

Book Review

INHERITED DISORDERS OF THE THYROID SYSTEM

Geraldo Medeiros-Neto and John Bruton Stanbury
Boca Raton, FL: CRC Press, Inc., 1994, pp. 218.

Let me start by saying that I am no expert on inherited diseases of the thyroid nor do I have any particular qualifications to be a "book reviewer." I elected to review this book to learn more about thyroid disease and, I will admit, to get a free copy of a book that I probably would not buy—not because it is not well written (because it is), but simply because my professional interests and activities steer me toward other publications. I do have a background in cancer genetics and was anxious to get this book in hopes of reading good review chapters on familial medullary thyroid carcinoma and MEN II. What a disappointment it was to find that only 4 sentences about these inherited thyroid conditions (page 200, Chapter 11, "Other Inherited Diseases of the Thyroid") are included in a book titled "Inherited Disorders of the Thyroid System," written by internationally recognized endocrinologists Drs. Geraldo Medeiros-Neto and John B. Stanbury. The only other chapter that focuses at all on thyroid malignancies is the 12 page last chapter, Chapter 12—"Thyroid Malignancy and the Dys hormonogenic Goiter," which discusses the risks and pathogenesis of malignancies in congenital goiter associated with hypothyroidism. However, after my initial dismay, I was pleasantly surprised when I sat down to read this well-organized textbook that presented excellent written discussions of several inherited thyroid conditions about which I, because of this book, now know more.

After Chapter 1, which presents "A Retrospective View of the Inherited Errors of the Thyroid System" (a concise and interesting historical background of our understanding of these conditions), the remaining 11 chapters provide easy-to-follow reviews of several conditions that include sections on the clinical aspects, pathophysiology, animal models, and molecular biology (only as current as 1993) as appropriate. Chapters 2–10 are, respectively, titled "Familial Inherited TSH Deficiency," "Congenital Hypothyroidism Associated with Thyrotropin Unresponsiveness," "The Iodide Transport Defect," "Defective Organification of Iodide," "Pendred's Syndrome: Association of Congenital Deafness with Sporadic Goiter," "Defects in Tg Gene Expression and Tg Secretion," "The Iodotyrosine Deiodinase Defect,"

"The Syndromes of Reduced Responsiveness to Thyroid Hormone," and "Inherited Abnormalities in Thyroid Hormone Transport Proteins." All of the chapters are well referenced, are clearly written, and contain appropriate, abundant helpful figures and tables. Any of these chapters could securely stand alone as an excellent, independent, review paper. Each chapter follows essentially the same easy-to-follow format with excellent introductions, outlined summaries of key points, and complete reference lists which include full titles. These organizational features, along with the numerous illustrations, help make this book one can easily pick up and read intermittently as time allows. Even if one only has a few moments to look through a chapter, the way this book is structured, it is possible to absorb some useful information in a short amount of time.

In summary, for those of us who have little expertise in endocrinology, this is an easy-to-read, well-illustrated, informative book focusing on several inherited disorders of the thyroid—largely those disorders associated with abnormal iodide utilization and/or hormonal function. For those with expertise in this area, the content of this book may be largely of a review nature. I do wish that I would have had the excellent chapter reviewing Pendred syndrome available to me during my first year as a clinical genetics fellow when (never having before heard of the syndrome) I had the opportunity to evaluate a girl who presented with deafness, short stature, and hypothyroidism. As I was busy thinking about a differential diagnoses that, in my naiveté, focused largely on chromosomal abnormalities, I was called by the girl's endocrinologist who wondered why I was looking for a chromosome abnormality in a girl who clearly had Pendred syndrome, an autosomal recessive disorder. Well, needless to say, I have since read a lot about Pendred syndrome and really appreciated the great review of it in this book. Despite the fact that I do think it is a shortcoming of the book to omit any significant discussion of MEN II and medullary thyroid carcinoma, I would highly recommend this little book to individuals who are looking for comprehensive, concise reviews of any of the topics included in it.

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