ORIGINAL ARTICLE

Role of anorectal manometry in the differential diagnosis of chronic constipation in children

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Abstract

Objectives 1. To evaluate the role of anorectal manometry in recognizing patients with histological abnormalities of the myenteric plexus as the cause of chronic constipation. 2. To correlate clinical features with the etiology of the constipation.

Methods: the etiology of the constipation was identified retrospectively from the medical records of 57 patients submitted to anorectal manometry. The test was performed in the Pediatric Gastroenterology Laboratory of the University Hospital of the State University of Campinas Medical School. The procedure was performed using a single-balloon system in which a water-perfused pressure transducer was interfaced with a polygraph and displayed on a computer screen. Manometry was performed in patients who, after 6 to 23 months of intense medical treatment using osmotic laxatives and dietary fibres (20 g/d), continued to develop fecal impaction and/or needed evacuatory enema. Regularization of bowel movements during at least 24 months follow-up or the visualization of abnormalities in myenteric plexus in rectal biopsy were used as reference points for the definitive diagnosis.

Results: rectoanal reflex was present in 44 of the 57 patients (77%). Thirteen patients with a negative reflex underwent rectal biopsy which showed abnormalities of the ganglion cells in 10. Subsequent manometry performed on those three patients revealed a normal rectoanal reflex. Sensitivity, specificity, positive and negative predictive values of the manometric test were 1.0, 0.94, 0.77 and 1.0, respectively.

Conclusions: anorectal manometry is a diagnostic technique with very small possibility of error in differential diagnosis between constipation of a chronic functional nature and that which is secondary to ganglion cell abnormalities.

J Pediatr (Rio J) 2000; 76(3): 227-32: child, constipation, anorectal manometry.

Introduction

Most pediatric constipation cases may be treated with an adequate diet and moderate use of laxatives and enemas. A significant proportion of patients, however, does not show improvement with these therapeutic measures.[1-3] In these cases, it is necessary to establish the differential diagnosis between functional and structural constipation. The most difficult differential diagnosis is that of ultrashort-segment Hirschsprung's disease. This is due to the similarity of clinical manifestations and to the fact that in the ultrashort variety, the barium enema does not reveal a spastic aganglionic segment and intestinal dilatation, identifiable in the most frequent form of Hirschsprung's disease (congenital aganglionic megacolon). Thus, a functional constipation diagnosis based solely on a single radiological examination where the "transition zone" has not been visualized may delay the solution of a condition for which surgery is required.

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Anorectal manometry is useful for identifying ultrashort-segment Hirschsprung's disease. One of the aims of the manometric examination in the investigation of constipated patients is to demonstrate the presence of a rectosphincteric inhibitory reflex. The observation of internal sphincter relaxation in response to the distension of the rectal wall eliminates the possibility of aganglionosis and makes the diagnosis of some of the variable forms of neuronal dysplasia of the myenteric plexuses improbable.[4]

As is the case with the barium enema, the differential diagnosis with anorectal manometry is not 100% accurate,[5-9] with false-negative[10-11] and false-positive[12-14] results. In the past decade, the literature has reported variable rates of reliability for the identification of Hirschsprung's disease and of neuronal dysplasias by manometry.[15-18]

The objective of this study was to evaluate the power of anorectal manometry to identify patients who carry morpho-functional alterations of the ganglionic and myenteric plexuses, and, at the same time, to correlate clinical features with the definitive etiologic diagnosis of constipation. The clinical response during 24 months of follow-up or the identification of histologic abnormalities on rectal biopsy were used as reference for the definitive diagnosis.

Patients and Methods

From April 1996 to June 1997, 57 children presenting chronic constipation were submitted to anorectal manometry at the Pediatric Gastroenterology Laboratory, Hospital das Clínicas, Universidade Estadual de Campinas.

The histories of these 57 patients were retrospectively analyzed in search of data such as age at the moment of the examination, beginning of constipation complaints, soiling complaints, opaque enema findings, anorectal manometry results, rectal biopsy, and clinical evolution.

All the patients had been referred by other services to the Pediatric Gastroenterology and/or Pediatric Surgery ambulatories because their condition had not improved with the previous clinical treatment.

At the ambulatory, the patients went through the following treatment protocol: 1. initial hospitalization for removal of fecaloma through intestinal lavage with 20 ml/kg of glycerine physiologic solution at 12%; 2. ambulatorial follow-up with prescription of osmotic laxatives (lactulose or magnesium hydroxide) in a dose of 0.5 to 2 ml/kg once daily, and orientation to ingest 20 g of fiber per day; the use of enemas was allowed only if evacuation did not occur for 72 hours with the use of oral medication.

Patients underwent manometry when, after periods varying from 1 to 6 months of treatment, they presented relapse of fecal impaction and/or could not reestablish spontaneous bowel movements, that is, if they required the help of an enema.

The equipment that measures anorectal pressure includes a low-compliance pneumohydraulic capillary perfusion system (Arndorfer™, INC, Greenvale, WI) connected to a computer (IBM™ PC) with a specific software (Gastrosoft™, Polygram Lower GI™, version 6.40, Sinetics Medical™). In order to investigate the rectosphincteric reflex, we employed a six-channel catheter distributed along the same axis (axial catheter), with perfusion orifices at 5 mm intervals, perfused with 25 microliters of distilled water per minute and with a latex balloon at its end.[19] Investigation of the rectosphincteric reflex was carried out through quick insufflation and deflation of the intra-rectal balloon with 5 to a maximum of 60 ml of air. The reflex was considered present if there was a fall of $\geq 20\%$ in anal pressure in relation to the resting pressure. Figure 1 shows the manometric tracing obtained after insufflation of air into the intra-rectal balloon in a patient with morphologic and functional integrity of the myenteric plexus. As can be seen, soon after the stimulus, once the voluntary contraction of the external anal sphincter is over, there is a decrease of resting pressure in the anal canal. This relaxation characterizes the rectosphincteric inhibitory reflex. Figure 2 shows anal canal pressures in a patient with aganglionosis. An elevation in the pressure is identified in response to the stimulus, corresponding to the voluntary contraction of the external anal sphincter; yet, the relaxation of the internal sphincter was not triggered. The presence of this reflex signals the integrity of the myenteric plexus.[20] Patients who did not present this reflex were submitted to rectal biopsy.

Rectal biopsy was performed under general anesthesia in all patients. A progressive dilatation of the anal canal was initially performed with the patient in the lithotomy position. After having identified the pectineal line, the biopsy site was chosen on the lateral wall of the rectum, at least 2 cm above the pectineal line to avoid the physiologic aganglionic zone. Two repair sutures were placed, and a fragment of the total rectum wall (approximately 0.5 cm in diameter) was removed. After that, hemostasia was performed, and the defect on the rectal wall was closed with an absorbent suture. The fragment was referred to the anatomopathologic laboratory for processing in paraffin and for staining with hematoxylin-eosin. The slides were examined in search of ganglionic cells in the submucous and muscular plexuses.

In patients who underwent intestinal biopsy, the definitive diagnosis of the constipation cause was based on biopsy findings. In patients without rectal biopsy, the diagnosis of functional constipation was based on an adequate response to the subsequent clinical treatment after at least 24 months of follow-up.

Statistical Analysis

Student's t test and the chi-square (χ^2) test[21] were used for statistical analyses. The t test was used to assess differences in mean age, median duration of symptoms, and median Z scores for the relationships between weight/age, height/age and weight/height in the functional constipation
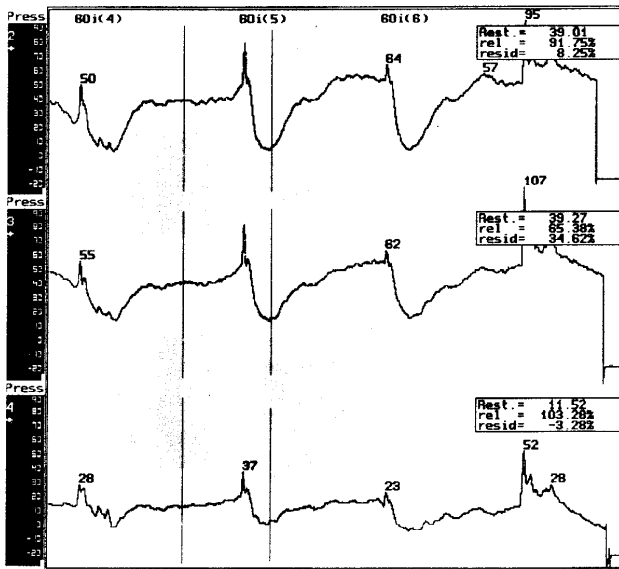
Figure 1 - Anorectal manometry tracing showing rectosphincteric reflex triggered after insufflation of intrarectal balloon with 60 ml of air

Table 1 - Radiologic signs identified in the opaque enema of 51 children with functional or secondary constipation (n=41) the anomaly of the myenteric plexus (n=10), assisted in the Pediatric Gastroenterology Clinic, Hospital das Clínicas, Universidade Estadual de Campinas, 1996/1997

	Functional constipation (n=41)	Malformation of the myenteric plexus (n=10)	P
Rectum dilatation	39	9	>0.05*
Sigmoid and rectum dilatation	34	7	>0.05*
Transition zone	0	0	>0.05*
Absence of haustrations in the dilated segment	18	5	>0.05*

* chi-square

and myenteric neural plexus disease groups. The χ^2 test was applied to determine the differences between the groups in the prevalence of clinical data (soiling complaint, beginning constipation, presence of abdominal distension on physical examination, and barium enema findings).

The manometric results for the diagnosis of Hirschsprung's disease or neuronal dysplasia were analyzed in terms of sensitivity, specificity and negative and positive-predictive values.[22] A significance level of 0.05 was adopted.

Results

Between April 1996 and June 1997, 57 children were submitted to manometry as part of the investigation of intestinal constipation of difficult clinical treatment. The indications for the exam included inability to evacuate spontaneously (without the help of an enema) in 18 out of 57 patients, and fecal impaction relapse (verified by the presence of palpable fecal mass in the abdomen and/or by reported reappearance of fecal soiling) in 41 patients. From 57 children, 51 had undergone a barium enema that did not define the differential diagnosis in any of the patients (Table 1).

The rectosphincteric inhibitory reflex was demonstrated in 44 of the 57 patients (77%). The 13 patients (23%) without a reflex were submitted to rectal biopsy, which revealed myenteric plexus aganglionosis in eight patients, intestinal neuronal dysplasia in two, and presence of morphologically normal ganglionic cells in three. These

last patients were submitted to a second manometry that demonstrated, after adequate preparation of the colon, the presence of the inhibitory reflex. Thus, a definitive diagnosis of morphologic and functional disorder of the myenteric plexus was established in 10 of the 57 patients.
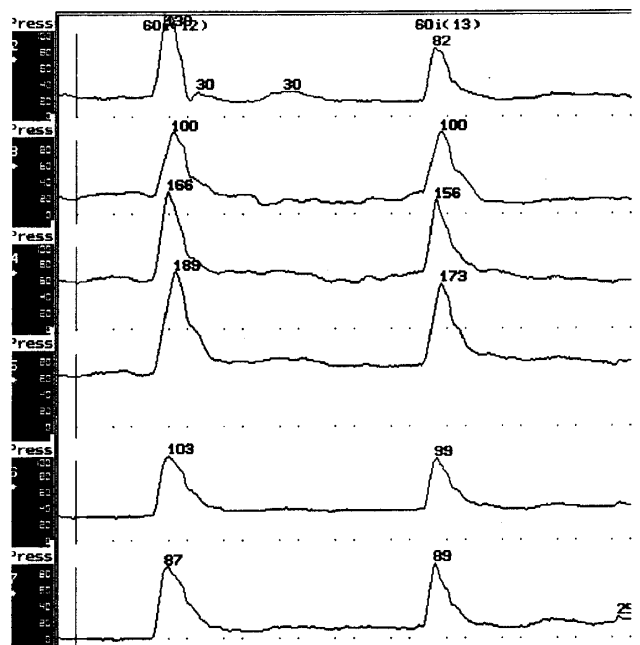

Figure 2 - Manometric tracing in child with Hirschsprung's disease. The rectosphincteric reflex could not be triggered

Table 2 presents clinical data about the 47 patients with functional constipation in comparison to the group of patients with myenteric plexuses anomalies. The duration of the constipation complaint and the average age in which manometry was performed were similar in all groups. The data in Table 2 indicate that beginning of constipation in the 1st week of life was significantly more frequent among children with myenteric plexus dysfunctions (P<0.05). None of the patients with intrinsic nervous system dysfunctions presented soiling, while 31 of the 47 subjects with functional constipation reported soiling; the difference between the groups is statistically significant (P<0.001).

There were three false-positive and no false-negative results for manometry (Table 3), corresponding to a sensitivity of 100%, specificity of 94%, positive-predictive value of 77% and negative-predictive value of 100%.

Discussion

Chronic constipation in childhood is a symptom of several diseases. A detailed clinical history associated with a careful physical examination may, in most of the patients, differentiate functional constipation and aganglionosis from other causes of constipation. A diagnosis of Hirschsprung's disease must be considered in newborns with intestinal obstruction, in children with diarrhea and abdominal distension, and in constipated and malnourished children.[23] In the present sample, the assessment of clinical features suggests that the evolution of children with malformations

of the myenteric nervous system may be very similar to that of children with functional constipation until they reach pre-school and school age. Abdominal distension and malnutrition, signs that are classically associated with aganglionosis, were not significantly associated with this diagnosis in the present study. The median age of patients with myenteric plexus dysfunction reveals an undesirable delay in the diagnosis of a condition that can be resolved with surgery. The appearance of symptoms in the neonatal period and the absence of soiling were statistically related to aganglionic constipation, although each of these signs in isolation is not sufficient to eliminate a functional cause.

The precision of the barium enema for diagnosing aganglionic congenital megacolon is considered to be poor in newborns, in the presence of total aganglionosis, and in cases of ultrashort aganglionosis.[7-9] In a group of 58 constipated children, the presence of a rectosigmoid transition zone, usually associated with aganglionosis, was identified in nine children with functional constipation. In this same group, 20% of the enemas that were positive for Hirschsprung's disease did not present a transition zone.[24] The present results suggest that the barium enema was not adequate to establish a diagnosis of Hirschsprung' disease. The typical findings of stenosed rectum with transition zone were not identified in the radiological examinations of children with a histological diagnosis of aganglionosis. The low accuracy of the opaque enema is mainly due to technical errors during the examination process, the most common being high-pressure injection of the contrast liquid, leading to a dilatation in the terminal section of the rectum, and consequently to a false diagnosis of megarectum.

Table 2 - Clinical features in children with functional constipation (n=47) and secondary constipation and innate neuronal dysfunction of the digestive tube (n=10) assisted in the ambulatory of Pediatric Gastroenterology at Hospital das Clínicas, Universidade Estadual de Campinas, in 1996/1997

	Functional constipation (n=47)	**Malformation of the myenteric plexus (n=10)**	**P**
Age (months)[‡]	53.1±14	50.6±12.1	>0.05*
Beginning of complaint on the 1st week of life	21	08	<0.05[†]
Duration of the symptoms (months)[‡]	49.5±27.3	49.1±47.2	>0.05*
Soiling	31	0	<0.001[†]
Abdominal distention (n=43)	27	7	>0.05†
Height/age Z score	0.981±0.941	0.803±1.243	>0.05*
Weight/age Z score	1.217±1.173	1.326±0.841	>0.05*
Weight/height Z score	0.124±1.004	0.201±1.187	>0.05*

* t Student   [†] chi-square   [‡] median±SD

Table 3 - Results of manometric studies versus results of the rectum biopsy or clinical evolution of constipation. Sensibility, specificity and positive and negative-predictive values of the examination were calculated with basis on these data

		Anorectal manometry		
		Absence of reflex	Presence of reflex	Total
Definitive	Malformation of the myenteric plexus	10	0	10
diagnosis	Functional constipation	3	44	47
	Total	13	44	57

Anorectal manometry is based on the relaxation of the internal anal sphincter as a reaction to the distension of the rectum wall. This reflex is identified in normal individuals and is absent in cases of Hirschsprung's disease, regardless of disease extension, and in all the diseases in which a dysfunction of the myenteric plexus occurs.[20]

Assessments of anorectal manometry accuracy for diagnosis of Hirschsprung's disease with ultrashort segment, performed in both neonates and older children, have identified a sensitivity varying from 75 to 100%, and a specificity varying from 95 to 98%.[5-6] False-positive results (incorrect diagnoses of aganglionosis) have been more frequently found in neonates and attributed to the immaturity of the myenteric nervous system.[14]

In the present study, rectal biopsy revealed the presence of aganglionic cells in the myenteric plexuses of three children without the rectosphincteric reflex. In these cases, the physician must be alert to the possibility of congenital megacolon in its ultrashort form, which requires (as other forms of megacolon) surgical treatment. In the case of an ultrashort megacolon, anorectal manometry is the only diagnostic tool available. In turn, the presence of rectum dilatation (megarectum) in functional constipation may interfere with the rectosphincteric reflex, so that diagnosis requires caution.[25] This probably explains the diagnosis of absent reflex in children with a normal biopsy, since a later examination revealed the presence of internal sphincter relaxation in the three cases. The favorable clinical evolution of these patients confirmed the diagnosis of functional constipation.

The research of rectosphincteric reflex in the group of patients analyzed in this study did not reveal false-negative results, that is, the test was sensitive to identify all the patients affected (sensitivity of 100%).

Rectum biopsy was not performed in patients whose rectosphincteric reflex was demonstrated. The diagnostic criterion for confirmation of functional constipation was the re-establishment of regular intestinal function in the 24 months after the manometric study.

An important information resulting from the analysis of the present sample is that 17.5% of the patients who were under clinical treatment presented forms of intestinal constipation that could be surgically corrected. Anticipating the diagnosis in these children means shortening the course of the disease, sparing the patient a long treatment based on bowel irrigation. Clinical assessment of this group of patients showed that children without soiling and whose history begins in the 1st week of life are at higher risk for carrying anomalies in the myenteric neural plexuses, and consequently should be submitted to manometry earlier.

According to the literature, the proportion of children with chronic constipation and absence of rectosphincteric reflex ranges widely depending on the type of sample studied; variations from 5%[26,27] to 10% have been reported.[28] It is possible that the relatively high percentage identified in the present sample reflects the severity of the disease in this group of patients referred to a tertiary hospital. The prevalence of constipated children with dysfunction of the myenteric plexuses in less specific samples is probably lower.

In short, the presented data allow us to conclude that the anorectal manometry is an adequate diagnostic test for the identification of Hirschsprung's disease or of intestinal neuronal dysplasias in patients with chronic constipation. Manometry should be included in the investigation of patients who do not respond satisfactorily to the initial clinical treatment. Given its high sensitivity, we believe that rectal biopsy is unnecessary in cases in which manometry demonstrates the presence of rectosphincteric inhibitory reflex.

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