INCREASED INTRACRANIAL PRESSURE IN CHILDREN

SECOND EDITION

Volume VIII in the Series

MAJOR PROBLEMS IN CLINICAL PEDIATRICS

BELL and McCORMICK

INCREASED INTRACRANIAL PRESSURE IN CHILDREN

Diagnosis and Treatment

Second Edition

by

William E. Bell, M.D.

Professor, Departments of Pediatrics and Neurology
Director, Section of Pediatric Neurology
The University of Iowa College of Medicine
Iowa City, Iowa

and

William F. McCormick, M.D.

Professor, Departments of Pathology, Neurology
and Neurosurgery; Chief, Division of Neuropathology
University of Texas Medical Branch
Galveston, Texas

Volume VIII in the Series
MAJOR PROBLEMS IN CLINICAL PEDIATRICS

ALEXANDER J. SCHAFFER
Consulting Editor
MILTON MARKOWITZ
Associate Consulting Editor

W. B. Saunders Company, Philadelphia, London, Toronto 1978

W. B. Saunders Company: West Washington Square
Philadelphia, PA 19105

1 St. Anne's Road
Eastbourne, East Sussex BN21 3UN, England

1 Goldthorne Avenue
Toronto, Ontario M8Z 5T9, Canada

Listed here is the latest translated edition of this book together with the language of the translation and the publisher.

Spanish—Editorial Cientifico-Medica, Barcelona, Spain

Increased Intracranial Pressure in Children:
Diagnosis and Treatment

ISBN 0-7216-1708-5

Last digit is the print number: 9 8 7 6 5 4 3 2 1

Forthcoming Monographs

Altman and Schwartz: *Malignant Diseases of Infancy, Childhood and Adolescence*

Belman and Kaplan: *Urologic Problems in Pediatrics*

Bluestone and Klein: *Otitis Media in Infants and Children*

Drash: *Juvenile Diabetes*

Glader: *Anemia in Children*

Griffin: *Children's Orthopaedics*

Harrison and Harrison: *Disorders of Calcium and Phosphate Metabolism in Childhood and Adolescence*

Solomon, Esterly, Loeffel: *Adolescent Dermatology*

Wannamaker: *Streptococcal Infections*

Other monographs in the series, Major Problems in Clinical Pediatrics:

Avery and Fletcher: *The Lung and Its Disorders in the Newborn Infant*—Third Edition, 1974

Bell and McCormick: *Neurologic Infections in Children*—1975

Brewer: *Juvenile Rheumatoid Arthritis*—1970

Cornblath and Schwartz: *Disorders of Carbohydrate Metabolism in Infancy*—Second Edition, 1976

Dubowitz: *Muscle Disorders in Childhood*—1978

Gryboski: *Gastrointestinal Problems in the Infant*—1975

Hanshaw and Dudgeon: *Viral Diseases of the Fetus and Newborn*—1978

Lubchenco: *The High Risk Infant*—1976

Markowitz and Gordis: *Rheumatic Fever*—Second Edition, 1972

Oski and Naiman: *Hematologic Problems in the Newborn*—Second Edition, 1972

Rowe and Mehrizi: *The Neonate with Congenital Heart Disease*—1968

Royer, et al: *Pediatric Nephrology*—1974

Scriver and Rosenberg: *Amino Acid Metabolism and Its Disorders*—1973

Smith: *Recognizable Patterns of Human Malformation*—Second Edition, 1976

Smith: *Growth and Its Disorders*—1977

Solomon and Esterly: *Neonatal Dermatology*—1973

Foreword

The first edition of Bell and McCormick's *Increased Intracranial Pressure in Children* appeared in 1972 and was received by the pediatric community with appreciation. In all my years of editing, I remember few volumes whose reviews were so consistently laudatory and so free of derogatory comment. Now the authors have revised their material drastically. They have updated all of it, with special attention focused upon the importance of the CAT scanner in diagnosis. They have enhanced the discussion of central nervous system physiology and have made us aware of the clinical importance of intracranical pressure recordings, among many other improvements. We do not doubt that this edition, too, will be accepted with enthusiasm.

Alexander J. Schaffer, M.D.

Preface

In the interval since the publication of the first edition of this book in 1972, certain previously held concepts regarding intracranial pressure have been abandoned, others have been expanded, and new methods of diagnostic examination have been developed. The most important advance in the clinical management of intracranial disorders in this period has been the technique of computerized axial tomography. The value of this remarkable procedure is beyond doubt in regard to diagnosis and management of many different disorders which involve the intracranial contents. Indeed, the procedure is so definitive in many instances that its availability can lead one to the dangerous temptation of minimizing the importance of the time-honored history and neurological examination, the basic methods of inquiry which provide the foundation for the selection of the appropriate diagnostic procedures and the most logical method of treatment.

We stress again the need for the clinician to be constantly aware of the possible adverse effects of some diagnostic procedures, as well as certain types of treatment, when dealing with the child having one of the many causes of increased intracranial pressure. It is as important to avoid unnecessary and potentially dangerous procedures as it is to select those studies most likely to yield the needed information.

We have attempted in this volume to outline logical and reasonably safe methods of diagnostic analysis of the disorders described. It must be added, however, that our suggestions concerning selection of diagnostic tests and plans for therapeutic approach are not necessarily the only acceptable ways by which these problems can be handled.

We wish to acknowledge the resources and help provided by Dr. M. W. Van Allen and Dr. Fred Smith which enabled the accomplishment of this volume. Dr. Steve Cornell kindly provided many of the computed tomograms that appear in these pages. We also thank Mr. John J. Hanley and Susan G. Hunter of the W. B. Saunders Company for their encouragement and skillful guidance.

William E. Bell
William F. McCormick

Contents

PART
I

Increased Intracranial Pressure in Children – General Concepts

Chapter One

SYMPTOMS, SIGNS, AND PHYSIOLOGIC CONCEPTS

Increased intracranial pressure in infants and children occurs with a great variety of disorders, is of variable degree from patient to patient, and is manifested differently according to the age of the child and other factors. Intracranial hypertension can itself become hazardous, regardless of the cause, and can sometimes be forestalled with appropriate therapeutic measures. Awareness of the existence of an abnormal elevation of intracranial pressure is also necessary so that certain potentially harmful measures may be avoided. For example, it is generally unwise to administer enemas to children with increased intracranial pressure because straining may markedly elevate the intracranial venous pressure, possibly precipitating transtentorial herniation with brain stem compression and death. A tap water enema can result in expansion of the vascular volume due to absorption of water from the rectum, producing further expansion of the intracranial contents with potential disaster. Narcotic drugs, such as meperidine or morphine, may also be hazardous in patients with increased intracranial pressure and must be used only with caution. Acute respiratory compromise followed by hypoxia and hypercapnia is now recognized as an event that can provoke a precipitous and disastrous rise in pressure in a child with pre-existent intracranial hypertension. For this reason, diligent respiratory care represents one of the critical aspects of management of the child with many acute neurologic and neurosurgical conditions. An additional potentially dangerous problem in conditions involving increased pressure concerns the possible effects of lumbar puncture, a consideration to be discussed later.

There is normally a rather delicate balance between the volume of the intracranial space and the volume of the contents occupying the intracranial space. After the fontanels have closed and the skull sutures have fused, the cranium is a rigid container with a limited capability for effective adaptation to the development of mass lesions within the skull or significant enlargement of any of the contained structures. The intracranial contents include the meninges, the brain parenchyma, cerebrospinal fluid within the ventricular and subarachnoid spaces, and blood vessels and their contents. For pressure relationships to remain unaltered, enlargement of

any one of these intracranial structures must be accompanied by a reduction in volume of one or more of the others, a principle that is constructed on the basis of modifications of the Monro-Kellie doctrine. The margin of safety afforded by this adaptive mechanism is limited and consists primarily of the ability of displacement of cerebrospinal fluid from the intracranial cavity. Once this has occurred to the maximal limit, the anticipated response to further enlargement of any of the intracranial compartments is the appearance of the various manifestations of increased intracranial pressure. Enlargement of the cerebral ventricles resulting from cerebral atrophy is clearly a different matter in most instances, since the ventricular dilatation in this instance is a secondary phenomenon resulting from an atrophic state of the brain.

SYMPTOMS AND SIGNS—INCREASED INTRACRANIAL PRESSURE

Recognition of the presence of increased intracranial pressure is usually dependent on the existence of certain symptoms and signs (Table 1–1), the finding of characteristic roentgenographic abnormalities, or the demonstration of an abnormally high pressure at the time of lumbar puncture. Ordinarily, it is a combination of these findings that establishes the presence of increased intracranial pressure, since any one isolated abnormality may be misleading because of possible alternative explanations. Thus, headache alone is rather nonspecific, but the presence of both headache and papilledema is virtually diagnostic of a syndrome associated with increased intracranial pressure, assuming that the blood pressure is normal. Also, there is great variation in the expression of the various manifestations of increased pressure from patient to patient.

Table 1–1

Symptoms—Increased Intracranial Pressure
Headache
Nausea and vomiting
Diplopia
Transient obscurations (unusual)
Lethargy, drowsiness
(None)
Signs—Increased Intracranial Pressure
Papilledema
Sixth nerve palsy
Decreased awareness
Bulging fontanel (infant)
Palpable suture spread (infant)
Progressive head enlargement (infant, child)
(Signs of internal herniation)
(None)

The rigidity and lack of expansile characteristics of the skulls of older children and adults are not qualities of the skulls of infants or young children. The potential ability of the intracranial volume to increase in the infant as an adaptation to increasing pressure within the head accounts for certain differences in the clinical manifestations at various ages. The anterior fontanel usually remains patent until 15 to 18 months of age in the normal infant. While this may serve only slightly as a mechanism for compensation for increasing pressure within the skull, it does provide a valuable clinical sign indicative of abnormal elevation of intracranial pressure. The tense, bulging fontanel of the infant under one year of age is indicative of an increased intracranial pressure syndrome and should correlate with other findings consistent with this impression. For proper assessment of the anterior fontanel, the baby should be relaxed and not crying or straining. The child should be held in the sitting or upright position, with the examiner's hand then making an estimate of the degree of elevation and the resistance to gentle pressure. In the normal, quiet infant in the upright position, the anterior fontanel is usually either flat or slightly concave compared with the surrounding scalp. There is a great deal of variation in the size of the anterior fontanel in the normal newborn infant (Popich and Smith, 1972; Tan, 1976), and it customarily enlarges somewhat in the first few months after birth (Scammon and Adair, 1930). Pulsations can be felt by pal-

pation of the normal anterior fontanel but are also frequently noted in the full fontanel in the infant with increased intracranial pressure (Blaauw et al, 1974). Pulsations are usually lost in the very tense fontanel secondary to marked intracranial pressure elevation. One may observe a bulging fontanel, perhaps secondary to increased venous pressure, in an infant with congestive heart failure without other evidence of significant cerebral disease.

The cranial sutures rapidly become separated with increased intracranial pressure in infants and young children. In young infants this is easily identified by palpation of the skull, while in older children skull roentgenography is usually necessary for its demonstration. Suture spread on x-ray may occur in children at least up until 12 years of age, but with considerable variation from case to case. Percussion of the head of the young child with abnormal suture spread may cause a "cracked-pot" sound referred to as the Macewen's sign. The ability of the cranial sutures to spread in children may delay or abate some of the symptoms or signs ordinarily associated with the various disorders in question. For example, in infantile hydrocephalus due to aqueductal atresia, marked head enlargement often occurs, while papilledema is unusual. The presence of an open fontanel and the ability of cranial sutures to spread have been assumed to be protective mechanisms in the young infant, and thus to his advantage. This is no doubt true from the standpoint of symptoms and signs of certain conditions involving increased intracranial pressure. In the infant with progressive hydrocephalus, however, physiologic concepts now suggest that cranial sutural separation can actually serve to his disadvantage, in that it prevents the possible development of cerebrospinal fluid absorptive capabilities by bringing about a natural lowering of intracranial pressure. The hypothesis, if correct, explains one of the main disadvantages of shunting procedures for treatment of the infant with hydrocephalus. The lowered pressure discourages the potential function of absorptive pathways, and the child remains shunt-dependent. This is the basis for the concept of head-wrapping for treatment of certain types of infantile hydrocephalus (Epstein et al., 1973, 1975).

The commonest and most consistent symptoms and signs of increased intracranial pressure in the child beyond infancy include headaches, nausea, vomiting, diplopia, and papilledema. Children in the toddler age group or younger usually express the presence of headache in the form of irritability and anorexia. Older children with intracranial hypertension describe headaches of variable intensity, usually generalized but with a bifrontal predominance. The headache may be persistent but is more often intermittent and frequently is associated with vomiting. When the increased pressure syndrome is due to an intracranial tumor, headaches and vomiting sometimes have a marked tendency to recur in the early morning hours soon after arising but before breakfast. This can also occur with other causes of increased pressure but is most often described in children with posterior fossa neoplasms. The cause of these early morning symptoms is not entirely clear, although continuous intracranial pressure monitoring with a subdural transducer has revealed large, intermittent pressure increases during the rapid eye movement phase of sleep (Cooper and Hulme, 1966). It is assumed that such pressure changes during certain stages of sleep are secondary to cerebral vasodilatation associated with metabolic changes during sleep. Vomiting in children with abnormal increase in the intracranial pressure is not different from vomiting due to other causes. The projectile component has been overemphasized in this regard, as it is less common than generally believed. In addition, vomiting of a projectile nature can occur with a number of disorders that do not involve the brain.

Personality and behavioral changes are additional common symptomatic expressions of intracranial hypertension. Infants and toddlers become irritable, increasing the complexities of physical examination.

Older children are frequently noted to become indifferent and drowsy or to show loss of interest. School performance may decline, and physical activity diminishes. A previously active youngster may now come home from school and prefer to lie down or play alone within the house rather than to be outside with his playmates. Moodiness and ease of tearfulness over trivial matters may become evident. These personality changes secondary to increased pressure pass through stages of subtle indifference to lassitude and eventually lead to lethargy or drowsiness. Parents often observe progressively increasing complaints of ease of fatigue and tiredness, with more and more time being devoted to sleep. As a result of loss of appetite compounded by recurrent attacks of vomiting, significant weight loss supervenes. If persistent and if of sufficient degree, increased intracranial pressure can lead to significant memory loss, eventually with evidence of progressive dementia. An unusual complication of certain conditions associated with sudden, severe increase in intracranial pressure is pulmonary edema, probably secondary to an excessive autonomic discharge of central origin (Ducker et al., 1968, 1969). In the later stages of intracranial hypertension, alterations of vital signs appear, with slowing of the pulse, elevation of the blood pressure, and irregularity of the respiratory rhythm. These signs, sometimes associated with periodic episodes of decerebrate posturing, indicate the probable occurrence of transtentorial herniation secondary to increased intracranial pressure and imply the possibility of impending death if the process cannot be reversed.

PAPILLEDEMA

Papilledema is the single most reliable physical sign indicative of increased intracranial pressure. The possibility of intracranial pressure elevation is not excluded by the absence of papilledema, however, as some patients will show no funduscopic abnormalities, even in the presence of marked increase of the intracranial pressure. Originally, optic disc swelling regardless of the cause was called optic neuritis, but in 1908 Parsons proposed the term papilledema for edema of the optic nerve head associated with intracranial tumors. The term has been variously used since that time, with some authors equating papilledema to any type of disc swelling in which visual acuity remains unaffected. Recent authors have logically divided disc edema into a variety of categories and have emphasized the importance of reserving the designation of papilledema for that type of optic disc swelling that is secondary to increased intracranial pressure (Lubow, 1973; Hedges, 1975) (Table 1–2).

Papilledema associated with intracranial hypertension is subdivided into incipient (early) papilledema, fully developed papilledema, chronic papilledema, and chronic atrophic papilledema (Lubow, 1973). The latter is the result of long-standing disc edema in which degenerative changes have resulted in glial proliferation, giving rise to a pale gray discoloration of the disc with narrowing of the retinal arteries and accompanied by visual acuity loss and visual field changes.

Table 1–2. Classification of Optic Disc Edema*

TYPE	PATHOGENESIS
Papilledema	Increased intracranial pressure
Papillitis	Inflammatory—demyelinating
Ischemic optic neuropathy	Arterial insufficiency (hypertension, cranial arteritis, embolic thrombosis, acute blood loss, anemia)
Stasis optic neuropathy	Central retinal vein thrombosis
Compression optic neuropathy	Mass lesions or infiltrative lesions at distal end of optic nerve

*Modified from Lubow, M.: Optic disc edema, revisited. *In* Smith, J. L.: Neuro-ophthalmology. Vol. VII, C. V. Mosby, St. Louis, 1973.

Disc edema of inflammatory origin is appropriately termed papillitis, and although it may be identical in ophthalmoscopic appearance to papilledema, it is accompanied by significant loss of visual acuity and visual field deficits. Ischemic optic neuropathy is a type of disc edema secondary to arterial insufficiency, and thus it is far more often observed in adults than in children. Hayreh (1969) has shown that the arterial supply of the optic disc is not via the central retinal artery but is from the short posterior ciliary arteries which form the circle of Zinn. It is occlusion of segments of the posterior ciliary arteries which gives rise to ischemic optic neuropathy. The area of the disc affected depends upon the extent of occlusion of its blood supply. Occlusion of one small branch of the posterior ciliary artery leads to a sectoral disc lesion, while thrombosis of the main posterior ciliary artery produces extensive infarction of the entire disc. Hypertension, cranial arteritis, atherosclerosis, and embolic thrombosis are the most common causes (Boghen and Glaser, 1975). Visual loss is common although not always present, and visual field defects are frequently of the altitudinal type or involve sectors of retinal function. Other unusual types of disc edema in children include stasis optic neuropathy which follows central retinal vein thrombosis, and compression optic neuropathy which is the result of a mass lesion or infiltrative lesion implicating the distal part of the optic nerve.

The period of time required for papilledema to develop following the occurrence of increased intracranial pressure is not clear and certainly must be quite variable, depending especially on the rate of rise and also on the degree of intracranial hypertension. The earliest ophthalmoscopic signs can occur within 24 to 48 hours, and perhaps a week or more is required for fully developed papilledema to become evident (Hedges, 1975). Following return of intracranial pressure to normal levels, several weeks, perhaps six to eight in many cases, are usually required for complete resolution of fully developed papilledema. An additional factor that influences the occurrence of papilledema is pre-existing primary optic atrophy. Although it is difficult to document, children with optic atrophy who subsequently develop intracranial hypertension appear less likely to reflect its presence by the development of papilledema. Experimental studies have shown that traumatic atrophy of half the fibers of the optic nerve prevents papilledema in the atrophic half of the disc (Hayreh, 1964, 1968.) The ability of cranial sutures to become widely separated in early infancy is another obvious limiting factor in this regard. The young infant with hydrocephalus rarely exhibits papilledema.

When all age groups are considered, it has been estimated that papilledema is observed in approximately 70 percent of patients with infratentorial tumors and 50 percent of patients harboring supratentorial neoplasms (Petrohelos and Henderson, 1950; Huber, 1971). As papilledema develops, the degree of disc swelling is often asymmetrical, regardless of the cause of increased intracranial pressure. With progression, hemorrhages can be entirely unilateral, even though the disc swelling may appear to be equal in the two eyes. With a localized supratentorial mass lesion, papilledema is usually bilateral. When it is asymmetrical, the side with the greatest degree of edema *may* reflect the side of the lesion. Experimental studies by Hayreh have demonstrated that when increased intracranial pressure is induced in monkeys by inflation of a balloon on one side above the tentorium, papilledema is usually more pronounced on the side of the balloon.

Because of the importance of optic disc abnormalities in providing evidence of increased intracranial pressure as well as many other pathologic states, practitioners should become experienced at funduscopic examination by including it as part of every physical examination. Ophthalmoscopic examination should be done with the room partially darkened but with enough illumination available so that the patient can fixate on a designated object. If the pupils are small or the patient is

uncooperative, it is advisable to dilate the pupils with a short-acting mydriatic before the fundus examination is attempted. Adequate fundus examination is indispensable, and its importance far outweighs the disadvantages of temporarily obscuring certain signs by the use of drugs to produce pupillary dilatation. Although induction of pupillary dilatation in the lethargic or comatose patient should not be done indiscriminately, it is entirely acceptable when a short-acting agent is used and when adequate funduscopy cannot otherwise be accomplished. The patient's eye to be examined should be rotated slightly medially to facilitate observation of the optic disc. Older children of normal intellect will usually sit quietly with eyes directed toward an object, permitting funduscopic examination. Retarded children or those in the younger age group are often more easily examined while in the supine position. This position results in stabilization of the child's head and trunk, thus eliminating the usual fidgety body movements characteristic of this group of children. A flashlight held by the parent and directed at the ceiling immediately above will usually be watched by the child, providing ocular fixation. Funduscopic examination in the infant occasionally can be done without restraint of the child, but assistance may be needed to immobilize the head and arms.

Papilledema in its early stages is ordinarily an objective finding identified by the examiner and usually does not cause deficits of which the patient is aware. An occasional patient with high-grade papilledema will describe recurrent, brief periods of momentary disturbance of vision of both eyes. These amblyopic attacks are referred to as transient obscurations and usually last only a few seconds. Some patients will describe a sensation resembling a heavy fog or veil transiently obscuring the visual environment. Others will simply describe the sudden appearance of dim vision as though the lights were turned down, with complete recovery following seconds later. These episodes are associated with advanced papilledema due to marked increase in intracranial pressure but under any circumstances cannot be considered common. The pathogenesis of such attacks remains undetermined, although they are believed to be related to spontaneous fluctuations in cerebrospinal fluid pressure and the A pressure waves described by Lundberg (1960).

With the exception of the infrequent occurrence of the transient episodes described above, visual acuity remains unaffected in most individuals with early or mild papilledema. The persistence of the disorder for a long period of time can be followed by secondary atrophic changes in the nerve head, which is accompanied by visual acuity loss and visual field changes. The prevention of this late complication of papilledema must always be given due consideration when one is dealing with patients with increased intracranial pressure, regardless of the cause. The visual field abnormality in the patient with papilledema consists of a concentric enlargement of the blind spots, best identified by tangent screen or Goldmann perimetric examination. As a rule, ophthalmoscopic evidence of edema of the nerve head is present by the time the blind spot is significantly enlarged on formal visual field examination. Thus, it can be assumed to be consistent with the diagnosis of papilledema but as an isolated finding should not be relied upon as diagnostic. As swelling of the optic disc advances, the edema spreads into the retina toward the macula. This leads to further blind spot enlargement toward the point of fixation, eventually converting an enlarged blind spot into a cecocentral scotoma with slanting margins. The identification of enlarged blind spots has its greatest usefulness in excluding other conditions that may resemble papilledema, such as optic papillitis.

The ophthalmoscopic appearance of the nerve head with papilledema varies depending on the degree of swelling and the duration of the process (Fig. 1–1). Findings in the adjacent retina also are somewhat variable, depending on the nature of the disorder producing increased intracranial pressure. For example, retinal