persisted in twin A but was modified in twin B by secondary loss of the terminal ileum and large bowel, due to progressive ischemia caused by their extra abdominal location and the increasing pull exerted by the returning small gut of twin A." Histology suggested that the fusion length may have resulted from the conjunction to appendixes.

In this case, twin 2 had both a colon and BSA, and died shortly after birth. Similar reports were gathered for comparison to this case (► **Table 1**). The difference between the infant with a colon and the infant without a colon was not determined. The

Table 1 Clinical characteristics of minimally conjoined omphalopagus twins

Author, Y	Appendix (es)	Intestinal conjunction	Characteristics of the twin without a colon (twin A)	Characteristics of the twin with a colon (twin B)
Weston et al, 1990	N/A	The ileum of twin A entered the abdomen of twin B to achieve a Y-shaped conjunction, just proximal to the Meckel diverticulum of twin B. There was a side-to-side fistula, with the blind gut of twin A decompressing into the ileum of twin B.	Plagiocephaly, a dysplastic left acetabulum, small descended testes, mild glandular hypospadias, anal atresia, and three vessels in cord.	Amyoplasia-related arthrogryposis multiplex congenita with plagiocephaly, hypoplasia, and diastasis of the pubic symphysis, dysplasia of the acetabula, patent urachus, right cryptorchidism, anal atresia, and two vessels in cord.
Walton et al, 1991	2	Twin B had an apparently normal small bowel that entered a common large cecum. From this, short segment of colon passed down into the pelvis behind the vagina. Twin A's small bowel connected to the cecum by a fibrous band.	Didelphian uterus. Anal atresia.	Gastroschisis. Splayed symphysis pubis. Didelphian uterus. Patent urachal structure.
Poenaru et al, 1994	N/A	The short small bowel of twin A ended in the terminal ileum of twin B. The colon of twin B had triplication area and ended in a rectovaginal fistula.	Urachal bridge, dead because of hyaline membrane disease.	Urachal bridge
	2	The shortened small bowel of twin A inserted on the terminal ileum of twin B. The intestinal junction area was vascularized primarily by an anomalous vessel believed to be the vitelline artery. The intestinal tract of twin A was lengthened with 10 cm of twin B's colon.	Urachal bridge, anal atresia	Urachal bridge, anal atresia
Kapur et al, 1994	N/A	The small intestines were largely separate except for the distal ileum, which came together to form a single shared terminal ileum, cecum, and proximal colon. The latter was not fixed, and ended in a shared persistent cloaca.	Dead, posteriorly angulated right ear, talipes equinovarus, anal atresia. A portion of the cloaca of twin A had no connection with the intestinal tract.	Talipes equinovarus
Koltuksuz et al, 1998	N/A	The terminal ileum of twin A lay in the omphalocele sac, and was joined to the terminal ileum of twin B at ~ 5 cm from the ileocecal junction.	Anal atresia, dead in 6 months.	Cloacal anomaly, hydrometocolpos, and a bicornuate distended uterus, septic shock on day 2. The colon of twin B ended at the cloaca.
Karnak et al, 2008	N/A	Twin A had an apparently normal intestinal tract with small intestine and colon, which opened into the cloacal cavity. Twin B had a normal length of small intestine with the terminal ileum joined to that of twin A at a point 10 cm proximal to the cecum. The endocolostomy was created for twin A.	Large defect of abdominal wall, anal atresia, bicornuate uterus, dead within 3 hours after operation.	Large defect of abdominal wall, anal atresia, bicornuate uterus, dead at admission. Twin B had no colon, but the colon of twin A was located in the abdomen of twin B.
Tihtonen et al 2009	2	United at the terminal part of the ileum. Colon opening was at the cloaca.	Cloacal exstrophy, omphalocele. Medical interruption of pregnancy was induced.	Cloacal exstrophy, omphalocele. Medical interruption of pregnancy was induced.
Maruyama et al, 2015 (present case)	2	The colon of twin A was joined to the small intestine of twin B.	Anal atresia, omphalocele	Body stalk anomaly, dead at birth

Abbreviation: N/A, not applicable.

presence of a colon increased the abdominal size, potentially displacing other organs. However, the infant with a colon would have the advantage of nutrient absorption.

There are many considerations for treatment of MCOTs. Poenaru et al⁶ reported on a case in which bowel tissue was transplanted to the twin who had a short bowel. Karnak et al¹ reported on a case in which the colon, which had wandered into the other twin's abdomen, was pulled back and used for a colostomy. In both the cases, maintenance of the feeding artery was important. In this case, if the twin 1-derived colon in twin 2's abdomen was fed by twin 1, this section of colon could be used for colostomy.

Conclusion

This report details a rare case of MCOTs with a BSA. Histological findings showed colon–small intestine conjunction, which was conflicted by the presence of two appendixes in the same place.

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