be considered, particularly when the patient's symptoms persist, as described in this case.

Seung Han Kim, MD
Hoon Jai Chun, MD, PhD
Division of Gastroenterology and Hepatology, Department of Internal Medicine, Korea University College of Medicine, Seoul, Korea

REFERENCES

1. Kochman ML, McClave SA, Boyce HW. The refractory and the recurrent esophageal stricture: a definition. *Gastrointest Endosc*. 2005;62:474–475.
2. de Moura EGH, Toma K, Goh KL, et al. Stents for benign and malignant esophageal strictures. *Ann Ny Acad Sci*. 2013;1300:119–143.
3. Didden P, Spaander MC, Bruno MJ, et al. Esophageal stents in malignant and benign disorders. *Curr Gastroenterol Rep*. 2013;15:319.
4. Siersema PD, de Wijkerslooth LRH. Dilation of refractory benign esophageal strictures. *Gastrointest Endosc*. 2009;70:1000–1012.
5. Kim JH, Song HY, Choi EK, et al. Temporary metallic stent placement in the treatment of refractory benign esophageal strictures: results and factors associated with outcome in 55 patients. *Eur Radiol*. 2009;19:384–390.
6. van Halsema EE, Song LMWK, Baron TH, et al. Safety of endoscopic removal of self-expandable stents after treatment of benign esophageal diseases. *Gastrointest Endosc*. 2013;77:18–28.
7. Siersema PD. Treatment options for esophageal strictures. *Nat Clin Pract Gastr*. 2008;5:142–152.
8. Siersema PD. Stenting for benign esophageal strictures. *Endoscopy*. 2009;41:363–373.
9. de Wijkerslooth LRH, Vleggaar FP, Siersema PD. Endoscopic management of difficult or recurrent esophageal strictures. *Am J Gastroenterol*. 2011;106:2080–2091.
10. Siersema PD, Hirdes MMC. What is the optimal duration of stent placement for refractory, benign esophageal strictures? *Nat Clin Pract Gastr*. 2009;6:146–147.
11. Vleggaar FP, Siersema PD. Stents for benign esophageal strictures. *Tec Gastrointest Endosc*. 2010;12:231–236.
12. Eloubeidi MA, Lopes TL. Novel removable internally fully covered self-expanding metal esophageal stent: feasibility, technique of removal, and tissue response in humans. *Am J Gastroenterol*. 2009;104:1374–1381.

Role of Prucalopride in Treatment of Chronic Constipation and Recurrent Functional Obstruction in a Patient With Steinert Myotonic Dystrophy

To the Editor:

Steinert myotonic dystrophy (dystrophia myotonica type 1 or DM type 1) is an autosomic dominant disorder caused by a trinucleotide repeat expansion, in the 3′-untraslated region of the *DMPK* gene (dystrophia myotonica protein kinase) on chromosome 19q13.3.[1] This disorder is characterized by myotonic phenomena, progressive muscular weakness, and wasting, involving predominantly facial, semidistal, and distal compartments. Nevertheless, patients may show a multiorgan involvement with abnormalities of endocrine system (diabetes, thyroid dysfunction, hypogonadism), heart system (arrhythmias, ischemic disease, mitral valve prolapse), cataract, cognitive impairment, and mental retardation.[2] Gastrointestinal (GI) tract may also be involved with motility abnormalities that can occur at any level. Small and large bowel may be affected by a wide range of symptoms, from chronic constipation due to "inertia coli" to intestinal pseudoobstruction, sigmoid volvulus, and megacolon.[3]

A 37-year-old female patient with diagnosis of DM type 1 was seen as an outpatient in our Department for chronic constipation and recurrent intestinal pseudoobstruction. Myotonic dystrophy was first diagnosed when the patient was 25, onset symptoms being represented by distal muscle weakness (orbicularis and perioral muscles) and myotonic phenomena involving hand muscles. EMG reports, at the time of diagnosis, showed electrical myotonia and conduction delay. More, the presence of a familiar pattern (a brother diagnosed as DM type 1 based on genetic test) led to the definitive diagnosis. However, no significant worsening of neuromuscular skills has been noted from the time of diagnosis to date. When she was 27, she developed a symptomatic hypothyroidism that was controlled by replacement therapy. No involvement of cardiovascular system has been found during these years: she has never had clinical symptoms and ECG and echocardiogram, practiced, respectively, every 6 and 12 months since diagnosis, have never shown any abnormality. However, GI system involvement might be suspected: since she was 18, she had recurrent period of chronic constipation (bowel movements < 3/wk). In 2010, she had an admission in a surgical department cause of severe abdominal pain; abdomen was distented and x-ray showed colonic dilatation with multiple air-fluid levels. Emergent surgery was required: a volvulus of sigmoid colon was found and a loop-descending colostomy was performed. Colon biopsy showed only submucosa edema and a mild muscular hyperplasia. Colostomy was reversed after 1 month with a small partial sigmoid resection and side to side anastomosis. A severe chronic constipation appeared again 1 year after the operation (bowel movements < 1/wk) and she experienced 12 episodes of intestinal pseudoobstructions in a 2-year time: all of them needed admission in surgical capacities and were treated with conservative measures: restriction of oral intake, nasogastric tube, intravenous fluid, and decompressive endoscopy or rectal probes. Maintenance therapy with daily enemas, daily assumption of polyethylene glycol preparations and prokinetic agents (trimebutine) failed to avoid new episodes of intestinal pseudoobstruction. Finally, she was seen as outpatient at our Department asking for a total colectomy. However, a second-line treatment with a new prokinetic agent (Prucalopride 2 mg/die) was attempted. The woman was reviewed in clinic after 3 weeks. She reported that a normal defecation, with daily stool frequency, was restored a couple of weeks after the beginning of the treatment. Patient was suggested to keeping up with the therapy and no changes in the new defecation habitus have been noted in a 6-month follow-up time, with no more episodes of intestinal pseudoobstruction.

The authors declare that they have nothing to disclose.

DISCUSSION

GI tract is frequently involved in Steinert myotonic dystrophy (up to

28% of patients may be affected). It may also be the first symptom of the disease and sometimes it precedes skeletal muscle involvement by 10 years; moreover, there is a low correlation among the degree of GI disturbances, the severity of muscle involvement, and duration of the disease.[3]

Clinical manifestations may involve GI tract at any level[3]; in particular small and large bowel may be involved with reduced or absent peristaltic and/or segmentary activity with delayed intestinal transit[4] leading to chronic constipation; the clinical scenario may change from diarrhea (bacterial overgrowth due to reduced peristaltic activity) to megacolon; recurrent intestinal pseudoobstruction and sigmoid volvulus due to myotonic contractions may also occur.

The pathophysiological mechanism of these digestive motor disorders is not clear.

This has generally been attributed to smooth muscle damage,[5] but evidence is poor and not always supported by histologic patterns.[6]

Recent studies suggest that neurological alterations might play a role[7]: in particular, neural dysfunctions, such as alteration in the nonadrenergic noncholinergic neuronal control of the GI tract, have been suggested[7] to explain symptoms and instrumental findings that may be present even in the absence of definite histologic damage; nevertheless, the degeneration of argyrophilic neurons in the colonic myoenteric plexus has also been found in a case of Steinert myotonic dystrophy complicated by megacolon.[6]

Despite its pathogenesis, chronic constipation in these patients is usually treated with laxative, enemas, and prokinetics.[8] Cisapride, a nonselective 5-HT4 receptor agonist, has shown to improve intestinal functions in these patients.[9] However, its arythmogenic effect has led to its withdrawal; more, the possibility of heart involvement in Steinert Myotonia patients[2] was also considered an additional risk for adverse event. Prucalopride belongs to the class of benzofurancarboxamide agonists, which have high affinity and selectivity for the 5-HT4 receptor.[10] The action on this receptors leads to an activation of cholinergic and nonadrenergic, noncholinergic neurotransmission by entheric neurons[11]: this might explain its effect on GI disorders in DM type I, considering the pathogenic mechanisms which were described earlier. Safety profile regarding cardiovascular adverse effects has been assessed in several trials.[10] This may be due to lack of interaction with the hERG channel or 5ht1 receptors, which were considered to be responsible for the development of adverse cardiovascular effects with other 5-HT4 receptor agonist.[12]

This is the first report, to our knowledge, that clearly shows the effect of prucalopride in prompt resumption of GI symptoms in a Steinert disease patient. Bigger series are probably needed to confirm this finding. This might also represent an important step in a better understanding of the physiopathology and GI disorders in Steinert myotonic dystrophy.

Mariano Cesare Giglio, MD*
Gaetano Luglio, MD*
Rachele Tarquini, MD*
Domenico Cerbone, MD*
Paolo Bucci, MD†
Luigi Bucci, MD*
*Dipartimento di Medicina Clinica e Chirurgia
†Dipartimento di Neuroscienze e Scienze della Riproduzione e Odontostomatologia
Università degli Studi di Napoli "Federico II," Napoli, Italy

REFERENCES

1. Brook JD, McCurrach ME, Harley HG, et al. "Molecular basis of myotonic dystrophy: expansion of a trinucleotide (CTG) repeat at the 3′ end of a transcript encoding a protein kinase family member". *Cell*. 1992;68(no. 4): 799–808.
2. Romeo V. Myotonic dystrophy type 1 or Steinert's disease. *Adv Exp Med Biol*. 2012;724:239–257.
3. Bellini M, Biagi S, Stasi C, et al. Gastrointestinal manifestations in myotonic muscular dystrophy. *World J Gastroenterol*. 2006;12(no. 12): 1821–1828.
4. Goldberg HI, Sheft DJ. Esophageal and colon changes in myotonia dystrophica. *Gastroenterology*. 1972; 63(no. 1):134–139.
5. Nowak TV, Ionasescu V, Anuras S. Gastrointestinal manifestations of the muscular dystrophies. *Gastroenterology*. 1982;82(no. 4):800–810.
6. Yoshida MM, Krishnamurthy S, Wattchow DA, et al. Megacolon in myotonic dystrophy caused by a degenerative neuropathy of the myenteric plexus. *Gastroenterology*. 1988; 95(no. 3):820–827.
7. Lecointe-Besancon I, Leroy F, Devroede G, et al. A comparative study of esophageal and anorectal motility in myotonic dystrophy. *Dig Dis Sci*. 1999;44(no. 6):1090–1099.
8. Bujanda L, López de Munain A, Alcón A, et al. The gastrointestinal changes in dystrophia myotonica. *Rev Esp Enferm Dig*. 1997;89(no. 9): 711–714.
9. Brunner HG, Hamel BC, Rieu P, et al. Intestinal pseudo-obstruction in myotonic dystrophy. *J Med Genet*. 1992; 29(no. 11):791–793.
10. Tack J, Camilleri M, Chang L, et al. Systematic review: cardiovascular safety profile of 5-HT(4) agonists developed for gastrointestinal disorders. *Aliment Pharmacol Ther*. 2012;35(no. 7):745–767.
11. Wong BS, Manabe N, Camilleri M. Role of prucalopride, a serotonin (5-HT(4)) receptor agonist, for the treatment of chronic constipation. *Clin Exp Gastroenterol*. 2010;3:49–56.
12. Potet F, Bouyssou T, Escande D, et al. Gastrointestinal prokinetic drugs have different affinity for the human cardiac human ether-à-gogo K(+) channel. *J Pharmacol Exp Ther*. 2001; 299(no. 3):1007–1012.

Chronic Hepatitis E Virus Infection

Challenges in Diagnosis and Recognition in the United States

To the Editor:

Chronic hepatitis E virus (HEV) infection in immunocompromised patients due to HIV infection, cancer chemotherapy, and solid-organ transplantation is unusual and is a particularly underrecognized condition.[1] The lack of commercially available, FDA-approved serologic tests can hinder timely diagnosis. This presents a challenge and may result in underrecognition of chronic HEV in populations at risk. Herein, we report a case of chronic HEV in a renal transplant recipient who had indeterminate chronic hepatitis that was

K.R.R.: concept/design, data analysis/interpretation, drafting article, critical revision of the article, and approval of the article; T.M.P.: drafting the article, critical revision of the article, and approval of the article; and D.A.H.: drafting the article, data collection, approval of the article, and critical revision of the article.The authors declare that they have nothing to disclose.