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Anorectal Malformations in Monozygotic Twins: An Illustration of Management of Anorectal Malformations in A Tertiary Hospital in Indonesia

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Abstract

A case of monozygotic twin boys with anorectal malformation with delayed presentation of bowel perforation. Both are managed surgically with a colostomy; after a definite diagnosis of anorectal malformation with a urethral fistula based on distal colostography, both boys underwent anoplasty with a posterior sagittal approach. Stoma closure was performed, and we found no complication during eight months of follow-up. Delay in the management of the twins was due to a lack of anal screening of the newborn. The unavailability of the neonatal intensive care unit became a significant concern. Distal colostography was performed for the definitive procedure. Despite that limitation, anorectoplasty was performed with a posterior sagittal approach. Anorectal malformations in twins are a rare case requiring thoughtful surgical care. A thorough understanding of the limitation of our resources could help in planning management for anorectal malformation, especially in twins.

Key words: anorectal malformation, sigmoid perforation, twins, limited resources

Introduction

Anorectal malformations in twins infrequently reported. Anorectal malformations are a multifactorial disorder caused by abnormal development of the hindgut, allantois, and Mullerian duct, leading to incomplete or partial urorectal septal malformations.¹

The genetic component is thought to be responsible for the etiology of anorectal malformations due to the report of familial recurrence and the association with other congenital anomalies.² We reported a rare case of anorectal malformations with urethral fistula in monozygotic twins in this article. Even though in a tertiary hospital in Indonesia, we found some commonly met limitations while managing this complex case.

Case illustration

We reported a case of naturally conceived identical twin boys born by Caesarean section from a primigravida mother with a gestational age of 36 weeks. The birth weight of the twins was 2.600 g and 2.750 g, respectively. Both showed no anus, abdominal distention, and vomiting during the presentation three days after birth. Meconium was found in the urine of both boys. The AP and lateral abdominal X-ray showed intraabdominal free air. They both suspected perforation of the hollow viscus due to anorectal malformation with the urethral fistula.

Baby A underwent surgery immediately after admission. Laparotomy was performed, we found perforation of the distal sigmoid and closed with a primary suture. The proximal sigmoid was treated as a loop colostomy. Postoperatively, he was taken care of at the neonatal intensive care unit (NICU) for four days. He was discharged on day seven postoperatively with no complications.

Baby B underwent a delay due to the unavailability of the NICU. In this newborn, intraabdominal drainage was performed, whereas ten mL of enteric juice and intraperitoneal free air was drained.

The newborns fell to sepsis with WBC 21.200/uL and a total bilirubin of 5.1 g/dL during the care. Laparotomy was performed on the seventh day after the presentation, perforated the distal sigmoid, and intraoperatively found adhesion. Adhesiolysis was performed, and the perforation was closed by suture. The sigmoid proximal to the perforation was exteriorized as a loop colostomy. The newborn was taken care of in the NICU for five days postoperatively. He was discharged on the 22nd postoperative days, despite wound dehiscence, no other complication.

These twins were screened for other associated anomalies during the admission and follow-up period, including echocardiography and abdominal ultrasound. In the screening, we found no associated anomalies.

Distal colostography proceeded on both boys four months postoperatively. Both showed the most distal parts of the rectum were below the pubococcygeal line, denoting a low level of anomaly. No fistulas were found on the examinations.

Figure 1. Postoperative illustration of anorectoplasty in twins (baby A, left and baby B, right)

The definitive surgery proceeded eight months postoperatively. Baby A weighed 7.9 kg, and baby B weighed 8 kg at the time of surgery. Intraoperatively, both urethral fistulas were ligated, and anorectoplasty proceeded with a posterior sagittal approach. Both were discharged on day six postoperatively.

At 15 months of age, both babies underwent anastomosis. Oral feeding started on POD 4, and they were discharged on postoperative days 7. No complication was reported until eight months following anastomosis.

Discussion

Although the anorectal malformation is among the most frequent congenital anomalies in paediatric surgery, occurring in 1 out of every 2000-5000 live births, anorectal malformation in twins is a rare case with approximately 3 per 10,000 twin pairs.^{1,3} The spectrum of the diseases varies from relatively minor lesions to complex anomalies, and while about one-third of the cases were isolated, the remainder associated with other congenital abnormalities.^{4,5} It is a multifactorial disorder, and the current understanding of the cause of the diseases is incomplete.

The descending process of the urorectal septum towards the cloacal membrane during the 4th and the 8th weeks of gestation arrested. The phenomenon is believed to be responsible for this anomaly.⁶ The anomaly may manifest as the only pathology or as a part of a more complex syndrome. It may affect a single individual (sporadic) or occur in more than one individual from the same family (familial), as seen in twins.⁶ While little has been reported, anorectal malformation in twins showed the genetic component of its etiologic. This complex phenomenon could result from chromosomal, monogenic, or even teratogenic syndrome.^{6,7} Several chromosomal abnormalities impairing the development of an anorectal canal and associated structures have been described associated with this anomaly.⁸ Two of the most frequent is trisomy 21 and a microdeletion of the chromosome 22q11.^{2,4}

A wide spectrum of anomalies associated with anorectal malformations had been reported, ranging from sequences, VACTERL associations, and multiple congenital anomalies.^{6,7} Twins had been reported to have higher frequencies of having associated anomalies.⁷

In this report, we presented a twin with anorectal malformation with a urethral fistula. Both were diagnosed three days after birth with suspected perforation of the hollow organ as a complication. The delay in diagnosis was attributed to a lack of policy in screening the anus of the newborn. Lack of maternal education could also contribute to ignorance of antenatal care and prenatal ultrasound to screen the anomaly, even though the prenatal diagnosis is rare.⁹ Negligence defecation status of the twins was also a factor in late diagnosis.

Even though both were diagnosed with suspicion of hollow organ perforation, only one could go immediately to the operating theatre. The limited NICU bed was a major problem in our centre, and it might describe an even more significant similar problem in the whole country. Intraabdominal drainage was performed on baby B. The procedure addressed to reduce the intraabdominal pressure and drain the faeces intraperitoneal as a temporary source control along with broad-spectrum antibiotics. He survived for ten days without an anus and underwent surgery as soon as NICU available, then was discharged without significant complications other than superficial wound dehiscence.

Distal colostography was examined to plan for definitive treatment. The study is essential to delineate the most distal part of the rectum, the distance between rectum and perineum, the relation of rectum and sacrum, the level of the fistulous tract, and the degree of faecal impaction and ectasia of the blind end of the rectum.⁹⁻¹¹ Both boys showed no

fistula in distal colostography. It might due to lack of pressure injected to open the striated muscle that keeps the rectum collapsed. The information about the distance between the rectum and perineum was valuable to determine the approach. We proceeded with the surgery with a posterior sagittal approach due to a low level of an anomaly regarding the pubococcygeal line.

A definitive procedure might be performed as early as 4-8 weeks after the colostomy procedure, assuming the baby had average growth.¹² Early definitive repair leads to less time with an abdominal stoma, minor size discrepancy between the proximal and distal bowel at the time of stoma closure, and avoidance of psychological sequelae from painful perineal surgery.¹² In our cases, the delay in a definitive procedure referred to the patient delay, which is ignorance.

Both boys had average growth and development and undergone anastomosis seven months following definitive surgery. The procedure proceeded after anal dilatation using dilator number 15. We monitor the babies for eight months and no complications such as constipation after the anastomosis.

Conclusions

Although anorectal malformation is a common anomaly in paediatric surgery, anorectal malformation in twins is a rare case requiring thoughtful surgical care. Some limitations in the management are lack of parental education, intensive care facilities, and distal colostography performed without sufficient pressure. A thorough understanding of the limitation of our resources could help in planning management for anorectal malformation, especially in twins

Disclosure

Authors declare no conflict of interest

References

1. Moore SW. Genetics, pathogenesis and epidemiology of anorectal malformations and caudal regression syndrome. In: Holschneider AM, Hutson JM., editors. Anorectal malformations in children. Berlin: Springer; 2006. p 31-48.
2. Van Gelder DW, Kloepfer HW. Familial anorectal anomalies. *Pediatrics* 1961; 27:334-336.
<https://pediatrics.aappublications.org/content/pediatrics/27/2/334.full.pdf>
3. Gangopadhyay AN, Pandey V. Anorectal malformations. *J Indian Assoc Pediatr Surg* 2015;20:10-5.
<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4268748/>
4. Herman RS, Teitelbaum DH. Anorectal malformations. *Clin Perinatol* 2012;39:403-22. <https://pubmed.ncbi.nlm.nih.gov/22682388/>
5. Cuschieri A, EUROCAT Working Group. Descriptive epidemiology of isolated anal anomalies: a survey of 4.6 million births in Europe. *Am J Med Genet* 2001;103:207-15.
<https://onlinelibrary.wiley.com/doi/abs/10.1002/ajmg.1532>
6. Martucciello G. Genetics of anorectal malformations. In: Holschneider AM, Hutson JM., editors. Anorectal malformations in children. Berlin: Springer; 2006. p 17-30.
7. Cuschieri A, EUROCAT Working Group. Anorectal anomalies associated with or as part of other anomalies. *Am J Med Genet* 2002; 110(2):122-30.
<https://onlinelibrary.wiley.com/doi/full/10.1002/ajmg.10371>
8. Marcelis C, de Blaauw I, Brunner H. Chromosomal anomalies in the etiology of anorectal malformations: a review. *Am J Med Genet Part A* 2011;155:2692-704. <https://pubmed.ncbi.nlm.nih.gov/21990113/>
9. Pandya S. Anorectal malformations. *NeoReviews* 2016;17:251-62.
<https://doi.org/10.1542/neo.17-5-e251>

10. Levitt MA, Bischoff A, Pena A. Anorectal malformations. In: Spitz L, Coran A, ed. *Operative pediatric surgery*. 7th ed. New York: Taylor & Francis Group. 2013;p508.
11. Rahalkar MD, Rahalkar AM, Phadke DM. Pictorial essay: Distal colostography. *Indian J Radiol Imaging* 2010;20(2):122-5. <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC2890919/>
12. Levitt MA, Peña A. Management in the newborn period. In: Holschneider AM, Hutson JM., editors. *Anorectal malformations in children*. Berlin: Springer; 2006. p 289-93.