SHORT COMMUNICATION

Cough suppression disorders spectrum

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Summary
Volitional cough suppression, identified exclusively in females, is an unusual causal mechanism for instances of lobar atalectasis and bronchiectasis. It is a postulated mechanism for the genesis of Lady Windermere Syndrome.

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Introduction

Cough. A convulsion of the lung vellicated by some sharp serosity.

Samuel Johnson. Dictionary, 1755.

The superaddition of cough to mucociliary clearance is critically important in preserving pulmonary sterility and ventilation. Volitional cough suppression (VCS) as a pathogenetic pathway for a variety of pulmonary disorders is infrequently encountered. It may be more common than is currently recognized.

Cough suppression disorders

Harris appears to be the first person to assign responsibility to VCS for instances of pulmonary disease. He reported a series of five otherwise healthy females, ages 8–66, with recurring pulmonary atelectasis, each of whom acknowledged VCS. Bronchoscopy revealed inspissated sputum at proximal levels. Further episodes were averted by author’s admonition to cough freely [1].

Wells et al. characterized the syndrome of bronchiectasis attributable to VCS under the rubric, “atussis nervosa”, alluding to the gender and psychological parallels with anorexia nervosa. The authors described four adolescent girls with chronic purulent sputum production who were free of evident predisposing host defense defects. Bronchiectasis was evident on CT in two. Bronchoscopy, undertaken in three, showed copious secretions in the central airways; in two, CT showed a tracheal air/fluid level. Tracheobronchial clearance, assessed in three, showed a hold-up of particles in the trachea. Cough responses to inhaled capsaicin were normal in all. Flow volume curves showed poor cooperation, extremely low reproducibility and, in two, a pronounced expiratory flutter. The authors concluded that there was no organic cause for their inability to expectorate secretions, noting that their illnesses conferred (unspecified) psychiatric gain [2].

Marsh et al. reported a similar disorder in three girls, each of whom demonstrated a tracheal air/fluid level.
Age of onset of symptoms was 18-months, 3-years and <13-years in one. Bronchoscopy showed copious tracheal and proximal bronchial secretions in each; bronchiectasis was evident on CT in two. The authors found no evidence of cystic fibrosis or an underlying ciliary, immunological or neurological deficit, and concluded that VCS of undetermined cause was the likely source of the syndrome [3]. They cited "rattling" breathing as a clinical feature, and concluded, "Where there is a suspicion of cough suppression, chest CT scan showing air/liquid levels in the main airways or bronchoscopy showing markedly excessive upper airway and tracheal secretions are highly suggestive of the diagnosis". All 12 individuals comprising these three case series were female (P=1/2² = 0.00024).

Reich and Johnson reported on 29 non-immunocompromised individuals with pulmonary disease due to Mycobacterium avium complex in a case series originating in a non-referral setting [4]. Six (one-fifth) exhibited a distinctive, unusual clinical pattern: disease limited to the lingula or middle lobe, exclusively in elderly females free of a predisposing pulmonary disorder. The authors hypothesized that VCS sequentially led to: selective retention of secretions in regions most dependent on cough for clearance due to their limited collateral ventilation and their lengthy, narrow and dependent subserving bronchi → low grade suppuration → localized bronchiectasis → colonization by M. avium complex. To convey the fastidiousness resulting in VCS, they suggested the appellation, Lady Windermere Syndrome [5] (LWS) after the title role character in a 19th century British play. In support of VCS as a pathogenetic mechanism, they instanced two cases with acknowledged VCS, both in females, one with predominantly bibasal tuberculosis and a second with bilobar atelectasis [5]. The author (JR, unreported case) identified a young female nurse with cavitary pulmonary tuberculosis who developed ileoceleal tuberculosis, inferring that this complication was a consequence of swallowing respiratory secretions to suppress her cough.

**Discussion**

That VCS is the underlying pathogenetic mechanism of pulmonary disorders in the cases reported by Harris, Wells and Marsh et al. appears indisputable. The coinage "ataussis nervosa" conveys the idea that, like inferring appetite suppression in the genesis of anorexia nervosa, one can infer VCS from ineffective or absent cough in neuromuscularly unimpaired individuals exhibiting copious, proximal tracheobronchial secretions.

In contrast, the pathogenesis of LWS is disputed. Pomcrantz et al. [6] reported on the resection of the right middle lobe and/or lingula in 13 persons, all females, for treatment of isolated M. avium disease. The authors emphasized phenotypic features frequently associated with this disorder – asthenic body habitus, straight back, and pectus deformity – and emphasized the frequent observation of elongation the lingula and the intact nature of the adjacent fissures (signaling diminished collateral ventilation). Iseman et al. [7] emphasized the existence of thoracic abnormalities – pectus excavatum, scoliosis, straight back syndrome – as predisposing features. On the basis of this commonly encountered phenotype and a frequent association with mitral valve prolapse, Iseman hypothesized that LWS reflected a connective tissue disorder.

Kim et al. undertook a systematic evaluation of potential predisposing features in 63 subjects with severe, refractory, non-tuberculous mycobacterial pulmonary diseases [8]. Morphotype, immunophenotype, and cystic fibrosis transmembrane conductance regulator (CFTR) genotypic characterization failed to account for the constellation of defining features of LWS – female exclusivity, advanced age at onset, absence of predisposing pulmonary disease, distinctive restricted disease localization, and M. avium susceptibility.

The inference that cough suppression was the prime mechanism is supported by reports of elderly women with LWS who acknowledged lifelong VCS [9,10]. Bronchiectasis confined to the right middle lobe preceded the diagnosis of M. avium disease by 3-years in the first case [9]. The complaint of coughing in advanced cases (possibly overcoming the ability to suppress it) does not exclude VCS in its genesis; among individuals unaccustomed to coughing, its presence may be excessively disturbing. (Notably, cigarette smokers rarely complain of cough.) These alternative explanations are not mutually exclusive: the frequently encountered phenotypic and anatomic features would be expected to potentiate the effect of VCS in causing stasis, and the effect of pectus excavatum in compressing and elongating the lingula would appear to be sufficient in itself to generate the syndrome.

In summary, VCS appears responsible for a variety of uncommonly encountered respiratory disorders. Its female exclusivity has not been accounted for. "Rattling" breathing and absence of a percussive note of glottal decompression initiating the cough are useful clinical indicators of VCS. Demonstration of a tracheal air/fluid level on CT, absent an accountable neuromuscular disorder, furnishes confirmation. The consequences of VCS can often be reversed or ameliorated by encouraging subjects to cough vigorously.

**Conflict of interest**

None.

**References**


