The Cause of Rectal Prolapse in Children

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• Fifty-four pediatric patients with rectal prolapse (RP) were identified by review of medical records from 1977 to 1987. Rectal prolapse was attributed to chronic constipation (15 patients), acute diarrheal disease (11 patients), cystic fibrosis (CF) (six patients), and neurologic/anatomic abnormalities (13 patients). In nine patients, no underlying cause was identified. The patients with CF did not differ from the other groups in terms of age at time of onset of prolapse, growth measurements, or number of episodes of prolapse. All patients with CF had a history of abnormalities or presented with signs and symptoms consistent with this diagnosis; none had a history of constipation. Although physicians can be reassured that CF is not a likely diagnosis in patients with RP and acute diarrheal disease or a clear history of constipation, a sweat test is indicated in all such cases as well as in those in which there is no apparent underlying cause. A sweat test is not usually indicated in patients with RP in association with underlying anatomic abnormalities.

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Rectal prolapse (RP) is a common complication in patients with cystic fibrosis (CF), occurring in 18% to 23% of patients before definitive diagnosis.[1,2] Rectal prolapse has also been reported in association with diarrheal disease,[3] ulcerative colitis,[4] chronic constipation,[5] malnutrition,[3] Hirschsprung's disease,[6] Ehlers-Danlos syndrome,[7] meningomyelocele,[3] pertussis,[8] rectal polyps,[3] and following surgical repair of anorectal anomalies.[8] A sweat test is recommended to rule out CF in all patients presenting with RP.[1,2] We attempted to determine the cause of RP in pediatric patients presenting with this complaint and to focus on the associated clinical findings in patients with an eventual diagnosis of CF.

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PATIENTS AND METHODS

We attempted to retrieve the charts of all patients younger than 18 years of age who presented to The Johns Hopkins Hospital, Baltimore, from 1977 to 1987 with a complaint of RP. Records were obtained from the following sources: hospital medical records (ten patients), pediatric gastroenterology clinic (four patients), general pediatric surgery clinic (two patients), sweat test referrals (22 patients), and the pediatric emergency room and walk-in clinics (16 patients). The diagnoses of all patients seen in the pediatric emergency room and walk-in clinics are coded and entered into a computerized data retrieval system. The charts were reviewed and abstracted for demographic information, symptoms, physical findings, laboratory results, associated problems, and diagnoses. Follow-up information was obtained from three sources: review of medical records, communication with primary care physicians, and telephone contact with patients' families.

RESULTS

We were able to find 54 patients who presented with RP. At the time of RP, the patients ranged in age from 5 days to 11 years (mean, 32 months). Nine patients were under 6 months of age, five were 7 to 12 months, ten were 13 to 24 months, 13 were 25 to 36 months, six were 37 to 48 months, and 11 were over 48 months. Among the patients, 61% were female, 67% were white, 31% were black, and 2% were Oriental. The associated diagnoses in these patients are shown in the Table.

The most frequent underlying problems were chronic constipation (15 patients), acute diarrheal disease (11 patients), and CF (six patients). In 13 patients, RP occurred in association with underlying anatomic defects, including meningomyelocele (six patients), imperforate anus repair (four patients), and rectal polyps (three patients). In nine patients, the condition was considered idiopathic, as no underlying cause was established either during the acute episode or during follow-up. Quantitative pilocarpine iontophoresis sweat tests were performed in 33 patients, including 12 of 15 with constipation, seven of 11 with diarrhea, two of 13 with anatomic defects, six of six with CF, and six of nine with idiopathic RP. Follow-up (duration, six months to 9.5 years; mean, 37 months) data were available on the ten patients without anatomic defects who did not receive sweat tests; nine were well and one continued to have episodes of RP secondary to constipation.

In patients with diarrhea and constipation, RP resolved when the stool pattern returned to normal. In patients with CF, episodes of RP were not seen after the initiation of pancreatic enzyme supplementation. Surgical intervention was not required for any of the patients with RP.

Among the 41 patients with RP who did not have an underlying anatomic abnormality, CF was confirmed in six (14.6%). The patients with CF did not differ from the other patients in mean age at time of RP (27.5 vs 31.9 months). Of the patients with CF, four of six were equal to or less than the 25th percentile for weight compared with 11

Associated Diagnoses in Patients With Rectal Prolapse

Diagnosis	No. (%) of Patients
Chronic constipation	15 (27.8)
Acute diarrheal disease	11 (20.4)
Cystic fibrosis	6 (11.1)
Meningomyelocele	6 (11.1)
Imperforate anus (postrepair status)	4 (7.4)
Rectal polyps	3 (5.5)
No known cause	9 (16.7)
Total	**54 (100)**

of 35 patients without CF.

The six patients who were eventually confirmed to have CF all had a history of abnormal (loose) stools at the time of presentation with RP. None had a history of constipation. We therefore compared the six patients with CF with the 11 patients without CF who had acute diarrhea at the time of RP. The two groups were similar for age at presentation with RP, growth variables, and number of episodes of RP. Repeated episodes of RP were seen in all six patients with CF and in eight of the 11 patients with acute diarrhea.

However, there were clinical clues to the underlying diagnosis in all six patients with CF, including loose stools (six patients), poor growth pattern (four patients), wheezing (one patient), history of meconium plug syndrome (one patient), family history of CF (one patient), and digital clubbing (one patient). Among the 11 patients with acute diarrhea, one had a history of wheezing and one had a history of recurrent upper respiratory tract infections. Respiratory symptoms (wheezing) were prominent in only one of the patients with CF.

COMMENT

Cystic fibrosis has been cited as the most common cause of RP in children in the United States.[3,9] The diagnosis of CF needs to be considered in all children who present with RP, and a diagnostic sweat test is recommended for all such patients.[1,2] These recommendations were based on studies that approached RP by examining a population of patients with CF rather than examining a group of patients presenting with RP. In other countries, particularly developing countries, most cases of RP have been attributed to acute diarrheal disease and intestinal parasitic infestation, usually in association with malnutrition. In a study from India, among 80 children with RP, diarrhea was present in 84%, two thirds of whom had amebiasis.[10]

The results of our study suggest that, at least in the United States, most instances of RP are not related to CF but rather to stool abnormalities, such as acute diarrhea and chronic constipation, or neurologic or anatomic defects. Among 41 children who presented with RP and who did not have an anatomic defect, a diagnosis of CF was eventually established in only six patients (14.6%). These results are similar to those reported in a study from the Soviet Union.[11] Among 418 children with RP, underlying problems included colitis (39.6%) and chronic constipation (27%), along with a variety of other problems, including anorectal anomalies and respiratory problems. Cystic fibrosis was not specifically mentioned.

The patients with and without CF in our study did not differ in terms of age at time of initial RP, growth status, or frequency of episodes of RP. However, all of the patients with CF had a history of chronic stool abnormalities, including the passage of greasy, oily, malodorous, and/or floating stools. None of the patients with CF had a history of constipation. Careful review of the records of the patients with CF revealed that clues to the underlying diagnosis were present in all six patients. Most helpful was a stool history consistent with fat malabsorption. It is of interest, and possibly misleading, that only one of the patients with CF had a history of significant respiratory symptoms. Among the 11 patients with acute diarrhea, only one had a history that was at all suggestive of CF.

On the basis of our results, it is clear that CF needs to be considered in patients presenting with RP. In this regard, it is important to obtain a careful history to identify stool and respiratory problems, family history, and history of neonatal stool problems, and to evaluate for the presence of growth failure and digital clubbing. However, it is important not to be dissuaded from the diagnosis of CF by the absence of respiratory symptoms or normal results of a physical examination.

Our results indicate that the majority of cases of RP in children are not related to CF but rather to constipation or acute diarrheal disease. In such instances, physicians can usually be reassured that CF is not a likely diagnosis. Because of the difficulty in differentiating diarrhea from steatorrhea by history, we recommend that a quantitative pilocarpine iontophoresis sweat test be carried out in all patients with RP who report loose stools. A sweat test is also indicated in all instances of RP in which there is no apparent underlying cause. In patients with RP in association with underlying anatomic abnormalities, a sweat test is not usually indicated. In patients with a clear history of constipation and no other abnormalities according to history or on physical examination, the likelihood of CF is probably very small. However, in view of the ease of performing a sweat test and the potentially disastrous consequences of missing the diagnosis of CF, a sweat test is indicated in all such patients.

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