

Dyskeratosis Congenita

A 5 year old boy presented with history of progressively increasingly pallor and abnormality of nails for one year. On examination, the child had severe pallor with petechiae and purpura. There was dystrophy of nails in all the fingers and toes (**Fig.1**), lacey reticulated pigmentation over chest (**Fig.2**) and oral leukoplakia. There was no hepatosplenomegaly. Hemoglobin (Hb) was 2.0 g/dL, total leucocyte count 2450/cmm with a differential count of N19L80M1 and platelet count of 16,000/ μ L. General blood picture, bone marrow aspiration and biopsy were suggestive of hypoplastic anemia. In view of the typical physical and hematologic findings a diagnosis of Dyskeratosis congenita was made. The child was started on androgens.

Dyskeratosis congenita (DC) (also known as Zinsser – cole – Engmak syndrome) is an inherited bone marrow failure syndrome which presents with bone marrow failure and diagnostic triad of lacey reticulated pigmentation, dysplastic nails and oral leukoplakia. The diagnosis is often made in adulthood and most of the physical findings appear with increasing age. Treatment is initiated when Hb is <8g/dL, platelets <30,000/ μ l and absolute neutrophil count <500/ μ L. Androgen therapy may improve but not cure aplastic anemia. Stem cell transplantation is recommended if there is an HLA-matched sibling donor. There is an increased incidence of carcinomas particularly of the head and neck.



FIG. 1 *Dysplasia of nails.*



FIG. 2 *Lacey reticulated pigmentation.*

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BOOK REVIEWS



Manual of Pediatric Therapeutics
GRAEF JW, WOLFSORF JI,
GREENES DS
*7th Edition. New Delhi:
Lippincott Williams & Wilkins:
2008. Pages: 716.
Price: Rs. 495/-.*

This is a very useful book for all categories of pediatricians, be it a naïve medical student stepping into a realm of pediatrics, practicing pediatrician or a pediatric consultant dealing with complex child health issues. It can rightly be termed a ready reckoner for management of not so familiar diseases. Lucid and crisp presentation, the updates and the precise format are its endearing features. 14 years wait for the new

edition, though long has resulted in a prized book. A highlight is the emergency medication and their dosage details, provided in the cover pages. A neat and complete drug formulary of commonly used drugs is an added benefit. Brief and well written covering neonatology makes the book complete. Little more care in the quality of paper and the lay out of the pages particularly margins, could have meant more comfort for the readers. Beauty of the book is that most of the topics are written in 2-3 pages and can be

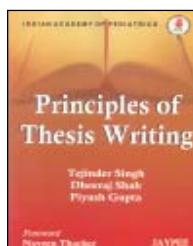
read and clearly understood within 10 minutes. Format of the book is very much readable, because the material is in salient words and not in full sentences. This is a manual worthy of possession by every pediatrician.

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Principles of Thesis Writing

TEJINDER SINGH, DHEERAJ SHAH
AND PIYUSH GUPTA

*New Delhi: Jaypee Brothers;
2008.*

Pages: 92.

Price: Rs. 150/-.

Principles of thesis writing is an excellent guide book written by three eminent teachers. Thesis writing is a must for all postgraduate students in India and neighboring countries. It is a beginning for the students to learn about research methodology. This book highlights the importance of thesis as a tool for beginners in the field of research in medicine. The book covers all steps for writing thesis in a very simplified manner starting from thinking process of hypothesis, writing protocol, reviewing literature, internet browsing, study design, analysis and compiling the data, and finally the process of writing. The book emphasizes the key components of a research and also highlights various types of studies that can be undertaken by the students in a resource poor settings. All the chapters are written in very simple language which is easy for average students to

understand. The methods of sample size collection, statistical analysis of different types and the ten steps of good presentations are very useful for students and also for teachers. Every chapter has been written well with effective illustrations and examples. The final chapter on submitting the thesis has summarized all important checklists for the students at the end of their hard work to compile and make the final presentation in a scientific manner. The chapter on basics of medical statistics as applicable for thesis writing has been elaborated in an excellent manner. However, if authors would have provided the simplified table as ready reckoner for calculation of sample size as per WHO recommendations, it would have been very useful for the learners. The book will be a useful companion for all postgraduate students. I am sure this book will find a place in the libraries of all medical institutions in the country as ready reference for guiding students in the art of thesis writing.

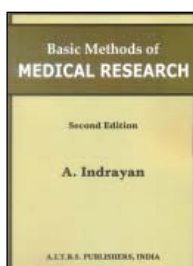
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Basic Methods of Medical Research, 2nd edition

ABHAYA INDRAYAN

New Delhi: AITBS Publishers;

Pages: 400.

Price: Rs. 299/-.

Epidemiology, biostatistics and research

methodology are generally considered to be mundane subjects by medical students and mainstream pediatricians. However, one has to be aware of their basic aspects to write thesis and also to comprehend research for practical application. Prof. Indrayan, an accomplished biostatistician with an exceptional gift of teaching, integrates the basics of all three subjects in an easy to read text, which is devoid of polemics and equations. This second

BOOK REVIEWS

edition represents an improved and slightly expanded version while retaining its charm of simplicity and readability.

The monograph provides step-by-step approach to all aspects of empirical research, beginning with selecting and writing a protocol, to framing an appropriate design of study, method of collection of valid data, their collation, analysis and interpretation, and preparing an evidence-based report. Due emphasis is given to practical aspects and the text has a liberal sprinkling of real-life examples for easy comprehension.

Each chapter begins with a list of key terms and concepts and ends with a summary. The shaded boxes

in the text summarize important information. A comprehensive glossary of terms at the end of the book represents a unique value addition.

The book is recommended for pediatricians interested in research methodology, and post-graduate students. Established researchers would also find it to be a handy adjunct.

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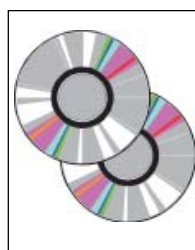
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CD-Rom on HIV – Nutrition: A Teaching Learning Module

TEJINDER SINGH AND MONIKA SHARMA

Ludhiana 2008.

Media for imparting education has changed over a period of time. With more people having access to computers and with its inherent attractiveness, they are becoming the logical choice as educational media. This CD on Nutrition and HIV is a good attempt to impart the knowledge on the concerned subject as a self learning module.

The module has been developed as part of a project 'Information technology (IT) with human face- a collaborative research project to improve nutritional training' and also gives useful websites which can be accessed on the internet. The material is locally relevant, interactive and suitable for self study. It takes about 1 hour of self study to complete all the modules. There are case studies and quizzes at the end of each module to facilitate learning.

This CD has 7 modules: introduction to HIV/AIDS; counseling of the patients; nutrition and infection complex; nutrition and HIV/AIDS; nutritional status assessment of people living with HIV/AIDS using ABCD (anthropometry, biochemical, clinical and dietary) parameters; nutritional care and management and caring for HIV/AIDS in pregnancy, infants, adults, elderly and contacts. The users do not need to install anything new on their computer. All the necessary files are included with the module. For ease of navigation, a user manual on VTS is also included.

The module provides relevant information in a simple language. It could be made more user friendly with less of text and more of flow charts and pictures. On the whole the module gives detailed information on all aspects of HIV with particular emphasis on nutritional requirements and will be useful for undergraduate, postgraduate students of nutrition and dieticians involved in the care of HIV/AIDS patients.

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