

Book Reviews

Adult Congenital Heart Disease: A Practical Guide.

Michael A. Gatzoulis, Lorna Swan, Judith Therrien, and George A. Pantely. Malden, MA: Blackwell Publishing; 2005. 228 pp. \$54.95. ISBN-0-7279-1668-8.

INTERNISTS AND CARDIOLOGISTS, many of whom are highly skilled in managing adults with an impressive array of serious cardiac problems, are often reluctant to treat an adult patient born with a complex cardiac malformation. If the adult cardiologist has had little or no exposure to the unique anatomy and pathophysiology associated with congenital cardiac malformations, there is little chance that he or she will feel comfortable managing the multitude of problems associated with this condition. The pediatric cardiologist, on the other hand, whose skills are focused on diagnosis and management of complex cardiac malformations, more often than not loses his role as the care provider when the patient becomes an adult and turns to the internist or the adult cardiologist for ongoing cardiac care. A few centers, however, bridge this gap and produce cardiologists fully capable of managing the problems that crop up in adults born with cardiac malformations.

For the internist or cardiologist who needs a guide for the diagnosis and treatment of adult congenital heart disease, there is a new softcover, relatively inexpensive 274-page book that can be particularly helpful. The book, entitled *Adult Congenital Heart Disease: a Practical Guide*, authored by Michael A. Gatzoulis, Lorna Swan, Judith Therrien, and George A. Pantely, is concise and informative. There is a wealth of detail available, more than found in any other "pocket edition" published on this subject to date. Although the print is small, the text is clear enough to help the reader to fully understand the anatomy and pathophysiology of cardiac malformations. In addition, there are informative chapters on various services for adults with congenital heart disease, as well as such complicating factors as pregnancy and contraception; anticoagulation; and lifestyle issues, which are important subjects rarely discussed in standard textbooks.

One illustration, Fig. 13.1, is somewhat misleading. The relationship of the main pulmonary artery to the ascending aorta in congenitally corrected transposition is misrepresented. The illustration shows the ascending aorta medial to the main pulmonary artery, but in fact a hallmark of

congenitally corrected transposition is an ascending aorta lateral to the main pulmonary artery. This one error, however, does not significantly detract from the value of this edition. As someone who has spent more than four decades dealing with patients who have congenital heart disease, I would still find it of value to have this volume on my bookshelf.

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Cardiovascular Development and Congenital Malformations.

Michael Artman, D. Woodrow Benson, Deepak Srivastava, and Makoto Nakazawa, editors. Malden, Oxford, Victoria: Blackwell Publishing; 2005. 292 pp. \$120.00. ISBN-13: 978-1-4051-3128-5 and ISBN-10: 1-4051-3128-4.

My first exposure to patients with congenital heart disease was in my third year of medical school, in 1952, when I elected to join Dr. Ludwig Eichna's Adult Congenital Heart Disease Clinic at Bellevue Hospital. I attended every Thursday evening throughout my third and fourth year of medical school. It took me a while to realize that the patients at this clinic were highly unusual, since in those days most infants and young children with congenital heart disease did not survive. It was a tribute to the relatively primitive but expert surgery and care provided for these patients that they lived and functioned as well as they did. Of course, aside from the anatomical aspects of their disease, not much was known, and most interventions that we now take for granted did not exist.

The last 50 years have greatly improved the outlook of babies born with congenital heart disease. A major reason for this change is that we are learning more and more about the causes of these defects, and (as has become the rule) investigation of these abnormalities has taught us an enormous amount about the normal embryology of the cardiovascular system. And so the book covers both normal and abnormal cardiovascular development.

Cardiovascular Development and Congenital Malformations, edited by four experts in the field, is a compilation of 70 contributions divided into 13 sections. Each of these contributions, many written by the scientists responsible for the ideas

and experiments leading to these advances, is a concise, well-written and well-referenced paper averaging two to four pages in length (although some are longer and more detailed). The brevity of the contributions does not detract from their usefulness, and, if anything, makes them easier to read and understand. Each of the 13 sections (except one) is introduced by an instructive summary of the subject, written by one of the editors.

The first 11 of these sections concern what is known about the developmental biology of cardiovascular embryology. The field is fully covered, including establishment of left-right patterning, cardiogenesis, formation of valves, chambers and conduction tissues, the development of coronary arteries, the role of neural crest cells, formation of outflow tracts and physiological aspects during development. Also included are animal models of congenital cardiovascular malformations and imaging techniques in embryos. The last two sections of the volume are more clinically oriented and deal with patent ductus and human clinical genetics and epidemiology as they concern congenital heart disease. While one could highlight many of the individual contributions, I was particularly impressed by two papers (provided by Bruce Gelb), which typify the new molecular approaches to an understanding of these complex disorders, and an impressive set of contributions concerning various aspects of animal models of DiGeorge syndrome. There are numerous illustrations and a truly wonderful collection of color plates that illustrate many of the points made in the papers.

The book is quite easy to read, and each of the contributions provides a comprehensive bibliography for those who wish to delve more deeply into the subject. If I were to find one thing to quibble about, it is that many of the bibliographies end in 2001, with some going to 2002 and rarely into 2003. However, by using PubMed, one can probably update these bibliographies, something that hopefully will be done in a future edition. That such an update of the field is already necessary is shown by the acknowledgments by the various authors as to what is not yet known. This comprehensive collection should be of great interest to all pediatric cardiologists, embryologists and practitioners of fetal medicine and belongs on their shelves for ready reference.

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The Brugada Syndrome: From Bench to Bedside.

Charles Antzelevitch, Pedro Brugada, Josep Brugada, and Ramon Brugada, editors.

Malden, MA: Blackwell Publishing; 2005.
248 pp. \$99.50. ISBN-1405127783.

FIRST DESCRIBED as a clinical entity in 1992, the Brugada syndrome has since attracted great attention from the medical community because of its association with a high risk for sudden death in young adults, particularly men. The Brugada syndrome is characterized by ST segment elevation in the right precordial EKG leads in patients with structurally normal hearts. The syndrome is believed to be responsible for 4–12% of all sudden deaths and at least 20% of deaths in patients with structurally normal hearts. Investigation into the etiology of the Brugada syndrome has revealed that it is characterized by an abnormality in phase 2 of the action potential, leading to fatal ventricular arrhythmias. Although the etiology remains controversial, several risk factors have been identified for a Brugada-type EKG and the associated ventricular arrhythmias and sudden death.

In *The Brugada Syndrome: From Bench to Bedside*, edited by Charles Antzelevitch, the authors comprehensively describe the many aspects of Brugada syndrome. The book begins with an overview of the clinical characteristics, definition, mechanism, and therapy of the disease by Dr. Antzelevitch, the lead author of the current consensus statement for the management of patients with the syndrome. From there, after a brief description of the history of Brugada syndrome, we get an in-depth review of the genetics of the syndrome, the make-up and mutations of the sodium channel attributed to the syndrome, and the underlying cellular mechanisms believed to underlie the arrhythmic manifestations. The book then moves into more clinical aspects of the disorder. The diagnostic criteria are described with distinguishing characteristics relative to other disorders linked with sudden death. The role of genetics in clinical practice is reviewed, including the gender differences inherent to the disorder. Finally, the issue of prognosis and risk stratification of individuals with the Brugada EKG is examined, with recommendations for deciding who should be treated with an implantable cardioverter defibrillator.

In summary, *The Brugada Syndrome: From Bench to Bedside* is a comprehensive review of all aspects of the syndrome, although little attention is paid to the ongoing controversies on the role of

electrophysiologic testing for risk stratification. It is an excellent read not only for cardiologists and electrophysiologists but also for those interested in how to approach a disorder comprehensively, through molecular, genetic, and clinical aspects. Indeed, the methodology used to explore the features of Brugada syndrome in this book could serve as a paradigm for the approach to various clinical syndromes in medicine.

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Textbook of Hemophilia.

Christine A. Lee, Erik E. Berntorp, and W. Keith Hoots, editors. Malden, MA: Blackwell Publishing; 2005. 416 pp. \$199.00. ISBN-1-405-12769-4.

HEMOPHILIA and the related bleeding disorders have been the subject of numerous monographs. This text is a compilation of 63 chapters written by 90 world authorities. Topics covered include the basic mechanisms of hemostasis, biochemistry of clotting factors, relevant genetics, and current and evolving treatments (specifically, gene therapy).

The book also contains an up-to-date presentation of the basic science of hemophilia, the clinical and laboratory evaluation of patients, the therapies, and the pharmacoeconomics of treatment. There is an appropriate mix of the basic science of hemophilia A and B (factor IX and VIII deficiency, respectively) with information on the different factor products available for treatment. Development of auto-antibodies or inhibitors, a major complication to treatment, is clearly described. Other complications are chronic joint disease due to recurrent joint bleeding and potential exposure to infectious agents in factor preparations. All these problems are discussed, as well as the care of the bleeding infant/child and obstetrical/gynecologic patients.

Although the bulk of the text is devoted to hemophilia A and B, any text worth its salt must include chapters on von Willebrand disease and the rare bleeding disorders (factors II, V, XI etc.)—and this one does.

The writing style is straightforward and the tables and illustrations (black and white) are good companions to the text. For those not familiar with hemophilia treatment options, a foldout with explicit treatment guidelines would have been a nice touch.

Overall, this is a concise (each chapter ranges from 4–6 pages) but thorough treatment of bleeding

disorders. Certainly a worthwhile monograph for hematologists, surgeons, gynecologists, and dentists.

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Clinical Obesity in Adults and Children.

Peter G. Kopelman, Ian D. Caterson, and William H. Dietz, editors. 2nd edition. Malden, MA: Blackwell Publishing; 2005. 493 pp. \$199.95. ISBN-10:1-4051-1672-2, ISBN-13: 978-1-4051-1672-5.

THE EPIDEMIC OF OBESITY in Western societies has stimulated considerable research on the effects of obesity on public health. One major example: It is estimated that over 800,000 residents of New York City have diabetes, with the vast majority having type 2 diabetes, associated with obesity. Multidisciplinary approaches are necessary to tackle this emerging epidemic, which is now afflicting tens of thousands of children in New York City alone.

This book is comprehensive and easy to read. There are three distinct sections: epidemiology, associated morbidities and management. All are well written and current. Of particular interest to the general practitioner are the chapters dealing with management. Often clinicians experience great frustration when trying to persuade their patients to modify their behavior. The section on the management of obesity provides realistic data that I believe can help us with this challenge.

The book also considers the social consequences of obesity and the differences between cultures in how they deal with obesity.

Proper emphasis is placed on the medical evaluation of the obese patient as well as the important co-morbidities of sleep apnea, hypertension and dyslipidemia. The message is clear—evaluate your patient for the consequences of obesity and treat these syndromes by treating the obesity. This book could serve as an excellent resource to educate patients about the potential risks of obesity. I would also recommend it highly to clinician nurse specialists.

Overall, this excellent review is evidence-based and user friendly.

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