component of the storage material (R D Jolly and coworkers) is an exciting new advance. At least two
groups of researchers are applying linkage analysis
using DNA markers in an effort to map the disease
gene or genes.

This volume is a must for anyone involved with
the ceroid-lipofuscinoses at the bedside or in the
laboratory. For any paediatric neurologist, genetic-
ist, biochemist, or pathologist with only the haziest
notions concerning this group of diseases, persual
will make you the local expert and provide the best
introduction to one of the most intriguing enigmas in
medicine.

R M Gardiner

Recent Advances in Ectodermal Dysplasias
Edited by C F Salinas, J M Opitz, and N W Paul.

This book consists of the papers presented at a
symposium sponsored by the Society of Craniofacial
Genetics in 1985. As such, the topics chosen
represent the personal interests of the authors; the
quality and quantity of content and presentation is
variable.

In the first section (Nosology and basic science), I
found two papers of interest. Holbrook’s contribution
on the histological and ultrastructural study of the
early development of skin and its appendages was
valuable; Jorgenson and others discussed techniques
for studying palmar sweat pore density.

In the next section (Clinical manifestations), there
were 10 papers. Opitz discussed several congenital
anomalies, in which a dysplasia manifesting in
embryonic or fetal life may have caused ectodermal
and other anomalies to develop as a sequence rather
than a series of primary malformations. Freire-Maia
and Pinheiro describe in outline the clinical appear-
ance of 15 ectodermal dysplasias (EDs) described
since their 1984 book went to press. Gorlin reviews
some interesting EDs with hearing loss. Levin
provides a helpful description of the oral abnor-
malities in a number of EDs.

In addition to these reviews, there were also
contributions on the clinical features of single ED
syndromes. One very useful paper described two
extensive Rapp-Hodgkin families, and extended the
reported spectrum of manifestation of this disorder.
Other papers described a restrictive dermatopathy,
one form of autosomal recessive hypohidrotic ED
(HED), and a trichothiodystrophy in which the
hair shaft is dystrophic and there may be an
underlying metabolic disorder amenable to chemical
analysis. The paper on cephalometry in X linked
HED reported only very preliminary results; it
seems that this technique is unlikely to help diagnose
carrier status. The paper on speech in HED makes
the non-contentious assertion that dentures help
children with hypodontia to articulate with clarity.

The third section (Management) is polysyllabic,
but low on content. There are hortatory comments
on team care and regionalisation, on the advantages
of early diagnosis, and on the benefits of visiting a
dentist if you have few teeth. One useful tip is
included: that nasal crusting can be helped by
sniffing water or saline into or through the naso-
pharynx. Tanner discusses the emotional adapta-
tion of those with ED syndromes to their disorder and
the effect on the family as a whole. This is a difficult
topic, with no easy answers.

This book will be of interest to two groups. The
reviews and the scientific papers will interest those
with a very special interest in the ectodermal
dysplasias. Gorlin’s review and the papers describing
Rapp-Hodgkin disease and trichothiodystrophy will
also interest some dysmorphologists. Only very few
will want a personal copy, but access to a library
copy of this volume might be helpful for dysmorpho-
logists and for paediatric dentists interested in
 genetics.

A Clarke