form, is discussed under the heading of primary
diseases of cementum. These are, however, small
criticisms of a generally excellent text which should be
of considerable value to clinicians in many branches of
medicine and dentistry.

G. B. WINTER

The Eye in Chromosome Duplications and
Deficiencies
By Marcella Jay. (Pp. xi + 249; Figures + Tables.

Ophthalmologists have long been interested in
genetics. One of the first textbooks on genetics was
written in 1934 by an ophthalmologist, Waardenburg,
and the tradition has been carried on by Franceschetti
and, in this country, by Sorsby. Some ophthalmic
hospitals have a genetic clinic (the author helps her
husband at the one at Moorfields, London), so that
this book, though not written by a clinician, has its
origins in the clinic.

Since the advent of banding techniques, many new
chromosomal syndromes have been described. Clinici-
ans are aware that eye anomalies appear in many
different syndromes, so that the presence of hyper-
telorism, ptosis, strabismus, or epicanthus are not
helpful diagnostic criteria. If less nonspecific features,
such as optic atrophy or colobomata, were to be found
in certain of the aneuploids, these would be more
useful clinical signs. The main purpose of this book is
to describe and tabulate the clinical findings, with
particular emphasis on the ocular anomalies in the
partial deletions and trisomies. As the book has its
origin in a thesis, the detail is excellent and there is a
comprehensive coverage of references since the intro-
duction of banding in 1972. In addition, there is a
concise first chapter on the chromosome and a useful
glossary of terms. The interesting association of retino-
blastoma with partial deletion of chromosome 13 is
also dealt with. Perhaps potential readers should be
warned that duplications and deletions referred to in
the title do not include whole chromosomes, so that
trisomy 13, trisomy 18, and even Down's syndrome
are not described. Surprisingly, an exception is made
for Turner's syndrome. Nevertheless, this is a most
useful reference work for those dealing with the 'funny
looking kid' syndromes.

M. BARAITSER

Myotonia Congenita and Syndromes Associated with
Myotonia. Clinical-Gene Genetic Studies of the Nondystro-
phic Myotonias
By Peter Emil Becker, with contributions by Rainer
Knussmann and Erick Kuhn. Translated by Mary F.
Passarge. (Pp. x + 181; 146 Figures + 40 Tables.

Myotonia refers to the phenomenon of delayed relaxa-
tion after either voluntary contraction or contraction
induced by percussion. The first clear and thorough
description of a dominantly inherited form of myo-
tonia was by Dr Asmus Julius Thomsen 100 years
ago, who observed the disease in himself as well as in
other members of his family. Since then it has become
increasingly clear that the inherited myotonic syn-
dromes are a very heterogeneous group of disorders.
In fact some 15 clinically distinct forms have now been
described. Professor Becker has been particularly
interested in these disorders, and this detailed mono-
graph concerns his studies of 142 probands with
established myotonia congenita and their relatives,
including Thomsen's original family. His major
contribution has been to show the clear distinction
between dominant and recessive forms of myotonia
congenita. The former tend to have an earlier onset,
to be more variable, less severe, and less often associ-
ated with persistent weakness than the latter. There
appear to be at least 3 clinically distinct forms of
dominant myotonia congenita: Thomsen's disease
itself, a form associated with myalgia, and another
with marked cold dependence.

Becker observed the prevalence of dominant
myotonia congenita in West Germany to be about 1 in
500 000, though this is clearly an underestimate for
often symptoms are mild and ascertaining therefore
not complete. In fact, he thinks the true prevalence
may be as high as 1 in 23 000, about twice the pre-
valece of recessive myotonia congenita. Interestingly,
malignant hyperthermia was not observed in any of
the families he studied.

Besides presenting detailed clinical findings, the
monograph concludes with 2 brief sections on bio-
chemistry (by Kuhn) and multivariate analysis (by
Knussmann). The results of the biochemical studies
(serum and muscle enzymes, serum electrolytes, serum
and muscle lipids) are unrevealing, apart from
abnormalities in the lipid composition of muscle in
dominant myotonia congenita, the relevance of which
is not yet clear. With regard to the statistical analyses,
an attempt was made to verify Becker's classification
firstly by testing the significance between the
myotonic types he described, and secondly by
attempting to establish an independent classification
from the original data. In general, both approaches
tended to confirm Becker's intuitive classification.

Altogether this is a very valuable monograph and
can be unreservedly recommended to anyone with an
interest in this complex group of disorders. It also pro-
vides an excellent model for anyone embarking on a
clinical-genetic study. It is gratifying to those with
primarily clinical interests that this approach is still
very worth while and can produce valuable results as
well as being intellectually satisfying.

ALAN E. H. EMERY