

Speech Pathology Management of Chronic Refractory Cough and Related Disorders

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Anne E. Vertigan

Peter G. Gibson

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Foreword

“I am so frustrated.” This is a phrase that we hear very frequently in our clinics when evaluating patients with chronic cough. It is also frequently mentioned by our colleagues, health care professionals in many specialties who often feel helpless when treating these patients. This welcome book alleviates the frustration as it helps to provide a better understanding of the diagnosis and treatment of this condition, the cough that has no clear etiology, the cough that persists after most common causes of cough have been ruled out, the cough that lingers for years after traditional medical management options have been exhausted.

Imagine yourself avoiding going out to a restaurant or to a movie, missing important family gatherings, staying home instead of going out with friends, being embarrassed to go to church. Imagine further, having coughing fits so strong that your chest hurts from breaking a rib, fear of becoming incontinent, throwing up, or passing out, often without triggers or knowing how to prevent it. This is the reality that countless patients who suffer from chronic refractory cough, their families and coworkers experience on a daily basis. This condition can become debilitating, leading to depression and social isolation. The health-care community is often at a loss to understand the causes and provide relief to patients who suffer from it.

The last decade has seen a surge in the interest in this type of cough, from research attempting to better characterize the pathophysiology, to development of new therapeutic modalities, including drug trials and behavioral interventions. Anne Vertigan and Peter Gibson have been at the forefront of this important work.

I met Anne Vertigan at the Chronic Cough Conference in New York in 2013, and later at the 2nd Australasian and Asia Pacific Laryngology Congress in Hobart, Australia. I was impressed with her body of work, and appreciated her passion for the pursuit of answers to help cough patients. She has highlighted the critical role of Speech Pathology in the evaluation and treatment of patients with chronic cough and related conditions.

Speech Pathologists are now routinely key players in interdisciplinary teams and work closely with allergy, pulmonary and ear, nose and throat specialists

interested in these patients. This collaboration is due in no small part because of Anne Vertigan's work.

The nature of the book is comprehensive. It summarizes the current understanding of the pathophysiology of laryngeal conditions that include refractory cough, laryngeal hypersensitivity and hyperreactivity, and paradoxical vocal fold motion. It includes the perspectives from pulmonary and ear-nose and throat specialties, covers current treatment modalities, and highlights a detailed protocol for behavioral treatment of these disorders. The group of collaborators is top notch, and all share a similar interest and dedication in expanding knowledge and evidence based approaches for cough management.

To date, there have been few resources to help Speech Pathologists who have an interest in chronic cough. This book fills this void. I am sure it will become the reference standard for years to come. It clearly achieves its goal to describe and define chronic refractory cough, to help clinicians in their interdisciplinary efforts to address the condition, and most importantly, to provide Speech Pathologists with a comprehensive guide to understand and treat this difficult patient population.

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Preface

We started working with patients with chronic refractory cough and paradoxical vocal fold movement over twenty years ago. At that stage, there was little published information about speech pathology treatment for these conditions, with the majority of the evidence reported in small case series. In fact, we even questioned whether speech pathology treatment was appropriate for these patients.

Over the last twelve years, there has been growing interest amongst speech pathologists in the assessment and treatment of these conditions and an expansion in the evidence for successful treatment. While management of patients with chronic refractory cough is an expanding area of speech-language pathology practice, clinical exposure to the treatment of this condition is variable. Details regarding the theory and treatment of chronic cough have been published in several different fields including respiratory physiology, respiratory medicine, speech pathology, and otolaryngology. This book aims to consolidate this information in order to inform speech pathologists who manage patients with chronic refractory cough and related laryngeal conditions.

This work has developed from our own clinical research and experience working with patients who have chronic refractory cough, however we are constantly learning from other clinicians and researchers as new developments are made in the treatment of these conditions. Three renowned authors, Tom Murry, Ken Altman and Marc Haxer, have contributed chapters to this textbook.

This book has been designed to assist speech pathologists managing patients with chronic refractory cough from the receipt of the initial referral, through to discharge. The book combines theoretical information about cough, with practical advice and tools. The first seven chapters of the book describe the physiology of cough, medical treatment, related laryngeal conditions and hypersensitivity. The remaining chapters describe the treatment approach for speech pathology management of chronic refractory cough and related laryngeal conditions. Although it may be tempting to begin reading this book with assessment and treatment chapters, we believe that it is beneficial to commence with the earlier chapters in order to understand the etiology and physiology of cough and associated medical conditions prior to commencing treatment. It is hoped that this

information will make speech pathology treatment for chronic refractory cough more accessible for patients.

This work has only been made possible with the support of our colleagues. We would particularly like to acknowledge the invaluable assistance of Sarah Kapela, speech pathologist at John Hunter Hospital, for her invaluable work as a research assistant and for advice regarding photography and layout. There are several other people we would like to thank, including Dr Josephine Smith from the University of Newcastle for her expertise in cranial nerves, Sienna Tuckerman, speech pathologist at John Hunter Hospital for assistance regarding cough physiology, Mark Rothfield from the Hunter Medical Research Institute, and Mary Aldrich and for photography and formatting. We would like to thank the Hunter New England Speech Pathology Evidence Based Practice group for designing the format upon which we based our clinical observation form. We appreciate the assistance of Professor Deborah Theodoros and Dr Alison Winkworth for their expertise in supervising the early research that underpinned this work. We would like to thank our colleagues in speech pathology, respiratory medicine and otolaryngology at John Hunter Hospital who have provided ongoing support and continued to raise questions to expand the work in this important area. Finally we would like to thank our patients who have taught us so much over the years.

Dr. Anne Vertigan
Prof. Peter Gibson

Dedicated to Jennifer Thomas, whose generosity in providing a grant to support our initial research has led to the development of our treatment program and paved the way for new developments in the field.

Introduction

Coughing is such a common experience that it in fact accounts for the largest number of outpatient health care visits.^{1,2} Yet because most cases of acute cough resolve spontaneously or with a single course of medical treatment, we barely give it a second thought. But chronic cough is another matter. This is defined as a cough that lasts for longer than eight weeks, and it is a significant health problem for many people.¹ Often lasting for months or even years, chronic cough has debilitating side effects including stress urinary incontinence, depression, poor sleep, headaches and reduced quality of life.^{3,4} Some patients resign from work. Many avoid social situations. Even simple activities such as conversing or using the telephone can become distressing because talking can trigger coughing episodes.⁵

The history of cough

The recent history of cough in the modern medical era was succinctly described by Song.⁶ Cough was previously thought to be associated with respiratory infection and chronic bronchitis resulting from transmitted infection, or exposure to airborne pollutants or tobacco smoke. It was subsequently found that not all patients fitted the respiratory infection paradigm and that cough may be due to other diseases such as asthma, rhinitis, and gastroesophageal reflux disease.

In 1990, Irwin^{7,8} described the Anatomic Diagnostic Protocol. This protocol recognised the wide sensory distribution of the vagus nerve, and proposed that sensory stimulation of any of the organs innervated by this nerve could lead to hyperstimulation and cough. This insight was then applied to diagnostics and the goal was to identify the associated disease causing cough in an individual patient. For example, asthma results in airway irritation and cough, rhinitis leads

to stimulation of vagal nerve endings in the upper airway, and gastroesophageal reflux causes acid induced esophageal inflammation and neural activation. The result of these diverse diseases is a common pathway of vagal sensory activation and cough. The Anatomic Diagnostic Protocol then directs treatment to the associated disease, as a means to resolve the cough.

This approach was reported to be successful in virtually all cases. Idiopathic or unexplained cough was diagnosed when cough persists despite complete workup and appropriate therapeutic trials.⁹ This entity has been questioned as a failure to systematically apply the Anatomic Diagnostic Protocol. However, McGarvey¹⁰ and Haque *et al.*¹¹ postulated that idiopathic cough was a distinct entity with unknown underlying pathophysiology and no treatment options. Idiopathic cough is more likely preceded by an upper respiratory tract infection, is usually longer duration of cough and shows increased sensitivity to capsaicin.

More recently, in 2010, the notion of Cough Hypersensitivity Syndrome was introduced.¹² This hypothesis views chronic cough as a single syndrome with the underlying etiology of cough hypersensitivity. The Cough Hypersensitivity Syndrome theory reconceptualises commonly associated diseases, such as asthma, which were previously thought to cause cough, as cough triggers in an individual with underlying hypersensitivity. Another concept is the existence of a second lesion which causes cough persistence despite treatment. This could be (1) central sensitisation of the cough reflex, or (2) extrathoracic airway hyper-responsiveness, or both. These paradigm shifts are summarised in Table 1.1.

The role of speech pathology

Speech pathologists have an important role to play in the treatment of patients with chronic cough. It is an emerging area of speech pathology practice that provides a valuable treatment option for patients who may have exhausted medical treatment for their condition. There may also be a role for speech pathologists in non-pharmacological management of cough in a range of other conditions. However, further research is required to establish this.

Speech pathology treatment for chronic cough improves the individual's control over cough, reduces cough symptoms and associated symptoms of dysphonia, and paradoxical vocal fold movement (described in Chapters 7 and 11). This treatment results in improved cough symptom ratings,¹³ acoustic and auditory perceptual voice results,¹⁴ cough frequency, and capsaicin cough reflex sensitivity.¹⁵

	< 1990	1990	2010
Prevailing concept	Cough as a response to environmental irritants.	Stimulation of afferent limb of cough reflex.	1. Central reflex sensitization. 2. Laryngeal hypersensitivity.
Cause	Disease. Infection. Tobacco. Smoke.	Asthma. Gastroesophageal reflux. Rhinosinusitis	Sensory neuropathy Central sensitization
Treatment	Antibiotics. Smoking cessation.	Proton pump inhibitors. Inhaled corticosteroids. Antihistamines. Topical steroids.	Centrally acting neuromodulators. Speech pathology.

Table 1.1: Incremental concept developments in chronic refractory cough

There are several reasons why speech pathology treatment is appropriate for chronic cough. Speech pathology management of chronic cough employs techniques adapted from those used to treat hyperfunctional voice disorders^{16,17} and involves teaching individuals over a number of sessions to control a function previously considered automatic and outside of their control. Speech pathology management of hyperfunctional voice disorders is effective in teaching patients to reduce phonotraumatic behaviours such as coughing and throat clearing.¹⁸ This is important because such behaviours may lead to ongoing tissue damage and cough persistence. Other techniques used in the management of chronic cough and paradoxical vocal fold movement have included pitch change, diaphragmatic or abdominal breathing, focusing on expiration, reducing extrinsic muscle tension, relaxation and sniffing to reduce improper vocal fold adduction.¹⁷ Stemple claimed that the combination of a detailed knowledge of the upper airways and proven ability to effect behavioural change make speech pathologists excellent professionals to treat this condition. Speech pathologists have extensive experience in education and training in respiratory physiology and modifying laryngeal behaviour.¹⁹ Mathers-Schmidt²⁰ argued that the speech pathologist's knowledge in the areas of voice, swallowing, and motor speech disorders

prepares clinicians to detect abnormalities in laryngeal and respiratory functions and to teach laryngeal and respiratory control techniques. Finally, laryngeal dysfunction is implicated in the pathogenesis of chronic cough. This concept will be elaborated on in Chapter 7.

The aim and outline of the book

The aim of this book is to equip speech pathologists to manage patients presenting with chronic cough. Management of chronic cough is an expanding area for speech pathologists, yet many have limited clinical exposure to the treatment of this condition. Furthermore, while the comprehensive details of the theory and treatment of chronic cough are known, they have been published in a diverse range of separate documents in the fields of respiratory physiology, respiratory medicine, speech pathology, and otolaryngology, and as such they are not readily available in a combined form. Knowledge of the etiology and physiology of cough and associated medical conditions is essential for speech pathologists.

This book aims to describe the theoretical basis of this growing area of clinical practice, to describe a new approach to the problem, and provide clinicians with tools that can aid their practice. It will outline the assessment and treatment process to manage the condition. While knowledge of voice disorders is invaluable in the treatment of chronic cough, a specific framework for assessing and treating chronic cough is frequently required. These concepts will be outlined in this book.

Laryngeal hypersensitivity is emerging as an important concept in chronic refractory cough. It is the target of mechanisms studies, novel therapies and also novel pharmacological developments. Recently published evidence for laryngeal hypersensitivity in chronic refractory cough will be outlined along with new approaches for measuring this component of the disorder.

Speech pathologists, referring medical specialists and patients have expressed a need for more specialized speech pathology services for the management of chronic cough. In particular, speech pathologists are requesting more comprehensive and cohesive information to guide their patient management. As more speech pathologists are required to treat patients with chronic refractory cough, there is a need for more information regarding treatment of this disorder.

This book describes the pathophysiology of chronic cough and the speech pathology assessment and treatment the condition. The first seven chapters provide theoretical information about chronic cough which is not easily accessible

to most speech pathologists. This information includes the definition and clinical presentation of the patient with chronic cough, associated medical conditions and laryngeal disorders associated with chronic refractory cough, and the otolaryngology and respiratory medicine management of the condition. A chapter on pulmonary function testing has been included to assist the speech pathologist interpret and understand pulmonary function test results.

The remaining chapters (8 to 13) include a detailed outline of the speech pathology assessment and treatment of chronic cough. It describes the evidence base for behavioural treatment, detailed instructions regarding selection and use of treatment techniques along with user friendly resources. Information about the management of related laryngeal conditions such as paradoxical vocal fold movement and globus pharyngeus has been included as these conditions frequently co-occur with chronic cough and there are several overlaps in the treatment of the conditions. It may be tempting for the speech pathologist to commence reading at this latter section. However we argue that the speech pathologist should be familiar with the physiology of cough and associated medical conditions in order to effectively treat the conditions. The anatomy and physiology of speech, phonation and respiration at an undergraduate speech pathology level is presumed as is competency in the assessment and treatment of speech, voice and swallowing disorders.

Terminology and definitions

Cough

A cough can be defined as “an airway defensive reflex consisting of an inspiratory phase followed by a forced expiratory effort initially against a closed glottis, followed by active glottal opening and rapid expiratory flow”^{21 (page S3)}. Cough has an important role in protecting the airway from foreign bodies and aspiration, removing noxious substances and increasing mucociliary clearance. Cough is particularly important in protecting the airway during swallowing in individuals with aspiration resulting from oropharyngeal dysphagia.

Cough can become a troublesome symptom leading people to seek medical assistance. Under this situation, temporal definitions are applied to the classification of cough, such that cough is defined as acute, or chronic. Cough may also be classified as subacute when it lasts for an intermediate period of time. This classification is more frequently used in paediatrics. It is discussed in more detail

in Chapter 5. These terms are generally restricted to clinical definitions such as when cough becomes a symptom that the individual complains about.

Acute cough

An acute cough is defined as a cough that resolves within three weeks.²² Most cases of acute cough are due to upper or lower respiratory tract infection. These cases may resolve spontaneously or following appropriate pharmacotherapy. Speech pathologists are seldom required to provide intervention for patients with acute cough.

Chronic cough

Chronic cough is simply defined as a cough persisting for longer than eight weeks.^{23,9} Chronic dry cough without associated disease is considered to have no benefit to the respiratory system or the body in general.⁸ The majority of cases (80%) of chronic cough resolve after medical treatment. Medical treatment for chronic cough typically involves withdrawing Angiotensin Converting Enzyme-I inhibitors, if used, and treating coexisting asthma, gastroesophageal reflux, rhinosinusitis, or lung pathology.

Chronic refractory cough

Chronic refractory cough is a cough that has lasted for longer than eight weeks **and** is refractory to appropriate medical management. While the majority of cases of chronic cough respond to medical management, cough remains refractory to medical treatment in between 12% and 42% of cases.^{24,25,11} Some patients with chronic cough fail to respond to medical treatment despite extensive investigation.²⁶ It is this subset of patients with chronic refractory cough that requires speech pathology intervention.

It is important to distinguish between chronic cough that is responsive to medical treatment and chronic cough that persists despite medical treatment. Figure 1.1 outlines the model for conceptualising cough that does not respond to medical treatment. In this model, cough is classified as either acute or chronic depending upon the duration of symptoms. Chronic cough is then further classified as either cough that responds to medical treatment or cough that is refractory to medical treatment. Cough that fails to respond to medical treatment may be referred to as chronic refractory cough.

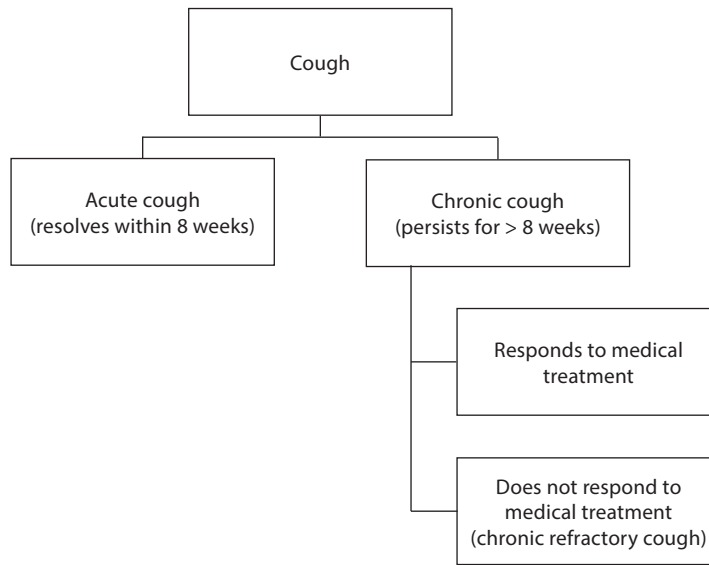


Figure 1.1: Model for the conceptualisation of chronic cough that does not respond to medical intervention.

A variety of terms have been used to describe chronic refractory cough in the literature. These terms include chronic idiopathic cough, chronic unexplained cough, cough hypersensitivity syndrome and chronic non-specific cough. These latter terms suggest that a cause for a cough cannot be found whereas the term chronic refractory cough suggests that the cause for cough is known but it remains refractory to medical treatment.

The difference in terminology makes research and common understanding of the condition difficult. Despite a lack of consensus around terminology to be used we have used the term *chronic refractory cough* to refer to cough that has lasted for longer than 8 weeks and which has either no known etiology, or has persisted despite medical management of a known etiology.

Chronic refractory cough may co-occur with paradoxical vocal fold movement and globus pharyngeus.²⁷ These conditions may be associated with laryngeal dysfunction and have an increased prevalence of impaired laryngeal sensation²⁸, voice disorders,²⁹ and paradoxical vocal fold movement. The etiology of chronic refractory cough is unknown. Research has shown it is not responsive to medical treatment including treatment of lung pathology, asthma, rhinosinusitis, and gastroesophageal reflux disease.¹¹

The burden of cough

Cough accounts for between 10% and 38% of respiratory outpatient visits in the USA.³⁰ The prevalence of chronic cough varies between studies: in the general population it is between 3% and 33%^{30,31,32} and as high as 40% in patients attending specialist cough clinics. In the general population, cough severe enough to interfere with every day activities is estimated to be 7%.³³ Although under-recognised, chronic cough is a significant health problem for many individuals. The incidence of chronic cough and the duration of the condition result in a significant disease burden.

Physical side effects of chronic cough

Physical side effects of cough include syncope, musculoskeletal chest wall pain, headache, difficulty speaking on the telephone, increased pressure on lumbar discs, laryngeal trauma, stress urinary incontinence, fatigue, damage to airway mucosa, exhaustion, vomiting, and rib fractures.^{34,35} Although not explicitly studied, cough may also contribute to the development of paradoxical vocal fold movement and laryngeal hypersensitivity.

Quality of life in patients with chronic cough

In addition to physical side effects, cough has a negative impact on quality of life. These effects include disruption to normal activity, interference with work and social relationships, worry about serious medical illness, frequent retching, exhaustion, embarrassment, self-consciousness, social isolation, prolonged frustration, guilt, depression, sleep deprivation, and lethargy.^{4,31,36-44} Effective treatment of chronic cough therefore has the potential to significantly improve the quality of everyday life for these patients.

Financial impacts of chronic cough

Chronic refractory cough also has significant financial implications. For the individual patient, the cost of medical consultations along with over the counter and prescribed medications is considerable. Reduced capacity for employment and work attendance can also have financial ramifications for some individuals. At the health service level, the costs of cough are frequently underestimated as patients rarely die and are seldom admitted to hospital. Nevertheless, the cost of physician consultations and emergency department visits is significant. There-

fore, it is important to have adequate treatment options available for individuals with chronic refractory cough.

Fortunately, the side effects of chronic cough are often relieved following successful medical treatment of this condition.⁴ However when cough persists despite medical treatment, the side effects are likely to continue thus exacerbating the impact of the condition. Further, the pathway to a final diagnosis of chronic refractory cough may be extended, and potentially lead to expensive or inappropriate medical treatments.⁴⁵ Murry reports that patients with chronic refractory cough have often been examined by several specialists before commencing speech pathology treatment.

Summary

Chronic cough is a significant clinical problem that is being increasingly managed by speech pathologists. Although cough resolves spontaneously or with medical treatment, it persists in a subset of patients. The development of the Anatomic Diagnostic Protocol and Cough Hypersensitivity Syndrome were significant developments in cough, and provided new avenues for the assessment and treatment of patients with the condition. Recently, the role of laryngeal hypersensitivity in cough has been recognised as an important component. These developments are significant as chronic refractory cough is a debilitating condition with significant impacts on quality of life.

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