

Salomón Hakim: the man behind normal pressure hydrocephalus

Salomón Hakim: o homem por trás da hidrocefalia de pressão normal

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ABSTRACT

The illustrious Colombian Professor Salomón Hakim provided the annals of neurology with one of the most brilliant and original bodies of research on record, developing the concept of normal pressure hydrocephalus, as well as proving that ventricular shunting is an effective treatment. Thus, Professor Hakim proved that some of the dementias, at that time considered senile, could be successfully treated. Here the authors present an historical review of his main contributions, which continue to influence the study of dementia to this day.

Keywords: Salomón Hakim; hydrocephalus, normal pressure; ventriculoperitoneal shunt.

RESUMO

O ilustre professor colombiano Salomón Hakim deixou como legado nos anais da neurologia uma das mais brilhantes e originais séries de pesquisa da história, desenvolvendo o conceito de hidrocefalia de pressão normal, bem como introduzindo a derivação ventricular como tratamento efetivo. Assim, Hakim provou que algumas das demências até então consideradas senis tinham possibilidade de tratamento bem-sucedido. Aqui os autores apresentarão uma revisão histórica de suas maiores contribuições, que continuam a influenciar o estudo de demências até os nossos dias.

Palavras-chave: Salomón Hakim; hidrocefalia de pressão normal; derivação ventriculoperitoneal.

Normal pressure hydrocephalus (NPH) is considered an insidious and progressive onset syndrome characterized by the classic triad of gait disturbance, urinary incontinence, and dementia. In NPH patients, the ventricular system has been shown to be enlarged on neuroimaging; however, the pressure of the cerebrospinal fluid (CSF) is within normal limits, with ventricular dilatation not attributable to cerebral atrophy; hence the name of the disease. The prevalence of NPH has been established at around 21.9 per 100,000 individuals¹. Some authors prefer the term chronic hydrocephalus in the adult, considering that in its early stages, intermittent increases in intracranial pressure can be detected with continuous monitoring².

Despite the controversies regarding the nomenclature, the authors will use the name coined by the Colombian neurosurgeon Salomón Hakim Dow (1922–2011) and pay homage to this great physician, who provided a body of work that continues to influence neurology.

Early life








Born to a family of Lebanese immigrants in 1922, Hakim enrolled in medical school in 1944 and, after graduation, became a neurosurgeon. Meanwhile, Hakim developed a passion for physics, performing side experiments on electricity. Soon after completing his studies in Bogotá, Hakim left for the United States and accepted a fellowship

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offer in Boston in 1950. In 1954, Hakim returned to Boston for another fellowship, this time at the Massachusetts General Hospital. However, as noted by Wallenstein in his seminal review, Hakim's great breakthrough, took place back in his homeland³.

Hakim and normal pressure hydrocephalus

In 1957, while working at the San Juan de Dios Hospital in Colombia, Hakim first faced the entity he originally named symptomatic occult hydrocephalus. A 16-year-old comatose male patient, who had suffered severe cranio-encephalic trauma in an automotive accident, developed a subdural hematoma that needed to be drained. After the surgery, there was no discernible clinical improvement, which led to a pneumoencephalography (Figure 1). To Hakim's surprise, the examination showed ventriculomegaly even though the CSF pressure was normal (150 mmH₂O). The patient improved after collecting 15 ml of CSF, being able to speak the following day. This led to repeated lumbar punctures and the placement of a ventriculoatrial shunt, followed by a steady improvement in the patient's condition in the following months, until he recovered completely, returning to school within three. This case was first reported in a thesis published in 1964⁴ and in the *Journal of Neurological Sciences* in 1965⁵.

The search for an explanation for this surprising phenomenon led Hakim to return to his physics notes; he remembered Pascal's law, according to which pressure remains constant in any closed system, thus amplifying the force imposed on a small column of fluid to another, larger column. Elegantly summed up by the formula $F = P \times A$ (F being force, P being pressure, and A being area), Pascal's law implied that even though the intracranial pressure may be normal in a patient with enlarged ventricles, the increased area of the ventricular surface leads to an abnormally large force upon the same surface – the aptly-named (by Hakim himself) hydraulic press effect^{5,6} (Figure 2).

This theory was met with skepticism by the medical establishment of the time, particularly among his former

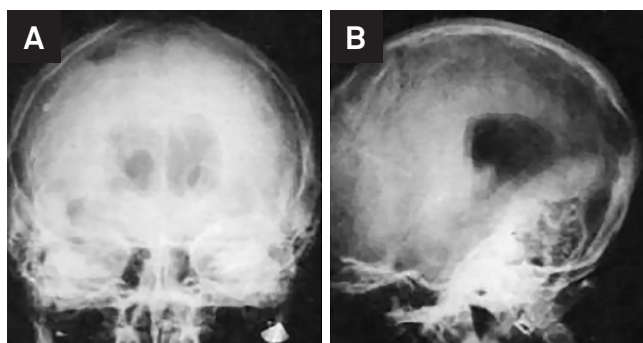


Figure 1. Dilated ventricles in the pneumoencephalogram of Hakim's original patient.

colleagues in Boston. However, after an American official stationed in Colombia with a similar condition was flown to the US accompanied by Hakim, the neurosurgeon insisted on the procedure. The patient's improvement led to yet another publication on the *New England Journal of Medicine*, co-authored by Adams and Fisher⁷ among others, which summed up the essential features of the condition, leading to a wider acknowledgment of its features, and ultimately becoming known, at least initially, as Hakim-Adams syndrome.

The path was not without opposition, with perhaps the most vocal critic being Professor Merritt, from the Columbia University College of Physicians and Surgeons. Ironically, Merritt eventually developed symptoms compatible with NPH and passed away due to complications following shunt placement surgery³.

Other endeavors

Over the course of his life, Hakim delivered lectures in over 30 countries and received numerous honors and awards. He also became a professor in his own alma-mater, the Universidad Nacional de Colombia. Hakim was a prolific inventor and made seminal advancements in the creation of various valve models. The most notable of these was developed in 1966: a unidirectional valve with spring-loaded pressure control, which set the standard for all future valves for treatment of the entity Hakim first described, a legacy continued by his son, Carlos Hakim³.

In conclusion, although he passed away from a hemorrhagic stroke in 2011, Hakim's embattled defense of the pathology of NPH led to the diagnosis and treatment of thousands of cases of this prevalent entity, within a context that still deems dementia incurable in most cases. As his seminal article reads, "Recognition and treatment of these cases is of great importance since it will result in what amounts to a 'cure' of a clinical condition that closely resembles presenile or senile dementia"⁷.

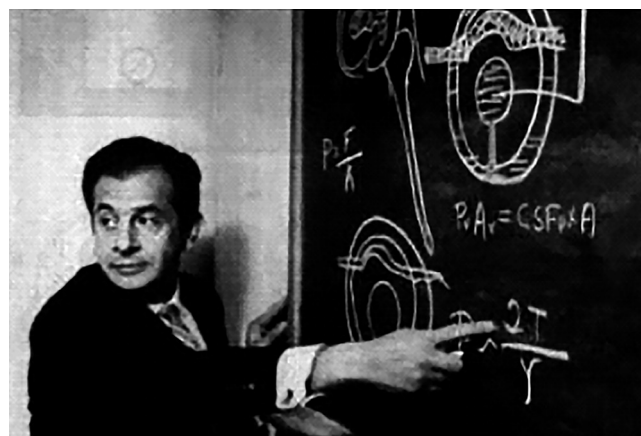


Figure 2. Salomón Hakim and Pascal's law.

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