Current significance of meconium plug syndrome

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Received 28 November 2007; accepted 3 December 2007

Key words:
Meconium plug;
Hirschsprung’s disease;
Cystic fibrosis

Abstract
Background: The significance of meconium plug syndrome is dependent on the underlying diagnosis. The incidence of pathologic finding, particularly Hirschsprung’s disease, contributing to the presence of these plugs, has been debated. However, there are little recent data in the literature. Therefore, we reviewed our experience with meconium plugs as a cause of abdominal distension to evaluate the associated conditions and incidence of Hirschsprung’s disease.

Methods: We reviewed the records of newborns with meconium plugs found in the distal colon on contrast enema from 1994 to 2007. Demographics, radiologic findings, histologic findings, operative findings, and clinical courses were reviewed.

Results: During the study period, 77 patients were identified. Mean gestational age was 37.4 weeks and birth weight, 2977 g. Hirschsprung’s disease was found in 10 patients (13%). One had ultrashort segment disease and another had total colonic aganglionosis. Maternal diabetes was identified in 6 patients. No patients were diagnosed with cystic fibrosis, meconium ileus, malrotation, or intestinal atresia.

Conclusion: Meconium plugs found on contrast enema are associated with a 13% incidence of Hirschsprung’s disease in our experience. Although all patients with plugs and persistent abnormal stooling patterns should prompt a rectal biopsy and genetic probe, the incidence of Hirschsprung’s and cystic fibrosis may not be as high as previously reported.

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Neonatal bowel obstruction typically presents with abdominal distension, emesis, and failure to pass meconium within the first 24 hours. Differential diagnosis includes Hirschsprung’s disease, malrotation, meconium disease, and intestinal atresia. Meconium plug syndrome is most often a benign cause of obstruction that clears after rectal stimulation or administration of contrast enema; however, the presence of meconium plugs may herald underlying intestinal pathologic nature. Literature on the topic is scattered and widely variant in the reported incidence of significant pathologic finding. The initial reports describing meconium plug syndrome documented a 13% incidence of Hirschsprung’s disease and no association with cystic fibrosis [1,2]. However, for the past 40 years, the literature has reflected different results with Hirschsprung’s in as many as 38% and cystic fibrosis in as many as 43% of patients with meconium plug syndrome [3,4]. Therefore, we performed a retrospective analysis to review the coexistence of meconium plug syndrome.
plugs and their relationship with Hirschsprung’s disease meconium ileus and other pathologic finding.

1. Methods

Retrospective review of the medical record was performed on all patients diagnosed with a meconium plug identified in the rectum on contrast enema from January 1994 through February 2007. During the study timeframe, 77 patients were identified. Gestational age, birth weight, passage of meconium, bilious vomiting, abdominal distension, associated anomalies, and maternal diabetes were recorded. The presence of meconium ileus, small descending colon, a transition zone, malrotation, and intestinal atresias on contrast enema were recorded. Results of suction rectal biopsies and studies performed to diagnose cystic fibrosis were recorded. Operative interventions, total length of stay, and outcome were reviewed.

2. Results

Mean estimated gestational age was 37.4 ± 2.4 weeks with a mean birth weight of 2977 ± 693 g. Sex distribution included 48 males and 29 females. Abdominal distension followed by bilious emesis was the most common clinical presentation. A history of maternal diabetes was present in 6 patients (7.8%). Mean age at presentation was 1.49 ± 0.92 days. Spontaneous passage of meconium in the first 24 hours occurred in 15 patients (19.5%). No cases of meconium ileus or intestinal atresia were identified. At the initial diagnostic contrast enema, a diagnosis of small descending colon was made in 27 patients (35.1%). A transition zone was identified in 13 patients (16.9%), of which 9 (11.7%) were diagnosed with both small descending colon and a transition zone.

Patients with a persistent abnormal stooling pattern underwent suction rectal biopsy resulting in 22 biopsies in the same number of patients (28.6%). Thirteen of these patients had a transition zone identified on the contrast study. Hirschsprung’s disease was confirmed histologically in 9 patients from the suction rectal biopsy performed in the neonatal period, of which 4 patients had a transition zone identified on the contrast study. One additional patient was identified as having Hirschsprung’s disease by balloon manometry at 4 years of age. Thus a total of 10 patients were found to have an aganglionic segment in this series representing 13.0% of the study population. Of these 10 patients, 8 had the typical rectosigmoid transition, 1 patient had ultrashort segment disease, and 1 patient had total colonic aganglionosis. All have undergone definitive procedures with no mortality, and all are currently doing well. The patient with ultrashort segment underwent internal sphincterotomy that has alleviated the constipation.

A total of 25 patients underwent investigation for cystic fibrosis, with 22 gene probe analysis and 3 sweat chloride tests, of which none had positive results.

3. Discussion

The term meconium plug syndrome was first reported by Clatworthy [1] in 1956 to describe the colonic obstruction because of inspissated meconium. Initial nonsurgical management was recommended as most of these patients were treated effectively with rectal stimulation or contrast enema. This original report was later updated with additional patients 10 years later, in which the authors recommended rectal biopsy to rule out Hirschsprung’s disease if normal bowel function did not occur after passage of the plug [2].

Small descending colon syndrome has been associated with meconium plug syndrome since the initial report [1]. The association of diabetic mothers giving birth to babies with small descending colon syndrome was subsequently recognized with an incidence of 40% to 50% [5,6]. Although our findings superficially support these studies as 3 of 6 diabetic progeny had small descending colon seen on contrast enema, this study has only a subset of diabetic mothers and does not address the entire denominator of these mothers.

The clinical presentation of meconium plug syndrome and meconium ileus is similar, although the source of meconium obstruction is at 2 separate locations. In our series, patients with typical meconium ileus appearing as ileal obstruction with proximal small bowel dilatation and microcolon were not included as meconium plug syndrome patients. In the past, cystic fibrosis, the usual cause of meconium ileus, has been reported to be associated with meconium plug syndrome [4,7]. One series of 25 patients with meconium plug syndrome found an impressive 24% who were found to have the cystic fibrosis mutation [7]. Recently, an even higher incidence (43%) appeared in the literature [4]. We found no association with cystic fibrosis and meconium plug syndrome in our series. This contrasting result may relate to the definition of meconium plug syndrome. None of the contrast enemas in our series had meconium plugs in the small intestine, only in the colon. In the previous report with a 24% incidence of cystic fibrosis, no mention of the anatomical location of the meconium is stated [7]. This implies that patients may have been included with ileal plugs. In the other report, meconium plug syndrome is in fact defined as meconium located in the colon but does not speak of whether the population is meconium exclusively in the colon [4]. That we had no patients with cystic fibrosis supports a theory that a meconium plug obstruction in the colon is a different disease entity than one located in the distal small bowel as previously reported [2].

The recognition that Hirschsprung’s disease may be the cause of the meconium plugs has been recognized since the
initial description was published (Table 1). That initial report of 30 patients found 13.3% had an aganglionic segment, which is identical to our series and similar to another small series of 8 patients with meconium plugs of which the result of 2 suction rectal biopsies were aganglionic [8]. The low incidence of Hirschsprung’s disease historically reported was recently contradicted in a report of 21 patients with meconium plugs, of whom 38% were diagnosed with Hirschsprung’s disease concluding that this association may be higher than has been historically reported [3]. This study precipitated our investigation. Our series represents the largest series ever compiled on meconium plug syndrome by more than 2-fold. In addition, the dataset is recently compiled thus representing current diagnostic knowledge and technical expertise of radiologists at a high-volume dedicated children’s hospital. Of the 77 patients in our series with meconium plug syndrome, 10 patients (13.0%) were ultimately diagnosed with Hirschsprung’s disease, a number identical to the initial series published 40 years ago [2]. The distribution of disease is about what would be expected from any 10 Hirschsprung’s patients with most demonstrating rectosigmoid transition with one very short and one very long (total colonic). This distribution implies that Hirschsprung’s diagnosed after meconium plug syndrome may be any level of disease that is an important fact for the counseling surgeon. The favorable outcomes after the definitive operation from our series have also been reported previously [2,7].

Based on this large, contemporary review, the presence of a meconium plug on contrast enema most often correlates with a benign clinical course. However, in the presence of an abnormal stooling pattern, further workup is indicated. When meconium plug syndrome is identified carefully as pellets only present in the colon, then a diagnosis of cystic fibrosis may be a rare event. It appears that the incidence of Hirschsprung’s disease remains around 13% as initially reported when the syndrome was defined.

References