Title
Normal Pressure Hydrocephalus

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even when $PaCO_2$ is not high. The family should be made to understand this so that oxygen will not be used outside the hospital in an emergency.

APPENDIX

Organizations devoted to education and research in ALS, from whom newsletters and other useful information for patients and physicians may be obtained.

Amyotrophic Lateral Sclerosis Society of America
15300 Ventura Blvd., Suite 315
Sherman Oaks, CA 91403

Muscular Dystrophy Association
810 Seventh Ave.
New York, NY 10019

National ALS Foundation Inc.
185 Madison Ave.
New York, NY 10010

Manufacturers of products and clothing for the physically disabled: .

Sears, Roebuck and Co., Home Health Care Catalog

FashionAble
Box S
Rocky Hill, NJ 08553

Fred Sammons Inc.,
Professional Self-help Aids Catalog
Box 32
Brookfield, IL 60513-0032

A recent book of interest to patients as well as physicians and other care providers: The Diagnosis and Treatment of Amyotrophic Lateral Sclerosis. Mulder DW. Boston: Houghton-Mifflin, 1980.

NORMAL PRESSURE HYDROCEPHALUS

CLAUDIA KAWAS, M.D.
LESLIE WOLFSON, M.D.

The syndrome of normal pressure hydrocephalus (NPH) was described in 1965 by Hakim and Adams. The clinical presentation consists of a gait disturbance dementia and incontinence, accompanied by cerebral ventricular enlargement with a normal cerebrospinal fluid pressure. In the Hakim and Adams series of three patients, there was improvement with a shunting procedure. Enthusiasm for another potentially correctable dementia led to broad application of diagnostic criteria; many patients with dementia and prominent ventricular enlargement underwent surgery, with variable, often unsatisfactory, response. Subsequently, there was an attempt to define physiologically and tighten the original diagnostic criteria. Emphasis was focused on the gait abnormality and an attempt was made to demonstrate a CSF absorptive defect which was produced by a block within the basal cisterns. In a syndrome with widely ranging diagnostic criteria, it was not surprising that there was a variable response to therapy in the reported series. Individual series reported clinical improvement ranging from 10 to 80 percent of patients (median 40%). Frequently, in patients who vigorously fulfilled diagnostic criteria, there was no improvement following shunting. Who should be considered for shunting? No single clinical feature or diagnostic procedure can determine this selection or predict improvement as a result of shunting. The frequent occurrence of significant, even life-threatening, com-

plications in these patients dictates that the physician's major responsibility is to select those patients who are most likely to benefit from the procedure as well as those patients who will tolerate it best. The selection must be made after careful consideration of the history and clinical status of each patient.

History

Patients with normal pressure hydrocephalus may manifest progressive symptoms months to years prior to presentation. Those patients with a progressive gait disturbance as an initial, or at least early and prominent, symptom are more likely to improve, sometimes remarkably, with shunting. The dementia is variable and most frequently is noticed by the family as a generalized slowing or even depression. Careful questioning regarding bladder function may reveal, if not frank incontinence, difficulties which may have been attributed to other causes (e.g., prostatism, prolapsed bladder). Many patients in fact have undergone surgical procedures without improvement. Patients with a remote history of subarachnoid hemorrhage, meningitis, or significant head trauma have been reported to have a higher rate of response to shunting (about 65%).

General Medical History

Surgical morbidity and mortality may be minimized by consideration of the patient's overall health (e.g., presence of significant cardiac, pulmonary or other disease). Age is not a contraindication to therapy, but the risks of surgery and general anesthesia do increase somewhat in the senium. Unfortunately, many complications of shunting are not predictable on this basis.

Physical Examination

The gait disturbance of NPH is often difficult to characterize and has been referred to as a gait apraxia, presumably a manifestation of bilateral frontal lobe dysfunction. The gait is characterized as slow, hesitant, with the feet moving as if they were attached to the floor. Steps are small and the patient cannot easily lift his feet over obstacles. Lower extremity spasticity and Babinski signs may be present. The severity of dementia ranges from mild psychomotor retardation to akinetic mutism. In our experience, the most likely candidate for improvement is the patient with a mild degree of dementia accompanied by psychomotor retardation or the patient with no discernible dementia. The clinician should look for signs of incontinence in addition to seeking historical evidence. No other significant deficits should be present on physical examination. Signs such as papilledema, significant extrapyramidal disease, or a hemiparesis should make the clinician consider alternative diagnoses such as multiple cerebral infarcts, Parkinson's disease, or alcoholism.

Differential Diagnosis

Other diseases that may be confused with normal pressure hydrocephalus include: Parkinson's disease, which can be differentiated by its clinical features and the response to Sinemet; multi-infarct dementia, which can be differentiated by clinical and CT or nuclear magnetic resonance (NMR) features suggestive of multiple cerebral infarctions; bifrontal structural lesions, which include both primary brain tumors (e.g., butterfly gliomas) and metastatic lesions; obstructive hydrocephalus produced by mass lesions or aqueductal stenosis which may result in a rapidly evolving gait abnormality and dementia; and metabolic illnesses such as pernicious anemia and hypothyroidism.

Diagnostic Results

Ventricular size is best determined by CT scan. We look for the pattern of ventricular enlargement without sulcal enlargement, although almost one-third of the patients who respond to shunting actually have some degree of cortical atrophy. The CT scan should be carefully evaluated for the presence of small infarcts and other structural lesions which may produce bifrontal dysfunction.

Further evaluation should be undertaken only if the patient is considered a candidate for surgery. Although various infusion and radiographic techniques have been developed, we further evaluate appropriate patients with radioisotope cisternography, a relatively safe index of the integrity of cerebral subarachnoid pathways which may not predict the therapeutic response or clarify the etiology of the illness. For example, 10 percent of patients with Alzheimer's disease have a positive cisternogram but would presumably not benefit from shunting.

Cerebrospinal fluid pressure can be measured at the time of isotope injection; the fluid should be sent for diagnostic studies which include cells, protein, glucose, cytology, bacterial and fungal cultures, and cryptococcal antigen.

Demonstrating abnormal intracranial pressure phenomena using invasive pressure monitoring is carried out in some centers. It is unclear how much this procedure improves patient selection although figures

of therapeutic response in the range of 70 to 80 percent have been reported in at least two of these series.

Ideally, the patient selected for surgical intervention will be in relatively good health, complaining of a gait disorder (with or without dementia and incontinence), whose CT scan shows communicating ventriculomegaly (which should be markedly out of proportion to sulcal atrophy), and whose additional diagnostic studies are compatible with the diagnosis of normal pressure hydrocephalus.

THERAPY

Currently, shunting is the only type of definitive treatment available. Most commonly, this consists of ventricular shunting to the peritoneal cavity which carries less morbidity than does shunting to other body cavities such as the atrial or pleural cavity.

A consideration at the time of shunting is simultaneous brain biopsy. This additional procedure may increase the complication rate somewhat but should be considered as a possible ancillary diagnostic procedure for selected patients, especially for those who have a prominent dementia. The biopsies often produce definitive diagnoses (e.g., Alzheimer's disease, hypertensive vascular disease) which can explain a failure of therapy. The feasibility of this procedure should be determined with the neurosurgical consultant and is dependent in measure upon the availability of neuropathologic facilities.

Complications

Forty percent of the patients sent to surgery will suffer major complications which include subdural and intracerebral hematoma, cerebral infarction, infection (including the central nervous system), seizures, shunt malfunction, pulmonary embolus, and death. This complication rate obligates the practitioner to a careful selection process in order to optimize the risk-benefit ratio.

Outcome

Improvement often occurs relatively promptly (especially in patients with only a gait disorder), but it should be noted that in some patients, improvement has been reported weeks to months later. This improvement may not necessarily correlate with change in ventricular size after shunting. Follow-up scans should be routinely done in order to assess catheter placement as well as any changes in cerebral appearance such as infarcts or subdural hematomas.

Deterioration subsequent to an initial response to shunting frequently occurs and should be evaluated by the physician as possible shunt failure. Despite this consideration, many patients are found to have normal shunt function and seem to enjoy only a transient improvement. This transient improvement following shunting has not been satisfactorily explained. Since no other specific therapeutic options exist, these patients should be treated supportively as are other dementia patients.

Ancillary Care

For those patients who are not treated with shunting, or who do not respond to surgery, management of the gait disorder should be accomplished with any necessary ambulatory aides (cane, walker, or even wheelchair) in conjunction with physical therapy. Attention should be given to making the patient's environment as safe and manageable as possible. As the patient's gait deteriorates and dementia progresses, protective services may be necessary. Supportive care for the families, as well as the patient, is an essential part of treatment.

Management of incontinence and urinary infections should include culture-appropriate antibiotics, adult diapers, and, occasionally, various types of catheters. Patients with behavioral symptoms such as severe agitation or psychosis may benefit from low-dose neuroleptic agents such as Haldol; concurrent depressions may require treatment. Care should be taken not to oversedate patients, many of whom will be exquisitely sensitive to these drugs.

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