"Paratrigeminal" paralysis of the oculopupillary sympathetic system

P J Goadsby

Raeder described five patients with mixed features of trigeminal nerve pathology and oculosympathetic impairment, with or without other cranial nerve lesions. This constellation of clinical features drew the original author’s attention to the paratrigeminal region as a likely site for the causative lesion in this syndrome. An analysis of the anatomy of the oculosympathetic innervation supports the view that a restricted lesion in the middle cranial fossa might cause the syndrome of trigeminal nerve involvement, neuralgic pain or sensory change, with ptosis or miosis, or both, but no anhidrosis. Such a paratrigeminal oculosympathetic syndrome (POSS) usefully reminds clinicians to pursue vigorously possible lesions of the middle cranial fossa with careful, and possibly repeated, imaging studies. Attaching the eponym Raeder’s syndrome or Raeder’s paratrigeminal neuralgia to this syndrome adds nothing valuable to the anatomical description (POSS), which might be preferred for clarity.

In 1924 Raeder wrote a classic clinical-anatomical colocalisation paper that described five patients, one of whom had been reported in 1918. The patients had two key features: involvement of the trigeminal nerve and the oculosympathetic nerves. He sought to differentiate the restricted oculosympathetic findings from the classic Horner’s syndrome: cervical sympathetic dysfunction characterised by ptosis, miosis, anhidrosis and enophthalmos. Since that time various terms have been employed, meanings defined, and classifications developed. It is against this background that one ventures into the Coliseum that is the history of medicine to play out the academic battle. An eponym is useful if it conveys a very clear meaning, or perhaps acknowledges some contribution that was so pivotal as to provide insight well beyond the describer’s demise. Raeder made an interesting clinical observation that pointed out the likely localisation of a lesion adjacent to the trigeminal nerve in the middle cranial fossa. In the era before cranial imaging this was a wonderful piece of neurology; in the modern era the eponym is much less useful. Solomon and Lustig recently set out the clinical cases that in many respects have illustrated the trigeminosympathetic anatomy of the cranial nerves. Conditions such as carotid disease, particularly dissection, may give rise to pain and a Horner’s syndrome, and cluster headache may lead to oculosympathetic loss and impaired sympathetic facial sweating. In both situations forehead sweating may be impaired. I will set out the relevant anatomy of the paratrigeminal region, then review Raeder’s patients, and finally make some suggestions for the postimaging era based on an anatomical/pathophysiological designation for such patients.

ANATOMY

The key anatomical feature to be understood for Raeder’s syndrome is the relation between the trigeminal nerve and the oculopupillary sympathetic fibres. The trigeminal nerve lies in the middle cranial fossa and in close proximity, paratrigeminally as Monrad-Krohn suggested, there are other cranial nerves. Most particularly for a short course the fibres that will innervate the levator palpebrae superioris, specifically Müller’s muscle, and the pupillodilator fibres without the sudomotor fibres for the forehead. Therein is the anatomical lesson of Raeder.

Cranial sympathetic innervation

The sympathetic outflow ultimately arises in the hypothalamus and can be modulated by brain stem neurons. Preganglionic fibres arise from neurons in the lateral column of grey matter of the thoracic and upper two to three lumbar segments. The fibres emerge through the ventral roots of the corresponding spinal roots, passing into the spinal nerve trunks and their ventral rami. The fibres leave the ventral rami in the white rami communicantes to synapse in the corresponding ganglion or, in the circumstance of interest for the cervical sympathetic ganglia, to ascend before synapsing. The sympathetic ganglia give rise to non-myelinated postganglionic fibres, which, for the head, arise as the internal carotid nerve from the superior cervical ganglion, which is, in effect, the rostralmost section of the sympathetic trunk. The cervical sympathetic ganglia destined to innervate the eye derive their input from the upper thoracic (T1) sympathetic white rami communicantes.

Oculosympathetic innervation

The internal carotid nerve ascends with the internal carotid artery dividing into medial and lateral branches in the bony carotid canal, and forming a plexus known as the carotid plexus. The medial plexus communicates with the trigeminal ganglion and abducens nerve, and the lateral plexus...
parasympathetic neurons may reinnervate the sweat glands, oculosympathetic fibres. It must be borne in mind that carotid plexus relatively laterally when compared with the is, therefore, normally mediated by fibres that exit the internal carotid plexus, whereas those for the remainder of the face traverse the external carotid plexus, excepting those for the cheek, which may take either route\(^8\). Sudomotor fibres destined for the forehead join the ophthalmic nerve in the region of the cavernous sinus reaching the skin through the supraorbital nerve\(^11\). Sweating of the forehead is, therefore, normally mediated by fibres that exit the internal carotid plexus relatively laterally when compared with the oculosympathetic fibres. It must be borne in mind that parasympathetic neurons may reinnervate the sweat glands, so that intact sweating may not necessarily imply sympathetic sudomotor integrity.

**Trigeminal nerve and oculosympathetic outflow**

As the internal carotid artery passes from being inferior to the trigeminal ganglion and oculomotor nerve, and from there it lies medial to the anterior clinoid process, it must distribute from the lateral internal carotid plexus the fibres for the dilator pupillae and levator palpebrae superioris; the sympathetic fibres destined for the trigeminal ganglion being delivered from the medial plexus before that point, as described above. Thus a lesion restricted to the middle cranial fossa medial to the trigeminal ganglion and effectively lateral to the anterior clinoid, would produce trigeminal pain by direct irritation of the anteromedial ganglion or initial section of the ophthalmic division, and could interrupt oculosympathetic outflow. There would be no expected effect on facial sweating because the sudomotor fibres have exited before this point. Thus a paratrigeminal oculosympathetic syndrome arises.

**RAEDER’S PARATRIGEMINAL SYNDROME**

Set against the anatomy it is appropriate to consider Raeder’s patients. Table 1 summarises the patients by dividing up the clinical symptoms and signs to highlight the trigeminal and oculosympathetic involvement, as well as other relevant clinical features. Several features emerge from such an analysis.

**Trigeminal involvement**

Considering three possible types of trigeminal involvement—pain, sensory change and motor signs—what is constant in Raeder’s patients? Only three patients had pain and only two had neuralgia. Thus the term Raeder’s paratrigeminal neuralgia is fanciful. Three patients had sensory change; one patient had pantrigeminal loss indicating that the lesion had moved lateral to the unique paratrigeminal region, and similarly patient 1 had motor loss indicating a more extensive lesion.

**Oculosympathetic loss**

All five of Raeder’s patients had miosis, and four of five had ptosis. Four of five also had normal sudomotor function. Thus oculosympathetic loss which must be peripheral was demonstrated but without anhidrosis.

**Other signs**

Four out of five of the original patients had other cranial nerves involved. It is interesting that one had cranial parasympathetic activation, most likely simply as a result of pain and activation of the trigeminal-autonomic reflex\(^12\).

**Status of Raeder’s patients**

An analysis of Raeder’s patients demonstrates several issues. It is notable that only two of the patients had neuralgia and two had lesions not restricted to the paratrigeminal region. Thus perhaps one patient at most, patient 4, really illustrated in a pure fashion the anatomical construct that Raeder seemed to illustrate. An analysis of the patients and the relevant anatomy suggests that Raeder’s paratrigeminal neuralgia might usefully be dropped from the textbooks with no substantive loss as the patients described did not really, perhaps save one, fulfil the anatomical requirements strictly.

### ANATOMICAL APPROACH: PARATRIGEMINAL OCULOSYMPATHETIC SYNDROME

How might one usefully draw on the lesson that Raeder sought to teach, essentially one of functional anatomy, and make a useful clinical rule? It could be suggested that Raeder made a first, and very creditable, attempt to describe a clinically useful syndrome based on the relevant anatomy (fig 1). Patients with oculosympathetic loss, miosis or ptosis, or both, with normal forehead sweating, and evidence of trigeminal involvement, either sensory change or neuralgia pain are highly likely to have a lesion in the middle cranial fossa that is medial to the trigeminal ganglion (paratrigeminal). The anatomy of this syndrome is clear with dissociation.

---

**Table 1 Raeder’s cases**

<table>
<thead>
<tr>
<th>Patient (age, sex: pathology)</th>
<th>Trigeminal sensory</th>
<th>Oculosympathetic</th>
<th>Other cranial nerves</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Pain†</td>
<td>Sensory change</td>
<td>Ptosis</td>
</tr>
<tr>
<td>18, male: parasellar endothelioma</td>
<td>+</td>
<td>–</td>
<td>+</td>
</tr>
<tr>
<td>65, male: not described</td>
<td>+[*]</td>
<td>+ (V1 loss)</td>
<td>+</td>
</tr>
<tr>
<td>48, male: run over by a car</td>
<td>–</td>
<td>+ (hypoaesthesia)</td>
<td>+</td>
</tr>
<tr>
<td>28, male: not described</td>
<td>+[*]</td>
<td>–</td>
<td>+</td>
</tr>
<tr>
<td>48, male: injured by runaway horse</td>
<td>–</td>
<td>+ (V1–V3 loss)</td>
<td>–</td>
</tr>
</tbody>
</table>

\(^†\)Although often described as a neuralgia only the patients marked * had clear neuralgia. 
\(^*\) Indicates data not described in case.
of the oculosympathetic outflow from forehead sweating in the middle cranial fossa; the only caveat being that if there is parasympathetic innervation of sympathetically denervated sweat glands, as seen in cluster headache,\(^8\) then the sweating in Raeder’s patients would be pathological and much of the argument would be lost. Given that Raeder was unaware of this and, therefore, could not have tested for it, the patients do not provide absolute evidence for his argument. Perhaps the name of the syndrome should simply reflect the anatomy—paratrigeminal oculosympathetic syndrome—and thus fulfil the useful role of teaching. One might suggest that the term Raeder’s paratrigeminal neuralgia is inaccurate, and Raeder’s (paratrigeminal) syndrome does not teach the essential lesson in its name. Certainly, the unique combination of symptoms and signs should trigger an exhaustive search for a lesion with MRI. When no lesion is found the patient must be followed up, and imaging should be repeated at least once. Neurology evolves with the lessons of history to better modern practice; the lesson of Raeder is learnt and requires more transparent dissemination, including dropping the eponym. One might simply argue that to learn one thing—that is, the paratrigeminal oculosympathetic syndrome—is to learn the entire lesson, to learn a person’s name adds nothing more in clinical terms. The use of such eponyms remains, however, a matter of taste and practice; and for Raeder should be tightly coupled to the anatomy.

ACKNOWLEDGEMENTS

I thank Dr Peter Drummond for helpful comments on the manuscript. PJG is a Wellcome Trust senior research fellow.

REFERENCES

1 Raeder JG. “Paratrigeminal” paralysis of the oculo-pupillary sympathetic. Brain 1924; 47:149–58.
"Paratrigeminal" paralysis of the oculopupillary sympathetic system

P J Goadsby

J Neurol Neurosurg Psychiatry 2002 72: 297-299
doi: 10.1136/jnnp.72.3.297

Updated information and services can be found at:
http://jnnp.bmj.com/content/72/3/297

These include:

Supplementary material can be found at:
http://jnnp.bmj.com/content/suppl/2002/03/04/72.3.297.DC1

This article cites 9 articles, 0 of which you can access for free at:
http://jnnp.bmj.com/content/72/3/297#BIBL

Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

An erratum has been published regarding this article. Please see next page or:
/content/72/5/684.full.pdf

Articles on similar topics can be found in the following collections

Cranial nerves (529)
Drugs: CNS (not psychiatric) (1945)
Neuromuscular disease (1311)
Ophthalmology (842)
Pain (neurology) (763)
Peripheral nerve disease (631)

Notes

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/
Conflict of intentions or inner negativism?

In a recent, fascinating article, Nishikawa et al describe their encounter with “three patients with callosal lesions who sometimes could not perform whole body actions as they intended because another intention emerged in competition with the original one.” Believing that “no specific term has yet been coined for this symptom” they “tentatively” named it “conflict of intentions.”

In fact, however, this symptom was described by Bleuler in his Textbook of psychiatry, which first appeared in English translation in 1924. Bleuler termed it “inner negativism,” and noted that when “patients make an effort to start an action . . . a counter-impulse, or only a mere blocking appears and hinders them in its execution.” Such inner negativism could prevent “the simplest acts like eating. The spoon is arrested half way up to the mouth and must finally be put down again.”

The great service of Nishikawa et al is to demonstrate the localising value of this symptom to the corpus callosum; it would be a disservice to medical history, however, to rename it.

J D O’Sullivan
Department of Psychiatry, University of Louisville School of Medicine, c/o Central Hospital, La Grange Road, Louisville, KY 40223, USA; dpmooremd@cs.com

References
demolished people. Practical psychiatry of old age, now in its 3rd edition, brings together the many fields of our discipline. It is liberally scattered with useful and interesting case histories and the advice on management is sensible and up to date.

The book is clearly written for a trainee and non-specialist audience and deals with most subjects with a fairly light touch. The references at the end of the chapters serve as useful reading lists, including as they do both recent and historical papers. For students and for trainees this book will provide a useful revision and summary aid although trainees will need also to have hand some of the works referred to in other disciplines may well find the book helpful to understand some of the classification and nomenclature issues of old age psychiatry.

Like the discipline itself, however, this book is very much a British affair. The sections on services have only limited international relevance and even the concept of a doctor who manages late onset psychosis, personality disorder, and dementia is not so common elsewhere. The concentration on the international classification of diseases has limited application in the United States. So for those in the UK who need an introductory text on medical disciplines, this book is a sensible choice. To those who have yet to appreciate the joys of being an old age psychiatrist, dip into a colleague’s copy—you may be pleasantly surprised.

Simon Lovestone

Brain Imaging in Schizophrenia, Insights and Applications

This comprehensive overview of brain imaging studies in schizophrenia is well illustrated with scan photographs. The first two chapters cover the techniques of brain imaging and include several tables summarising information. The structural imaging chapter describes the techniques of computed tomography and MRI, and introduces the novel methods of diffusion weighted imaging and magnetisation transfer imaging. Complex topics such as the underlying principals of MRI are tackled in a fairly accessible manner. The functional brain imaging chapter covers PET, SPECT, fMRI, and MRS. The next two chapters cover the results of structural and functional imaging studies. These chapters are thoughtfully subdivided, and papers up to and including the year 2000 are cited. The brevity of the volume of course restricts the range of studies discussed, but generally the selection is good.

Space also prevents areas of conflict from being fully resolved, for example into differing scan methodologies, data analysis protocols, and clinical populations. The penultimate chapter is titled “Brain and brain imaging” and describes imaging studies in twin pairs and members of multiply affected families. It includes discussion of the subtle abnormalities identified in presumed carriers. Finally there is a brief concluding chapter examining the current and future applications of the various imaging techniques in the study of schizophrenia. Overall, the results presented confirm that the complexity and heterogeneity of schizophrenia makes a simple uniform underlying pathology seem unlikely. Imaging studies, with their unique ability to examine the brains of living patients, have an important role in developing our increasingly sophisticated understanding of the disorder. I would recommend this well written monograph both to academics and to clinicians hoping to keep up with this fascinating and fast evolving area.

R Alexander Banick

Parkinson’s Disease in the Older Patient

This is a welcome addition to the literature. The book has been published with the help and support of the British Geriatrics Society special interest group on Parkinson’s disease and the Parkinson’s Disease Society of the United Kingdom. Both organisations have been at the forefront of increasing public and professional awareness of the need for a holistic approach to the care of older people with this condition. Understanding of the pathophysiology, therapeutics, and progression of the disease, as well as the management of late onset psychosis, personality disorders, and dementia, is very much a British affair. The sections on the classification and nomenclature issues of old age psychiatry.

are relatively rare, their management is generally not well discussed, but generally the selection is good.

With a book of this size, there is likely to be some variation in quality and the chapter on vasculitis and collagen vascular disorders is weaker than the rest. Though these conditions are relatively rare, their management is important, as it frequently vexes neurologists. It is simply inadequate to dismiss their classification as unsatisfactory and end the brief discussion of this topic with the implication that they can all be lumped together anyway, as the treatment is usually immunosuppression. In the same section, lupus and the anti-phospholipid syndrome are given as examples of the difficulty of accurate subclassification. But there is one situation where the authors have highlighted the different in treatment—that is, immunosuppression versus antithrombotic therapy and/or anticoagulation. Later in the same chapter, eosinophilia is given as a feature of Wegener’s granulomatosis yet is missing from the text. This is a pity that the one disease for which the British can claim special expertise—the human
form of bovine spongiform encephalopathy—
given under two names, “variant Creutzfeldt-
Jakob disease” in the section on dementia and
“new variant Creutzfeldt-Jakob disease” in that
on infection. Both are typographical errors,
which are too many for comfort, especially in
the tables, figures, and references, giving the
impression that the book was rushed in its final
production stages. Perhaps the most alarming
was the discovery of a new cranial nerve, the 13th, in table 1.4. Figure 8.5 shows
a retinal hamartoma, not haematoma. Figure 8.4
shows the optic fundus at an unusual angle. Figure 11.9 is anatomically incorrect. Figures 12.9, 13.10, and 29.11 are too small. The
caption to figure 2.23 is incomprehensible.
Many other examples could be given.
But these are mainly minor quibbles, easily
termed when the book is reprinted. Taken as a
whole, Big Brain is alive and well, and safe in
the hands of its new editor and his coauthors.
Lionel Ginsberg

Textbook of Clinical Neuropsychiatry
Edited by D P Moore (Pp 747, £69.50).

There is a certain logic to the system Moore
uses in his textbook of clinical neuropsychia-
try. The first half of the book essentially covers
all the various lists of causes of different
symptoms, signs, and syndromes. For example
lists are provided for causes of dementia
lacking distinctive features, demen-
tia associated with strokes, and dementia
with Parkinson’s syndrome or with parki-
sonism. Confronted with a patient with
dementia plus parkinsonism the reader has
quick access to conditions that need to be
covered. Or if the reader is looking for a list
of causes of catatonia he need look no further
than table 3.8. Having identified the potential
causes of the patient’s symptoms the reader
then goes to the second half of the book where
he will find up to date descriptions of the rel-
vant neuropsychiatric diagnoses.
The problem with such an approach is that it
leads to duplication. In the first half any
diagnosis has to appear as many times as
it is a symptom, syn, or syndrome that it can produce. The approach depends
heavily on the validity of the classification
of symptoms and syndromes; conditions with
different names often seem to share more in
common than they set them apart. For example it
is asserted that stupor can be distinguished
from akinetic mutism partly on the basis of
eye movements: in the former they are gener-
arly roving or disconjugate, whereas in
akinetic mutism they are conjugate tracking eye
movements are to be seen. I am not so sure and
would have preferred a critical discussion of
the nosological status of akinetic mutism
alone, non-stuporous, and catatonic frontal
lobe syndrome is given syndrome status,
but I could find no mention in the book on the
dysexecutive syndrome.
What is lacking in this book is a sense of
proportion. Three of the biggest suppliers of
referrals to a neuropsychiatric service—stroke,
head injury, and conversion disorders—hardly
get a mention. Given the huge range of condi-
tions that are covered it is not surprising that
the book sometimes gives a sense of having
been written in the library rather than from
clinical experience. But there are some excel-
ient sections; I was particularly impressed by
the chapters on epilepsy and the introduction to EEG.

It is a very comprehensive textbook. This is
its strength. The complete range of neuropsy-
chiatric conditions is described in a consist-
ent, easy to read, format. Large numbers of up
to date references are provided.

Overall Dr Moors is to be congratulated on
producing a useful textbook. Two neuro-
psychiatric colleagues gave this book the
thumbs up because Moore has achieved his aim of offering a ready reference for
established practitioners. It will be of inter-
est to both neurologists and psychiatrists.

Simon Fleminger

Wolf’s Headache and Other Head Pain, 7th edn.
Edited by S D Silberstein, R B Lipton, D J
dalesio (Pp 625, US$99.00). Oxford Univer-

There can be few people still alive who came
under the direct influence of Harold G Wolff
before his death in 1977 (Dona Dalesio
being one), but his influence on the whole of
neurology has been immense and still contin-
ues. His book soon became the classic—the two
editions that were written by him were now acquired
only with difficulty from antiquarian book-
sellers. Over the years it has become slowly
transformed, though perhaps some interme-
tiate editions were a less satisfactory hybrid
between the master and later developments,
“Wolf’s Headache” has now emerged as a
fully fledged multimedia text in its own right,
with less emphasis on the master’s own experimen-
tial work. We now have a 600 page authoritative
book, written largely by Ameri-
can authors, all clearly experienced clinicians.
It is comprehensive, but is perhaps
more manageable than its main competitors.
In the first 100 pages the classification,
anatomy, pathophysiology, genetics, and epi-
demiology of headache are covered, with dis-
cussion of imaging techniques and comorbid-
ity with other diseases. The core of the book
covers migraine, cluster headaches, and ten-
sion headaches, including a very comprehen-
sive review of every drug that has ever been
used to treat headache, including the obscure,
the ineffective, and the promising. This
section is also strong on the classification of
chronic headache syndromes and in discuss-
ing analgesic abuse. The third section
discusses every conceivable structural cause
of headache, including low and high CSF pres-
sure, metabolic disease, and disorders of the
neck, eyes, teeth, nose, and blood vessels,
including all the classic citations. The final
three chapters discuss headache in children,
behavioural management, and the consulta-
tion process itself.

This is an outstanding book; little of sig-
ificance is omitted, and yet one is not over-
whelmed with details. No doubt with the
trainee entering the field in mind, it is
particularly good when reviewing the litera-
ture, though some authors do occasionally
lapse into unidiomatic use of older
papers. It will prove to be a useful reference
text for more senior neurologists confronted
with a difficult patient, both for diagnostic
and therapeutic options, though these are
perhaps more from an American viewpoint.

Richard Peatfield

Multiple sclerosis: Tissue destruction and repair
Edited by L Kappos, K Johnson, J Kesselring,
and E W Radu (Pp 350, £65.00). Published by
872 7

The Martin Dunitz imprint produces high
quality books with catchy titles often built
around European congresses of neurology.
Brain disease: therapeutic strategies and repair
emerged from the European Neurology Soci-
ety meeting in Jerusalem (2000). Multiple sclero-
sis: tissue destruction and repair is the proceed-
ing of the joint meeting of ECTRIMS (European and American Commit-
tees for Treatment and Research in Multiple Sclerosis) held in Basel in 1999. Looked
at critically, neither book is much about repair.
Here, the 116 contributors were 34% editors
edited by a team from Switzerland and Balti-
more write on central nervous system-tissue-
immune interactions; in vivo assessment of tis-
ue destruction and its consequences; mul-
tiple sclerosis fatigue; new immunological
concepts and their therapeutic consequences;
treatment of relapse; modern concepts of
therapeutic immunomodulation; and an up-
date of therapeutic trials. Many of the usual
suspects are rounded up: magnetic resonance
surrogates for various histological compo-
nents of the disease process in multiple sclerosis; markers of demyelination in body
fluids; treatment effects of interferon beta and
its mechanisms of action; and strategies for
transplantation in multiple sclerosis. Some
authors take up old ideas: the use of steroids in acute episodes; and disease modi-
fying effects of non-specific immunosuppres-
sants. But there are also some new or emerg-
ing stories: inflammation and neuronal activity; interactions between interleu-
kin and growth promoting molecules; fMRI
 evidence for plasticity in multiple sclerosis; T helper and T regulatory activity; bone
marrow transplantation in multiple sclerosis;
prophylactic treatment of periperal disease
activity with intravenous immunoglobulin; and a brace of preliminary clinical trials
with hitherto unknown agents offering space to watch. Multiple sclerosis: tissue
destruction and repair succeeds as a statement from experts on where selected aspects of research
stood in 1999 and as testimony to the deserved and sustained success of ECTRIMS
(and ACTRIMS) but as a lasting statement on limiting and repairing the damage in multiple sclerosis, perhaps less so.
Katrina Dedman

Current management in child neuro-
ology, 2nd edn
Edited by Bernard L. Maria (Pp 562,
US$74.95). Published by BC Decker Inc,

Management includes assessment, diagnosis,
and treatment. What emerges therefore is a
book of clinical paediatric neurology—not a
book on treatment in paediatric neurology. It
is divided into outpatient and inpatient
sections and priority within these areas is
apportioned by incidence. The top four out-
patient neurological conditions presenting to
paediatricians in Florida are attention deficit
hyperactivity disorder (ADHD), seizures and
epilepsy, developmental delay, and headache.
The top four discharge diagnoses from hospi-
tal on the other hand are enotlevior meningi-
ts, epilepsy, hypokinetic syndrome (which
the author explains by the presence of comor-
bid conditions requiring hospital treatment),
and concussion.
The aim of this book is to provide “primary
care physicians, neurologists and house staff
with factual information on how to treat chil-
dren with the most common disorders of the
nervous system”. There are some surprising omissions in-
cluding spinal dysraphism. Movement disor-
ders generally get short shrift. Of the 550

www.jnnp.com
pages, cerebral palsy gets five (biomechanics gets five lines, prevention of secondary deformity is ignored), although there are a further eight on spasticity. There is nothing on chorea or dystonic syndromes—the latter omission is particularly surprising in view of the treatment implications.

In these days of economic scrutiny the evidence base for treatment recommendations should be referenced but is not for cerebral palsy, language disorders, or learning disability.

One hundred and nine authors contributed to this book. That so many have been induced to contribute may be because few provide more than seven pages. Thus, the most extensively treated topic is that of epilepsy with 86 pages from 13 separate authors. This leads to redundancy (treatment with antiepileptic drugs in most chapters but especially those on first choice antiepileptic drugs and recurrent seizures) and surprising omissions. A diagnostic approach to Lennox-Gastaut syndrome and progressive myoclonic epilepsies would have been useful. Nowhere are the implications of the genetics of familial epilepsies described. Genetic counselling generally is mentioned only in the chapters on neurofibromatosis and tuberous sclerosis. The concept of channelopathies is absent throughout.

The target audience for this book see a lot of headlines; hence, 46 pages and seven authors. Again, redundancy and gaps. No one mentions taking the blood pressure of a child with headache. Neuroimaging is thought unnecessary unless there are abnormal neurological signs. A slightly more sensible discussion is found on page 491 in the inpatient chapter dealing with acute headache.

Prominent also is ADHD with 26 pages and five authors, reflecting the American referral patterns described above. Another curious (to the European paediatric neurologist) area of practice is outlined in the chapter “Is my child ready for school?” (by which is meant for normal school since all American children are entitled to education). At the end of four pages, which include a list 14 tests—seven of which require special training and at least five of which seem specifically designed to address the question—it is concluded that “the paediatrician or family physician can assess school readiness using a thorough, careful medical history and physical examination”.

In contrast there are five pages on inborn areas of metabolism and eight on neurodegenerative disorders. Both tend to give lists of conditions but not the screening tests including DNA analysis for those conditions. Statements such as the value of increased cerebrospinal fluid lactate are of limited value unless normal concentrations are given. Curiously phenylketonuria is not mentioned. Half a page is given to treatment of inborn errors. Enzyme replacement is not mentioned under the neurodegenerative conditions. While these conditions are individually rare, their collective burden is considerable. Many, particularly the inborn errors, are both treatable and susceptible to prenatal diagnosis. Similar comments may be made for the hereditary neuropathies (eight pages) and muscular dystrophies and myopathies (eight pages).

Muscle histology gets five lines.

Some omissions may be considered dangerous. Meningoencephalitis is not mentioned as a cause of neonatal fits, optic neuritis as a cause of visual loss, and dystrophia myotonica as a cause of neonatal hypotonia. Step 2 in the treatment of status epilepticus is to give fosphenytoin. But what if the child is already on phenytoin? Step 3 has the child on either a midazolam or a pentobarbitone infusion achieving burst suppression pattern on electroencephalography—but no advice is provided on what to do if either of these drugs fails or, if they succeed, what to do next.

No doubt there are areas of the American physician will find useful—particularly, for example, the chapters on the economics of the health care system in the United States and advice on practice business management. Nevertheless, I think that this book sits uneasily between the needs of the general paediatrician and the needs of the neurologist. For the former there is more information—or not enough in a useable form—than is useful and for the latter the text is just not up to the standard already provided elsewhere. With the book is provided a CD-ROM, which has the text plus links to child neurology websites and the National Library of Medicine. Those who purchase this book are advised to avail themselves fully of these facilities.

Richard O Robinson

CORRECTIONS
