Raynaud's Phenomenon

Fredrick M. Wigley • Ariane L. Herrick
Nicholas A. Flavahan
Editors

Raynaud's Phenomenon

A Guide to Pathogenesis
and Treatment

Editors

Fredrick M. Wigley, M.D.
Martha McCrory Professor of Medicine
Associate Director Division
of Rheumatology
Director Johns Hopkins
Scleroderma Center
Johns Hopkins University School
of Medicine
Baltimore, MD, USA

Ariane L. Herrick, M.D., F.R.C.P.
Professor of Rheumatology
Centre for Musculoskeletal Research
Institute of Inflammation and Repair
The University of Manchester
Manchester Academic Health Science
Centre
Manchester, UK

Nicholas A. Flavahan, Ph.D.
Edward D. Miller Professor
Director of Research, Vice Chair
Department of Anesthesiology
and Critical Care Medicine
Johns Hopkins University
School of Medicine
Baltimore, MD, USA

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Foreword: United Kingdom

This is a most welcome publication, which fully illustrates every aspect of Raynaud's phenomenon, from epidemiology and pathophysiology to tests, treatments and clinical trials. It is an excellent resource for anyone who is interested in learning more about Raynaud's and associated conditions. I have had Raynaud's for almost 40 years and scleroderma for most of that time. During this period I have personally experienced the devastating effects, which these conditions can cause, including digital ulcers and amputation, in addition to internal organ involvement. I founded a national charity that provided me with the opportunity to understand the suffering of others, and I became a patient advocate, supporting people with these conditions. Literally millions of people worldwide have to cope with Raynaud's on a daily basis, not just in the cold but when faced with changes of temperature or stress. The pain can be excruciating and almost impossible to control. When I was first diagnosed, very little was known about the condition or how to treat it, and I faced a long journey of discovery, searching every possible pathway in pursuit of finding ways to help others and myself with similar problems. Being in touch with other sufferers gave me the incentive to continue my search for information. I formed sound relationships with consultants and researchers, and this bond enabled me to understand just how hard they were working and that research takes a long time and requires substantial financial support. Research and treatments have developed considerably over recent years, thanks to everyone who has devoted time and effort into trying to find a cure and to those who have helped to raise funds in order to finance the research.

I am delighted to be able to recommend this book, which not only gives valuable information but also clearly indicates how research has advanced and offers hope for the future.

Anne H. Mawdsley, M.B.E.
Founder Raynaud's &
Scleroderma Association (UK)

Post-script. Anne Mawdsley: May 1942 to October 2014. She made enormous contributions to the care of people with Raynaud's and Scleroderma, and to Raynaud's and Scleroderma research. She will be very much missed.

Foreword: United States of America

The orderly and regulated movement of blood through blood vessels has been recognized for centuries. In addition to its obvious physiological relevance, blood flow through vessels has been ascribed an almost spiritual significance. The color changes in the digits of the extremities, easily observed by patients and physicians, report on the function of small blood vessels and provide unique insights into age-old diseases. This extraordinary book focuses on the physiology and pathology of blood vessels, seen through the lens of the characteristic reversible changes in perfusion observed as Raynaud's phenomenon. The editors are leaders in the field, and they bring entire careers of knowledge about blood vessels and how they may be affected in disease. They have fashioned a resource which is as outstanding as it is timely.

Antony Rosen, M.B. Ch.B., B.Sc. (Hons.).
Mary Betty Professor of Medicine,
Professor of Cell Biology,
Professor of Pathology,
Director, Division of Rheumatology
Vice Dean for Research,
Johns Hopkins University School of Medicine,
Baltimore, MD, USA

Preface

The remarkable clinical and observational skills of Maurice Raynaud defined a previously unappreciated cause of digital gangrene. He recognized that digital and cutaneous blood vessels have the capacity to react to provocation and that compromise to tissue blood flow can occur in the absence of vessel obstruction or vessel wall disease. He described reversible vasoconstriction of digital blood flow triggered by cold that resulted in a “deadly white” pallor or “cyanotic color” of the skin. It is now known that this phenomenon occurs because the skin has specialized thermoregulatory vessels that play a major role in normal physiological responses to the environment in order to maintain stable core body temperature. Raynaud’s phenomenon (RP) is an inappropriate and exaggerated response of the digital and cutaneous circulation to cold environmental temperatures. Figure 1 depicts a dramatic clinical picture of the pallor phase of Raynaud’s (a); while the adjacent figure demonstrates how low blood flow alters the temperature as measured by thermography (b). Soon after Raynaud’s thesis was presented, it was recognized that the phenomenon was not caused by one process but could occur secondary to a variety of disorders that affect the peripheral circulation. We now appreciate that RP is a common disorder that is encountered both in otherwise healthy individuals and as part of a disease process altering the regulation of cutaneous blood flow.

Fig. 1 (a) Clinical picture of the pallor phase of Raynaud’s. (b) Altered temperature resulting from low blood flow, as measured by thermography

A comprehensive review of Raynaud's is now timely because of the incredible progress in understanding the molecular mechanisms of the normal regulation of cutaneous blood flow and how disease can disrupt the function of these specialized vessels. Likewise, the clinical implications of RP are better appreciated with studies defining its prevalence, the associated diseases, and new treatment approaches for patients suffering from it. We have structured the book to cover every aspect of RP from the history to the current treatment approach. The nomenclature use to classify cases has changed over the years and is important to understand these terms both for clinical care and research. Epidemiological studies have shown that RP is common, is influenced by environmental factors, is seen in both men and women, and is seen around the world in both as primary and secondary forms. A review of our current understanding of normal physiology and regulation of cutaneous blood flow sets the scene for an update of the pathology of primary and secondary RP. While primary RP is likely the most common clinical presentation, we wanted to comprehensively discuss various causes of RP including childhood RP, RP secondary to connective tissue disease, occupational causes, and a variety of other associated disorders. The many diseases with vascular perturbation that mimic RP and others that associate with RP are also reviewed.

We know that one of the main challenges facing a clinician who encounters a patient with RP is to define the underlying cause. We therefore included discussions of the pathogenesis as well as the methods and rationale for evaluating a patient with RP. Chapters are included that review the state of the art of nailfold capillary examination, noninvasive imaging, angiography, and appropriate serological testing. The concept that an RP is associated with systemic disease or the manifestation of a systemic vasospastic disorder is also presented.

Another challenge is to document the severity of RP for both clinical care and research toward developing effective treatment. Both outcome measures and the proper approach to study design for clinical trials in RP is reviewed. The final chapters of the book approach both nondrug and drug therapy for RP. We felt it essential to review evidence for and against current approaches to the treatment of RP. The management of the consequences of severe secondary RP is covered in chapters on how to care for digital ulcerations and surgical treatment options. We decided to provide our practical approach in specific clinical situations by presenting problematic cases and our expert opinion on therapy.

In an effort to have each chapter stand-alone, we recognize that some information in one chapter may be repeated in another chapter. We also note that much of the research and clinical trials in RP focuses on primary RP or RP associated with systemic sclerosis. However, we included comments to clearly define the design and population involved in investigations. We made a special effort to include photos of clinical situations and graphics to illustrate points. Each chapter is preceded by a list "key points" to highlight the major message of the chapter. Each author also provided an "expert opinion" to complement his or her review of published evidence.

It is clear that there are many challenges for the future. More studies are needed to better understand the molecular mechanisms of both primary and secondary RP. Appreciation of the full spectrum of diseases that associate or cause RP opens the opportunity to develop specific approaches for prevention and treatment. New diagnostic tools that could provide ambulatory monitoring would enhance our ability to test new treatment in a real-life situation. Better and safer medications are needed. Progress will come about only by applying novel thinking about the phenomenon; much like Maurice Raynaud's did years ago when he debated the usual concepts about digital gangrene of his time.

Baltimore, MD, USA
Manchester, UK
Baltimore, MD, USA

Fredrick M. Wigley
Ariane L. Herrick
Nicholas A. Flavahan

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Contributors

Rasidat Adeduntan, B.A. Department of Dermatology, Stanford University School of Medicine, Redwood City, CA, USA

Yannick Allanore, M.D., Ph.D. Service de Rhumatologie A, Hôpital Cochin, Paris, France

Marina Anderson, Ph.D. M.R.C.P. Institute of Ageing and Chronic Disease, University of Liverpool, Liverpool, UK

Francesco Boin, M.D. Department of Medicine, Division of Rheumatology, Johns Hopkins University School of Medicine, Baltimore, MD, USA

Marco Matucci-Cerinic, M.D., Ph.D. Division of Rheumatology, Department of Experimental and Clinical Medicine, University of Florence, Florence, Italy

Lorinda Chung, M.D., M.S. Department of Medicine, Division of Immunology and Rheumatology, Stanford University School of Medicine, Palo Alto, CA, USA

Maurizio Cutolo Research Laboratories and Academic Division of Clinical Rheumatology, Department of Internal Medicine, University of Genova, Genoa, Italy

Christopher P. Denton, Ph.D., F.R.C.P. Department of Inflammation, Division of Medicine, Royal Free Hospital and UCL Medical School, London, UK

Jean-Luc Drappé, M.D., Ph.D. Service de Rhumatologie B, Hôpital Cochin, Paris, France

Andrea Fava, M.D. Department of Medicine, Division of Rheumatology, Johns Hopkins University School of Medicine, Baltimore, MD, USA

Nicholas A. Flavahan, Ph.D. Department of Anesthesiology and Critical Care Medicine, Johns Hopkins University School of Medicine, Baltimore, MD, USA

Victoria Flower, M.B.B.S., B.Sc. Hons., M.R.C.P. Department of Rheumatology, Royal National Hospital for Rheumatic Diseases, Bath, UK

Heather Gladue, D.O. Department of Medicine, Emory University, Atlanta, GA, USA

Serena Guiducci, M.D., Ph.D. Department of Experimental and Clinical Medicine, Division of Rheumatology, University of Florence, Florence, Italy

Ariane L. Herrick, M.D., F.R.C.P. Centre for Musculoskeletal Research, Institute of Inflammation and Repair, The University of Manchester, Manchester Academic Health Science Centre, Manchester, UK

Michael Hughes, M.Sc., M.R.C.P. Centre for Musculoskeletal Research, Salford Royal NHS Foundation Trust, Manchester Academic Health Science Centre, The University of Manchester, Manchester, UK

Laura K. Hummers, M.D., Sc.M. Department of Medicine, Division of Rheumatology, Johns Hopkins University School of Medicine, Baltimore, MD, USA

Dinesh Khanna, M.D., M.S. Department of Medicine, Division of Rheumatology, University of Michigan School of Medicine, Ann Arbor, MI, USA

Linda Kwakkenbos, Ph.D. Lady Davis Institute for Medical Research, McGill University, Jewish General Hospital, Montréal, QC, Canada

Adam Maundrell Department of Rheumatology, Royal Adelaide Hospital, North Terrace, Adelaide, SA, Australia

Neil McHugh BM.B.Ch.B., M.D.(Thesis), F.R.A.C.P., F.R.C.P., F.R.C.Path., Division of Rheumatology, Royal National Hospital for Rheumatic Diseases, Department of Pharmacy and Pharmacology, University of Bath, Bath, UK

Zsuzsanna H. McMahan, M.D. Department of Medicine, Division of Rheumatology, Johns Hopkins School of Medicine, Baltimore, MD, USA

Lindsay Muir, M.B., M.Ch.(Orth.), F.R.C.S. (Orth.). Department of Orthopedic Surgery, Salford Royal NHS Foundation Trust, Salford, UK

Andrea Murray, M.phys., Ph.D. Centre for Musculoskeletal Research, Salford Royal NHS Foundation Trust, The University of Manchester, Manchester Academic Health Science Centre, Manchester, UK

Voon H. Ong, Ph.D. M.R.C.P. Royal Free Hospital and UCL Medical School, London, UK

Julie J. Paik, M.D. Department of Medicine, Division of Rheumatology, John Hopkins School of Medicine, Baltimore, MD, USA

Clare Pain Department of Paediatric Rheumatology, Alder Hey Children's NHS Foundation Trust, Liverpool, UK

John D. Pauling, B.Med.Sci., M.R.C.P.(Rheumatology), Ph.D. Department of Rheumatology, Royal National Hospital for Rheumatic Diseases, Bath, UK

Janet E. Pope, M.D., M.P.H., F.R.C.P.C. Division of Rheumatology, St. Joseph's Health Care London, University of Western Ontario, London, ON, Canada

Susanna M. Proudman Department of Rheumatology, Royal Adelaide Hospital, North Terrace, Adelaide, SA, Australia

Thomas Reifsnyder, M.D. Department of Surgery, Johns Hopkins University School of Medicine, Baltimore, MD, USA

James R. Seibold, M.D. Scleroderma Research Consultants, LLC, Litchfield, CT, USA

Ami A. Shah, M.D. Department of Medicine, Division of Rheumatology, Johns Hopkins School of Medicine, Baltimore, MD, USA

Vanessa Smith, M.D. Department of Rheumatology, Ghent University Hospital, Ghent, Belgium

Brett D. Thombs, Ph.D. Lady Davis Institute for Medical Research, McGill University, Montréal Jewish General Hospital, Montréal, QC, Canada

Antonia Valenzuela M.D., M.S. Department of Medicine, Division of Immunology and Rheumatology, Stanford University School of Medicine, Palo Alto, CA, USA

Fredrick M. Wigley, M.D. Department of Medicine, Division of Rheumatology, Johns Hopkins School of Medicine, Baltimore, MD, USA

Jack Wilkinson, M.Sc., B.Sc. Research and Development, Salford Royal NHS Foundation Trust, Salford, UK

Francesco Zulian Pediatric Rheumatology Unit, Department of Pediatrics, University of Padua, Padua, Italy