

# The Pigmentary System: Physiology and Pathophysiology

Edited by

**James J. Nordlund**

*Dermatologist and Professor Emeritus,  
Group Health Associates, Cincinnati, OH, USA*

**Raymond E. Boissy**

*Professor of Dermatology and Cell Biology,  
University of Cincinnati College of Medicine,  
Cincinnati, OH, USA*

**Vincent J. Hearing**

*Chief, Pigment Cell Biology Section,  
Laboratory of Cell Biology,  
National Institutes of Health, Bethesda, MD, USA*

**Richard A. King**

*Director, Genetics Division, Department of Medicine,  
Institute of Human Genetics, University of Minnesota,  
Minneapolis, MN, USA*

**William S. Oetting**

*Associate Professor, Genetics Division, Department of Medicine  
Institute of Human Genetics, University of Minnesota, Minneapolis, MN, USA*

**Jean-Paul Ortonne**

*Professor of Dermatology and Chairman, Department of Dermatology,  
University of Nice-Sophia Antipolis,  
Nice, France*

SECOND EDITION



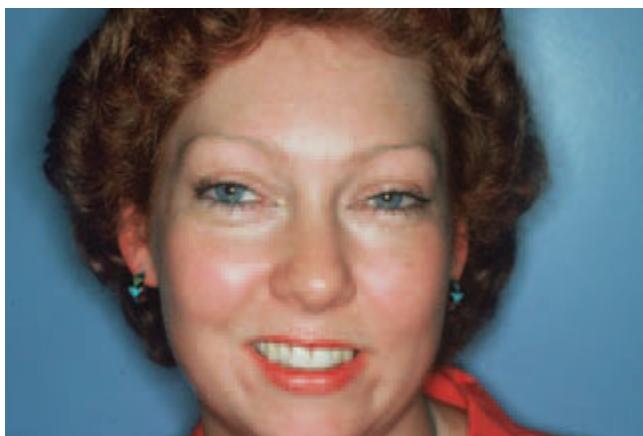


# The Pigmentary System: Physiology and Pathophysiology





A



B



D



C



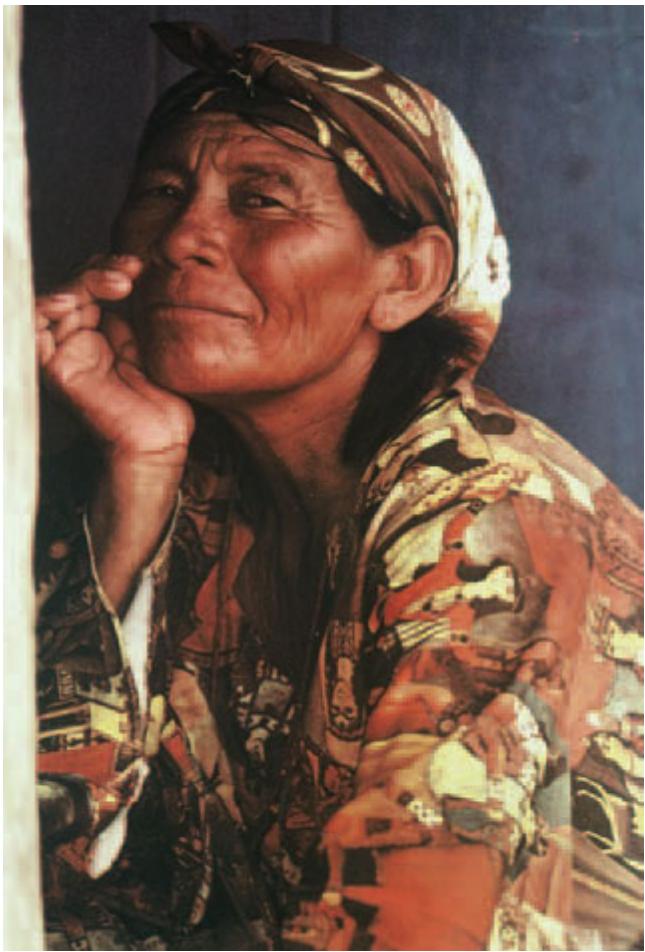
E



F



H



G



I

A: These two individuals illustrate the wide range of skin and hair color. [Marianne Greenwood (right) graciously shared the photographs (from her book *Varför Gråter Puman?*) that compose this frontispiece.] B: Classical Celtic women with blue eyes and red hair. C: Typical Scandinavian with blue eyes. D: Native American. E: A Peruvian girl. F: Himalayan woman and child. G: A Venezuelan woman. H: Two teenagers from New Guinea. I: Man from the New Hebrides Islands. See Chapter 27 for further information.

# The Pigmentary System: Physiology and Pathophysiology

Edited by

**James J. Nordlund**

*Dermatologist and Professor Emeritus,  
Group Health Associates, Cincinnati, OH, USA*

**Raymond E. Boissy**

*Professor of Dermatology and Cell Biology,  
University of Cincinnati College of Medicine,  
Cincinnati, OH, USA*

**Vincent J. Hearing**

*Chief, Pigment Cell Biology Section,  
Laboratory of Cell Biology,  
National Institutes of Health, Bethesda, MD, USA*

**Richard A. King**

*Director, Genetics Division, Department of Medicine,  
Institute of Human Genetics, University of Minnesota,  
Minneapolis, MN, USA*

**William S. Oetting**

*Associate Professor, Genetics Division, Department of Medicine  
Institute of Human Genetics, University of Minnesota, Minneapolis, MN, USA*

**Jean-Paul Ortonne**

*Professor of Dermatology and Chairman, Department of Dermatology,  
University of Nice-Sophia Antipolis,  
Nice, France*

SECOND EDITION



© 2006 Blackwell Publishing Ltd  
Blackwell Publishing, Inc., 350 Main Street, Malden, Massachusetts 02148-5020, USA  
Blackwell Publishing Ltd, 9600 Garsington Road, Oxford OX4 2DQ, UK  
Blackwell Publishing Asia Pty Ltd, 550 Swanston Street, Carlton, Victoria 3053, Australia

The right of the Author to be identified as the Author of this Work has been asserted in accordance with the Copyright, Designs and Patents Act 1988.

All rights reserved. No part of this publication may be reproduced, stored in a retrieval system, or transmitted, in any form or by any means, electronic, mechanical, photocopying, recording or otherwise, except as permitted by the UK Copyright, Designs and Patents Act 1988, without the prior permission of the publisher.

First published 1998 by Oxford University Press  
Second edition 2006  
1 2006

Library of Congress Cataloging-in-Publication Data

The pigmentary system : physiology and pathophysiology / edited by James J. Nordlund . . . [et al.].—2nd ed.  
p. ; cm.  
Includes bibliographical references and index.  
1. Pigmentation disorders. 2. Melanocytes. I. Nordlund, James J.  
[DNLM: 1. Pigmentation Disorders—physiopathology. 2. Melanocytes.  
3. Pigmentation—physiology. WR 265 P6309 2006]  
RL790.P53 2006  
616.5'5—dc22

2005030369

A catalogue record for this title is available from the British Library

Set in 9/12 pt Sabon by SNP Best-set Typesetter Ltd, Hong Kong  
Printed and bound in India by Replika Press PVT Ltd

ISBN-13: 978-1-4051-2034-0  
ISBN-10: 1-4051-2034-7

Commissioning Editor: Stuart Taylor  
Editorial Assistant: Saskia Van der Linden  
Development Editor: Rob Blundell  
Production Controller: Kate Charman

For further information on Blackwell Publishing, visit our website:  
<http://www.blackwellpublishing.com>

The publisher's policy is to use permanent paper from mills that operate a sustainable forestry policy, and which has been manufactured from pulp processed using acid-free and elementary chlorine-free practices. Furthermore, the publisher ensures that the text paper and cover board used have met acceptable environmental accreditation standards.

Blackwell Publishing makes no representation, express or implied, that the drug dosages in this book are correct. Readers must therefore always check that any product mentioned in this publication is used in accordance with the prescribing information prepared by the manufacturers. The author and the publishers do not accept responsibility or legal liability for any errors in the text or for the misuse or misapplication of material in this book.

# Contents

- Contributors, x  
Foreword, xv  
Preface, xvii  
Acknowledgements, xviii  
*Frontispiece can be found between pages ii and iii*
- Part I: The Physiology of the Pigmentary System**
- Section 1: Historical and Comparative Perspectives of the Pigmentary System, 3**
- 1 A History of the Science of Pigmentation, 5  
*Sidney N. Klaus*
  - 2 Comparative Anatomy and Physiology of Pigment Cells in Nonmammalian Tissues, 11  
*Joseph T. Bagnara & Jiro Matsumoto*
- Section 2: The Science of Pigmentation, 61**
- 3 General Biology of Mammalian Pigmentation, 63  
*Walter C. Quevedo Jr. & Thomas J. Holstein*
  - 4 Extracutaneous Melanocytes, 91  
*Raymond E. Boissy & Thomas J. Hornyak*
  - 5 Regulation of Melanoblast Migration and Differentiation, 108  
*David M. Parichy, Mark V. Reedy, & Carol A. Erickson*
  - 6 Melanoblast Development and Associated Disorders, 140  
*Richard A. Spritz*
  - 7 Biogenesis of Melanosomes, 155  
*Raymond E. Boissy, Marjan Huizing, & William A. Gahl*
  - 8 Melanosome Trafficking and Transfer, 171  
*Glynis A. Scott*
  - 9 Melanosome Processing in Keratinocytes, 181  
*H. Randolph Byers*
  - 10 The Regulation of Melanin Formation, 191  
*Vincent J. Hearing*
  - 11 The Tyrosinase Gene Family, 213  
*William S. Oetting & Vijayasaradhi Setaluri*
  - 12 Molecular Regulation of Melanin Formation: Melanosome Transporter Proteins, 230  
*Murray H. Brilliant*
  - 13 Transcriptional Regulation of Melanocyte Function, 242  
*Kazuhisa Takeda & Shigeki Shibahara*
  - 14 Enzymology of Melanin Formation, 261  
*Francisco Solano & José C. García-Borrón*

- 15 Chemistry of Melanins, 282  
*Shosuke Ito & Kazumasa Wakamatsu*
- 16 The Physical Properties of Melanins, 311  
*Tadeusz Sarna & Harold A. Swartz*
- 17 Photobiology of Melanins, 342  
*Antony R. Young*
- 18 Toxicological Aspects of Melanin and Melanogenesis, 354  
*Edward J. Land, Christopher A. Ramsden, & Patrick A. Riley*
- 19 Regulation of Pigment Type Switching by Agouti, Melanocortin Signaling, Attractin, and Mahoganoid, 395  
*Gregory S. Barsh*
- 20 Human Pigmentation: Its Regulation by Ultraviolet Light and by Endocrine, Paracrine, and Autocrine Factors, 410  
*Zalfa Abdel-Malek & Ana Luisa Kadekaro*
- 21 Paracrine Interactions of Melanocytes in Pigmentary Disorders, 421  
*Genji Imokawa*
- 22 Growth Factor Receptors and Signal Transduction Regulating the Proliferation and Differentiation of Melanocytes, 445  
*Ruth Halaban & Gisela Moellmann*
- 23 Aging and Senescence of Melanocytes, 464  
*Debdutta Bandyopadhyay & Estela E. Medrano*
- 24 The Genetics of Melanoma, 472  
*Vanessa C. Gray-Schopfer & Dorothy C. Bennett*
- 25 The Transformed Phenotype of Melanocytes, 489  
*Dong Fang & Meenhard Herlyn*

**Part II: The Pathophysiology of Pigmentary Disorders**

**Section 3: An Overview of Human Skin Color and its Disorders, 497**

- 26 A More Precise Lexicon for Pigmentation, Pigmentary Disorders, and “Chromatic” Abnormalities, 499  
*James J. Nordlund, Tania Cestari, Pearl Grimes, Henry Chan, & Jean-Paul Ortonne*
- 27 The Normal Color of Human Skin, 504  
*James J. Nordlund & Jean-Paul Ortonne*
- 28 Mechanisms that Cause Abnormal Skin Color, 521  
*Jean-Paul Ortonne & James J. Nordlund*

## CONTENTS

- Section 4: Disorders of Hypopigmentation, Depigmentation and Hypochromia, 539**
- 29 Genetic Hypomelanoses: Disorders Characterized by Congenital White Spotting—Piebaldism, Waardenburg Syndrome, and Related Genetic Disorders of Melanocyte Development—Clinical Aspects, 541  
*Richard A. Spritz*
- 30 Genetic Hypomelanoses: Acquired Depigmentation, 551  
Rozycki Syndrome, 551  
*Jean L. Bolognia*  
Vitiligo Vulgaris, 551  
*James J. Nordlund, Jean-Paul Ortonne, & I. Caroline Le Poole*
- 31 Genetic Hypomelanoses: Generalized Hypopigmentation, 599  
Oculocutaneous Albinism, 599  
*Richard A. King & William S. Oetting*  
Albinoid Disorders, 613  
*Philippe Bahadoran & Jean-Paul Ortonne*  
Ataxia Telangiectasia, 621  
*Anne-Sophie Gadenne*  
Hallerman–Streiff Syndrome, 623  
*James J. Nordlund*  
Histidinemia, 623  
*Marnie D. Titsch*  
Homocystinuria, 625  
*Allan D. Mineroff*  
Oculocerebral Syndrome with Hypopigmentation, 626  
*Jean L. Bolognia*  
Tietz Syndrome, 630  
*Jean-Paul Ortonne*  
Kappa-Chain Deficiency, 631  
*Jean-Paul Ortonne*  
Menkes' Kinky Hair Syndrome, 631  
*Tanusin Ploysangam*  
Phenylketonuria, 634  
*Allan D. Mineroff*
- 32 Genetic Hypomelanoses: Localized Hypopigmentation, 636  
“Hypomelanosis of Ito” and Mosaicism, 636  
*Wolfgang Küster & Rudolf Happle*  
Focal Dermal Hypoplasia, 645  
*James J. Nordlund*  
Hypomelanosis with Punctate Keratosis of the Palms and Soles, 646  
*Jean L. Bolognia*  
Darier–White Disease (Keratosis Follicularis; 124200), 647  
*Jean L. Bolognia*  
Nevus Depigmentosus, 649  
*Stella D. Calobrisi*  
Tuberous Sclerosis Complex, 652  
*Pranav B. Sheth*
- 33 Genetic Hypomelanoses: Disorders Characterized by Hypopigmentation of Hair, 657
- Bird-Headed Dwarfism (Seckel Syndrome), 657  
*Stan P. Hill*  
Down Syndrome, 658  
*Rosemary Geary*  
Fisch Syndrome, 659  
*Stan P. Hill*  
Premature Canities, 659  
*James J. Nordlund*  
Mandibulofacial Dysostosis (Treacher Collins Syndrome), 660  
*Rosemary Geary*  
Myotonic Dystrophy, 660  
*Peggy Tong*  
PHC Syndrome (Böök Syndrome), 661  
*Stan P. Hill*  
Pierre Robin Syndrome, 661  
*James J. Nordlund*  
Prolidase Deficiency, 662  
*Pranav B. Sheth*
- 34 Metabolic, Nutritional, and Endocrine Disorders, 664  
Metabolic and Nutritional Hypomelanoses, 664  
*Peter S. Friedmann*  
Hypomelanosis Associated with Endocrine Disorders, 667  
*Peter S. Friedmann*
- 35 Chemical, Pharmacologic, and Physical Agents Causing Hypomelanoses, 669  
Chemical and Pharmacologic Agents Causing Hypomelanoses, 669  
*Stefania Briganti, Monica Ottaviani, & Mauro Picardo*  
Physical Agents Causing Hypomelanoses, 683  
*Jean-Philippe Lacour*
- 36 Infectious Hypomelanoses, 686  
*Jean-Philippe Lacour*
- 37 Inflammatory Hypomelanoses, 699  
*Jean-Philippe Lacour*
- 38 Hypomelanoses Associated with Melanocytic Neoplasia, 705  
*Lieve Brochez, Barbara Boone, & Jean-Marie Naeyaert*
- 39 Miscellaneous Hypomelanoses: Depigmentation, 725  
Alezzandrini Syndrome, 725  
*Wiete Westerhof, David Njoo, & Henk E. Menke*  
Idiopathic Guttate Hypomelanosis, 726  
*Wiete Westerhof, David Njoo, & Henk E. Menke*  
Leukoderma Punctata, 729  
*Wiete Westerhof, David Njoo, & Henk E. Menke*  
Lichen Sclerosus et Atrophicus, 731  
*Philippe Bahadoran*  
Vagabond Leukomelanoderma, 732  
*Wiete Westerhof, David Njoo, & Henk E. Menke*  
Vogt–Koyanagi–Harada Syndrome, 734  
*Wiete Westerhof, David Njoo, & Henk E. Menke*  
Westerhof Syndrome, 741  
*Wiete Westerhof, David Njoo, & Henk E. Menke*

- 40 Miscellaneous Hypomelanoses: Hypopigmentation, 745  
 Disseminated Hypopigmented Keratoses, 745  
*Wiete Westerhof, David Njoo, & Henk E. Menke*  
 Hypermelanocytic Punctata et Guttata  
 Hypomelanosis, 746  
*Wiete Westerhof, David Njoo, & Henk E. Menke*  
 Progressive Macular Hypomelanosis, 748  
*Henk E. Menke, Germaine Relyveld, David Njoo, & Wiete Westerhof*  
 Sarcoidosis, 751  
*Wiete Westerhof, David Njoo, & Henk E. Menke*
- 41 Miscellaneous Hypomelanoses: Extracutaneous Loss of Pigmentation, 754  
 Alopecia Areata, 754  
*Wiete Westerhof*  
 Heterochromia Irides, 756  
*Wiete Westerhof, David Njoo, & Henk E. Menke*  
 Senile Canities, 760  
*Wiete Westerhof, David Njoo, & Henk E. Menke*  
 Sudden Whitening of Hair, 764  
*Wiete Westerhof, David Njoo, & Henk E. Menke*
- 42 Hypochromia without Hypomelanosis, 767  
*Jean-Philippe Lacour*
- Section 5: Disorders of Hyperpigmentation and Hyperchromia, 769**
- 43 Genetic Epidermal Syndromes: Disorders Characterized by Generalized Hyperpigmentation, 771  
 Adrenoleukodystrophy, 771  
*Sheila S. Galbraith & Nancy Burton Esterly*  
 Familial Progressive Hyperpigmentation, 774  
*Nancy Burton Esterly, Eulalia Baselga, Beth A. Drolet, Susan Bayliss Mallory, & Sharon A. Foley*  
 Fanconi Anemia, 776  
*Sheila S. Galbraith & Nancy Burton Esterly*  
 Gaucher Disease, 778  
*Sheila S. Galbraith & Nancy Burton Esterly*
- 44 Genetic Epidermal Syndromes: Disorders Characterized by Reticulated Hyperpigmentation, 780  
 Berlin Syndrome, 780  
*Eulalia Baselga & Nancy Burton Esterly*  
 Cantu Syndrome, 781  
*Eulalia Baselga & Nancy Burton Esterly*  
 Kindler Syndrome, 781  
*Eulalia Baselga & Nancy Burton Esterly*  
 Dermatopathia Pigmentosa Reticularis, 784  
*Eulalia Baselga & Nancy Burton Esterly*  
 Dyschromatosis Universalis Hereditaria, 786  
*Eulalia Baselga & Nancy Burton Esterly*  
 Epidermolysis Bullosa with Mottled Pigmentation, 788  
*Eulalia Baselga & Nancy Burton Esterly*  
 Familial Mandibuloacral Dysplasia, 790  
*Eulalia Baselga & Nancy Burton Esterly*  
 Hereditary Acrokeratotic Poikiloderma, 792  
*Eulalia Baselga & Nancy Burton Esterly*
- Hereditary Sclerosing Poikiloderma, 795  
*Eulalia Baselga & Nancy Burton Esterly*  
 Mendes Da Costa Disease, 796  
*Eulalia Baselga & Nancy Burton Esterly*  
 Naegeli–Franceschetti–Jadassohn Syndrome, 798  
*Eulalia Baselga & Nancy Burton Esterly*  
 Reticulated Acropigmentation of Dohi (Dyschromatosis Symmetrica Hereditaria), 799  
*Eulalia Baselga & Nancy Burton Esterly*  
 Reticulate Acropigmentation of Kitamura, 802  
*Eulalia Baselga & Nancy Burton Esterly*  
 Rothmund–Thomson Syndrome, 804  
*Eulalia Baselga & Nancy Burton Esterly*
- 45 Genetic Epidermal Syndromes with *Café-au-lait* Macules, 809  
 Familial Multiple *Café-au-lait* Spots, 809  
*Nancy Burton Esterly*  
 Neurofibromatosis, 809  
*Nancy Burton Esterly, Eulalia Baselga, & Sheila S. Galbraith*  
 Neurofibromatosis 1 with Noonan Syndrome, 816  
*Nancy Burton Esterly*  
 McCune–Albright Syndrome, 817  
*Sheila S. Galbraith & Nancy Burton Esterly*  
 Segmental Neurofibromatosis, 819  
*Nancy Burton Esterly & Eulalia Baselga*  
 Silver–Russell Syndrome, 820  
*Nancy Burton Esterly, Eulalia Baselga, & Sheila S. Galbraith*  
 Watson Syndrome, 823  
*Nancy Burton Esterly*
- 46 Genetic Epidermal Pigmentation with Lentigines, 824  
 Lentigo Simplex, 824  
*Mary K. Cullen*  
 Lentigo Senilis et Actinicus, 829  
*Mary K. Cullen*  
 Centrofacial Lentiginosis, 837  
*Mary K. Cullen*  
 LEOPARD Syndrome, 842  
*Mary K. Cullen*  
 Carney Complex, 851  
*Mary K. Cullen*  
 Other Lentiginoses, 863  
*Mary K. Cullen*
- 47 Genetic Epidermal Syndromes: Localized Hyperpigmentation, 873  
 Anonychia with Flexural Pigmentation, 873  
*Nancy Burton Esterly, Eulalia Baselga, & Beth A. Drolet*  
 Incontinentia Pigmenti, 873  
*Sheila S. Galbraith & Nancy Burton Esterly*  
 Periorbital Hyperpigmentation, 879  
*Nancy Burton Esterly, Eulalia Baselga, & Beth A. Drolet*

## CONTENTS

- Pigmentary Demarcation Lines, 880  
*Sheila S. Galbraith & Nancy Burton Esterly*
- Dowling–Degos Disease, 882  
*Sheila S. Galbraith & Nancy Burton Esterly*
- 48 Genetic Epidermal Syndromes: Disorders of Aging, 884  
Acrogeria, 884  
*Nancy Burton Esterly, Eulalia Baselga, Peter M. H. Chan, & Beth A. Drolet*
- Metageria, 886  
*Nancy Burton Esterly, Eulalia Baselga, Peter M. H. Chan, & Beth A. Drolet*
- Progeria, 886  
*Nancy Burton Esterly, Eulalia Baselga, Peter M. H. Chan, & Beth A. Drolet*
- Xeroderma Pigmentosum, 889  
*Anita P. Sheth, Nancy Burton Esterly, Eulalia Baselga, Peter M. H. Chan, & Beth A. Drolet*
- Werner Syndrome, 894  
*Nancy Burton Esterly, Eulalia Baselga, Peter M. H. Chan, Beth A. Drolet, & Cindy L. Lamerson*
- 49 Congenital Epidermal Hypermelanoses, 898  
Dyskeratosis Congenita, 898  
*Susan Bayliss Mallory, Peggy L. Chern, & Sharon A. Foley*
- Ectodermal Dysplasias, 901  
*Susan Bayliss Mallory, Peggy L. Chern, & Sharon A. Foley*
- Transient Neonatal Pustular Melanosis, 905  
*Susan Bayliss Mallory, Peggy L. Chern, & Sharon A. Foley*
- Universal Acquired Melanosis, 906  
*Susan Bayliss Mallory, Peggy L. Chern, & Sharon A. Foley*
- 50 Acquired Epidermal Hypermelanoses, 907  
Acanthosis Nigricans, 907  
*Norman Levine & Cynthia Burk*
- Acromelanosis Progressiva, 914  
*Norman Levine & Cynthia Burk*
- Becker Nevus, 915  
*Norman Levine & Cynthia Burk*
- Café-au-lait Spots, 917  
*Norman Levine & Cynthia Burk*
- Carcinoid Syndrome, 919  
*Norman Levine & Cynthia Burk*
- Confluent and Reticulated Papillomatosis, 922  
*Norman Levine & Cynthia Burk*
- Cutaneous Amyloidosis, 924  
*Norman Levine & Cynthia Burk*
- Dermatosis Papulosa Nigra, 928  
*Norman Levine & Cynthia Burk*
- Ephelides (Freckles), 929  
*Norman Levine & Cynthia Burk*
- Erythema ab Igne, 931  
*Norman Levine & Cynthia Burk*
- Erythema Dyschromicum Perstans, 933  
*Norman Levine & Cynthia Burk*
- Erythromelanosits Follicularis Faciei et Colli, 935  
*Norman Levine & Cynthia Burk*
- Erythrose Péribuccale Pigmentaire of Brocq, 937  
*Norman Levine & Cynthia Burk*
- Extracutaneous Neuroendocrine Melanoderma, 938  
*Norman Levine & Cynthia Burk*
- Felty Syndrome and Rheumatoid Arthritis, 939  
*Norman Levine & Cynthia Burk*
- Hyperpigmentation Associated with Human Immunodeficiency Virus (HIV) Infection, 941  
*Philippe Bahadoran*
- Melanoacanthoma, 946  
*Norman Levine & Cynthia Burk*
- Phytophotodermatitis, 948  
*Norman Levine & Cynthia Burk*
- Polyneuropathy, Organomegaly, Endocrinopathy, M Protein, and Skin Changes: POEMS Syndrome, 951  
*James J. Nordlund*
- Urticaria Pigmentosum and Mastocytosis, 954  
*James J. Nordlund*
- Poikiloderma of Civatte, 959  
*Vlada Groysman & Norman Levine*
- Riehl's Melanosis, 961  
*Scott Bangert & Norman Levine*
- Atrophoderma of Pasini et Pierini, 963  
*James J. Nordlund, Norman Levine, Charles S. Fulk, & Randi Rubenzik*
- Hyperpigmentation Associated with Scleromyxedema and Gammopathy, 965  
*Kazunori Urabe, Juichiro Nakayama, & Yoshiaki Hori*
- Ichthyosis Nigricans, Keratoses, and Epidermal Hyperplasia, 965  
*James J. Nordlund*
- Morphea and Scleroderma, 967  
*James J. Nordlund*
- Pigmentary Changes Associated with Addison Disease, 969  
*Cindy L. Lamerson & James J. Nordlund*
- Pigmentary Changes Associated with Cutaneous Lymphomas, 972  
*Debra L. Breneman*
- 51 Hypermelanosis Associated with Gastrointestinal Disorders, 979  
Porphyria Cutanea Tarda, 979  
*Eun Ji Kwon & Victoria P. Werth*
- Cronkhite–Canada Syndrome, 983  
*Eun Ji Kwon, James J. Nordlund, & Victoria P. Werth*
- Hemochromatosis and Hemosiderosis, 986  
*Joerg Albrecht & Victoria P. Werth*
- Primary Biliary Cirrhosis, 992  
*Joerg Albrecht & Victoria P. Werth*
- Inflammatory Bowel Disease and Pigmentation, 995  
*Joerg Albrecht & Victoria P. Werth*

- Pellagra, 995  
*Eun Ji Kwon & Victoria P. Werth*
- Peutz–Jeghers Syndrome, 999  
*Nancy Burton Esterly, Eulalia Baselga, & Beth A. Drolet*
- 52 Acquired and Congenital Dermal Hypermelanosis, 1003  
 Sacral Spot of Infancy, 1003  
*Sang Ju Lee, Seung Kyung-Hann, & Sungbin Im*  
 Nevus of Ota, 1006  
*Sang Ju Lee, Seung Kyung-Hann, & Sungbin Im*  
 Nevus of Ito, 1012  
*Sang Ju Lee, Seung Kyung-Hann, & Sungbin Im*  
 Phakomatosis Pigmentovascularis, 1013  
*Sang Ju Lee, Seung Kyung-Hann, & Sungbin Im*  
 Other Congenital Dermal Melanocytosis, 1015  
*Sang Ju Lee, Seung Kyung-Hann, & Sungbin Im*  
 Acquired Dermal Melanocytosis, 1016  
*Sang Ju Lee, Seung Kyung-Hann, & Sungbin Im*  
 Acquired Bilateral Nevus of Ota-like Macules  
 (ABNOM), 1017  
*Sang Ju Lee, Seung Kyung-Hann, & Sungbin Im*  
 Blue Macules Associated with Progressive Systemic  
 Sclerosis, 1018  
*Sang Ju Lee, Seung Kyung-Hann, & Sungbin Im*
- 53 Mixed Epidermal and Dermal Hypermelanoses and  
 Hyperchromias, 1020  
 Melasma, 1020  
*Sang Ju Lee, Seung Kyung-Hann, & Sungbin Im*  
 Melanosis from Melanoma, 1023  
*Sang Ju Lee, Seung Kyung-Hann, & Sungbin Im*
- 54 Drug-induced or -related Pigmentation, 1026  
*Peter A. Lio & Arthur J. Sober*
- Section 6: Disorders of Pigmentation of the Nails and Mucous Membranes, 1055**
- 55 The Melanocyte System of the Nail and its Disorders,  
 1057  
*Robert Baran, Christophe Perrin, Luc Thomas, & Ralph Braun*
- 56 Pigmentary Abnormalities and Discolorations of the  
 Mucous Membranes, 1069  
*John C. Maize, Jr. & John C. Maize, Sr.*
- Section 7: Benign Neoplasms of Melanocytes, 1091**
- 57 Common Benign Neoplasms of Melanocytes, 1093  
 Pigmented Spindle Cell Nevi, 1093  
*Julie V. Schaffer & Jean L. Bolognia*  
 Speckled Lentiginous Nevus (Nevus Spilus), 1098  
*Julie V. Schaffer & Jean L. Bolognia*  
 Melanocytic (Nevocellular) Nevi and Their Biology,  
 1112  
*Julie V. Schaffer & Jean L. Bolognia*
- 58 Rare Benign Neoplasms of Melanocytes, 1148  
 Nevus Aversion Phenomenon, 1148  
*James J. Nordlund*  
 Melanotic Neuroectodermal Tumor of Infancy, 1148  
*Julie V. Schaffer & Jean L. Bolognia*  
 Pilar Neurocristic Hamartoma, 1157  
*Julie V. Schaffer & Jean L. Bolognia*
- Section 8: Treatment of Pigmentary Disorders, 1163**
- 59 Topical Treatment of Pigmentary Disorders, 1165  
*Rebat M. Halder & James J. Nordlund*
- 60 Chemophototherapy of Pigmentary Disorders, 1175  
*Rebat M. Halder & James J. Nordlund*
- 61 UVB Therapy for Pigmentary Disorders, 1183  
*Thierry Passeron & Jean-Paul Ortonne*
- 62 Sunscreens and Cosmetics, 1188  
*James J. Nordlund & Rebatt M. Halder*
- 63 Surgical Treatments of Pigmentary Disorders, 1191  
*Rebat M. Halder & James J. Nordlund*
- 64 Laser Treatment of Pigmentary Disorders, 1198  
*Rebat M. Halder, Lori M. Hobbs, & James J. Nordlund*
- Index, 1205  
*Plate section can be found between pages 494 and 495*

# Contributors

## Zalfa Abdel-Malek PhD

*Research Professor of Dermatology, Department of Dermatology,  
University of Cincinnati College of Medicine, Cincinnati, OH, USA*

## Joerg Albrecht MD

*Department of Dermatology, University of Pennsylvania,  
Philadelphia, PA, USA*

## Mayra Alvarez-Franco (deceased)

*Formerly of the Departments of Dermatology and Pathology, Yale  
University School of Medicine, New Haven, CT, USA*

## Joseph T. Bagnara PhD

*Professor Emeritus, Department Cell Biology and Anatomy,  
University of Arizona College of Medicine, Tucson, AZ, USA*

## Philippe Bahadoran MD PhD

*Assistant Professor of Medicine, Service de Dermatologie,  
Hôpital l'Archet, Nice, France*

## Debdutta Bandyopadhyay PhD

*Instructor of Dermatology, Huffington Center on Aging, Baylor  
College of Medicine, Houston, TX, USA*

## Scott D. Bangert MD

*Banner Good Samaritan Hospital, Department of Medical  
Education, Phoenix, AZ, USA*

## Robert Baran MD

*Head of Nail Disease Center, Cannes, France*

## Gregory S. Barsh MD PhD

*Professor of Genetics and Pediatrics, Beckman Center for Molecular  
and Genetic Medicine, Stanford University School of Medicine,  
Stanford, CA, USA*

## Eulalia Baselga MD

*Consultant in Pediatric Dermatology, Hospital de la Santa Creu i  
Sant Pau, Barcelona, Spain*

## Dorothy C. Bennett MA PhD

*Professor of Cell Biology, St George's Hospital, University of  
London, London, UK*

## Raymond E. Boissy PhD

*Professor of Dermatology and Cell Biology, Department of  
Dermatology, University of Cincinnati College of Medicine,  
Cincinnati, OH, USA*

## Jean L. Bolognia MD

*Professor of Dermatology, Department of Dermatology, Yale  
University School of Medicine, New Haven, CT, USA*

## Barbara Boone MD

*Resident in Dermatology, Universitair Ziekenhuis Ghent, Ghent,  
Belgium*

## Ralph P. Braun MD

*Assistant Professor, Pigmented Skin Lesion Unit, Department of  
Dermatology, University Hospital, Geneva, Switzerland*

## Debra L. Breneman MD

*Associate Professor of Dermatology, University of Cincinnati,  
Cincinnati, OH, USA*

## Stefania Briganti PhD

*San Gallicano Dermatological Institute, Rome, Italy*

## Murray H. Brilliant PhD

*Lindholm Professor of Genetics, Department of Pediatrics,  
University of Arizona College of Medicine, Tucson, AZ, USA*

## Lieve Brochez MD PhD

*Professor of Dermatology, Department of Dermatology, University  
Hospital of Ghent, Ghent, Belgium*

## Cynthia J. Burk MD

*Section of Dermatology, University of Arizona Health Sciences  
Center, Tucson, AZ, USA*

## Stella D. Calobrisi MD

*Medical Director, The Dermatology Clinic, Boca Raton, FL, USA*

## Tania Cestari MD PhD

*Associate Professor of Dermatology, Department of Dermatology,  
University of Rio Grande do Sul, Hospital de Clínicas de Porto  
Alegre, Brazil*

**Henry H. L. Chan** MBBS MSc (Clin Derm) MD FRCP FRCP(Ed) FRCP(Glasg) FHKCP FHKAM (Med)  
*Specialist in Dermatology and Honorary Clinical Associate Professor, University of Hong Kong and Chinese University of Hong Kong, Hong Kong*

**Peter Man Hon Chan** MD  
*Clinical Research Fellow, University of Minnesota, Eagan, MN, USA*

**Peggy L. Chern** MD  
*Department of Dermatology, University of Iowa Hospitals and Clinics, Iowa City, IA, USA*

**Mary K. Cullen** MD  
*Department of Cell Biology and Physiology, Clinical Instructor, Washington University School of Medicine, St Louis, MO, USA*

**Beth A. Drolet** MD  
*Department of Dermatology, The Medical College of Wisconsin, Milwaukee, WI, USA*

**Carol A. Erickson** PhD  
*Professor of Molecular Cellular Biology, University of California at Davis, Davis, CA, USA*

**Nancy B. Esterly** MD  
*Professor of Dermatology and Pediatrics, Department of Dermatology, The Medical College of Wisconsin, Milwaukee, WI, USA*

**Dong Fang** MD PhD  
*Staff Scientist, The Wistar Institute, Philadelphia, PA, USA*

**Sharon A. Foley** MD  
*Department of Dermatology, Washington University School of Medicine, St Louis, MO, USA*

**Peter S. Friedmann** MD FRCP FMedSci  
*Professor of Dermatology and Head of Division, Dermatopharmacology Unit, Southampton General Hospital, Southampton, UK*

**Charles S. Fulk** MD FACP  
*Department of Pathology, Vanderbilt University, Nashville, TN, USA*

**Anne-Sophie J. Gadenne** MD  
*Department of Dermatology, University of Cincinnati College of Medicine, Cincinnati, OH, USA*

**William A. Gahl** MD PhD  
*Clinical Director, National Human Genome Research Institute, National Institutes of Health, Bethesda, MD, USA*

**Sheila S. Galbraith** MD  
*Assistant Professor of Dermatology, Dermatology Department, Medical College of Wisconsin, Milwaukee, WI, USA*

**José C. García-Borrón** PhD  
*Professor of Biochemistry and Molecular Biology, School of Medicine, University of Murcia, Murcia, Spain*

**Rosemary J. Geary** MD  
*President, East Valley Dermatology Center, Chandler, AZ, USA*

**Vanessa C. Gray-Schopfer** PhD  
*Signal Transduction Team, Cellular and Molecular Biology Section, Institute of Cancer Research, London, UK*

**Pearl E. Grimes** MD  
*Vitiligo and Pigmentation Institute of Southern California, David Geffen School of Medicine, University of California at Los Angeles, Los Angeles, CA, USA*

**Vlada Groysman** MD  
*Section of Dermatology, University of Arizona Health Sciences Center, Tucson, AZ, USA*

**Ruth Halaban** PhD  
*Senior Research Scientist, Department of Dermatology, Yale University School of Medicine, New Haven, CT, USA*

**Rebat M. Halder** MD  
*Professor and Chairman, Department of Dermatology, Howard University, Washington, DC, USA*

**Seung-Kyung Hann** MD PhD  
*Director, Korea Institute of Vitiligo Research, Yongsan-Gu, Seoul, Korea*

**Rudolf Happle** MD  
*Professor of Dermatology, Department of Dermatology, Philipp's University of Marburg, Marburg, Germany*

**Vincent J. Hearing** PhD  
*Chief, Pigment Cell Biology Section, Laboratory of Cell Biology, National Institutes of Health, Bethesda, MD, USA*

**Meenhard Herlyn** DVM, DSc  
*Professor and Chair of Program of Molecular and Cellular Oncogenesis, The Wistar Institute, Philadelphia, PA, USA*

**Stan P. Hill** MD  
*Denver, CO, USA*

**Lori M. Hobbs** MD  
*Assistant Professor, Department of Dermatology, Howard University, Washington, DC, USA*

**Thomas J. Holstein** PhD  
*Professor Emeritus, Roger Williams University, Bristol, RI, USA*

**Yoshiaki Hori** MD PhD (deceased)  
*Formerly Honorary Professor, Department of Dermatology, Kyushu University, Fukuoka, Japan*