The cough without a cause: Habit cough syndrome

To the Editor:

Cough is a common symptom for which children and adolescents seek medical care. Although often self-limited, coughing can be prolonged. When present for 4 or more weeks, the cough is considered to be chronic. Although there is an extensive differential diagnosis of common and uncommon causes of chronic cough,[1] cough can be functional in nature (ie, without an organic basis).

In 1991, we described the clinical characteristics and outcome of a disorder called habit cough (HC) in 9 children seen in our pediatric allergy and pulmonary clinic.[2] Since then, physicians in our clinic have used a consistent approach to diagnosis and treatment of HC based on the observations of that initial report. Beginning in 1995, the diagnosis of HC was entered into the problem list of our electronic medical records. This permitted us to identify the demographics and outcomes of 140 such patients with this disorder at a single center over a 20-year period. Approval for this retrospective study of HC in our patient population was obtained from the University of Iowa Institutional Review Board.

The search for HC in the problem list of our electronic medical records from mid-1995 to mid-2014 initially resulted in 183 charts of children and adolescents. All clinic records for the visit when HC was initially entered into the problem list were then individually examined to determine the adequacy of information to support the diagnosis and provide details of treatment. Insufficient descriptive information resulted in exclusion of 43 patient records.

Diagnosis of the 140 pediatric patients with sufficient descriptive information had been consistently based on the history of repetitive coughing, which was defined as up to several times per minute for extended periods from several hours at a time to all day but with complete absence when asleep. In general, although initial spirometry was performed, diagnostic tests were not needed to support the diagnosis, which was based on history and observation. Fifty-eight percent of the 140 patients were male. Ages ranged from 4 to 18 years, with a median age of 10 years (Fig 1). Most had a cough characterized by a loud barking sound; a repetitive softer throat-clearing sound was present in 10% of the patients, and 11% exhibited both patterns of coughing at times. Duration of cough before the initial visit to our clinic averaged 4 months (median) and ranged from less than 1 month to periods in excess of 1 year (Fig 2). Frequent use of albuterol, oral corticosteroids, montelukast, inhaled corticosteroids, various antibiotics, gastric acid suppressants, and cough suppressants was described in the histories of the clinical notes. Many had frequent unscheduled medical care visits, and 4 had been hospitalized for the cough.

Repetitive cough was directly observed during the clinic visit in 85 of the patients.

Using the suggestion therapy approach described in Table I, complete cough cessation occurred during 15 to 30 minutes in 81 (95%) of 85 of those patients. Autosuggestion instruction was then provided by ensuring the patient that the same principles could be applied at home, if needed for recurrence

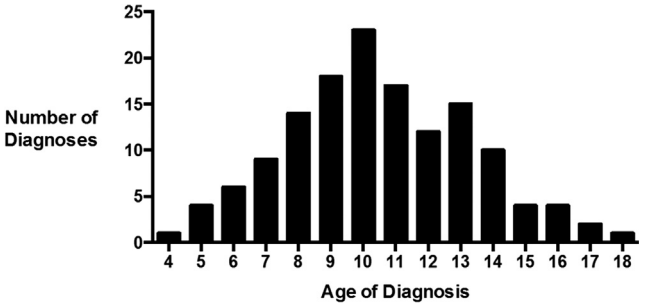

FIG 1. Age in years of the 140 children and adolescents at the time of HC diagnosis.

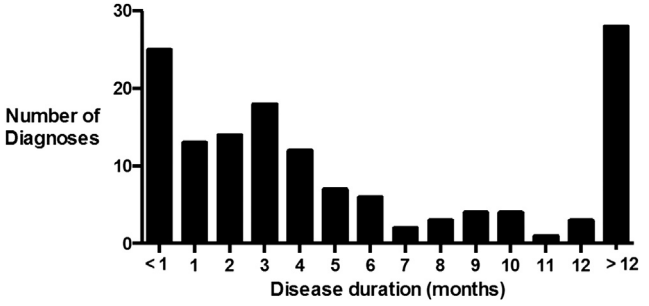

FIG 2. Duration of coughing before diagnosis of HC in our clinic among the 140 children and adolescents.

(see the footnote of Table I). Families were provided with specific contact information and encouraged to contact us if troublesome cough returned; none indicated return of the cough. Although we did not specifically survey these patients, sustained benefit with extended periods free of cough were seen in our previous report[2] and in 2 other case series.[3,4]

This study demonstrates that HC could be recognized, diagnosed, and treated promptly by several physicians in a pediatric allergy and pulmonary clinic over a 20-year period. The cough of this disorder is often associated with considerable morbidity. It is readily identified and stopped in most children with sustained effect by using a simple behavioral measure that should be used by any physician encountering this type of cough in a pediatric patient. In contrast to the successful cessation of cough in 95% of those coughing in this report, a study at the Mayo Clinic (Rochester, Minn) reported 60 patients given a diagnosis of "childhood involuntary cough syndrome" with a prior mean duration of 7.6 months of cough.[5] Because no behavioral treatment was made after diagnosis in these children, this was essentially a natural history outcome study. Cough in 44 (73%) of those patients persisted for an average of 6.1 additional months before spontaneous resolution. The other 16 (27%) were still coughing a mean duration of 5.9 years later.

The take-home message is that clinicians should recognize the unique characteristics of HC as distinguished from the many other causes of chronic cough[1] to avoid extensive and expensive evaluative measures and unnecessary pharmacotherapy. Clinical criteria, consisting of a repetitive cough up to several times per minute for extended periods of time that is totally absent once

TABLE I. Major elements of suggestion therapy

- Approach the patient with confidence that the coughing will be stopped.
- Explain the cough as a vicious cycle that started with an initial irritant that is now gone and that now the cough itself is causing irritation and more cough.
- Instruct the patient to concentrate solely on holding back the urge to cough for an initially brief timed period, such as 1 minute. Progressively increase this time period and use an alternative behavior, such as sipping lukewarm water or inhaling a soothing cool mist from a vaporizer, to "ease the irritation."
- Tell the patient that each second the cough is delayed makes it easier to suppress further coughing.
- Repeat expressions of confidence that the patient is developing the ability to resist the urge to cough: "It's becoming easier to hold back the cough, isn't it?" (Nodding affirmatively generally results in a similar affirmation movement by the patient.)
- When ability to suppress cough is observed (usually by about 10 minutes), ask in a rhetorical manner, "You're beginning to feel that you can resist the urge to cough, aren't you?" (said with an affirmative head nod)
- Discontinue the session when the patient can repeatedly respond positively to the question, "Do you feel that you can now resist the urge to cough on your own?" This question is only asked after the patient has gone 5 minutes without coughing.
- Express confidence that if the urge to cough recurs that the patient can do the same thing at home (autosuggestion).*

*Autosuggestion involved expressing confidence in 15-minute sessions at home concentrating on holding back the cough with sips of lukewarm water to "ease the irritation causing cough."

asleep is sufficient to make the diagnosis of this disorder. Once identified, a simple behavioral methodology can provide sustained cessation of cough for most children with this disorder.

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Dual response to dietary/topical steroid and proton pump inhibitor therapy in adult patients with eosinophilic esophagitis

To the Editor:

Eosinophilic esophagitis (EoE) is a common cause of chronic esophageal symptoms characterized by an eosinophil-rich inflammatory infiltrate limited to the esophagus.[1] Despite being first categorized as a distinct clinicopathological disorder 2 decades ago,[2,3] EoE has rapidly become recognized in recent years as the most prevalent cause of chronic dysphagia among children and young adults in Western countries.[4-6]

Increasing knowledge of the disease has led to gradual changes in interpreting the eosinophilic infiltration within the esophageal mucosa, and these changes are reflected in the differing guidelines categorizing EoE since 2007.[1,7-9] Until the early 1990s, a dense esophageal eosinophilia was mostly associated with gastroesophageal reflux disease (GERD).[E1,E2] The ineffectiveness of antireflux therapies in patients with characteristic EoE profile, however, led to the recognition of EoE as a new entity.[2,3,E3] Consensus guidelines published in 2007 rigidly separated EoE from GERD[7]: EoE was defined by either clinical and/or histologic unresponsiveness to proton pump inhibitor (PPI) therapy or a normal esophageal pH, whereas GERD was defined by either complete remission on PPI therapy or a pathological esophageal pH.

Soon after this distinction was made, a few retrospective studies suggested the existence of pediatric patients with clinicopathological features of EoE who fully responded to PPI therapy.[E4-E6] A large prospective adult series published in 2011 corroborated this finding, showing that PPIs effectively induced remission of both esophageal inflammation and accompanying symptoms in 50% of the patients with a presumptive diagnosis of EoE.[E7] Notably, most of these patients presented with an associated atopic background as well as symptoms of dysphagia and food impaction instead of heartburn. Furthermore, PPI responsiveness was independent of pH-monitoring results. These observations gave rise to the new "PPI-responsive esophageal eosinophilia (PPI-REE)" concept, which referred to patients who not only appeared to have EoE clinically but also achieved complete remission after PPI therapy. This novel phenotype was recognized in the 2011 updated consensus recommendations on EoE[1] and endorsed in all subsequent guidelines.[8,9]

Currently, several retrospective and prospective studies in both children and adults consistently show that at least one-third of the patients with suspected EoE eventually receive a PPI-REE diagnosis.[E8] Interestingly, PPI-REE and EoE remain indistinguishable based on clinical, endoscopic,[E9,E10] and histologic findings[E11]; pH monitoring[E17]; and the measurement of tissue markers,[E12,E13] cytokines related to eosinophilic inflammation,[E14,E15] and esophageal gene transcripts from esophageal tissue.[E16] In addition, PPI monotherapy completely reverses cytokine[E14,E15] and esophageal gene transcript[E16] levels in patients with PPI-REE, similar to the way that topical steroids do in EoE. Collectively, these data support the idea that PPI-REE may constitute a subphenotype of EoE rather than a distinct disease entity and that PPIs may be considered a therapeutic option to effectively manage a high proportion of patients with EoE.

Here, we provide additional evidence to support that PPI-REE and EoE should be considered within the spectrum of the same disease by showing that patients with PPI-REE also respond to dietary/topical steroid treatment and that some patients with EoE respond to PPI therapy. Databases containing information on patients with an EoE diagnosis who had prospectively attended 2