

## Book Reviews

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*Am. J. Hum. Genet.* 72:503, 2003

*Cleft Lip and Palate: From Origin to Treatment.* Edited by Diego F. Wyszynski. New York: Oxford University Press, 2002. Pp. 518. \$125.

Diego Wyszynski has done an admirable job of assembling a collection of reviews of various aspects of the etiology and treatment of oral clefts. This 500-page monograph is divided into three sections: "Basic Principles" (embryology, clinical features, epidemiology, and genetics); "Treatment" (surgery, speech care, dentistry, otolaryngology, etc.); and "Public Health Issues" (prevention, insurance coverage, ethical issues, and education). As with all edited monographs, some essays are more scholarly than others; a few are rather poor. Nevertheless, this book achieves its stated intention of "addressing issues that are relevant to clinicians, researchers, and family members." Of particular value to clinicians and well-informed family members is the section on treatment. The treatment of patients with oral clefts, from birth to adulthood, requires a well-coordinated team effort. This section does a good job of conveying the global nature and specific details of this comprehensive care. Thus, chapters 24–34 are highly recommended, as are chapters 36–39 in the section on public health issues.

One important misstep in this presentation is the lack of succinct clarity regarding our present knowledge of oral cleft etiology. In 1949, Curt Stern, writing in his text *Principles of Human Genetics*, noted: "Harelip and cleft palate are developmental abnormalities which have a genetic basis. In many pedigrees, they depend on the cooperation of specific alleles at several autosomal loci, and, in addition, require the presence of mostly uncontrolled environmental factors." In 2002, there is little else we can add. F. Clarke Fraser, in his foreword to Wyszynski's monograph, cogently tells us why: "Epidemiologic studies are hampered by the fact that, for an uncommon multifactorial trait, it is virtually impossible to assemble groups large enough to allow statistically valid comparisons of potentially significant factors such as prenatal events, susceptibility genes, social class, and race." Unfortunately, one is left to conclude that much of what is contained in chapter 3 and chapters 16–22 is much ado about very little. I hope that, someday, this will be different. Today, at bottom, we must rely entirely on the information in Carmella Stadter's chapter (31) on genetic counseling and empiric risk calculation. This message is lost amid the genomic hype. In a book "addressing issues ... relevant to clinicians ... and family members," this should have been in neon.

On balance, this monograph has more to recommend it than

not. It should be on the shelf of all those with a need to know about clefts.

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0002-9297/2003/7202-0036\$15.00

*Am. J. Hum. Genet.* 72:503–504, 2003

*Connective Tissue and Its Heritable Disorders: Molecular, Genetic, and Medical Aspects*, 2nd ed. Edited by Peter M. Royce and Beat Steinmann. New York: Wiley-Liss, 2002. Pp. 1201. \$350.

The second edition of *Connective Tissue and Its Heritable Disorders* has been vastly expanded, in comparison with the first edition, as is consistent with the rapid progress in the field over the last 10 years. The authors of individual chapters are all leaders in the field, and the book has been well edited to give it a consistent structure. This is a very dense book, with a relatively small font size, that is not designed for those seeking a quick review of the field but rather for those interested in all the facts as well as the controversies. It is relatively well illustrated, although many complex pathways are discussed without figures that would have made the relationships easier to understand for all but the mavens of the field.

The first third of *Connective Tissue and Its Heritable Disorders* covers the basic biology of connective tissue in great detail, although I found the treatment of the regulation of growth and development to be superficial compared with the review of the collagens. Chapters on clinical disorders constitute the remainder of the book. In addition to chapters on disorders traditionally placed under the heading of "connective tissue," such as osteogenesis imperfecta, Ehlers-Danlos syndrome, Marfan syndrome, cutis laxa, and pseudoxanthoma elasticum, there are chapters on epidermolysis bullosa, disorders of keratinization, Alport syndrome, defects in skeletal morphogenesis, and many other disorders. Those familiar with *The Metabolic and Molecular Bases of Inherited Disease* (MMBID) will recognize the format of the chapters in this book, with a summary of the important points at the beginning

of the chapter and with sections on the genetic, molecular, pathophysiologic, and clinical features of the disorders, animal models, and treatments. Most chapters have also appended any recent developments in the field, allowing this book to be as up to date as any book can be. In another similarity to MMBID, *Connective Tissue and Its Heritable Disorders* tends to emphasize the basic science over the clinical aspects of the conditions. Disorders without defined defects are covered well in some chapters but not in others. The only clinical area I felt was inadequately covered, in general, was the skeletal dysplasias, particularly those without defined defects. This is, perhaps, understandable, given that there are >200 recognized disorders, a proper treatment of which could fully occupy a volume of its own. A few disorders, such as van Buchem endosteal hyperostosis, sclerosteosis, and Camurati-Engelmann disease, that have recently defined defects escaped attention.

With many other books available and increasingly limited budgets, a comparison with other choices seems appropriate. Although recent chapters on many of the clinical entities covered in *Connective Tissue and Its Heritable Disorders* can be found in MMBID and *Emery and Rimoin's Principles and*

*Practice of Medical Genetics*, no one book covers all the disorders, along with offering an extensive review of the basic biology. I never bought the first edition of *Connective Tissue and Its Heritable Disorders*, because it was published about the same time as the fifth edition of *McKusick's Heritable Disorders of Connective Tissue*, a book that is more richly illustrated and more oriented toward the clinician. However, *McKusick's Heritable Disorders of Connective Tissue* is now out of date, and there are no current plans for a new edition. *Connective Tissue and Its Heritable Disorders* is, thus, a welcome addition to the library of the medical geneticist and is essential for workers in the connective tissue field.

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0002-9297/2003/7202-0037\$15.00