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978-0-521-86919-5 - The Pseudotumor Cerebri Syndrome: Pseudotumor Cerebri, Idiopathic Intracranial Hypertension, Benign Intracranial Hypertension and Related Conditions
Ian Johnston, Brian Owler and John Pickard
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The Pseudotumor Cerebri Syndrome

The condition known most widely as the pseudotumor cerebri syndrome is of diagnostic interest and clinical importance not just to neurosurgeons, but also to neurologists, ophthalmologists and headache specialists. In this book three clinicians with extensive experience of pseudotumor cerebri provide a comprehensive review of the condition, which has also been variously called idiopathic intracranial hypertension, benign intracranial hypertension, and other names over the century or so since it was first recognised. It argues for the grouping of all these conditions under the name of pseudotumor cerebri syndrome on the basis of a common underlying mechanism – an impairment of CSF absorption due to abnormalities at the CSF/venous interface.

Giving a detailed account of the history of the condition, the authors review the development of ideas around some of the more contentious issues, including mechanism, nosology and nomenclature. They then deal in depth with aetiology, investigative findings and strategies, treatment and outcome, based on an extensive patient series and a wide ranging review of the clinical literature. The book concludes with a chapter on experimental studies, considering the possibility of establishing a suitable experimental model to facilitate analysis of some of the unresolved issues, and pointing the way to a more complete understanding of this controversial condition.

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To
Alistair Paterson

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Preface

The syndrome we have termed the pseudotumor cerebri syndrome (PTCS) was first characterized as a distinct clinical entity in the papers by Quincke and Nonne, published a little over a hundred years ago. The condition has subsequently received a somewhat bewildering variety of names and its mechanism has also remained controversial. Moreover, it is probably not as rare as was originally thought. In addition, the insights gained by the study of its pathophysiology and management undoubtedly have more general implications for our understanding of intracranial dynamics in other conditions. For these reasons, and because there has been a considerable proliferation of literature on the subject in recent decades, we thought it would be timely to bring together the later observations with the extensive older literature.

The original intention was to have this book ready for publication in 2004 to mark the centenary of Nonne's paper which introduced the name 'pseudotumor cerebri' for a condition sporadically described during the four decades prior to that paper. Like many such endeavours, the present work took rather longer to complete than initially anticipated. Nonetheless, the belated acknowledgement of Nonne's paper does signify one of the central arguments of this monograph – that the name he proposed for the condition, a name that has endured despite many challenges, should be retained. The only proposed modification is the addition of the term 'syndrome' to embrace the collection of conditions which, in practice, share a common presentation, clinical picture, treatment strategy, and outcome, as well as, it is argued, a common mechanism. Whether this argument is successful must be left to readers to decide, but none would disagree that a consensus on nomenclature is desirable.

We believe that there is a close analogy between PTCS and hydrocephalus. Whether or not they do finally prove to have a similar mechanism, that of impaired CSF absorption, with differences being attributable to such factors as the site and cause of obstruction, the rigidity of the cranium, and other things as yet unidentified, remains to be seen. What is undeniable is that the two conditions

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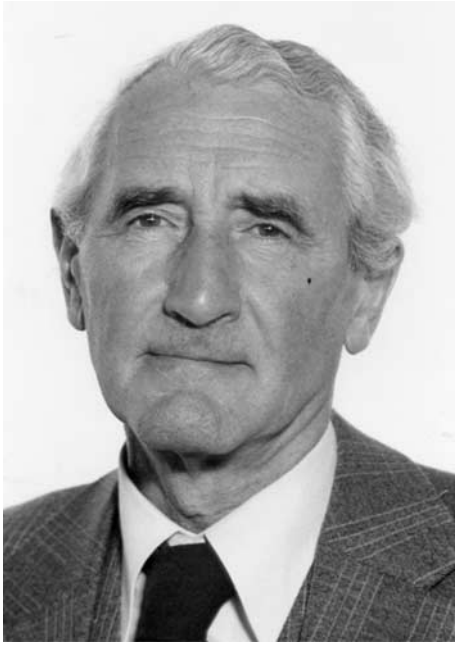

Alistair Paterson

do share a number of common aetiological factors, a similar significant proportion of cases for which there is no recognizable aetiological agent, similar clinical features insofar as these are the manifestations of intracranial hypertension without localization, and a similar dramatic therapeutic response to effective CSF drainage.

The book itself falls into three sections buttressed between brief introductory and concluding chapters. The first (Chapters 2–4) comprises the 'theoretical' section, dealing with the history of the condition, the theories on disease mechanism, and the vexed issues of nosology, nomenclature and classification. The second (Chapters 5–9) comprises the clinical section, and has two patient databases – a detailed study of two personal series of Ian Johnston covering approximately 60 years and 270 patients, and a comprehensive analysis of the burgeoning literature on the subject. The third section is a single chapter (Chapter 10) which examines experimental studies pertaining to the condition and has the underlying purpose of drawing attention to possibilities for establishing a satisfactory experimental model of PTCS which would surely help resolve some of the outstanding issues.

The three authors are closely linked, not only by their interest in the condition, but also personally, having worked together in different combinations in the three units whose patients are featured in the clinical chapters: the Institute of

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Neurological Sciences in Glasgow, the neurosurgical units associated with the University of Sydney, and the neurosurgical unit at the University of Cambridge. During our long association we have become indebted to many colleagues, both clinicians and researchers, within our own units and elsewhere. Because so many people have been involved we have decided, with regret, that they are too numerous to mention individually, but our debt is substantial. Individual mention must, however, be made of Alistair Paterson (pictured) who might justifiably be regarded as the instigator of this study which started more than thirty years ago and is still continuing. We are very happy to be able to dedicate the book to him as a mark of our enduring gratitude. We are also especially grateful to Peter McCluskey, Scott Dunkley, Marek Czosnyka, Nicholas Higgins and Nicholas Sarkies who have each made specific contributions to several of the chapters in relation to their respective specialties. Richard Barling and Rachael Lazenby at the Cambridge University Press deserve our thanks for their interest in this somewhat esoteric field, and their help generally with the project, not to mention their tolerance of the delay in delivery of the manuscript.

Finally, our hope is that this monograph might play a role in resolving some of the key issues in the continuing debate on this intriguing condition. At least it should provide an up-to-date summary of what has become a very substantial literature on the subject. To this end we have made the bibliography extensive, including papers not specifically referred to in the text but included in collected figures or influential in general analysis. We do hope that this monograph will stimulate new work and lead to further advances in the management of this distressing condition for the benefit of our patients.

Ian Johnston
Brian Owler
John Pickard

Life with Benign Intracranial Hypertension

What's in a name?
I'm angry, cross, annoyed
At a very misguided man
The one who names diseases
With inappropriate, ill suited titles.
Benign Intracranial Hypertension is the label
That doctors place on me.
If I met that man face to face
I would demand that he justify that name.
And tell me what's benign:

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I find the word an insult to my suffering.
It implies it's OK, harmless, curable,
Slight, superficial, easily treatable.
I know it's not life-threatening
In a mortal sense,
But it's killing my living.
I haven't worked for months
In the job I love,
Had countless lumbar punctures
And needles in other parts.
Operations with tubing and valves
Inserted in unsymmetrical patterns around my body.
Symptoms too numerous to list.
My marriage is under constant strain
And my children suffer,
That really hurts.
Will I be home next week or not?
I want to get on with living,
Have a routine or normality.
Yes, I'm angry all right
What right did he have to label all this benign.
I have a right to be exasperated, infuriated
With his lack of imagination and understanding.
Surely he could have come up with something,
Something just a little more grand,
Something to portray my distress,
To evoke a little understanding in people standing near,
To induce a little sympathy for me.
Come on someone please,
Start now with this disease
Let's have a renaming ceremony,
But please, remember, invite me.

Liz Galfskiy, Winchester, UK

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