

ARTS & HUMANITIES

What Do Physicians Know About Normal Pressure Hydrocephalus and When Did They Know It? A Survey of 284 Physicians

Harold O. Conn, MD^a, and Francis M. Lobo, MD^b

^a*Professor of Medicine, Emeritus, Yale University School of Medicine, New Haven, Connecticut;* ^b*Assistant Professor, Internal Medicine Immunology, Yale University School of Medicine, New Haven, Connecticut*

Normal pressure hydrocephalus (NPH)[†] is a relatively new neurologic disorder first described by Salamon Hakim of Bogotá, Colombia, in 1965. NPH is characterized by three symptoms — impaired gait, incontinence and dementia — and an anatomic abnormality, i.e., enlargement of the cerebral ventricles, which can be seen on computerized tomographic or magnetic resonance imaging. Surprisingly, the intracranial pressure is normal.

The first author of this article, a Yale Medical School faculty member, developed NPH over the decade from 1992 to 2002, during which it was erroneously diagnosed as cerebral atrophy and/or Parkinson's disease. On recognizing the lack of awareness of NPH by physicians, he initiated a survey to explore this problem. He interviewed 166 practicing physicians who graduated from 50 American and 33 foreign medical schools, using a one-page, 10-point questionnaire (Part I).

Almost one-third of the physicians had never heard of NPH. One-fifth had learned of NPH in medical school, and about half learned of it after medical school.

Because there were insufficient physicians surveyed from 1986 to 2005, we recruited 118 additional physicians from the 20 Yale Medical School graduating classes from 1986 through 2005 (Part II). Two-thirds of them had learned of NPH in medical school, and one-fourth during residency and fellowship. Seven percent had never heard of NPH.

The significance of these studies is discussed.

To whom all correspondence should be addressed: Harold O. Conn, MD. From May to September: 1 Mansfield Grove Road, Apt. 115, East Haven, CT 06512, Tele: 203-469-2007, Fax: 203-467-8988; from October to April: 1800 South Ocean Blvd., Apt. 1109, Lauderdale By The Sea, FL 33062, Tele: 954-876-0440, Fax: 954-876-0444, E-mail: halcon1109@aol.com. Francis M. Lobo, DCB 013 Int Med Immunology, 15 York Street, New Haven, CT 06510-3221, Tele: 203-688-6970, E-mail: francis.lobo@yale.edu.

[†]Abbreviations: NPH, normal pressure hydrocephalus; CSF, cerebrospinal fluid; VSS, ventriculosystemic shunting; YUSM, Yale University School of Medicine; PD, Parkinson's disease; VPS, ventriculoperitoneal shunt; HIC, Human Investigation Committee; AYAM, Association of Yale Alumni in Medicine.

INTRODUCTION

Normal pressure hydrocephalus (NPH) is a relatively new neurologic disorder of elderly patients described by Salamon Hakim in Spanish in a thesis in Bogotá, Colombia, in 1964 [1]. It first appeared in the medical literature in English in 1965 in articles by Hakim and Adams [2] and Adams, Hakim, et al [3]. NPH is characterized by an unusual triad of neurologic symptoms — impaired gait, urinary and/or fecal incontinence, and dementia — and an anatomic abnormality, i.e., enlargement of the cerebral ventricles that can be demonstrated by computerized tomography (CT) or magnetic resonance imaging (MRI) of the brain [4]. Recently, another anatomic abnormality was described in NPH — a decrease in midbrain diameter on MRI [5] that is restored to normal by ventriculosystemic shunting [6]. Surprisingly, the intracranial pressure of this unique type of hydrocephalus is normal, or nearly so.

The precise pathogenesis of NPH is not known, but it is well-known that despite the absence of increased intracranial pressure, the drainage of cerebrospinal fluid (CSF) regularly induces transient clinical improvement, and ventriculosystemic shunting (VSS) usually results in prolonged remissions [7]. For reasons that are not clear, some “experts” still question the reversibility of NPH — and even its very existence [8].

In 1992, after more than 50 years as a faculty member of the Yale University School of Medicine (YUSM), the first author of this article retired and soon thereafter began to develop progressive slowness and clumsiness of gait, which was diagnosed by one neurologist as cerebral atrophy and by others as variants of Parkinson’s disease (PD). He did not respond to pharmaceutical therapy of PD. The patient developed urinary urgency and incontinence in 2002, the second of the three classic symptoms of NPH, but the diagnosis of NPH was still not considered. In retrospect, by that time he had been having some subtle changes in mental state — loss of short term memory, slowing of responsiveness, a decrease in reaction

time and mental sharpness, and some loss of his sense of humor.

The correct diagnosis of NPH was made inadvertently in April 2003. By that time, the patient was virtually unable to walk at all. The patient, who spent winters in Florida, wrote his neurologist in Connecticut asking for authorization for an electric scooter, which Medicare provides without charge for all patients who can’t walk. The neurologist refused. He gave no explanation. The patient’s attending physician was so displeased by this refusal that he immediately referred the patient to another, much younger neurologist. He assured the patient that the new consultant would authorize the scooter and would give him a “second opinion.” The neurologist examined the patient and his MRI and approved the scooter. His “second opinion” was that he thought the patient had NPH, which is a reversible disease. The diagnosis was quickly confirmed by an almost miraculous, instantaneous improvement after removal of a large volume of CSF. Ironically, the refusal to approve the scooter led directly to the prompt restoration of the patient’s ability to walk.

A week later, a ventriculoperitoneal shunt (VPS) was implanted that virtually restored the patient to normal health. The author had never heard of NPH, and he realized that many of his medical colleagues also were unaware of NPH. To confirm and explain this observation, he started a survey to determine at what stage in their training physicians learned about NPH and whether the newness of NPH contributed to this lack of knowledge.

METHODS

The design of this investigation is simple. It consists of a one-page, 10-point questionnaire that can be completed within two minutes (Figure 1). The subjects of the survey were all the physicians with whom the first author came in contact during the succeeding two years (July 2003 through June 2005).

The questionnaire asks the name of the medical school from which the physician grad-

QUESTIONNAIRE FOR NPH SURVEY (PART 1)

Physician's Name: _____

- 1. I graduated from the _____ School of Medicine in 19 ____
- 2. My medical specialty (subspecialty is) _____
- 3. I first recognized the abbreviation "NPH" in
 medical school, residency, thereafter, I don't know the abbreviation
- 4. I learned about normal pressure hydrocephalus in the year _____
 medical school, residency, fellowship, never
- 5. I have seen about _____ patients with NPH in the past five years.
(number)
- 6. I have referred about _____ patients for confirmation of the diagnosis of NPH in the past 5 years.
(number)
- 7. I have referred about _____ patients for treatment of NPH in the past 5 years.
(number)
- 8. Do you believe it's ever too late to implant a shunt (when indicated)?
 Yes No
- 9. Do you believe that the diagnosis of NPH is frequently missed: Yes No
- 10. If "yes," why?

Signature _____ Date _____

Figure 1. Questionnaire for NPH survey (Part I). The questionnaires for Part I were identified by only a code number, e.g., 05-01, indicating the year and order in which the physician's questionnaire was received for that year. Questionnaires for Part II included one additional question.

uated, the year of graduation, and the medical specialty or subspecialty practiced. Before the disease under investigation was identified, the respondents were asked if they recognize the abbreviation "NPH," which is the term physicians who deal with NPH use to refer to it. We assumed that familiarity with the abbreviation "NPH" would indicate awareness of and knowledge about this syndrome. Respondents were asked the year they had first learned of NPH. In addition, each was asked to estimate the number of patients with NPH that they had seen during the preceding five years. Because clinical improvement usually follows the drainage of CSF and a sustained remission is usually induced by VSS, even when the patient appears terminal, the physicians were asked, "Is it ever too late to consider shunt therapy?" for patients with NPH. The final question was: "Is the diagnosis of NPH frequently missed? And, if so, why?"

During the allotted period, 166 physicians were interviewed. When the preliminary data were analyzed, it was obvious that the number of physicians who graduated in the most recent 20 years (1986 to 2005) was inadequate to answer definitively the questions asked. After considering ways of increasing the number of physicians surveyed without altering the conceptual simplicity of the survey, it was decided that these goals could be best achieved by adding randomly selected YUSM graduates from each of the classes from 1986 through 2005. The physicians who already had been interviewed were termed Part I, and the additional physicians, Part II. This modification of protocol was discussed with the Human Investigation Committee (HIC), the Dean of the YUSM, and the Association of Yale Alumni in Medicine (AYAM), all of whom ap-

Table 1. Stages of career at which surveyed physicians learned about NPH.

| Part I | | | | | |
|---------------------|----------------------|------------------------------|-------------------------------------|--------------------------------|-----------------------------|
| Group: Year-Grad | No. of Physicians | Medical School No. (%) | Residency- Fellowship No. (%) | Medical Practice No. (%) | Never Learned No. (%) |
| A 1935-1965 | 84 | 0 (0%) | 1 (1%) | 42 (50%) | 41 (49%) |
| B 1966-1975 | 32 | 3 (9%) | 13 (41%) | 10 (31%) | 6 (19%) |
| C 1976-1985 | 35 | 22 (63%) | 9 (26%) | 2 (6%) | 2 (6%) |
| D 1986-2005 | 15 | 12 (80%) | 1 (7%) | 1 (7%) | 1 (7%) |
| | 166 | 37 (22%) | 24 (14%) | 55 (33%) | 50 (30%) |
| Part II | | | | | |
| E 1986-1995 | 55 | 34 (62%) | 17 (31%) | 0 | 4 (7%) |
| F 1996-2005 | 63 | 46 (73%) | 13 (21%) | 0 | 8 (7%) |
| Subtotals | 118 | 80 (68%) | 30 (26%) | 0 | 8 (7%) |

proved the modification and agreed that HIC approval was not required, provided the survey be conducted voluntarily and anonymously. In the process, Frank M. Lobo, the president of the AYAM, became a co-investigator and supervised Part II of the survey. The investigators randomly selected 20 graduates from each of the last 20 graduating classes. It was recognized that including physicians from a single medical school might introduce bias into the survey, but because the nature of such bias is not known, it was assumed some random bias is inevitable and all medical schools teach medicine equally well. Randomization for Part II was achieved by choosing every fourth name from an alphabetical list of the graduates of each class until 20 names from each class had been accumulated. A letter from the investigators requesting completion of the enclosed research questionnaire, a copy of the questionnaire, and a stamped envelope addressed to the president of the AYAM was mailed to each of the selected physicians. Each was identified solely by the year of graduation and an accession number, e.g., 86-01, 05-07.

Origins of Physicians Surveyed

The physicians surveyed in Part I were encountered in the daily activities of the first author, including those who provided medical care for him and his family. They also included participants attending a monthly meeting of the Geezers, a group of Yale gastroenterologists, the Yale University emeritus professors, and a continuing medical education (CME) program about NPH conducted by Codman Inc., a medical device firm. Finally, a group of about 80 colleagues and friends of the first author who were recipients of the 2005 version of his annual New Year's greeting card were invited to participate. Each received, along with the greeting card, a questionnaire, an explanatory letter, and a stamped envelope addressed to the first author.

The patients are organized in six "decadal" groups by the year of graduation from medical school (Table 1): Part I: Group A, 1935-1965; Group B, 1966-1975, Group C, 1976-1985, Group D, 1986-2005.

Analysis of the backgrounds of the 166 respondents in Part I revealed that they had graduated from 83 medical schools, 50 of

Table 2. Origins of physicians surveyed.

| U.S. medical schools | No. of physicians |
|---------------------------------|-------------------|
| Yale University | 17 |
| Harvard University | 11 |
| Case Western Reserve University | 7 |
| Tufts University | 7 |
| University of Michigan | 6 |
| 45 Other Schools | 78 |
| Total | 126 |
| Foreign medical schools | No. of physicians |
| Great Britain (8) | 9 |
| Switzerland (3) | 4 |
| Australia (1) | 3 |
| 22 other countries (21) | 24 |
| Total (33) | 40 |

which were American and 33 foreign. Thus, 126 physicians (76 percent) were graduates of American and 40 (24 percent) of foreign medical schools in 26 countries (Table 2). These physicians had practiced almost all of the specialties and subspecialties of medicine. About half (49 percent) had graduated from medical school before NPH was described in the literature in 1965.

Analysis showed that the domestic- and foreign-educated physicians graduated from medical school at similar times, learned about NPH at similar educational levels, and similar percentages of both subgroups recognized the abbreviation "NPH." It is probable that the foreign physicians were about two years younger than the Americans, because their premedical training is about two years shorter.

The largest number of physicians who graduated from any one medical school was 17 from Yale, 11 from Harvard, seven from Tufts and Case Western Reserve, and six from the University of Michigan (Table 2). The largest number of foreign respondents from any one country (nine) had trained in Great Britain at eight different medical schools. Four graduated from three Swiss medical schools, and three from a single Australian medical school.

Three of the surveyed physicians stated that they had learned of NPH before 1965. One who attended the Codman CME program in Boston is a neurosurgeon and a son

(R. Hakim) of Salomon Hakim, the neurosurgeon who described NPH. Another is a colleague of Hakim's in Bogotá. The third was an American physician who trained at the Massachusetts General Hospital in Boston from 1960 to 1964 while Hakim was a postdoctoral fellow there in neurologic pathology. He had heard Hakim lecture on NPH at the MGH. Thus, all three of these physicians learned of NPH directly from Dr. Hakim.

RESULTS

Two hundred eighty-four physicians, 40 percent of whom had been encountered professionally and 60 percent socially, completed the questionnaire. In Part I, 166 physicians were interviewed between 2003 and 2005, and 118 physicians in Part II in 2006 (Table 2). Fifty of the 166 physicians (30 percent) were completely unaware of the existence of NPH. The large majority of this subgroup graduated from medical school before NPH was described in the literature. The remainder had heard of NPH during residency or fellowship training or while practicing medicine. The 118 physicians who comprise Part II all graduated from the YUSM. They were divided into Group E, 55 physicians who graduated from 1986 through 1995, and Group F, 63 physicians from 1996 through 2005. The physicians in

Table 3. Teaching tool by which physicians learned of NPH.

| Source of Information (Part II) | No % of Recalled Teaching Events |
|---------------------------------|----------------------------------|
| Lecture | 45 (45%) |
| Textbook | 21 (21%) |
| Ward Rounds | 17 (17%) |
| Grand Rounds | 7 (7%) |
| Medical Journals | 5 (5%) |
| Miscellaneous | 4 (4%) |
| Total | 99 |

Part II had practiced medicine in the same specialties and subspecialties as in Part I, but included several new subspecialties, such as emergency medicine, interventional radiology, organ transplantation, and medical imaging. Almost one-half (44 percent) had practiced primarily in the subspecialties of internal medicine, such as cardiology, endocrinology, hepatology, etc. Forty-one physicians, about one-fourth, had practiced neurological specialties, including 30 neurologists, seven neurosurgeons, and four neuroradiologists. Surgical subspecialties comprised about 15 percent, and the remainder practiced specialties such as dermatology, family practice, pediatrics, psychiatry, etc. There were fewer neurologically trained physicians in Part II. The physicians in Part II tended to be younger (30 to 50 years) than those in Part I (50 to 70 years).

Most physicians learn about new diseases in medical school. Among the physicians surveyed, J.F. was the first to have learned of NPH in medical school, and he graduated from an American university in June 1969. He did not remember the exact circumstances under which he learned about NPH. He recalls clearly that he had known about NPH since medical school, and his father was diagnosed with NPH in about 1990. The second and third physicians who learned about NPH in medical school graduated in 1970 from two other American medical schools and married each other. The husband (J.H.) remembered that he learned

about NPH from a textbook, which he believes was Merritt's *Neurology*. His wife (C.H.) does not recall any details about when or how she learned about NPH. They recall discussing NPH briefly when they received the Conn greeting card and questionnaire in 2005 and again in August 2007, when the first author requested follow-up information. These are the only three physicians surveyed who learned about NPH in medical school (9 percent) during the first decade after it had been described (Group B) (Table 2, Figure 2).

We added one new question to the questionnaire that asked how each physician learned about NPH, i.e., the source of information that made him or her aware of NPH ("textbook, journal article, lecture, grand rounds, other — what?"). Eighty (68 percent) answered this additional question. Eighteen physicians did not recall a specific event, and 11 did not respond to the question. Eight offered two responses, both of which are included in our analysis. Surprisingly, lectures, which are de-emphasized in the Yale system, were the single most effective teaching method. Almost half of the physicians credited lectures with making them aware of NPH (Table 3). About 20 percent credited textbooks and ward rounds. Less than 10 percent cited grand rounds. A single physician learned of NPH from a public television CME informational program sponsored by Codman Inc., which manufactures shunts to treat hydrocephalus.

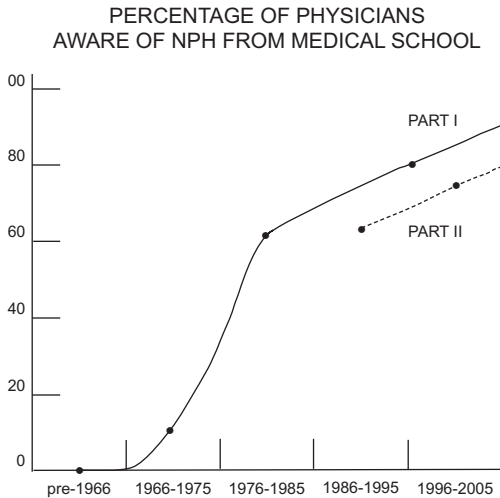


Figure 2. Learning about NPH in medical school 1935-2005. No physicians had learned of NPH in medical school before 1965, when NPH was first described, thus the percentage who learned of NPH in medical school was 0 percent. During the first decade thereafter, 9 percent learned about NPH in medical school. In the second decade, it increased to 63 percent. Although the percentage increased to 80 percent in the third and fourth decades, the slope decreased appreciably. The solid line shows the data for Part I. The dotted line shows data for Part II. Note that the curves for Part I and Part II are approximately parallel for the period from 1986 through 2005.

It is probable that some modifications in the teaching of NPH at Yale will be made, and the various medical subspecialties that encompass NPH — neurology, neurosurgery, radiology, geriatrics, and family medicine — will try to enhance the teaching of NPH. Perhaps the curriculum committee should reassess methods of teaching new, uncommon, and/or overlooked disorders.

We focused our analysis on the percentages of surveyed physicians who learned about NPH in medical school. The physicians in Group A all graduated before NPH was described, and, therefore, none could have learned about NPH in medical school. None did.

A sharp increase in the percentage of the surveyed physicians who learned about NPH in medical school occurred during the second decade after the description of NPH. Twenty-two of 35 physicians surveyed (63 percent) learned about NPH in that interval (Table 2, Figure 2). Although 80 percent of the 15 physicians in the third and fourth decades learned about NPH in medical school, the slope of the learning curve had sharply decreased and appears to have approached its asymptote at 85 to 90 percent. This surprising finding, however, is borne out in Part II, in which the recently graduated physicians in the third and fourth decades (Groups E and F) learned about NPH in medical school by only 56 percent and 70 percent, respectively (Table 2, Figure 2).

These data are shown graphically in Figure 2, where the slopes of the curves in the third and fourth decades after 1965 are parallel to that of the physicians in Part I and Part II. The slope decreased sharply during the third and fourth decades to approximately 80 percent in Part I and 70 percent in Part II (Figure 2, Table 2). The explanation for the decrease is not known.

Comparison of Neurologically Trained and Medically Trained Physicians

Initial examination of the data in Part I suggested that the neurologically trained physicians appeared to know more about NPH than the rest of the surveyed physicians. To determine if this impression was correct, 34 physicians in the neurologically trained group, which included neurologists, neurosurgeons, and neuroradiologists, were compared with 32 physicians in the “medical” group, which included gastroenterologists, hepatologists, general internists, cardiologists, endocrinologists, and miscellaneous subspecialists. These two subgroups were similar in the distribution of the dates of graduation (Groups B, C, D) and in the percentages of graduates from American and foreign medical schools (75 percent vs. 69 percent, respectively).

Ninety-seven percent of the “neurologic” group recognized the abbreviation “NPH” compared to 75 percent of the “medical” group. Every one of the neurologic

group (100 percent) was aware of NPH as compared to 88 percent of the medically trained internists. This difference is probably statistically significant. The group of non-internists, i.e., surgeons, anesthesiologists, and radiologists, all had higher percentages of physicians who were unaware of NPH. For example, 12 of 17 surgeons surveyed (70 percent) were completely unaware of NPH. It is not understood why these less common specialties seem to be especially unaware of NPH.

The neurologically trained physicians learned about NPH significantly earlier in their careers than the medically trained physicians, with a mean of 4.2 vs. 8.0 years, respectively, after graduation from medical school ($p < 0.01$).

Ninety-seven percent of the 34 neurologically trained physicians had observed patients with NPH in their practices. They ranged from having seen 1 to 250 per five years with a mean of 25. In comparison, only one medically trained physician had seen more than five NPH patients per five years, and the mean was 1.1 in the medically trained subspecialists ($p < 0.001$). This difference is probably the result of the referral of patients to neurosurgeons and neurologists.

The answers to the question "Is it ever too late to shunt?" are not analyzed. Fourteen percent said they didn't know or failed to answer the question. The remainder were evenly divided between "yes" and "no." We did not assess this question because it was ambiguously worded. To answer the question correctly would require that the physician to respond, in effect, "No, it's never too late." We believe that this double negative response may have confused some of the physicians, and, therefore, we excluded it from the analysis.

The Yale System

Three score and 17 years ago, our forefathers brought forth at this medical school a new curriculum, conceived in the maturity of its students and dedicated to the proposition that medical students are self-motivated adults who should be largely responsible for their own education. The curriculum, which

at that time was unique and is the model for a number of other such innovative curricula, is widely known as the Yale System (YS) [9]. Yale medical students are pre-selected, self-reliant individuals who are strongly motivated to become physicians. The selection process tends to accept students who already have demonstrated expertise in other areas such as science, literature, the arts, sports, etc. Presumably, such students do not require academic competition, such as grades, class ranking, or compulsive examinations, to achieve their goals. The students are free to use their time as they see fit. The faculty is to provide guidance.

In the Yale System, the acquisition of scientific principles and methodology are thought to be much more important than facts learned by memory. During the preclinical years, the students are expected to acquire biologic and biochemical knowledge and develop clinical skills. Attendance at basic science courses is expected, but is not recorded. Lectures are held to a minimum. This lecture policy is the only component of the YS with which the authors of this survey do not enthusiastically agree. We believe good lecturers should be nurtured as well and may be most effectively used to help students synthesize complex concepts and bring new, unusual, uncommon, or otherwise overlooked illnesses, such as NPH, to their attention.

At Yale, no examinations are administered except as anonymous exercises used as tools for self-evaluation by the students or faculty to assess their teaching techniques. The National Board of Medical Examination is considered to be a legal requirement for graduation rather than a component of the YS. Yale students almost invariably pass. The most outstanding aspect of the YS is the requirement that each student design and conduct a thesis project under the guidance of a faculty member. The project is in every way the equivalent of a Ph.D. thesis. Its purpose is to teach each student how to design, carry out, evaluate, and describe such investigations for publication. The experience of performing such a project in its entirety teaches students at the very least how to read scientific articles properly. Approximately two

thirds of the student theses are published in peer-reviewed journals each year. When Yale graduates are interviewed, they agree overwhelmingly that the thesis project is the single most difficult and rewarding part of the YS.

Is there any reason to think the YS can be responsible for either the relatively low rate of learning of NPH (93 percent) observed in recent YUSM graduates (Part II)? We find no reason to think so. We have considered the possibility that the de-emphasis of lectures might have conceivably contributed, but the recent Yale graduates in Part II reported that lectures were the single most effective teaching technique in making students aware of NPH, accounting for 45 percent (Table 3). We do not know whether a 7 percent rate of not learning about NPH in our survey is abnormally high. Conceivably, it may be normal. There are no data available with which our findings can be compared.

DISCUSSION

It seems clear to us that the primary explanation for the lack of awareness of physicians of NPH is the relative newness of this syndrome. Although 40 years seems like a long time, it may not be long enough to have a syndrome like NPH incorporated into the forefront of the diagnostic thought processes of several generations of practicing physicians. Our data indicate that awareness of NPH has increased markedly since the recognition of this disorder in 1965. However, the rate in the increase of awareness of NPH appears to have sharply decreased between the second and third decades after NPH was described. This decrease is shown dramatically in Figure 2, which depicts the percentage of physicians who learned about NPH during medical school. We interpret these data to mean that the learning curve peaked 20 to 25 years after NPH was introduced. The curve appears to have approached its asymptote (about 85 percent by 2005). We anticipated that the asymptote would be at a higher rate, e.g., 90 or 95 percent. Part II suggests that the asymptote might be slightly lower for graduates of Yale

than for other medical schools. (We are considering performing similar studies to determine the degree of awareness of other recent diseases.) Presumably, every disease has its own learning curve. We believe that the learning curve for AIDS, for example, would be much steeper, reaching an estimated asymptote of about 99 percent after about 10 years.

Perhaps peaks of 85 to 90 percent 20 to 35 years after introduction are all that can be expected for new disorders, certainly for uncommon diseases of elderly people like NPH. One could never expect 100 percent for any illness. Would 95 percent be acceptable? Ninety percent? We realize that every physician cannot be expected to know everything about every disease.

We also noted in this survey that the degree of awareness appeared to be lower for physicians in the less common medical specialties such as anesthesiology, oncology, pathology, and psychiatry, but the data are too few to be statistically significant. Clearly, however, awareness of NPH is highest in physicians who had been taught by neurologists, neurosurgeons, etc. The quality of the teaching is a key element.

Our data also suggest that classroom lectures appear to have been the most common means of making students aware of NPH. This finding reinforces the importance of the lecture format in medical education. Conceivably, the Yale curriculum can be modified to improve this deficiency.

Other factors must be considered. NPH is a complex, unique disease that may present in a surprisingly large number of ways. Although the classic case has three clinical components, it usually starts with one and progresses. Occasionally, it only displays two and, rarely, one of the classic three. Each of the components may present at unpredictable times in a continuum of severity in all possible combinations. Thus, patients with NPH may present in an almost infinite number of ways. Furthermore, there are many different senile gaits that are not easily differentiable. The incontinence may be urinary, fecal, or both, or may present as urinary urgency, rather than as overt incontinence.

Fecal incontinence may occur in NPH only in the presence of diarrhea. Dementia appears in many guises, ranging from mild recent memory loss to overt psychosis of a variety of types.

Moreover, like fingerprints, no two cerebral ventricular systems are identical. The degree of enlargement of the lateral ventricles varies greatly, and it is not clear whether the variable shapes of the ventricles can stretch or compress nerve tracts in discernible, predictable patterns, i.e., do symptoms vary with infinite variations in the shape and size of the ventricles? Is the appearance of diabetes mellitus associated with enlargement of the third and fourth ventricles as Hakim implied [1]? Although NPH is not a rare disease, it is uncommon, so that few physicians ever get to see many such patients. On the other hand, we believe it is far more common than the three to six per 100,000 shunt implantations per year that Swedish studies infer [10,11]. Only a few properly performed prevalence studies have been published. We know of only two such studies, both done in rural areas of Europe [12,13]. Both show that NPH appears to be present in one in 200 to one in 250 subjects 65 years of age, which is surprisingly high.

Furthermore, other disorders of elderly people such as Alzheimer's disease, Parkinson's disease, and cerebral atrophy may show enlarged ventricles, and demential disorders may be difficult to differentiate from each other. As a patient with NPH who had erroneously been thought by a competent neurologist to have one of these other disorders for almost a decade [14], I know this differential diagnosis is very difficult.

Moreover, the criteria to define cerebral ventriculomegaly precisely are vague and difficult to establish, and enlarged ventricles are surprisingly common. Many neuroradiologists are reluctant to report borderline or even moderate degrees of hydrocephalus because such diagnoses raise complex, medical, and socioeconomic issues and inadvertently may lead to the performance of brain surgery in less than optimal surgical candidates. Although shunt insertion is not

complex surgery and has low morbidity and mortality, it is not free of risk.

Finally, we believe strongly that elderly patients with these symptoms and their loved ones should have the right to make an informed decision as to whether they want shunt surgery, whatever the risk. The first author, having experienced end-stage NPH and having been essentially abandoned to his fate, did not feel that there was much to lose by choosing surgery. Happily, his dementia was subtle and his value center intact, which made the decision easy for him. Even though the decision may be more difficult for naive patients, they should be given the opportunity to make it after an unbiased presentation of the risks and benefits.

Acknowledgment: We are grateful to Kenny Marone, Jan Glover, and the staff of the John Hay Whitney Medical Library at Yale for their sustained efforts on our behalf in performing the endless literature searches that this project generated. We appreciate the assistance of Mary Meehan and Jean Peck of the AYAM who helped randomize and correspond anonymously with the 400 Yale medical school graduates in Part II. Finally, we thank Irene Cowern for her meticulous secretarial work and Marilyn Conn for helping us all work on the same page at the same time.

REFERENCES

1. Hakim S. Some observations on CSF pressure. Hydrocephalus syndrome in adults with 'normal' CSF pressure (Recognition of a new syndrome) [thesis]. Bogotá, Colombia, SA: Javeriana University School of Medicine; 1964. pp. 1-40.
2. Hakim S, Adams RD. The special clinical problem of symptomatic hydrocephalus with normal cerebrospinal fluid pressure. Observations on cerebrospinal fluid hydrodynamics. *J Neurol Sci.* 1965;2:307-29.
3. Adams RD, Fisher CM, Hakim S, et al. Symptomatic occult hydrocephalus with 'normal' cerebrospinal fluid pressure. A treatable syndrome. *N Engl J Med.* 1965;273:117-26.
4. Koenigsberg RA, Faro SH, Hershey BL, et al. Neuroimaging. In: *Textbook of Clinical Neurology*. 2nd ed. Philadelphia: Saunders. 2003. pp. 429-70.
5. Lee PH, Yong SW, Ahn YH, Huh K. Correlation of midbrain diameter and gait disorder in patients with idiopathic normal pressure hydrocephalus. *J Neurol.* 2005;252:958-63.
6. Mocco J, Tomey MI, Komotar RJ, et al. Ventriculoperitoneal shunting of idiopathic nor-

- mal pressure hydrocephalus with increased midbrain size. *Neurosurg.* 2006;59:851-4.
7. Hebb SDH, Cusimano MD. Idiopathic normal pressure hydrocephalus: a systemic review of diagnosis and outcome. *Neurosurg.* 2001;49:1166-86.
 8. Bret P, Guyotat J, Chazal J. Is normal pressure hydrocephalus a valid concept in 2002? A reappraisal in five questions and proposal for a new designation of the syndrome as 'chronic hydrocephalus.' *J Neurol Neurosurg Psychiatry.* 2002;73:9-12.
 9. Viseltear AJ. The Yale plan of medical education: the early years. *Yale J Biol Med.* 1986;59:627-48.
 10. Höglund M, Tisell M, Wikkelsø C. Incidence of surgery for hydrocephalus in adults surveyed: same number afflicted by hydrocephalus as by multiple sclerosis [Swedish]. *Lakartidningen.* 2001;98:1681-5.
 11. Tisell M, Höglund M, Wikkelsø C. National and regional incidence of surgery for adult hydrocephalus in Sweden. *Acta Neurol Scand.* 2005;112:72-5.
 12. Casmiro M, Benassi G, Cacciatore FM, et al. Frequency of idiopathic normal pressure hydrocephalus. *Arch Neurol.* 1989;48:101.
 13. Trenkwalder C, Schwarz J, Gebhard J, et al. Starnberg trial on epidemiology of Parkinsonism and hypertension in the elderly. Prevalence of Parkinson's disease and related disorders assessed by a door-to-door survey of inhabitants older than 65 years. *Arch Neurol.* 1995;52:1017-22.
 14. Conn HO. Normal pressure hydrocephalus: a case report by a physician who is the patient. *Clin Med.* 2007;7:296-9.