

# Review of *The Pigmentary System: Physiology and Pathophysiology* (2nd ed) by James J. Nordlund, Raymond E. Boissy, Vincent J. Hearing, et al.

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## Introduction

It is not without some apprehension that I submit my first book review to the readership of *Dermatology Practical & Conceptual*. The challenge of succeeding my colleague and friend, Mark Hurt, as Book Review Editor is great. In fact, I am certain that I will not be able to fulfill it alone for any length of time. Therefore I want to begin by inviting readers of the Journal to participate actively by suggesting titles they would like to see reviewed, submitting their own reviews and/or in any other way that might come to their mind. I would be extremely grateful to receive your contributions. I can be contacted at: francois.milette@cssspb.qc.ca.

The format that Mark used to apply to his reviews (submitting the books to two reviewers, obtaining responses from the authors when possible and only thereafter writing his own text) is ideal and should be maintained. However since I have accepted only very recently to the opportunity of Book Review Editor, I could not proceed that way this time. Rather, I have chosen to present a book that I had the pleasure to read and found unexpected and interesting surprises.

Let me now humbly present my first contribution as Book Review Editor of the Journal.

## Review by François Milette

I bought *The Pigmentary System* a few years ago because at the time I was wondering what was the scientific basis

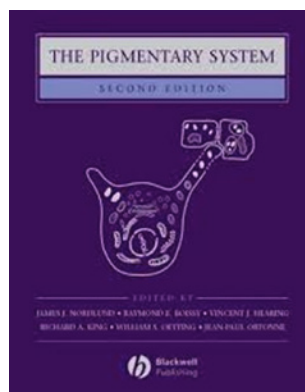


Figure 1. *The Pigmentary System* (2nd edition) by James J. Nordlund, Raymond Boissy, Vincent J. Hearing, Richard A. King, William Oetting (ed.), Jean-Paul Ortonne (ed.). Hardcover: 1229 pages. Publisher: Blackwell. ISBN: 1405120347. List Price: \$359.95.

that sustained the argument according that the presence of benign melanocytes in lymph nodes was the result of “migration arrest” during embryogenesis. The subtitle of this book: *Physiology and Pathophysiology* appeared promising.

Unfortunately, I did not find a satisfactory answer to my question, but after reading through its many pages I found, on page 1016, the following paragraph that I considered as a conclusion to my quest:

*The pathogenesis of persistent dermal melanocytes is uncertain. Dermal melanocytes are presumed by analogy with epidermal melanocytes to arise in the neural crest and migrate onto the skin. It has been suggested that dermal melanocytes are simply melanocytes destined for the epidermis that have remained in the dermis. Because of the lack of information on the*

*determinants of normal migration of epidermal melanocytes, our understanding of these lesions is poor.*

If this is true of dermal melanocytes, needless to say, it can only be truer for nodal melanocytes. My *a priori* intuition that the affirmation that nodal melanocytes were “migration arrested” was essentially speculative was confirmed.

However, as is often the case in life, the point of arrival was less interesting than the road followed. I had made a wonderful journey through the pages of this book that purported to be nothing less than the definitive text of its time! In fact this rather vain statement has some truth in it. The book is a team effort to which nearly 120 authors from all around the world collaborated. It covers all aspects of the “pigmentary system” that can come to mind. Even the contemptible notion of skin color based racism is evoked (on page 5):

*The furious pace of forward progress was slowed somewhat in the early part of the nineteenth century when skin color scientists in both Europe and America were drawn into acrimonious debates over social issues, especially slavery and the place of “peoples of color” in the family of man.*

As its subtitle indicates, the book is divided in two parts: I. Physiology and II. Pathophysiology.

Part I is particularly interesting for a dermatologist or a dermatopathologist curious to explore the basic science foundations of his specialty. Everything is addressed: the history of science, comparative anatomy, general biology, embryology, biochemistry and biogenesis of melanin and melanosomes, metabolism and regulation of melanin formation and “trafficking,” genetics, etc. In the various chapters of Part I, the reader will be amazed by the far-reaching extent the study of pigmentation can take.

Part II concerns pathophysiology. It is divided into six sections. The first section is interesting as it begins by an overview of the disorders of pigmentation in humans. In it are discussed the various mechanisms by which pigmentation can be altered. It also proposes a series of clear definitions supporting a coherent classification of pigmentary disorders (Tables 26.5 and 26.6 on p 502):

**TABLE 26.5.** *Some essential definitions.*

*Achromia: a type of leukoderma; totally white skin from any cause.*

*Amelanocytosis: total absence of all melanocytes in the epidermis that will result in amelanosis.*

*Depigmentation (amelanosis): a type of leukoderma also called amelanosis caused by total absence of melanin in the epidermis from any cause.*

*Dermatomal: following the distribution of a cutaneous sensory nerve.*

*Hyperchromia: skin color that is darker than normal from any cause.*

*Hypermelanocytosis: a higher than normal population density of melanocytes in the skin, in the epidermis, the dermis, or both, resulting typically in hypermelanosis.*

*Hypermelanosis: a type of hyperchromia that results from increased melanin in the skin, in the epidermis, dermis, or both.*

*Hypochromia: a type of leukoderma; skin color that is lighter than normal from any cause.*

*Hypomelanocytosis: a lower than normal population density of melanocytes in the epidermis typically resulting in hypomelanosis.*

*Hypomelanosis: a type of leukoderma caused by decreased melanin in the epidermis.*

*Hypopigmentation (hypomelanosis): a type of leukoderma caused by partial absence of melanin in the epidermis.*

*Leukoderma: skin with a white discoloration from any cause or by any mechanism.*

*Pigmentary system: all melanocytes and their product melanin at all sites within the body.*

*Pigmentation: of or pertaining to melanocytes or melanin.*

*Segmental: one portion of the integument, usually unilateral.*

*Skin color: the color of the skin is determined by two distinct groups of chromophores (cells, structural agents, chemicals that impart color to the skin), those of the pigmentary system (i.e. melanin and melanocytes) and those composed of other elements (chromatics) of the skin such as collagen, blood, carotenes, etc.*

**TABLE 26.4.** *A classification of the disorders of skin color.*

A) *Hyperchromia*

I) *Melanotic types of hyperchromia (hyperpigmentation)*

a) *Hypermelanosis (increased melanin only and normal population density of melanocytes)*

1) *Congenital*

i) *Localized, variable, or generalized*

ii) *Epidermal, dermal, or mixed*

2) *Acquired*

i) *Localized, variable, or generalized*

- ii) *Epidermal, dermal, or mixed*
- b) *Hypermelanocytosis (increased melanocytes and melanin)*
  - 1) *Congenital*
    - i) *Localized, variable, or generalized*
    - ii) *Epidermal, dermal, or mixed*
  - 2) *Acquired*
    - i) *Localized, variable or generalized*
    - ii) *Epidermal, dermal or mixed*
- II) *Nonmelanotic types of hyperchromia*
  - 1) *Congenital*
    - i) *Localized, variable, or generalized*
    - ii) *Epidermal, dermal, or mixed*
  - 2) *Acquired*
    - i) *Localized, variable, or generalized*
    - ii) *Epidermal, dermal, or mixed*
- B) *Hypo- or achromia (leukoderma)*
  - I) *Melanotic types of leukoderma (hypo- or depigmentation)*
    - a) *Hypomelanosis or amelanosis (decreased melanin only)*
      - 1) *Congenital*
        - i) *Localized, variable, or generalized*
        - ii) *Epidermal, dermal, or mixed*
      - 2) *Acquired*
        - i) *Localized, variable, or generalized*
        - ii) *Epidermal, dermal, or mixed*
    - b) *Hypo- or amelanocytosis (partial or total absence of melanocytes)*
      - 1) *Congenital*
        - i) *Localized, variable, or generalized*
        - ii) *Epidermal, dermal, or mixed*
      - 2) *Acquired*
        - i) *Localized, variable, or generalized*
        - ii) *Epidermal, dermal, or mixed*
  - II) *Nonmelanotic hypochromia*
    - 1) *Congenital*
      - i) *Localized, variable, or generalized*
      - ii) *Epidermal, dermal, or mixed*
    - 2) *Acquired*
      - i) *Localized, variable, or generalized*
      - ii) *Epidermal, dermal, or mixed*

The following two sections explore respectively hypopigmentation and hyper-pigmentation disorders from the most common to the rarest and exotic entities. Each is presented in a coherent manner going through historical perspective, terminology, epidemiology, clinical findings, associated disorders, histopathology, laboratory investigations, diagnostic criteria, differential diagnosis, pathogenesis, treatment and

prognosis. Iconography of these two sections can be considered remarkable especially if one considers the rarity of many entities treated.

The following section addresses the pigmentary disorders of nails and mucous membranes. It is not clear to me why placing those entities affecting nails and mucosae apart appeared necessary to the authors and editors of the book. In my opinion it creates some confusion, as in the preceding sections extracutaneous disorders of pigmentation affecting the meninges or the eyes were included with disorders of the skin.

The next section concerns benign neoplasms. This is probably, at least to the eye of a pathologist, the weakest part of the book. The section is divided in two parts devoted respectively to frequent and rare neoplasms. The designation "rare" should probably be replaced by "controversial," as at least some entities discussed in this section captioned "Rare benign neoplasms of melanocytes" are clearly malignant. Consider, for instance, the so-called "melanotic neuroectodermal tumor of infancy" described on pages 1148 to 1157. Can it be reasonably considered benign when it is acknowledged that it has significant metastatic potential and is lethal in a significant number of cases impossible to differentiate from nonmetastatic tumors? My late friend Bernie Ackerman would have cut short this debate: THIS IS MELANOMA!

Incidentally, it is perhaps regrettable and largely unexplainable that there is no chapter devoted to malignant tumors (that is, melanoma) of melanocytes in this book. Perhaps is it better that way, though, judging from the unconditional adoption of the dysplastic nevus theory presented in chapter 24 devoted to the genetics of melanoma. The mythology of precancers surrounding the concept of melanocytic neoplasia physiopathology could only have been spread by a chapter such as this.

The last section of the book concerns treatment of pigmentary disorders. I humbly admit that, since I am a pathologist, I skipped this section. Pardon me! As an excuse I may say that the book already is seven years old and I suspect that many things probably have evolved during these years so. Since I cannot judge, I leave it to the interested reader to decide.

I will conclude by admitting that this second edition is already dated and it is possible and even probable that some notions presented have evolved. To my knowledge there has been no further edition of this book. Nevertheless, this second edition is so large in its scope that I am convinced any curious scientist interested by the subject will still find valuable things in it.