Intestinal obstruction is the most common finding in patients with congenital intestinal atresia. Colonic atresia accounts for only 1.8-5% of intestinal atresia with the reported incidence of 1 in 66 000 births.\(^1\)\(^2\) Mortality rate of colonic atresia, which increases with delayed treatment, is 25.7%.\(^3\) Colonic atresia is mostly located in the right colon.\(^3\) Like other intestinal atresia colonic atresia is classified as: Type I, membranous atresia with continuous intestinal wall; Type II, blind ending intestines separated by fibrous cord (no continuous intestinal wall but intact mesentery); Type III, blind ending intestines with mesenteric defect between two ends; Type IV, multiple atresias.\(^4\) Most of the colonic atresia is type III (60.4%). The rate of type I (membranous) atresia is 15% and type IV (multiple) atresia is 10% among all colonic atresia.\(^3\)

Only ten cases of ileocecal valve atresia are found in the existing English literature.\(^5\)\(^-\)\(^13\) We present a neonatal baby with ileal perforation caused by a double membranous atresia (type IV atresia), one in the ileocecal valve, and one in the caecum.

A five-day-old girl born at 37 weeks as 2485 gr, was referred to our clinic with pre-diagnosis of necrotizing enterocolitis (NEC). Subdiaphragmatic free air was detected in the first abdominal graph taken in our clinic (Figure 1). In the emergent laparotomy there was a perforation in the distal ileum. We performed an end-ileostomy. During the second operation (5-month-old), the catheter placed to the distal of the ileostomy did not go further through the ileocecal valve. The catheter that passed from the appendix stump also stopped at a distance of 1 cm. An anastomosis was made between the ileum and the colon. The differential diagnosis of ileocecal valve atresia includes ileal atresia, meconium ileus and necrotizing enterocolitis. In the case of a perforation, we recommend opening an ileostomy initially and then starting a treatment for sepsis. We advise to check the distal colon for further atresia.

**Keywords:** Ileocecal valve; colonic atresia; atresia of small intestine; intestinal perforation; ileostomy; anastomosis, surgical;
it was unused. The patient was discharged without any complications. At the fifth month control, there was no passage of contrast dye beyond the proximal ascending colon with the contrast enema and there was a stenosis in the mid portion of the descending colon (Figure 2).

During the second operation (5-month-old), distal ileostomy extending to the ileocecal valve was identified and separated from the abdominal wall. The catheter placed inside the distal ileostomy could not be advanced further through the ileocecal valve (membranous atresia). Appendectomy was performed and we observed that the catheter that was passed through the appendix advanced only for 1 cm and did not go further, pointing to another atresia at the cecum. When cecum was excised, a membranous atresia was found on the edge of the excision (Figure 3). The mesentery was intact. After opening the membrane, the catheter easily advanced forward and the passage was controlled by injecting saline into the lumen; there was no other atresia. Neither malrotation nor an associated anomaly was detected. We performed a two-layer anastomosis between the proximal ileostomy end and the ascending colon. Also, the stenotic segment at the mid portion of the descending colon was freed from the adhesions. The fluid that was given to the lumen of the colon passed easily from the stenotic segment, so no further intervention was done. Postoperative period was uneventful.

During the contrast enema given one month after the operation, the contrast passed to the ileum segments, but there was still a stenosis at the mid portion of the descending colon (Figure 4). As there was no clinic of intestinal obstruction and no dilatation of the proximal segments, we decided to follow the stenotic colon segment without further intervention. Six months later, the control examination was normal. After five years of follow-up, no problem was observed.

FIGURE 1: Patient’s first plain abdominal graph: subdiaphragmatic free air was seen.

FIGURE 2: Contrast colon graph: no passage of contrast beyond proximal ascending colon.

FIGURE 3: Image at operation: Atretic cecum.
The presented case is the third case presenting with intestinal perforation due to ileocecal atresia.5,6 Shaoying et al. reported a postmortem examination of a case with intrauterine fetal demise. At the examination they found ileal perforation, meconium peritonitis, a small-caliber colon and a mucosal web at the ileocecal valve region.5 Cacciari et al reported a case with similar clinical examination and history to our patient, in which they performed a jejunostomy secondary to the jejunal and ileal perforations and discovered an ileocecal atresia in the second operation conducted 4 months later.6 They applied a different surgical technique and they created a new ileocecal valve by resecting a small circular portion from the center of the membranous atresia and suturing the wound lips with a few hemorrhagic stitches.6 Mousari et al. also reported a similar operation which involved opening the mucosal membrane between the ileum and the cecum.7 In the remaining 7 cases, the resection of the atretic segment and ileocolonic anastomosis were executed.8-13 In our patient a second atresia, 1 cm distal to the first one, complicated the case and the membranous atresia of the ileocecal valve was too narrow to allow such an operation. So, we had to remove the cecum (both atresias) and made an anastomosis between the ileum and the ascending colon.

Similar to our case, the common findings in all of the reported alive cases, except one, were bilious vomiting, abdominal distension, severe distention of bowel loops in abdominal x-ray scans and micro-colon (unused colon) without reflux to terminal ileum during colon enema.6,7,9-13 The only atypical presentation which was diagnosed in a patient one month of age, after two attacks of necrotizing enterocolitis, was reported by Kassira et al.8 In that case, the diameter of the colon was also normal, without reflux to the terminal ileum during colon enema.8 No data about meconium passage was noted.8 Although these data point out an acquired obstruction (atresia) of the ileocecal valve, the histopathological examination of the atretic segment has been reported to be compatible with congenital atresia.8

Some authors hypothesized, due to the pathological findings of the atresia, that the ileocecal atresia may be secondary to the intrauterine intussusception.9,11 The histopathologic examination of the resected atretic membrane was reported to have an ileal mucosa on the ileum side, colonic mucosa on the cecum side and muscular cells in between the mucosas.6,8,10,11,13 However, the polypoid mucosal protrusion that was assumed to be a remnant of the intrauterine intussusception,14 was reported in the pathological examination of only one of the cases.11 No other case was reported to have a polypoid mucosal protrusion in the pathological examination.5,6,8,10,13

The differential diagnosis of ileocecal atresia includes distal ileal atresia and meconium ileus.7,9,10 Even in the perforated cases, the first pre-diagnosis that came to mind was necrotizing enterocolitis similar to what we had thought for our patient.6

Only two among 224 colonic atresia cases had intestinal perforation of the distal microcolon as a result of the contrast enema, and all other colonic atresia cases presented with intestinal obstruction.3 Mortality rate in colonic atresia cases is reported as 25.7%, and it increases if the surgery is performed later than 72 hours.1 Even though our case was a more complicated one with a perforation, the patient who was operated on the fifth day of life survived without morbidity. The resection of the atresia and direct anastomosis can be done for the right colon.3,15 The presence of perforation and intra-abdominal meconium in our patient lead us to conduct an ileostomy without wasting time, as the patient’s general condition was not well enough. A review study reveals that Type IV atresia is seen in 10% (19/187) of colonic
atresia cases.\textsuperscript{3} Therefore, it is important to check the distal colon for another atresia.

Colonic atresia is associated with anomalies with a ratio of 47.3-58 \%.\textsuperscript{3,16} Colonic atresias associated with Hirschprung disease (HD) was reported at 31 cases so far.\textsuperscript{1,16-19} The ratio of Hirschprung disease among colonic atresia was reported as 8.5\% in the review study of Etensel et al.\textsuperscript{3} Although association of Hirschprung disease with colonic atresia is rare, if the combination was not diagnosed earlier, mortality and morbidity were elevated due to postoperative complications such as anastomosis leakage or dysfunction.\textsuperscript{1,16,17} Despite the fact that some authors claim rectal biopsy for HD screening is not necessary because the association of HD with colonic atresia is very rare, most of the authors suggest to perform rectal biopsy before the closure of the colostomy or ileostomy to prevent fatal complications.\textsuperscript{1,3,16,17} In our case, we diagnosed the ileocecal valve atresia and cecal atresia during the closure of the ileostomy and we decided to resect the atretic segments and perform an anastomosis preoperatively. As we thought that the perforation was due to NEC and did not suspect colonic atresia, we did not perform a rectal biopsy before. The postoperative clinical course was uneventful and no other intervention was needed. Although we have not performed one, we think that doing a rectal biopsy should be taken in consideration in patients with colonic atresia to rule out Hirschprung disease.

Ileocecal atresia is a very rare pathology. The common findings in these cases are bilious vomiting, abdominal distension, severe distention of bowel loops in abdominal x-ray scans and microcolon (unused colon) without reflux to the terminal ileum during colon enema. The differential diagnosis of ileocecal valve atresia includes distal ileal atresia, meconium ileus and necrotizing enterocolitis. For the treatment of ileocecal atresia, primary anastomosis or resection of the web are suggested in the literature. If a perforation is present, we recommend first opening an ostomy and then the treatment of sepsis. We think that rectal biopsy should be taken in patients with colonic atresia to rule out Hirschprung disease and further treatment should be managed according to avoid serious complications. We also advise to check the distal colon for further atresia.

Acknowledgements
We would like to thank Ülkü Ceren Köksöy for providing English language editing.

Source of Finance
During this study, no financial or spiritual support was received neither from any pharmaceutical company that has a direct connection with the research subject, nor from a company that provides or produces medical instruments and materials which may negatively affect the evaluation process of this study.

Conflict of Interest
No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions
\textbf{Idea/Concept:} Tuğba Acer Demir; \textbf{Design:} Tuğba Acer Demir, Esra Elif Arslan; \textbf{Control/Supervision:} Tuğba Acer Demir; \textbf{Data Collection and/or Processing:} Tuğba Acer Demir, Esra Elif Arslan; \textbf{Analysis and/or Interpretation:} Tuğba Acer Demir, Esra Elif Arslan; \textbf{Literature Review:} Tuğba Acer Demir, Esra Elif Arslan; \textbf{Writing the Article:} Tuğba Acer Demir, Esra Elif Arslan; \textbf{Critical Review:} Tuğba Acer Demir, Esra Elif Arslan.

\textbf{REFERENCES}