Endoscopic third ventriculostomy versus Ventriculoperitoneal shunt in patients with idiopathic normal pressure hydrocephalus.

A systematic review

A dissertation submitted for partial fulfillment of the conditions for the Master Degree in Neurosurgery.

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2016

Acknowledgements

In the beginning, I am very grateful to GOD ALMIGHTY for without His graces and blessings this review would not have been possible.

Immeasurable appreciation and deepest gratitude are extended to my advisor ***Prof.Dr. Khaled El-Bahy*** for his continuous support, his patience, motivation, and immense knowledge. His guidance helped me in all the time of research and writing of this review. I couldn't have imagined having a better advisor and mentor for me.

Beside my advisor, I would like to thank ***Asst.Prof.Dr. Ahmed Faisal Toubar*** for his insightful comments and encouragement, without his passionate participation and input, this review couldn't have been successfully conducted.

My sincere gratitude for ***Asst.Prof.Dr. Mohamed Nosseir*** for his unconditional support, absolute patience, continuous education and guidance which helped me in writing this review.

بسم الله الرحمن الرحيم

وقل رب زدني علما

صدق الله العظيم

(سورة طه - الآية ١١٤)

Table of Contents

List of Figures

List of Tables

List of abbreviations

Abbreviation	Full meaning
AC-PC Plane	Anterior Commissure- Posterior Commissure Plane
CBF	Cerebral Blood Flow
CNS	Central Nervous System
CSF	Cerebrospinal fluid
CSF-OP	Cerebrospinal fluid opening pressure
CT	Computed tomography
DESH	Disproportionate Enlarged Subarachnoid space Hydrocephalus
DM	Diabetes mellitus
ELD	External Lumbar Drain
EMG	Electromyography
ETV	Endoscopic third ventriculostomy
HTN	Hypertension
ICP	Intracranial pressure
INPH	Idiopathic Normal Pressure Hydrocephalus
ISF	Interstitial fluid
KS	Kiefer Scale
LP	Lumbar puncture
MMSE	Mini Mental status Examination
MRI	Magnetic resonance imaging
NPH	Normal Pressure Hydrocephalus
NPHRR	NPH Recovery Rate
RCT’s	Randomized controlled trials
SRINPH	Shunt Responsive Idiopathic Normal Pressure Hydrocephalus
VP shunt	Ventriculoperitoneal shunt

INTRODUCTION

I. Rationale and justification of the study

Idiopathic normal pressure hydrocephalus (INPH) is an adult onset syndrome of uncertain origin involving non-obstructive enlargement of the cerebral ventricles. In the absence of papilledema and with normal cerebrospinal fluid (CSF) opening pressure on lumbar puncture.(1)

Hakim and Adams first described the syndrome of NPH in 1965 as a syndrome characterized by a clinical triad of progressive gait disturbance, dementia and urinary incontinence. These symptoms vary in severity and appearance. Gait impairment is the most common clinical feature in INPH, with a frequency ranging from 80% to 100% and it is often the patient's initial complaint. The second most frequent symptom is cognitive impairment, which ranges from 42% to 100%. Urinary incontinence ranges from 34% to 82%, where full clinical triad is present in 38-82% of cases.(2)

The pathophysiology of INPH remains unknown, and the assessment of these patients is still debated, however suggested mechanisms include reduction of blood flow leading to reduced periventricular metabolism and axonal degeneration without significant cortical damage and stretching of the periventricular white matter.(3, 4)

Diagnosis of INPH can be achieved through detailed history, physical examination, and neuroimaging which is considered as an obligatory part in the evaluation of suspected INPH to document ventricular enlargement, to rule out macroscopic obstruction to CSF flow and other pathology. Magnetic resonance imaging (MRI) is considered the method of choice due to the vastness of information it provides, but computed tomography (CT) is an acceptable alternative. Prognostic tests like the CSF tap test, the lumbar infusion test and intracranial pressure (ICP) monitoring have made it easier to identify the patients who will most likely benefit from surgery. (1, 5)

No rule for conservative treatment in management of INPH. Surgical treatment is mandatory because it has been associated with a positive impact on the course of the disease, in terms of the quality of life of patients and caregivers.(6)

The treatment for INPH is surgical diversion of CSF where ventriculoperitoneal shunt (VP shunt) is the most commonly used method but more recent studies have suggested a positive effect of endoscopic third ventriculostomy (ETV).[5, 6]

The purpose of this review was to provide an overview of the current literature investigating the treatment and outcome in INPH patients.

II. Aim

To review and summarize available knowledge on the rule of endoscopic third ventriculostomy versus ventriculoperitoneal shunt in the management of patients with idiopathic normal pressure hydrocephalus.

III. Objectives

To compare the efficacy and effectiveness of VP shunt and ETV in management of INPH as regards improvement of clinical picture as a primary outcome and preservation of quality of life in terms of morbidity and mortality rates as a secondary outcome.

REVIEW OF LITERATURE

History of normal pressure hydrocephalus

Normal pressure hydrocephalus was first described by Salomón Hakim in his degree thesis-*Some Observations on C.S.F. Pressure: Hydrocephalic Syndrome in Adults with "Normal" C.S.F. Pressure* (Thesis No. 957, Javeriana University School of Medicine, Bogotá, Colombia, March 10, 1964).[7]

The first case was described in 1957 for a 16 years old boy with severe head injury after a motor car accident operated on for a subdural hematoma, and surgery was considered to be successful. However the patient remained in an impaired level of consciousness. As a diagnostic procedure, pneumoencephalography was performed and revealed ventricular dilatation. The pressure readings taken at that time displayed rather normal intracranial pressure.[8]

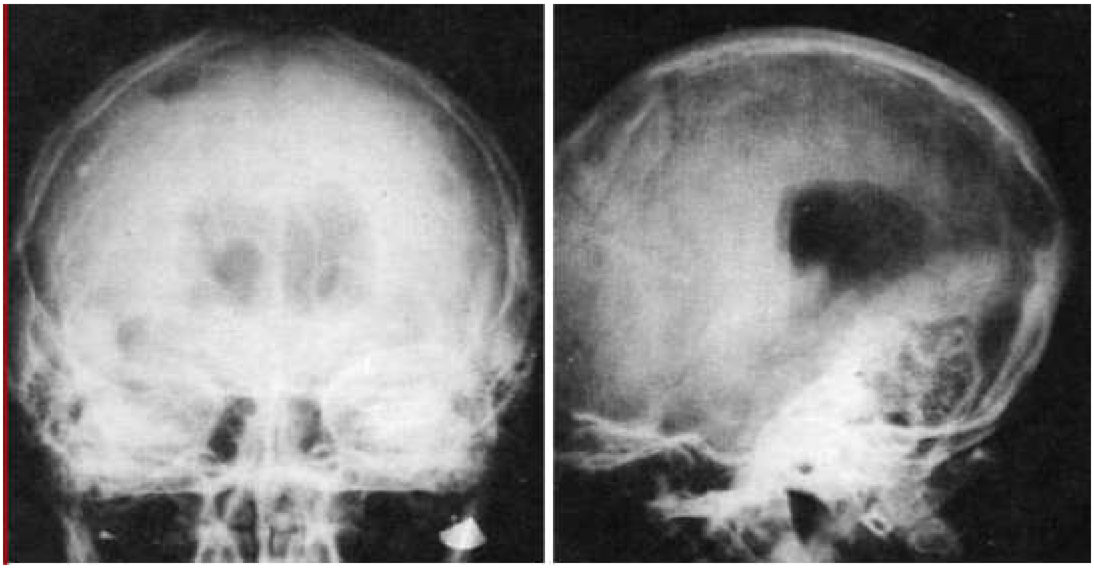

Figure 1: A pneumoencephalogram of Hakim's first normal-pressure hydrocephalus patient.[8]

Hakim removed 15 mL CSF for further laboratory investigation. After the CSF removal, the patient's level of consciousness improved the next day. His alertness subsequently declined over the following days and then improved again after a second lumbar puncture. Hakim decided to implant a ventricular atrial shunt. The patient improved significantly and the treatment was long lasting. The case was presented in Hakim's doctoral thesis in 1964.[8]

After Hakim's publication in 1965, the spinal tap test became the standard diagnostic test. Further tools to predict shunt responsiveness include lumbar spinal drainage, and intracranial pressure monitoring.[9]

In 1965, the syndrome was introduced by Adam et al. and Hakim et al. in an article published in *New England Journal of Medicine* were three case reports were described, one idiopathic and two post-traumatic. The syndrome was described as a triad of gait disturbance, dementia, and urinary incontinence associated with patients in whom ventricular enlargement occurred in the absence of elevated intracranial pressure.[10]

History of endoscopic third ventriculostomy

This first ETV was performed in 1923 by William J. Mixter in a 9 month old hydrocephalic patient. Utilizing a urethroscope, he was able to pass through the foramen of Monro, visualize the third ventricle and cerebral aqueduct, and make a hole in the floor of the third ventricle connecting it to the interpeduncular cistern. The next ETV did not appear in the literature until 1935, when Scharff reported his initial results using an endoscope. Scharff described several modifications including an irrigation system that kept the ventricles open, a mobile cautery tip, and a moveable operating tip that could perforate the floor of the third ventricle. However, the long term results were not rewarding yet and the morbidity and mortality rate not accepted, were poorly designed instruments and optical apparatus were the main causes of the disappointing outcomes.[11, 12]

The procedure disappeared till the development and improvement of fibro optic cables that occurred between 1950s to 1970s. In 1978, Vries et al. used fiber optic endoscope inserted through a small burr hole over the coronal suture. The technique has been successfully employed in five patients without complications. In 1990, Jones et al. reported successful outcome with ETV with low morbidity and no mortality. His work became a milestone for the indications and the evaluation of the results after ETV. [11, 12]

Neuroanatomy of the ventricular system

I. Lateral ventricle

Each lateral ventricle is a C-shaped cavity that winds around the thalamus and is situated deep within the cerebrum.

Each lateral ventricle has five parts:

- Frontal horn.
- Body.
- Atrium.
- Temporal horn.
- Occipital horn.

Each of these five parts has a roof, and a floor, medial and lateral walls. In addition, the frontal and temporal horns and the atrium have anterior walls.[13]

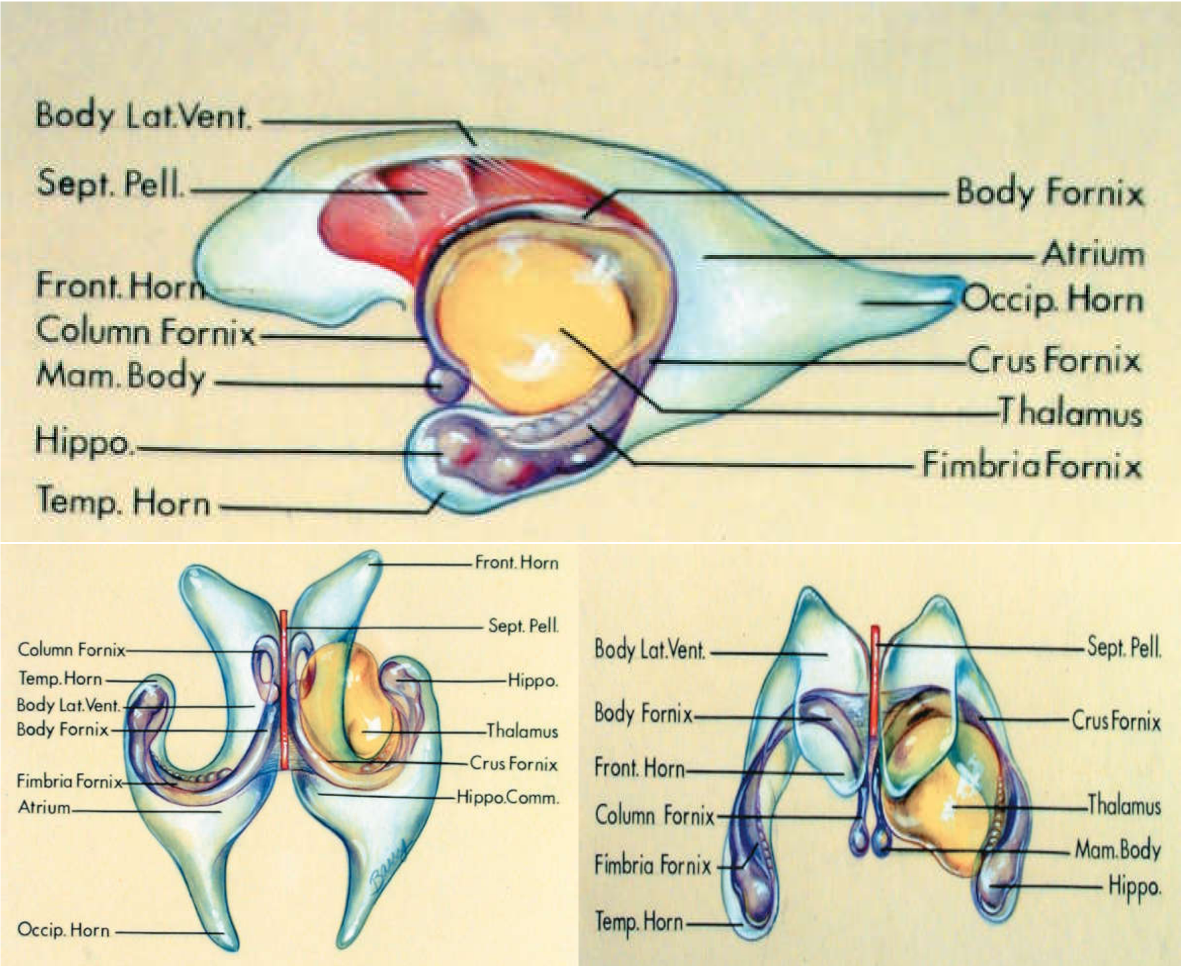

Figure 2: Lateral ventricle shape, parts and relation to surrounding structure.[13]