



ERS | *monograph*

Complex Breathlessness

Edited by James H. Hull and
Jemma Haines

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James H. Hull and Jemma Haines

Editor in Chief
John R. Hurst

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Preface

John R. Hurst 

It is a pleasure to introduce and recommend to you this latest edition of the *ERS Monograph*, which covers the important and neglected topic of complex breathlessness, here defined as breathlessness where the cause remains unclear or appears disproportionate to any underlying conditions. Complex breathlessness is challenging both to people living with this problem, and to clinicians leading assessment and management. And that's where this collection really excels – providing a clear, concise and yet authoritative, comprehensive and systematic approach to investigation and treatment. It's a must-read, and so I congratulate the Guest Editors James H. Hull and Jemma Haines on delivering this excellent collection, which deserves a place on the (e-)shelf of every practising respiratory clinician.



To end on a personal note, this is the last Preface I will write as Editor in Chief, as I hand over the *Monograph* to the experienced hands of Professor Peter M.A. Calverley. Thank you to everyone who has contributed as an Editorial Board member, Guest Editor, author and reviewer over the past few years, giving freely of your time and expertise to the benefit of our Society. Special thanks to the ERS Publications Office team, Caroline Ashford-Bentley and Rachel Gozzard without whom, quite literally, there would be no *Monograph*! Peter, over to you.

Disclosures: J.R. Hurst reports receiving grants, personal fees and non-financial support from pharmaceutical companies that make medicines to treat respiratory disease. This includes reimbursement for educational activities and advisory work, and support to attend meetings.

Guest Editors

James H. Hull



James H. Hull is a Consultant Respiratory Physician at the Royal Brompton Hospital (London, UK) and an Honorary Professor at University College London (London, UK). He qualified from St George's Hospital Medical School in 2000, with an intercalated degree in exercise physiology. His doctorate and subsequent position as a National Institute for Health and Care Research (NIHR) lecturer, with time spent at UCLA in Los Angeles (CA, USA), provided him with expertise in physiological assessment and, specifically, cardiopulmonary exercise testing.

He is currently clinical lead for the unexplained breathlessness service at the Royal Brompton Hospital, providing a one-stop assessment service for national- and international-based referrals of complex breathing issues. In this context and as President of the Association of Respiratory Technology and Physiology (ARTP), James is an advocate for the use of physiological investigations and particularly cardiopulmonary exercise testing, to help characterise reasons for exertional breathing difficulties.

Working closely with colleagues in the multidisciplinary team, James developed the now widely used breathing pattern assessment tool (BPAT). He established an upper airway service at the Royal Brompton Hospital and has published work arising from this service, outlining the benefits of assessing upper airway closure during physical activity with/without other forms of provocation, with the continuous laryngoscopy test.

James has a specialist clinical and research interest in helping athletic individuals with respiratory issues and in this area; he is a specialist advisor to the English Institute of Sport and Team GB, the International Olympic Committee, the British Thoracic Society (BTS) and several professional sporting organisations.

Jemma Haines



Jemma Haines MBE qualified in 2003 with a First-Class Honours in Speech and Language Sciences from Newcastle University (Newcastle upon Tyne, UK). In her early career she worked as a

voice clinician and developed a keen interest in laryngology. Acknowledging the significance of the larynx during respiration, she pioneered a clinical specialism to support patients suffering with complex breathlessness.

Jemma is now Chief Allied Health Professional at Manchester University NHS Foundation Trust (Manchester, UK) and a NIHR Manchester Biomedical Research Centre PhD Fellow. She developed and led the multidisciplinary team Manchester Airways Service, which is now established as a leading national referral centre for complex breathlessness management. Her specialist expertise includes managing patients with inducible laryngeal obstruction, refractory chronic cough and difficult-to-control asthma.

Jemma is a national Royal College of Speech and Language Therapists (RCSLT) professional respiratory advisor and an elected member of the BTS Cough Specialist Advisory Group. She has co-authored several national professional respiratory guidelines and has many peer-reviewed publications relating to her work. Jemma has attended numerous national and international conferences, presenting her clinical research findings. Currently, her research investigates standardised evaluation and treatment for inducible laryngeal obstruction.

In 2021, Jemma was made a Member of the Order of the British Empire in the Queen's birthday honours list and a Fellow of the RCSLT. These prestigious awards are in recognition of her leadership within the field of upper airway disorders in speech and language therapy, in addition to her significant contribution and response to supporting modifications to practice during the COVID-19 pandemic.

Introduction

James H. Hull ^{1,2} and Jemma Haines^{3,4,5}

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This *Monograph* provides a comprehensive guide to the assessment and management of individuals presenting with seemingly “unexplained” or complex respiratory symptoms, which are frequently overlooked and often associated with significant patient morbidity <https://bit.ly/3HbXEs6>

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Breathlessness is the key presenting symptom in many heart or lung diseases and thus its presence mandates a systematic and thorough search for any underlying pathological cause. In a proportion of individuals, however, despite extensive investigation, the origin and aetiology of their breathlessness may remain unclear or appear disproportionate to any underlying condition that is identified. In this context, the term “complex breathlessness” may be applied.

The assessment and management of complex breathlessness is challenging both from the patient’s and the clinician’s perspective. For patients suffering with this issue, there are usually three main concerns: 1) the need to be satisfied that no pathological explanation has been overlooked; 2) to obtain acknowledgement of the impact of symptoms; and 3) to be reassured that there is a logical approach to their treatment.

The overall aim of this *Monograph* is therefore to provide clinicians at all levels with a comprehensive guide to the assessment and management of individuals presenting with seemingly “unexplained” or complex breathlessness. Its 15 chapters cover all aspects needed to assess and successfully manage this challenging clinical scenario, providing clinicians with a useful reference when faced with a symptomatic breathless patient with no obvious underlying cause or with disproportionate symptoms. Specifically, the *Monograph* acts to highlight several potentially less commonly considered clinical entities and their associated features, to ensure that they are not overlooked. Secondly, it considers the symptom of breathlessness and how to approach its management, potentially in the absence of a clear diagnosis or set of abnormalities on investigation.

The introductory chapters provide the reader with a solid foundation from which to better understand the context of complex breathlessness. The epidemiology of breathlessness is considered in chapter 1, which explores persistent breathlessness symptom prevalence across

populations, pathophysiological models, risk factors and the adverse health consequences this brings [1]. Individuals suffering breathlessness not explained by objective measures of disease, prove challenging to clinicians. The neurocognitive basis of breathlessness is therefore important, and chapter 2 on the neuroscience of breathlessness offers a useful consideration of current theories of perception applied to the understanding of breathlessness [2]. To cement the foundations, chapter 3 examines the physiology of breathlessness and summarises how specific physiological factors may contribute to symptoms, offering guidance on how these should be measured [3].

Next, assessment guidance is provided in chapter 4, initially exploring the tools required and available, followed by a discussion on the appropriateness of tool selection, depending on which aspect of breathlessness is under review [4]. This then leads into a useful chapter detailing a systematic clinical approach to assessing complex breathlessness, which includes points for a comprehensive evaluation with a focus on the nature and impact of the patient's breathlessness symptom(s) [5].

Airway-focused chapters follow to educate the reader on specific disorders associated with complex breathlessness, including: laryngeal considerations in chapter 6, with detail on inducible laryngeal dysfunction; excessive airways collapse in chapter 7; BPD in chapter 8; and allergic factors in chapter 9 [6–9]. Each chapter provides a summary overview, associated assessment specifics and management approaches.

Cardiopulmonary vascular considerations are next addressed to provide detailed insight into uncommon but often overlooked causes of breathlessness. These include pulmonary vascular causes in chapter 10, cardiovascular drivers in chapter 11 and autonomic dysfunction presentations, including PoTS, in chapter 12 [10–12].

The final chapters of the *Monograph* are dedicated to overall holistic management [13–15]. Suboptimal treatment adherence, whether intentional or not, has the potential to play a role in amplifying the complexity of breathlessness and its management, so this is explored first [13]. The subsequent chapters then provide an invaluable lens on the latest evidence on treating unexplained breathlessness holistically [14, 15].

We are particularly grateful to our authors and reviewers, who have dedicated their time and expertise to enable this *Monograph* to come to fruition. We would also like to acknowledge the support given to us by the *ERS Monograph* team who have expertly guided us throughout. We hope readers will find this resource a useful guide to support the assessment and management of individuals who present with seemingly “unexplained” or complex breathlessness.

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List of abbreviations

BMI	body mass index
BPD	breathing pattern disorder
CPET	cardiopulmonary exercise testing
CT	computed tomography
D_{LCO}	diffusing capacity of the lung for carbon monoxide
EDAC	excessive dynamic airway collapse
FEV₁	forced expiratory volume in 1 s
FVC	forced vital capacity
ILD	interstitial lung disease
ILO	inducible laryngeal obstruction
IPF	idiopathic pulmonary fibrosis
MCID	minimal clinically important difference
MRI	magnetic resonance imaging
PFT	pulmonary function test
PoTS	postural tachycardia syndrome