The Turkish Journal of Pediatrics
Volume 27, No. 3, July - September 1985

THE MEGACYSTIS–MICROCOLON–INTESTINAL HYPOPERISTALSIS SYNDROME*

A case report

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Key words : bladder, colon, intestinal obstruction, megacystis-microcolon-intestinal hypoperistalsis syndrome, newborn

In 1976, Berdon et al[1] first described the fatal neonatal intestinal obstruction, now known as the megacystis-microcolon-intestinal hypoperistalsis syndrome, as it presented in five girls. This syndrome has to date been reported in 23 cases, nineteen of them girls and four of them boys[2-14]. We encountered another patient with this syndrome, and we believe that our patient is the twenty-fourth case in the medical literature.

Case Report

A 3000 gram, full-term, four-day-old newborn girl was admitted to the Hacettepe Children's Hospital with marked abdominal distention. The pregnancy had been complicated by polyhydramnios but the delivery had been normal. Shortly after delivery, the baby had begun to vomit a bile-stained material, and this had continued. According to the report no meconium had been passed since birth, and a low urine output had also been observed.

On physical examination the baby was found to have a very distended abdomen but with no tenderness or discrete masses. Intravenous fluids were started immediately, a number 8 Fr. nasogastric tube was passed into the stomach and 40 cc of a bile-stained material were aspirated. A small rectal tube was inserted but no meconium was obtained. Serum electrolytes, BUN and creatinine were found to be normal.

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Abdominal X-rays showed a small gastric bubble in the left upper part of the abdomen and intestinal gas in the subhepatic region. A barium enema revealed an unused colon as shown in Figure 1. The bladder was catheterized and

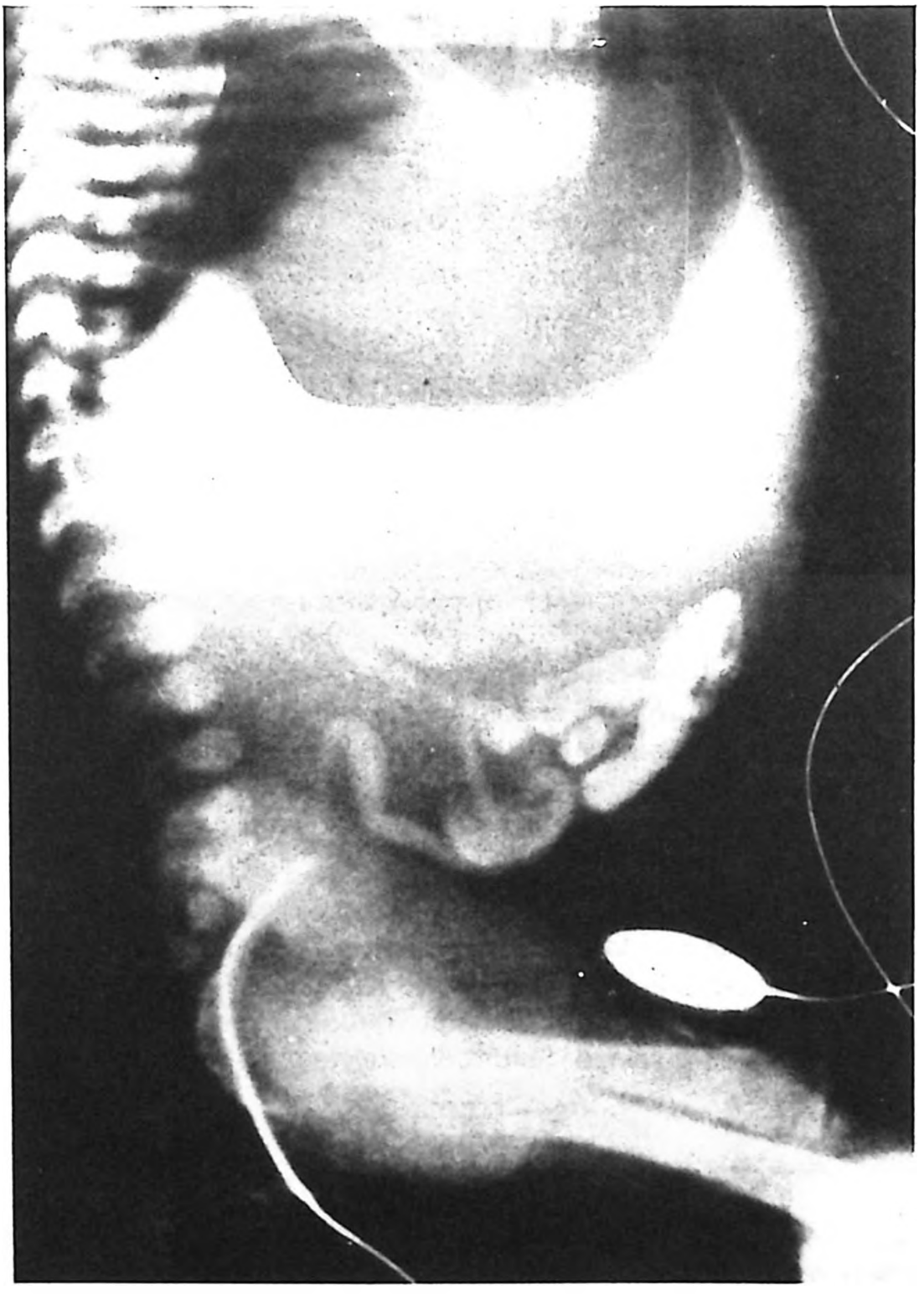


Figure 1

Barium enema showing unused colon (microcolon).

approximately 250 cc of urine obtained. Intravenous pyelography showed bilateral minimal hydronephrosis with dilated ureters (Figure 2). A cystogram revealed a large bladder with no reflux (Figure 3).

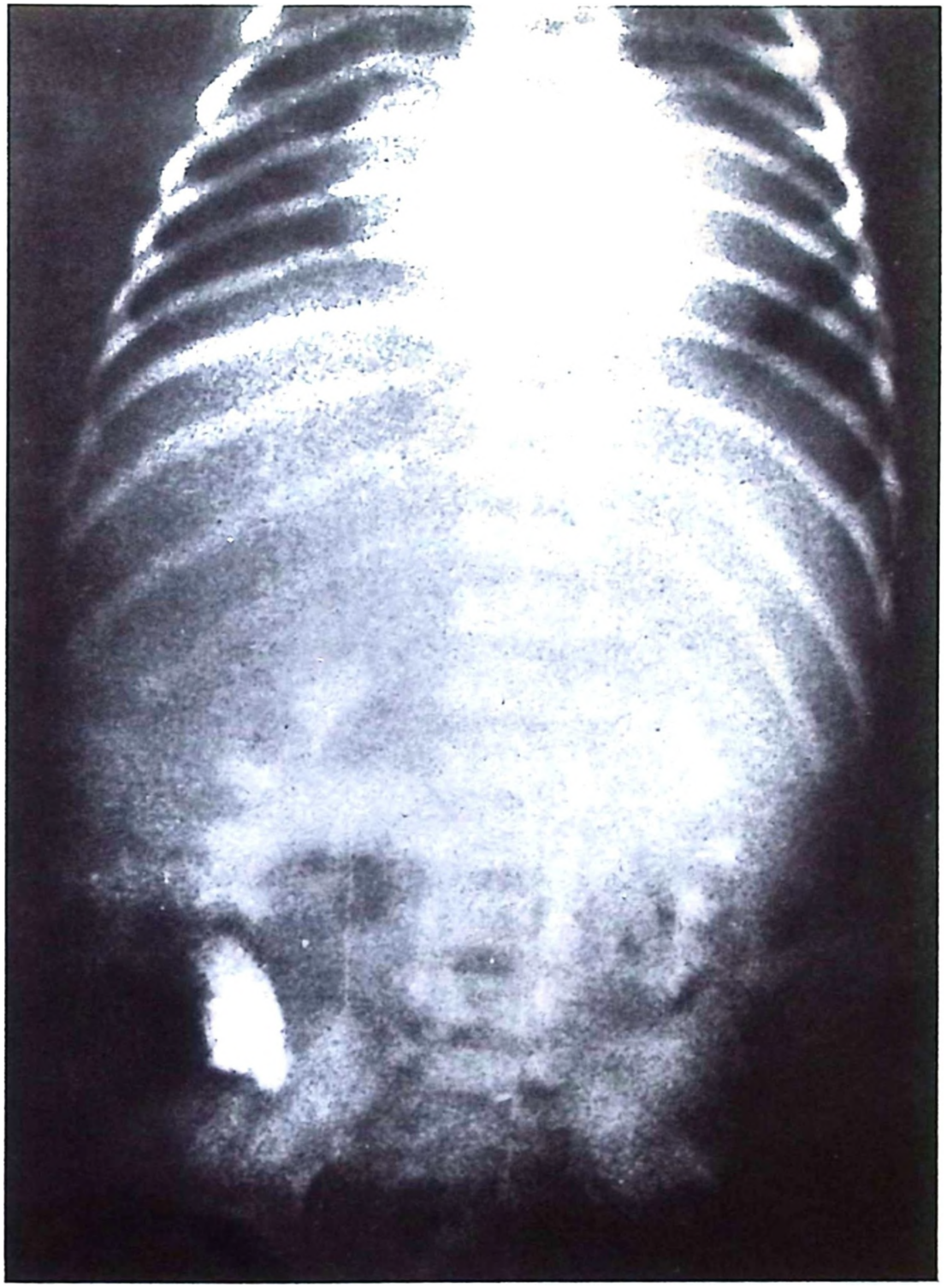


Figure 2

Intravenous pyelography after bladder drainage. There existe minimal hydronephrosis and residual barium persists in colon.

A laparotomy was performed under general anesthesia using a right transverse rectus incision. On exploration, the small intestine was found to be dilated, the

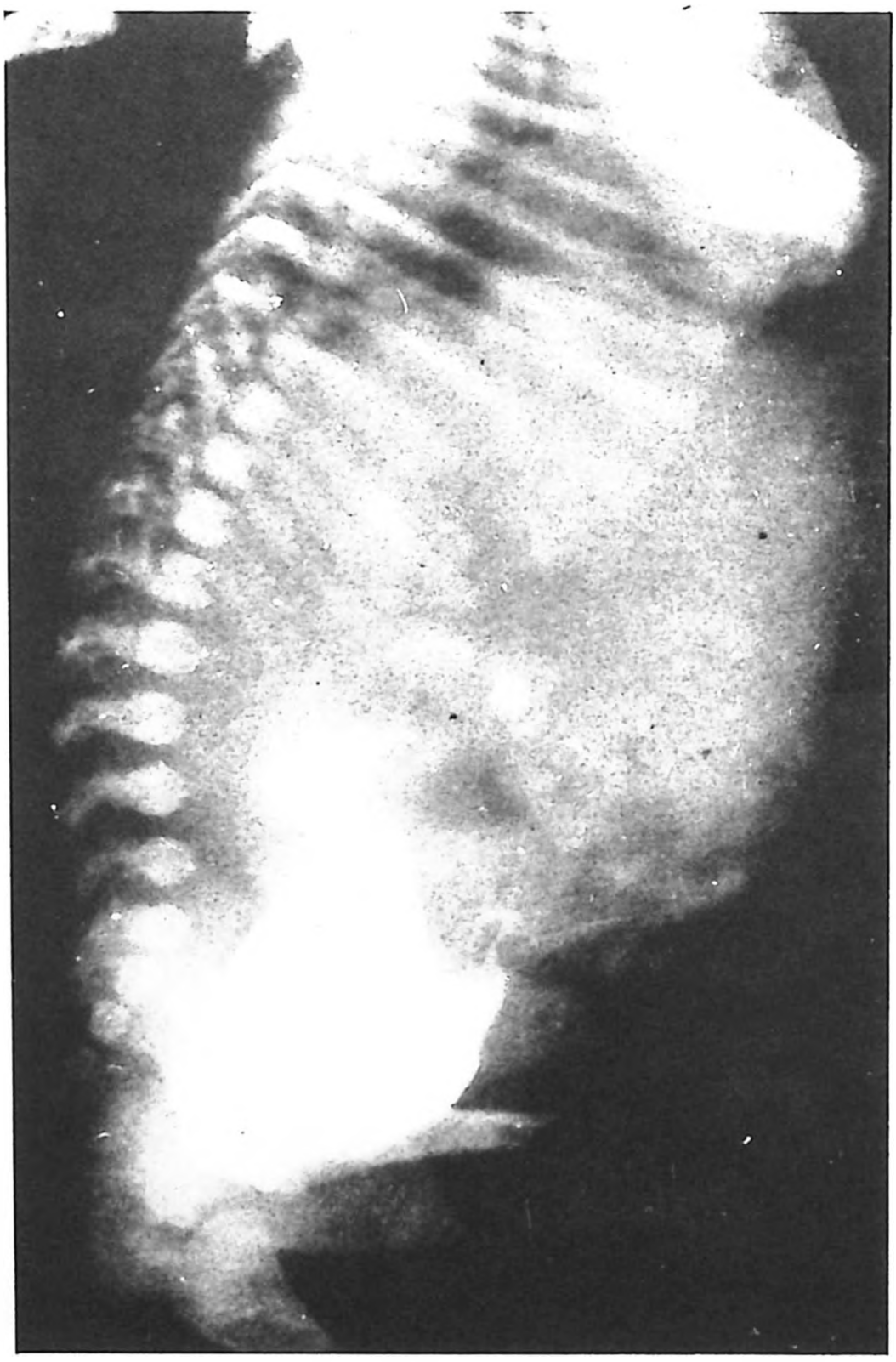


Figure 3

Cystogram showing a huge bladder.

large unused. A malrotation was also found but there were no signs of intestinal or duodenal obstruction. The urinary bladder was distended and measured 10x10x10 cm. Total aganglionosis was suspected but a frozen section of the removed appendix showed normal ganglion cells.

After the operation the patient did not pass any gas or meconium and could not tolerate feedings. A small amount of barium was given through a nasogastric tube and this was followed by a series of X-rays; the barium stayed 48 hours in the duodenum and the upper jejunum without any passage into the colon (Figure 4).

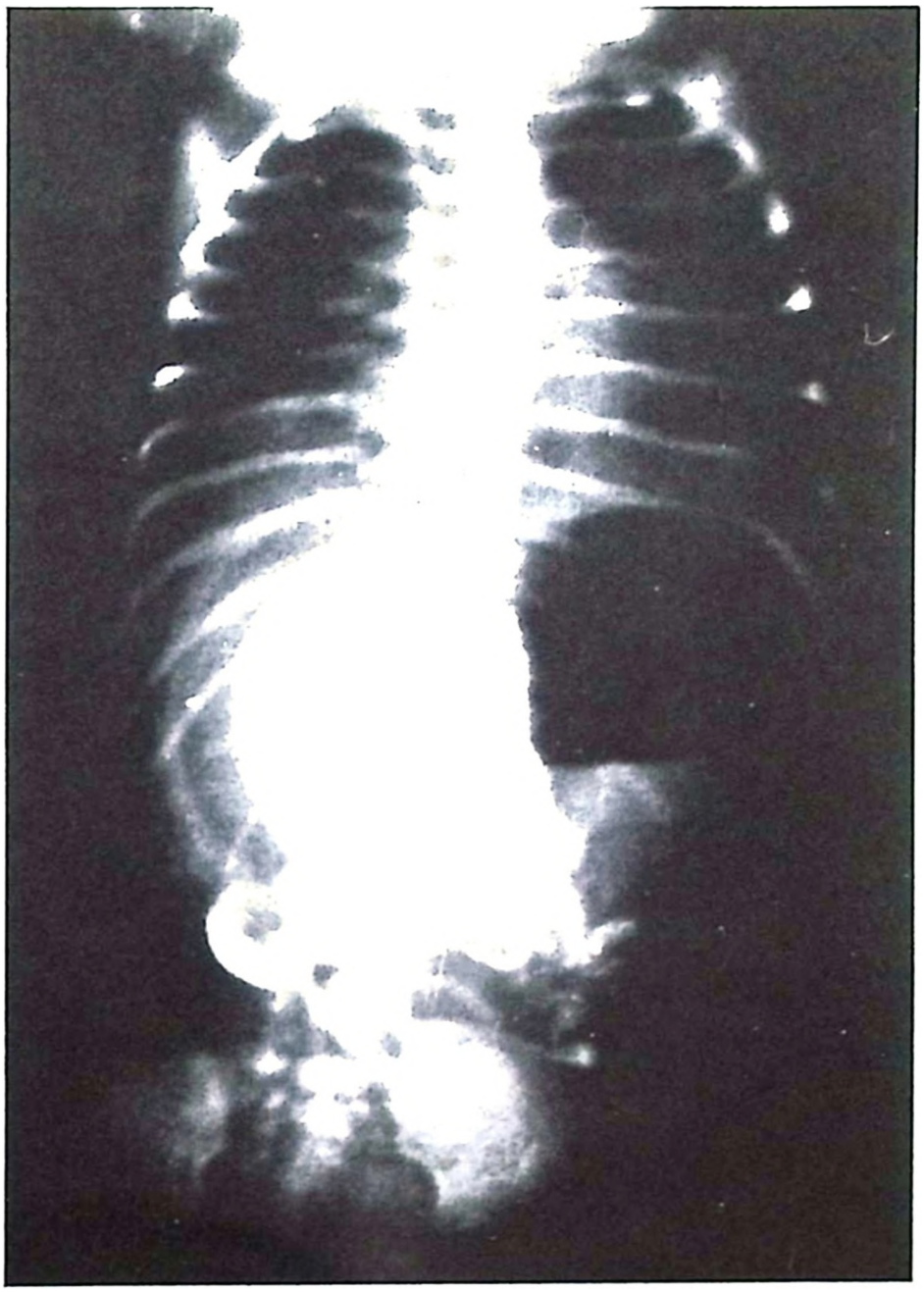
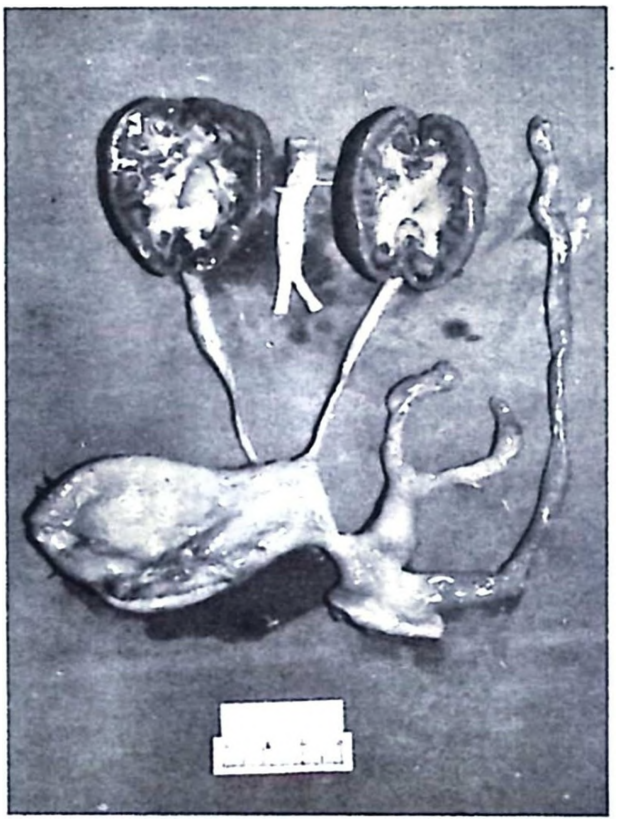


Figure 4

Contrast given through nasogastric tube stayed 48 hours in duodenum and upper jejunum and did not enter the colon.

Parenteral hyperalimentation was started but the patient became septic and died aged 23 days.

On autopsy, the small intestine measured 64 cm from the ligament of Treitz to the ileocecal valve. The colon was 24 cm in length. The distal ileum and the colon were 0.7 cm in diameter (Figure 5). There were ganglion cells and nerve fibers in the submucosal and myenteric plexuses throughout the intestine. The abdominal musculature was normal. There were dilated ureters, a huge bladder and minimal hydronephrosis, but there was no bladder neck obstruction. The ultimate cause of the death of the baby was nonsuppurative necrotizing pneumonitis and resulting sepsis from Pseudomonas species.



K : kidney
Ut : uterus
U : ureter
UB : urinary bladder
C : colon

Figure 5

Postmortem specimen. There are dilated ureters, a huge bladder and minimal hydronephrosis. The colon is unused.

Discussion

The megacystis-microcolon-intestinal hypoperistalsis syndrome (MMIHS) is a very rare disease which was first described in 1976[1]. In all, 23 cases have been reported[2-13]; 19 of the patients were girls but there is no explanation for this predominance.

The clinical symptoms of the neonatal MMIHS are similar to those of lower intestinal obstruction, namely vomiting, abdominal distention and absence of bowel movements. But in this case abdominal distention was secondary to dilated ureters and a distended urinary bladder. Intestinal malrotations are found in the majority of cases, but obstructive volvulus of the bowel was a complication in only one case[11].

With MMIHS there is urinary and intestinal obstruction but without any discernible organic lesions. The causes of the intestinal obstruction and the vesicoureteral dilatations of MMIHS are not known. This syndrome is usually fatal. Although one patient has survived now for 14 years, intestinal hypoperistalsis and megacystis have persisted[11]. In two of the cases followed urographically by Berdon et al[1], the urinary collecting systems in the subsequent excretory urograms appeared to be normal. Other types of neonatal functional intestinal obstruction are different from MMIHS because they do not present with urinary tract findings[11,13].

This fatal syndrome although rare, is one that should be taken into consideration in the newborn when there is difficulty in both urination and defecation.

Summary

The case of a newborn girl with the megacystis-microcolon-intestinal hypoperistalsis syndrome is described. The patient had a huge bladder without distal obstruction. Although a laparotomy revealed malrotation and a microcolon, the main cause of the intestinal obstruction was hypoperistalsis. Intestinal hypoperistalsis persisted without any improvement and hyperalimentation was required. The outcome was fatal as in most of the reported cases.

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