BOOK REVIEW

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Craniosynostosis. Diagnosis, evaluation and management. 2nd edition. Editors M Michael Cohen Jr, Ruth E MacLean. Oxford: Oxford University Press, 2000.

In 1986, Dr Cohen published the first edition of this book on craniosynostosis. As correctly identified by Dr Gorlin in the foreword to that volume, it proved to be a "truly signal text", so much so that, though inevitably dated by the passage of time, the first edition is still cited regularly as an essential reference in relation to craniosynostosis. In addition to providing a comprehensive approach to all aspects of diagnosis and management of craniosynostosis, the first edition reviewed the basis for clinical delineation between syndromes of craniosynostosis, bringing a degree of order to a somewhat chaotic body of publications. However, the passage of time, with the emergence of new clinical reports, advances in the identification of molecular causes of craniosynostosis, insights into pathogenicity, not to mention new perspectives in management of patients with craniosynostosis, have necessitated a complete revision of that original text.

The revised edition has eight constituent parts, addressing the biological background, aetiology and pathogenesis, topical subjects, radiological and other imaging studies, neurological and ophthalmological assessment, speech and hearing, surgical management, and the syndromology of craniosynostosis. Of the 31 chapters, Michael Cohen is sole author of 17 and is joint author of a further two. The breadth of expertise encompassed within these chapters is formidable, ranging from historical reviews of the former classification systems proposed for craniosynostosis through gene and mutational data relating to phenotypes of craniosynostosis to consideration of the optimal name for phenotypes associated with *FGFR3* pro250arg mutation, given the notoriously variable clinical manifestations of that disorder.

Clinical photographs form an important element of the didactic aspects of the book. These are invariably excellent, accompanied by comprehensive legends and augmented appropriately by radiological and diagrammatic aids. The text embraces all aspects of current knowledge with respect to craniosynostosis and Dr Cohen attempts to incorporate molecular findings into a modern approach to the classification of the craniosynostoses. This works well for the author; it facilitates the elimination of one or two minor errors from previously published views and, more importantly, allows that some cases may be unclassifiable on clinical features alone. This book, however, is much more than a dry recounting of literature. Dr Cohen's personality and trenchant views are reflected throughout, with the result that a somewhat strident tone can, just occasionally, prevail. Few readers will be surprised to hear his thoughts on the condition referred to in publications as "Crouzon syndrome with acanthosis nigricans" which, in his view, shows "that we are prisoners of our own conventional terminology." He coins the neologism "Crouzondermoskeletal syndrome" to reflect the skeletal and dermatological findings of this entity. In similar vein, his recounting of various key contributions to the craniosynostosis story represents an interesting, but not universally accepted, view.

The publication of this book is of monumental importance to all syndromologists, plastic surgeons, neurosurgeons, maxillofacial specialists, and everybody involved in the diagnosis and care of patients suffering from craniosynostosis. Dr Cohen reminds us in the foreword that his interest in this area of medicine stems from an early study of Apert syndrome in 1973. The current volume confirms that he has used the intervening 27 years wisely and well. In an age when many are predicting the end of the published textbook as we know it, I cannot believe that this important and authoritative book should not be on the shelf of every single department of clinical genetics, idiosyncrasies notwithstanding.

WILLIE REARDON