A rare association of anorectal malformation with bladder agenesis, bilateral ectopic ureter, and left pelviureteric junction obstruction

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1. Introduction
Urologic abnormalities (UA) have been reported in the literature but not bladder agenesis. The occurrence of genitourinary anomalies has been reported in association with numerous pathologic alterations classified as vertebral/skeletal, genital, gastrointestinal, and miscellaneous. The combination of symptoms is nonrandom. These malformations are presumed to result from a disturbed migration of abnormal hindgut development and to originate somewhere between the 4th and 10th weeks of gestation during early embryogenesis. Although a combination of symptoms associated with genitourinary anomalies and anorectal malformation (ARM) has been reported, there is no report of the bladder agenesis (BA) associated with bilateral ectopic ureters (EUs) and left PUJ obstruction in a newborn with ARM (imperforate anus).

2. Case presentation
A 1-day-old female infant was admitted to the Selcuk University Meram Medical Faculty Department of Pediatric Surgery with the diagnosis of ARM (imperforate anus and the vagina was normal). An abdominal exploration was performed after a physical examination and radiographic investigations. BA, bilateral UEs, and left pelviureteric junction (PUJ) obstruction were determined. The sacrum, spine, and right kidney were normal. No other associated gastrointestinal anomaly or tethered cord was identified. The parents were not related and the mother was not taking drugs during pregnancy. During surgery, bilateral ectopic ureters were found connected to the vagina (Fig. 1); bilateral cystic ovaries were identified. Colostomy was performed in newborn period, posterior sagittal anorectoplasty at the age of 6 months. Ileoceleal segment was considered as a good continent reservoir and thus a new bladder was performed at the age of 15 months. Ileal and colonic ends were anastomosed. To save the functioning bilateral kidneys, distal ends of both ureters were ligated and dissected from vagina. Excision of stenotic left (the left ureter was in the retroiliac position before pyelopyelostomy) ureter was performed and right ureter was anastomosed to the ileal end of ileocecal continent reservoir after pyelopyelostomy. The appendix was connected to the cecum with antireflux procedure and with its blood supply preserved. Mitrofanoff procedure was performed after anastomosing apex of appendix to umbilicus (Fig. 2). Postoperative evaluation documented no stone, infection, and malignancy in the continent reservoir. Fecal continence of the patient, who has fecal incontinence, was carried out with intestinal management programme. The patient followed up at 3 month intervals for 12 years and kidney function was moderately preserved (Urea: 76 mg/dl and creatinine: 3.22 mg/dl).

3. Discussion
Bladder agenesis has never been reported in the literature, and the incidence of such as a urologic abnormality has been estimated to be 1 in 10,000 to 1 in 50,000 live births. Variants of UA include pubic diastasis, female epispadias, male epispadias, superior vesical fistula, duplex exstrophy with imperforate anus, and cloacal exstrophy together with ARM. Our case is another example of an associated malformation in the newborn with bladder agenesis and raises again the discussion.*

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of possible etiologic factors. This may show that a solitary explanation for the pathogenesis is difficult to define. The one possibility of a disturbed migration of abnormal hindgut development will likely answer the questions concerning the etiology of both BA and EUs.

Ileocecal segment has been constructed to perform a good conduit and thus a new bladder. While ileocecal segment may be an alternative to the conventional ileal segment procedure, we performed the excision of stenotic left ureter after pyelopyelostomy in this case because the functionality of the left kidney was uncertain. Therefore, we think that the ileocecal segment should be preserved as a continent reservoir during the colostomy procedure in newborns with BA, EUs, and PUJ obstruction even though it is rarely seen. Thus, an artificial ileocecal segment continent reservoir may be necessary as a urinary tract reconstruction in newborns with a BA, EUs, and PUJ obstruction.

Fig. 1. The appearance of the bilateral ectopic ureters connected to the vagina, left pelviureteral junction (PUJ) obstruction, and bilateral cystic ovaries.

Fig. 2. The ileal end of ileocecal conduit after pyelopyelostomy, the radix of appendix was connected to the cecum, Mitrofanoff procedure (anastomosing apex of the appendix to the umbilicus).

4. Conclusion

Ileocecal segment has been constructed to perform a good continent reservoir and thus a new bladder in congenital genitourinary anomalies.

Declaration of interest

There are no conflicts of interest.

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