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Pediatric Hydrocephalus

Second Edition

Springer

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Editors

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Second Edition

With 956 Figures and 104 Tables

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Feci quod potui, faciant meliora potentes.
(Anonymous, Latin)
(I did what I could, let those who can do better.)

To Fabrizia, so much of you in this book...
Every moment spent to do it or learn the
things in it has been stolen from you.
To Francesco and Maria Allegra, for all the love and happiness you give to me, for being so beautiful, proud, and perfect in everything you do.
To my wonderful, incredible mother, who after so many years is still asking for a copy of the book to read.

GC

To my wife Eren and daughter Ceren

MÖ

To Federica, Elise, Georges, and Balthazar

CSR

Introduction

> Quidquid autem spatii est inter vaginam durae matris et medullam spinalem, id omne *plenum* etiam semper est: non medulla quidem ipsa in viventibus turgidiori, non nube vaporosa; quod in re adhuc obscura, suspicantur summi Viri; sed *aqua*, ei quidem simili. . ..
>
> Domenico Cotugno, *De Ischiade Nervosa Commentarii*, 1764
>
> ("Whatever the space between the dural sheath and the spinal cord, it is always *full*: It is not filled with the spinal cord that would be bigger in living person, or with an aeriform vapor; as strangely suspected by Very Famous Men; but *water*, or something similar. . ..")

Cerebrospinal fluid was first discovered and fully described in 1764 in Naples (at that time capital of the Kingdom of Naples) by Domenico Cotugno, a doctor working at the Ospedale degli Incurabili. Although the existence of fluid inside the brain and around the spinal cord had been incidentally described by several authors before, Cotugno was the first one to clearly understand that the spinal cord was surrounded by fluid, that the same fluid was also present inside the brain and surrounded the brain itself and for the first time understood and described the existence of cerebrospinal fluid (CSF) circulation. He was well aware of the importance of his finding: he modified the technique of autopsy to prove the existence of this fluid and calculate its amount with precision (see chapter ▶ "Cerebrospinal Fluid Circulation"), and the *corsivo* (italics) of the words *plenum* (full) and *aqua* (water) are present in the original edition of his book *De Ischiade Nervosa Commentarii*; that, by the way, is the first book to clearly classify the clinical findings of different distribution of radicular pain observed in lombosciatalgia. After his publication, the cerebrospinal fluid was called for a long time *Liquor Cotunnii* (Cotugno Fluid) and the Latin word *liquor* is still nowadays the medical and scientific term in Italy to indicate the cerebrospinal fluid.

The Ospedale degli Incurabili, founded between 1520 and 1522, has also been the earliest meeting point of the Council of Governors (Governo) of the Pio Monte della Misericordia, a nonreligious charity foundation of Catholic inspiration founded in 1602, still existing and very active in Naples today. The building and its associated churches work as a hospital, although as a very small one compared to more modern architecture. It remains especially famous for its fundamental role in the history of Italian medicine, for the presence of a museum of medicine and above all for the magnificent *Farmacia degli Incurabili*, a large room fully decorated with maiolica jars of drugs and herbal remedies and artistically structured according with Barocco-Rococò style following very suggestive masonry symbology.

Preface to Second Edition

This book is somehow a second edition. The first book was published in 2005, after a long gestation, and came in a very precise moment, at the end of the first phase of "enthusiasm" for the neuroendoscopic techniques in high-income countries and in the very first moments of burst of neuroendoscopy in low-income countries thanks to the revitalization of old techniques. The sudden awareness that thousands of hydrocephalic babies throughout the world could be treated in an effective and economically affordable way without shunt implant acted as a second injection of enthusiasm in pediatric neurosurgeons for neuroendoscopic techniques and, even more important, another wave of interest toward hydrocephalus, its pathophysiology, and its treatment. The first edition in that special moment, when considered retrospectively, looked especially incomplete and insufficient. Attention to low-income countries was only limited to a couple of chapters; very important subjects were missing completely (craniosynostosis, venous hypertension, arachnoid cysts, prenatal hydrocephalus, and others); and other CSF circulation disturbances like pseudotumor and syringomyelia were insufficiently treated. Finally, the number of pages and color figures was limited due to budget restrictions. Nevertheless, Springer accepted to publish it (and we are still grateful to Donatella Rizza from Springer Italy for that), and the book became a Springer best seller for 2005–2006, to witness the everlasting interest among pediatric neurosurgeons for the pathology they more frequently deal with.

This second edition has many advantages over that book. We understood quickly our mistakes and, like elephants, kept them hidden in memory during many years. Wirginia Maixner had to abandon the project due to her busy schedule in spite of our insistence to remain. We are very grateful to her for the major help in the first edition and for spontaneously denying authorship when she realized she could not fulfill her editor tasks. This is so special and rare nowadays that she would have deserved to remain among editors only for that. The advent of e-book revolution and the creation of the "Major Reference Works" category in Springer library allowed for the creation of a book that was able to fulfill most of our needs in terms of the number of pages, length and number of chapters, color figures, and number of references. This time, we had no excuses. In addition to this, we must consider as well the extraordinary enthusiasm of all the expert colleagues all over the world to whom we asked for a contribution. The percentage of acceptance to write the 71 chapters of the book was 100%. In only a couple of cases, we were obliged to change the authors of a chapter at a later stage due to impossibility of the original author to conclude in time. We are extraordinarily grateful to those few authors who accepted the very hard task to write a chapter in a much shorter time than their colleagues, especially for the incredibly high scientific level they were able to produce. This was for us the final confirmation that smart people work incredibly well even under pressure.

The final result is under our eyes. Well far from perfection, this book certainly represents a useful update to the first edition and partially fills the strange void on the publications on pediatric hydrocephalus, which still remains the most frequently treated condition by pediatric neurosurgeons and the most frequent disease of the pediatric central nervous system throughout the world, whatever the etiology. We tried to rationalize as much as possible the incredible variety of etiologies, pathophysiological mechanisms, clinical features, and treatment options of this condition that complicates an incredible number of diseases of the central nervous system, remaining for this reason extremely common and still highly enigmatic under many aspects.

We are wholeheartedly grateful to all the world's leading experts who contributed to this book. They did it; we just tried to give an arbitrary order to their knowledge. We remember the friends who contributed to the first edition and were not there anymore to help us with the second one: Jean Aicardi, the giant who had the first idea, Patrick Hanlo who was a great scientist and a fantastic friend, and Enrico Leone, fantastic example of how making neuroradiology with heart and culture.

Finally, we thank Veronika Mang, Shruti Datt, Jayasri Jayakrishnan, and the whole staff of Springer Nature for their patience, support, and professionalism in following this project.

Naples, Italy
Istanbul, Turkey
Fort-de-France, Martinique
December 2018

Giuseppe Cinalli
M. Memet Özek
Christian Sainte-Rose

Preface to First Edition

This book was conceived as a result of the weekly staff meetings and daily clinical rounds at the Department of Neurosurgery, Hôpital Necker-Enfants Malades in Paris. The understanding and management of pediatric hydrocephalus has been part of our routine work for years. As so often happens to those who are thus preoccupied, the first proposition for the book came not from the neurosurgical unit, but from our colleagues, in this instance Jean Aicardi in 1994. He understood that it was a period of "revolution" for pediatric hydrocephalus particularly as it pertained to neuroendoscopy. He therefore petitioned Christian Sainte-Rose to write a book on pediatric hydrocephalus addressed to pediatric neurosurgeons, pediatric neurologists, and pediatricians.

The gestational phase was long, but serendipitous for the project. This phase witnessed the publication of the final results of the shunt trial (1998 and 2000) and the evolution and definition of neuroendoscopy. In 1999, the time finally became ripe to look back and try to define the real impact of these two events in the treatment of pediatric hydrocephalus. Thus, the book was born.

We are deeply indebted to Springer-Verlag Italia, who agreed to publish the book, and we are particularly grateful to Dr. Donatella Rizza and to the whole editorial team for their patience, their professionalism and for the quality of their work.

It is necessary also to thank in this page all the people who have directly or indirectly contributed to the ideas that are included within – to all the permanent and temporary neurosurgical staff at Necker-Enfants Malades, the residents, the chefs de clinique, the students, and the consultants whose questions, criticisms, and comments have molded our thoughts. We are and always will be grateful to Jean-Franjçois Hirsch for his curiosity, his teaching, and his support. We thank our nurses for their care of our patients. Most of all, we thank our little patients, all of them, for bearing the consequences of our decisions.

January 14, 2004

Giuseppe Cinalli
Wirginia Maixner
Christian Sainte-Rose

Foreword

I am honored to have this opportunity to write a foreword for this outstanding text on hydrocephalus. As a pediatric neurosurgeon, I have been involved with hydrocephalus since the first days of my neurosurgical training. Hydrocephalus interacts with much of what we do in neurosurgery: trauma, intracranial hemorrhage, tumors, developmental disorders, spasticity, epilepsy, and infection. In almost every area of subspecialty, we deal with this disease.

Hydrocephalus as a disease has been recognized since the time of Hippocrates. As organized neurosurgery began in the early part of the twentieth century, hydrocephalus was a major component of neurosurgical disease and complicated attempts at treatment.

Walter Dandy attempted to remove the choroid plexus in an attempt to control hydrocephalus but was largely unsuccessful. Early attempts at third ventriculostomy were occasionally successful but no truly effective treatment for hydrocephalus was available until the development of silicone and the one-way flow shunt valve.

It has been 70 years since the first shunts were placed. Surprisingly, only a limited amount of progress has been made regarding shunt technology. Following the development of the shunt, there were a large number of neurosurgeons and neurologists that felt hydrocephalus was "cured" and a surprising number of our prominent neuroscientists still harbor this belief. It is now clear that shunting, although saves lives, brings a host of potential complications.

The original shunts were simple differential valves. By the 1970s, we began to see the development of anti-siphon devices and attempts at flow control. However, the seminal publication by Drake et al. in 1998 revealed that there was no statistical difference between the types of shunt valves available on the market at that time. Thankfully, over the past two decades, progress has been made. Programmable valves and gravity-compensating valves have given us increased choices, but no rigorous clinical trials have been performed to compare these more expensive options with each other or the standard valve data that we have from previous studies.

The Hydrocephalus Clinical Research Network (HCRN) has provided us with excellent data from clinical trials regarding shunt infection, shunt re-infection, shunting outcomes, risk factors for shunt malfunction, outcomes in posthemorragic hydrocephalus, and ventricular catheter entry sites. They have assembled a database with multiple prospectively collected data points that should prove valuable as we continue to increase our understanding and treat this disease. The Adult Hydrocephalus Clinical Research Network (AHCRN) is now undertaking a similar approach to adult hydrocephalus, including transition patients and idiopathic normal pressure hydrocephalus (iNPH).

Many focus groups, organizations, and foundations throughout the world are focusing on hydrocephalus. Here in the United States, the Hydrocephalus Association (HA) has become a major source of research funds for young researchers. After receiving startup funding, several of these young investigators have gone on to receive significant federal grants. HA has facilitated interaction within the neuroscience community between basic and clinical researchers by creating the Hydrocephalus Association for Discovery Network (HANDS). The Hydrocephalus Society (IHSCSF) has become a major focus for hydrocephalus researchers and clinicians. Their meetings now have over 300 international participants. An increased interest in hydrocephalus among our young trainees is critical for this movement to go forward. We must attract trainees to the many hydrocephalus research opportunities that are increasingly available. Hydrocephalus offers us an opportunity to study a novel approach to a potentially reversible neuronal injury.

Since the development of the shunt, there have been only a few books devoted to hydrocephalus. Each has served its purpose, but none are as comprehensive as this text. This collection is a much-needed addition to our study of hydrocephalus and should be recognized as an important reference. The authors are all recognized experts in their field of study. The comprehensive topics in 8 sections and 71 chapters cover all areas of interest. This text will be required reading for all interested in understanding, treating, and assessing outcomes for those afflicted with this chronic and very difficult disease.

Emeritus Professor of Neurological Surgery
University of Utah

Marion L. (Jack) Walker, M.D.

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About the Editors

Giuseppe Cinalli was born in Naples on June 16, 1961. He completed the secondary school in Naples and the medical studies at the "Federico II" University of Naples (1980–1986). He started the residency program in Neurosurgery on April 1, 1987, at the "Federico II" University of Naples, but he was rapidly attracted by the Pediatric Neurosurgery, so he asked and obtained to complete his residency at the Department of Pediatric Neurosurgery of the Hôpital Necker-Enfants Malades in Paris, France, directed by Professor Jean François Hirsch, with grants from French Government and the Italian Ministry of Public Instruction (Ministero della Pubblica Istruzione). He completed his residency in October 1991 and continued to work in Necker as assistant-attaché. He married Fabrizia in 1991 and they had Francesco and Maria Allegra. In 1993, he spent 5 months as visiting fellow in the Department of Pediatric Neurosurgery at the New York University Medical Center directed by Fred Epstein and 3 months in the Department of Pediatric Neurosurgery at the Primary Children's Medical Center of Salt Lake City directed by Marion "Jack" Walker.

On November 1, 1993, he started his university career in France first as chef de clinique-assistant (1993–1997) and later as praticien hospitalo-universitaire (1997–1998), before deciding to go back to Italy. He then worked in Terni (1998–1999) and in Naples at the Department of Pediatric Neurosurgery, Santobono-Pausilipon Children's Hospital, directed by Dr. Giuseppe Maggi, where he started to work in October 1999.

He became chief of the Department of Pediatric Neurosurgery on April 1, 2007, and head of the Department of Neurosciences in July 2016. He is active member of 7 scientific societies, editor of 3 books, and founding member in 2001 of the International Study Group of Neuroendoscopy (ISGNE), later transformed into International Federation of Neuroendoscopy (IFNE) of which he was elected president in November 2017 for a 2-year term, and author of 116 indexed papers.

M. Memet Özek was born in Tübingen on November 23, 1956. He did Abitur in German High School of Istanbul and graduated with honors from Istanbul University School of Medicine. He did his residency in Hacettepe University in Ankara followed by his mandatory military service in the Department of Neurosurgery at the Turkish Military Academy in Ankara.

In 1989, he started his academic career in Marmara University School of Medicine as an assistant professor. In 1990 and 1991, he was a fellow in the Department of Pediatric Neurosurgery at the New York University Medical Center directed by Fred Epstein. Soon after his return to Istanbul, he established the first independent Division of Pediatric Neurosurgery of his country and became an associate professor. In 1999 he achieved the position of full professor in his department. Between the years 2008 and 2010, he was the president of the European Society for Pediatric Neurosurgery. In 2011, he moved to Acıbadem University and became the head of the Department of Neurosurgery in 2018.

He is an active member of 8 scientific societies, editor of 4 books, and author of 105 indexed papers. He joined 82 neurosurgery courses as a member of the faculty and organized 18 courses on different topics of pediatric neurosurgery in his own institution to educate young neurosurgeons of his geographical region.

He is married to Prof. Dr. Eren Özek and is the father of Ceren Ozek Moore, Ph.D.

Besides his career in neurosurgery, he is a dive master and an enthusiastic collector of Anatolian carpets and seashells.

Christian Sainte-Rose was born in Fort-de-France (Martinique) in 1949. He attended school in Martinique and then moved to Grenoble, France, where he graduated in the medical school and had his first contact with Neurosurgery. During the last years of his medical school, he started to build a boat that was complete at the time of his graduation, with which he spent a sabbatical year cruising in the Mediterranean. On land again, he started his residency in Neurosurgery in Dakar, Senegal, where he spent 4 years learning Neurosurgery before going back to France, where he was recruited by Jean François Hirsch in 1978 in the Department of Pediatric Neurosurgery at Necker-Enfants Malades Hospital in Paris. After a few years, he spent 6 months of research fellowship at the Hospital for Sick Children in Toronto, Canada, in the department directed by Harold Hoffman. He concentrated his early interest of research on the mechanical complications of cerebrospinal fluid valves, writing a seminal paper on the long-term functioning of ventriculoperitoneal shunts, analyzing a series of several hundreds of patients treated over years in Paris and Toronto, describing for the first time the different modalities of shunt dysfunction. As a natural consequence, he directed his research to the creation of a different kind of valve, intended to reduce the CSF overdrainage and its consequences. At the same time, he pioneered many neuroendoscopic techniques, participated in the first robotic microscope project, designed several surgical instruments (subdural catheter, dumbbell catheter for neuroendoscopy), hosted in Paris in 1995 one of the very first meetings on minimally invasive neurosurgery, and wrote seminal papers on virtually every aspect of pediatric neurosurgery. He became chief of the Department of Pediatric Neurosurgery of Necker-Enfants Malades in 2005 and then moved to Martinique in 2015 where he is working currently.

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Part I

Basics, Anatomy, Embryology, Physics, and Morphology

History of Hydrocephalus and Its Surgical Treatment

James Tait Goodrich

Contents

Abstract
Hydrocephalus, as known as a disease process, is an ancient one. The disorder has described in cave paintings, in early manuscripts in antiquity, and in many clinical observations over the last 25 centuries. The treatment of hydrocephalus as a surgical disorder is very recent, only dating to the 1950s. Prior to this period, treatment surgically lead to the death of the patient in almost one hundred percent of the time. Our medical colleagues did not do much better. In this chapter, we review the medical and surgical treatment of hydrocephalus dating to antiquity using contemporary reference sources. The development of the anatomy of the ventricular system, often very confusing to early anatomists, is reviewed if only to show the many errors generated by physicians in its understanding. The history of hydrocephalus is a fascinating one, and in order to better understand the concepts, illustrations from contemporary sources and the individuals involved have been included.

Keywords
2 Hydrocephalus · History of hydrocephalus · Surgical history of hydrocephalus · Ventriculo

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peritoneal shunt history · Ventriculocisternostomy · Endoscope · Choroid plexectomy

Introduction

The surgical treatment of hydrocephalus has had a very long history dating back to antiquity, having said that the first successful surgical treatment of hydrocephalus only dates back to that of the 1890s. What we now consider successful treatment is really only a palliative treatment with the exception of the third ventriculocisternostomy. To treat what is a quite complex disorder, neurosurgeons simply do some plumbing and place a tube to divert fluid from one compartment to another. In this chapter, we review the history of this subject along with contributions to the surgical treatment of hydrocephalus. Our early surgical brethrens were quite inventive and sometimes offered novel treatments – unfortunately only a few of which actually worked (Figs. 1 and 2).

Early History

In museum and private collections, we have numerous examples of children (adults) with clinical findings consistent with hydrocephalus. Due to a lack of written material, we have no knowledge what these early healthcare givers thought of the disorder or surgical methods to treat it. The Mesoamerican cultures have provided many artistic examples of children with clinical findings of hydrocephalus. Illustrated in Figs. 3 and 4 are early examples from the Olmec and Colima cultures illustrating two examples of children with findings consistent with hydrocephalus. Figure 3 is a rare example from the Olmec culture of a child with achondroplasia with hydrocephalus. Figure 4 originates from the Mexican west coast, an area that includes the state of Colima. This terracotta figure dates from about 200 A.D and shows an individual with spina bifida and hydrocephalus (Figs. 3 and 4) (Goodrich and Ponce de Leon 2010; von Winning 1987).

In the writings of Hippocrates (ca. 460 BC–368 BC) and his schools of learning, we find some of the earliest medical writings on hydrocephalus (Fig. 5) (Hippocrates 1525, 1925). Hippocrates erroneously believed that the accumulation of cerebrospinal fluid (CSF) was due to the excess “pituita” of chronic epilepsy. As a clinician, he gave clear descriptions of the signs and symptoms associated with hydrocephalus: headache, vomiting, visual loss, diplopia, and squinting. Hippocrates also noted that outcome and prognosis of hydrocephalus were extremely poor, a theme that would remain constant throughout history until the nineteenth century, when several surgical innovations became available. When it came to treatment of a patient with hydrocephalus, the Hippocratic schools could only offer laxatives, diuretics, and dietary supplements. Over the years, a number of authors have suggested that Hippocrates advocated trephination over the anterior fontanelle to relieve pressure in hydrocephalus. A recent examination of the Hippocratic corpus by this author failed to disclose any recommendations of surgical treatment for internal hydrocephalus. It is possible that Hippocrates might have advocated a form of trephination in a case of “external” hydrocephalus, a situation where fluid collects above the pericranium (Hippocrates 1925). For Hippocrates, surgery was not an option; only medical treatment would be offered (Figs. 6 and 7).

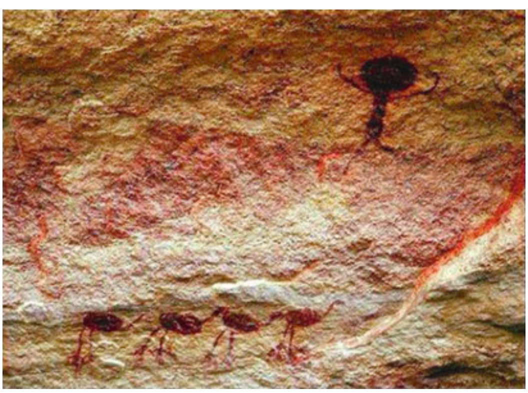


Fig. 1 A cave painting dating from the paleolithic period (10,000–4000 BC) – Piaui, Brazil. An early artist has painted in natural pigments a child with macrocephalic head, likely due to hydrocephalus. Courtesy of Bencio Oton de Lima, MD

Fig. 2 A burial dating from about 1200 AD discovered in Chancay, Peru. The individual is a child about 3 years of age with macrocephaly likely due to hydrocephalus

Fig. 3 An early Olmec figure dating from about 1500 BC showing a child with hydrocephalus and achondroplasia, likely a court jester or an attendant to a royal court. (Author's personal collection)

Fig. 4 A terracotta figure from the west coast of Mexico (the state of Colima) showing an individual with hydrocephalus and classic findings of an associated spina bifida with a hunchback (kyphosis) and "hands on knee" posture. (Author's personal collection)

One of the great figures in the history of medicine and surgery was Galen of Pergamon (130–200 A.D.). This great Alexandrian anatomist and surgeon provided several early descriptions of hydrocephalus (Galen of Pergamon 1576–1577). To help the surgeon understand this disorder, he categorized hydrocephalus into four types according to the site of fluid accumulation: between the brain and meninges, between the meninges and bone, between the bone and pericranium, and between the pericranium and skin. Following views originating with Hippocrates, Galen argued that in cases in which fluid accumulated below the meninges, this condition was not

Fig. 5 The earliest known painting of Hippocrates dating from the 8th century – image from the author's collection.

Fig. 6 A looseleaf printing dating from about 1820 showing a hypothetical portrait of Galen. From the author's collection

curable (Table 1). However, fluid that accumulated above the pericranium could be surgically drained. Galen did a number of studies on the brain and appreciated that CSF was in free communication within the ventricular system, i.e., flowed freely throughout the ventricles, interestingly an anatomical concept that lost in literature and not rediscovered until the eighteenth century (Figs. 8, 9, 10, and 11).

Following Greco-Roman medicine came the Byzantine period and a number of important eastern medical figures. The Byzantine Empire began with Constantine the Great and his ascent to the Roman throne in 324 A.D. The capital of the Roman Empire was moved from Rome to a small town, Byzantium, soon renamed Constantinople (now Istanbul, Turkey). This town was to remain the capital and the center of the Byzantine Empire till 1453, when the Ottoman Turks conquered it.

Several important Byzantine era surgeons contributed to the study of hydrocephalus: Oribasius (fourth century A.D.), Aetius (sixth century A.D.), and Paul of Aegina (seventh century AD). Oribasius (325–403), a surgeon, classified hydrocephalus into three types: fluid collections between the brain and meninges, which he deemed not operable, whereas fluid collections between the bone and pericranium and pericranium and scalp could be surgically drained (Oribasius 1557; Raeder 1931; Lascaratos et al. 2004). Oribasius did make an interesting technical suggestion, namely, that the surgeon should always place the lance behind the hairline so as not to leave a visible scar (Raeder 1931).

The classification of hydrocephalus and related views on the surgical treatment of hydrocephalus, like those discussed above, remained a dominant theme in the literature until very recent times. An inadequate understanding of CSF physiology plus the risk of surgical infection and meningitis

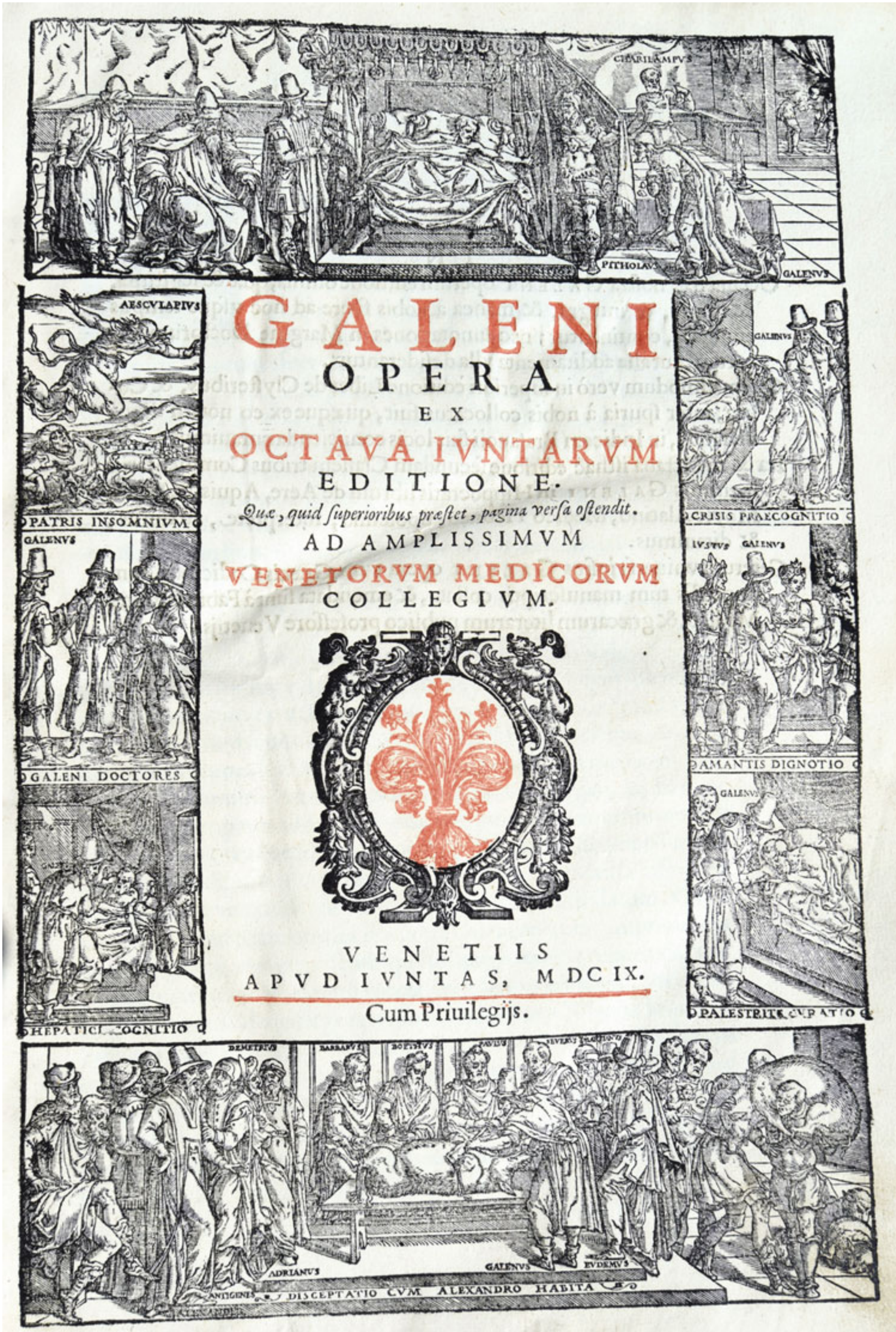


Fig. 7 The allegorical title page from Galen's collected works. Venice 1604 (Galen of Pergamon 1576–1577). In the panels surrounding the title are illustrated many of the famous figures of medicine and surgery. The panel at the bottom shows Galen performing a surgical transection of the recurrent laryngeal nerve in a pig. (Author's personal collection)

secondary to contaminated surgical instruments leads one to understand why the outcomes of surgery on hydrocephalus were almost invariably lethal. Such outcomes kept only the very rare surgeon from offering any form of surgical treatment.

Aetius of Amida (502–573), a prominent Byzantine physician, also wrote on hydrocephalus and offered surgical intervention as a form of treatment. Aetius studied in Alexandria and lived in Byzantium, where he was court physician to Emperor Justinian I. In his writings, he described the different types of fluids seen in the ventricles (Aetius of Amida 1542). He notes that the causes of this condition are not always known. For a classification system, he uses the same system as Galen, also asserting that surgery is indicated only in the external types of hydrocephalus. With regard to incisions, he declared that sometimes several are needed to adequately drain the fluid. He also believed that one of the causes of hydrocephalus was violent handling of the head by midwives, a common theme in the writing of physicians of this era. He also recommended different medications for treatment of hydrocephalus, mostly desiccants or diuretics "designed" to draw the fluid out.

Table 1 Galen of Pergamon (130–200 A.D.)

Types of hydrocephalus:
Between the brain and the meninges
Between the meninges and the bone
Between the bone and the pericranium
Between the bone and the skin

Hydrocephalus between the meninges and brain is *incurable*

Fig. 8 A collection of Roman era instruments scapels, probes and forceps – circa 200–400 AD. From the author's collection

Fig. 9 Roman era scaplel with sceptor point for stabbing the ventricles. Balkans region circa 200 AD. From the author's collection

Paul of Aegina (Paulus Aegineta Lt) (625–690?), among the last of the great Byzantine physicians and a skilled surgeon, also followed a similar theme of treating external hydrocephalus but not operating on the internal type. He did recommend a different type of incision that he felt offered a better solution, an "H" type of incision of his own design for the more extreme cases of hydrocephalus, reasoning that as more fluid was present in these cases, a larger incision was needed (Paul of Aegina 1534, 1844–1847).

A dominant figure in Islamic medicine was a learned Moorish Spaniard, Albucasis (Abu Al-Qasim or Al-Zahrawi; 936–1013) – a prolific writer and great compiler whose writings (some 30 volumes!) were focused mainly on surgery, dietetics, and materia medica (Albucasis [Abu Al-Quasim] 1519; Spink and Lewis 1973; Al-Rodhan and Fox 1986). He provides an interesting discussion on hydrocephalus and the prevalent fear of operating on this disorder:

> On the cure of hydrocephalus: This disease occurs most commonly in infants upon delivery when the midwife grasps the child's head roughly. It also sometimes happens from some hidden and unknown cause. I have never seen this disease except in very small children; and death very quickly overtook all those that I have seen; therefore I have preferred not to undertake operation in these cases. I have seen a child whose head was filled with fluid and daily growing in size, until the child could not sit upright on account of the size of his head, and the humidity increased till he died. (Spink and Lewis 1973)

Like previous surgeons, Albucasis would only operate on cases of "external hydrocephalus." Albucasis designed a "+"-type incision to drain off the "humidity" and in these cases claimed good results (Albucasis [Abu Al-Quasim] 1519). Albucasis also designed a series of special surgical instruments just for the treatment of hydrocephalus (Figs. 12 and 13). Albucasis noted that before considering an operation for hydrocephalus, he would offer the child a more conservative nonsurgical treatment. This treatment would include "binding" the head with a tight constricting wrap. Then the child would be placed on a "dry diet" with fluid restrictions (Spink and Lewis 1973). Albucasis' surgical writings would go onto be used extensively in the schools of Salerno and Montpellier and hence had an important influence in the Middle Ages.

Fig. 10 A cache of Roman era surgical instruments – circa 2nd–4th century AD. From the author's collection

Fig. 11 The instruments to the left are typical designs used for “stab and drain” for hydrocephalus

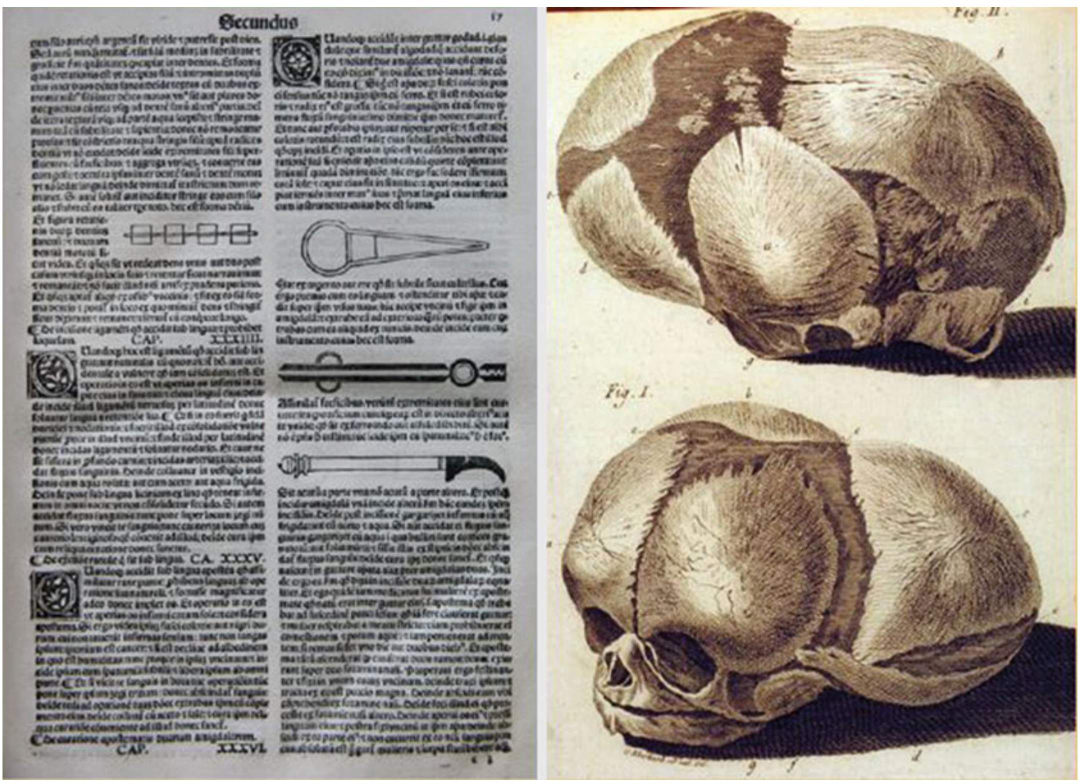


Fig. 12 Instruments designed by Albucasis for the treatment of hydrocephalus – “stab and drain” technique (Albucasis [Abu Al-Quasim] 1519) (Author’s personal collection)

Fig. 13 Surgical instruments from the author's collection that date to Byzantine era, circa 1000 AD. From the author's collection

Middle Ages

The Middle Ages was a period with little innovation in both the fields of surgery and medicine and hydrocephalus was no exception. The dominant writings and educational influences still came from the Hippocratic and Islamic schools. Due to a codification of earlier doctrines from these schools, surgeons were restricted in their practices to only using what was previously written by the likes of Hippocrates, Galen, Albucasis, and others. In actual fact, most surgeons of this era were "barber-surgeons" with very little if any formal surgical training. Training was by an apprenticeship to a senior surgeon. Even the academically trained surgeon offered no new insights into either the surgical or medical management of hydrocephalus.

Renaissance

Leonardo da Vinci (1452–1519) revolutionize the study of anatomy and made a number of contributions to cerebral anatomy among the most

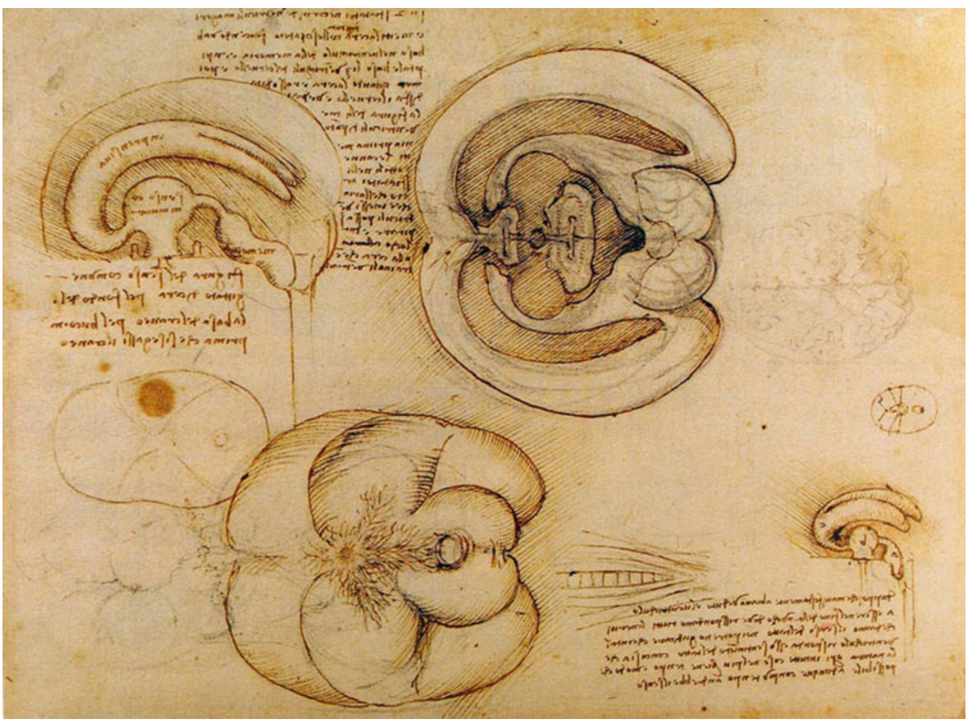
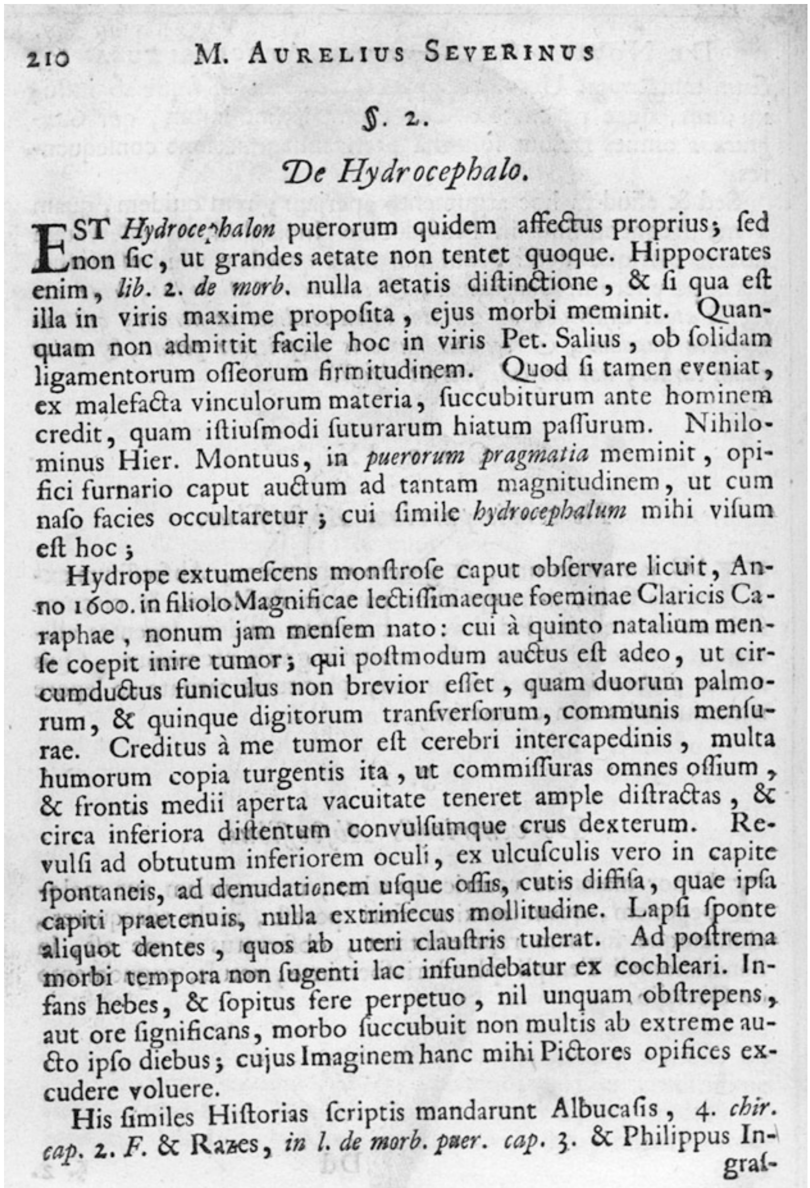


Fig. 14 The ventricular system using Leonardo da Vinci's wax casting technique

Fig. 15 Leonardo's view of the cell doctrine theory where brain function resides within the ventricular system as opposed to the brain parenchyma (da Vinci 1911–1926). (Author's personal collection)

important illustrating the ventricular system. He also offered some interesting but incorrect views on the anatomy of the brain and the ventricular system including invoking the "cellular doctrine," a doctrine that felt brain function existed in the ventricles not the brain itself. Leonardo had planned on compiling a large 120-volume work on anatomy but did not live long enough to complete it (da Vinci 1911–1926, 1979). Within this remarkable anatomical collection are several drawings dealing with the brain and the ventricles. Leonardo developed an ingenious system of injecting liquid wax into the ventricles to make a cast of the ventricular system (Figs. 14 and 15). Unfortunately Leonardo's drawings disappeared after his death in 1519 and did not to reappear until the eighteenth century, when they were "rediscovered" in England (da Vinci 1911–1926, 1979).

A physician and pathologist – Marco Aurelio Severino (1580–1656) – provided the first printed illustration of a child with hydrocephalus in a book published in 1632 (Figs. 16 and 17) (Severino 1632; Lyons 1995). This illustration appears in a section on the "obscure nature of tumors." Disorders of this type were often referred



210 M. AURELIUS SEVERINUS

§. 2.

De Hydrocephalo.

EST *Hydrocephalon* puerorum quidem affectus proprius; ſed non ſic, ut grandes aetate non tentet quoque. Hippocrates enim, *lib. 2. de morb.* nulla aetatis diſtinctione, & ſi qua eſt illa in viris maxime propoſita, ejus morbi meminit. Quanquam non admittit facile hoc in viris Pet. Salius, ob ſolidam ligamentorum oſſeorum firmitudinem. Quod ſi tamen eveniat, ex malefacta vinculorum materia, ſuccubiturum ante hominem credit, quam iſtiuſmodi ſuturarum hiatum paſſurum. Nihilominus Hier. Montuus, in *puerorum pragmatia* meminit, opifici furnario caput auctum ad tantam magnitudinem, ut cum naſo facies occultaretur; cui ſimile *hydrocephalum* mihi viſum eſt hoc;

Hydrope extumeſcens monſtroſe caput obſervare licuit, Anno 1600. in filiolo Magnificae lectiſſimaeque foeminae Claricis Caraphae, nonum jam menſem nato: cui à quinto natalium menſe coepit inire tumor; qui poſtmodum auctus eſt adeo, ut circumductus funiculus non brevior eſſet, quam duorum palmorum, & quinque digitorum tranſverſorum, communis menſurae. Creditus à me tumor eſt cerebri intercapedinis, multa humorum copia turgentis ita, ut commiſſuras omnes oſſium, & frontis medii aperta vacuitate teneret ample diſtractas, & circa inferiora diſtentum convulſumque crus dexterum. Revulſi ad obtutum inferiorem oculi, ex ulcuſculis vero in capite ſpontaneis, ad denudationem uſque oſſis, cutis diffiſa, quae ipſa capiti praetenuis, nulla extrinſecus mollitudine. Lapſi ſponte aliquot dentes, quos ab uteri clauſtris tulerat. Ad poſtrema morbi tempora non ſugenti lac infundebatur ex cochleari. Infans hebes, & ſopitus fere perpetuo, nil unquam obſtrepens, aut ore ſignificans, morbo ſuccubuit non multis ab extreme aucto ipſo diebus; cujus Imaginem hanc mihi Pictores opifices excudere voluere.

His ſimiles Hiſtorias ſcriptis mandarunt Albucaſis, 4. *chir. cap.* 2. *F.* & Razes, *in l. de morb. puer. cap.* 3. & Philippus Ingraſ-

Fig. 16 Severino's discussion of hydrocephalus from his book of 1632 (Langenbeck and Hahn 1969) (Author's personal collection)

to as "swellings," revealing what little understanding of what these disorders were all about. Unique to Severino was that he often added the patient's history, and if surgery was performed, the techniques and instruments used were outlined and discussed.

In the seventeenth and eighteenth centuries, initial steps were finally taken, though the path was circuitous, toward formulating a physiological understanding of hydrocephalus. In the fields of anatomy and physiology, much progress was being made especially in the understanding of the brain, its anatomy, and function. Still in treatment for hydrocephalus, surgeons were still not eager to do surgical intervention; they rather turned the care over to physicians. If surgical intervention was indicated, it was almost always a "stab and drain" technique where the anterior fontanelle was penetrated with a sharp lance, sometimes a wick or cannula was placed to assist in the drainage.

Fig. 17 The earliest known printed illustration of a child with hydrocephalus, from Serverino's text of 1632 (Langenbeck and Hahn 1969)

Unfortunately the outcome was nearly 100% mortal (Fig. 18).

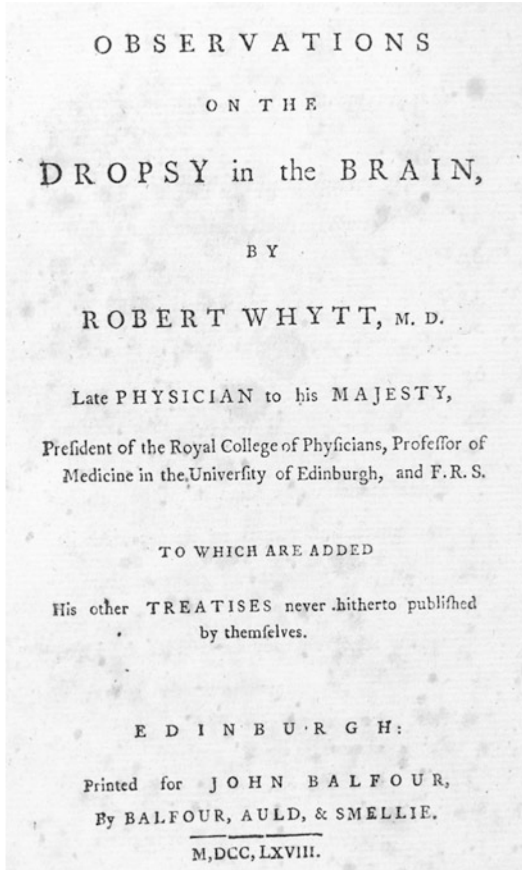
While this chapter is devoted to surgical treatments of hydrocephalus, it is useful to take a brief look at the medical treatments offered to children with hydrocephalus.

> The hydrocephalus, or dropsy of the head, is either external or internal. The former has its seat in the cellular substance, between the skin and the pericranium, or between this membrane and the skull. In the internal hydrocephalus, the water is sometimes collected between the cranium and dura mater, or between this last and the pia mater; but most commonly is found in the ventricles of the brain, immediately below the corpus callosum; And this is not only the most frequent and fatal species of the hydrocephalus, but also that with which medical writers seem to have been least acquainted. (Whytt 1768)

The above quotation comes from one of the first monographs devoted entirely to hydrocephalus. This book was published by an English physician – Robert Whytt (1714–1766). His

Fig. 18 A collage of medical images and scenes from this period. Circulation of blood was discovered by William Harvey (left), wound man (upper middle), a montebank traveling barber surgeon, and his advertising broadside, (lower middle), The King's Touch, laying on the hands to cure scrofula (Upper right), Physician making a diagnosis by examination of the urine – uroscopy (lower right)

monograph, *Observations on the Dropsy in the Brain*, (Whytt 1768) appeared in 1768. A review of his writings provides us with an extensive discussion of hydrocephalus along with a number of good clinical observations including descriptions of its various clinical stages of hydrocephalus. While the work is rich in clinical observations, Whytt offered little in the way of clinical treatment and actually was still adopting the views of the "ancients" as discussed above. It is Whytt's clinical findings regarding patients with hydrocephalus that we find of greater interest. These findings were divided into three stages of severity: stage 1, quickened pulse; stage 2, slow irregular pulse; and stage 3, rapid pulse; in the last stage, the pupils become dilated and are no longer responsive even "in the greatest light" (Payr 1911). Whytt offered the following treatments for hydrocephalus: emetics, purging, blisters, and leeches. It is interesting to note for that Whytt's surgical treatment was not even an option. When it came to the late stages of hydrocephalus, Whytt was quite pessimistic: "[When] so much water is accumulated as, by its pressure on the sides of the ventricles, to disturb the action of the brain, we have little to hope from medicine" – a clinical correlation that remained essentially correct for the next 150 years (Fig. 19) (Whytt 1768).



OBSERVATIONS

ON THE

DROPSY in the BRAIN,

BY

ROBERT WHYTT, M. D.

Late PHYSICIAN to his MAJESTY,

Preſident of the Royal College of Phyſicians, Profeſſor of Medicine in the Univerſity of Edinburgh, and F. R. S.

TO WHICH ARE ADDED

His other TREATISES never hitherto publiſhed by themſelves.

EDINBURGH:

Printed for JOHN BALFOUR, By BALFOUR, AULD, & SMELLIE.

M,DCC,LXVIII.

Fig. 19 The title page from Whytt's book, the first English work devoted solely to the treatment of hydrocephalus (Whytt 1768) (Author's personal collection)

In the nineteenth century, a number of physicians and anatomists began to formulate and understand the production, anatomical pathways, and in turn causes of CSF obstruction. With this new research, there came a better understanding of the physiology of hydrocephalus. With this new information by the end of the nineteenth century, innovative surgeons were beginning to come up with more relative surgical techniques for diverting CSF as a form of surgical treatment.

> Except for cases that cure themselves, i.e., those that undergo spontaneous arrest, the outlook is almost hopeless. The majority die within the first four years of life, the first year claiming most of them.... At the present time, even the best-intentioned surgery can hardly serve other than to delay to some few cases the possibility of a spontaneous recovery by subjecting them to an operation and a surgical death. (Bucy 1932)

When Paul Bucy (1904–1992) wrote the above statement in 1932 in a textbook on pediatrics, there were very few effective surgical treatments for hydrocephalus (Bucy 1932). In reviewing this early surgical period, it is of interest to consider what some of our surgical colleagues had attempted over time to find a "surgical cure" for hydrocephalus.

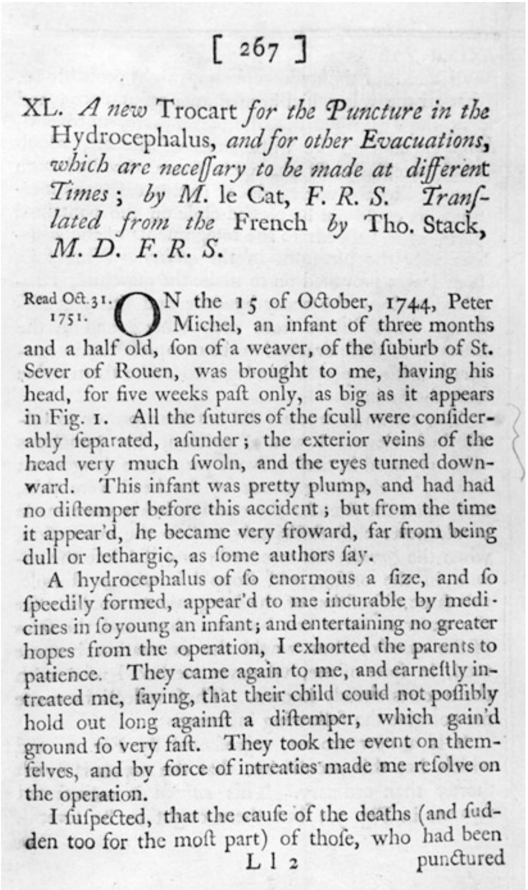
A common and standard technique for draining CSF was the use of trocars of various types and "stabbing" into the lateral ventricle via the anterior fontanelle. An early variation on this theme was published by an Italian surgeon, Dominico Galvani (d. 1649) (Galvani 1620). In 1620 Galvani developed a surgical drainage system to which he added to his trocar device. This drainage system included a device to hold the drainage tube in place and then various receptacles for collecting CSF. A specially designed trocar was used to

Fig. 20 Title page and illustrated surgical set designed by Galvani to treat hydrocephalus by a "stab and drain technique via the anterior fontanelle (Galvani 1620)

place the drain and a cap added to prevent entry of air into the ventricles (Fig. 20). Unfortunately, either because of over drainage or sepsis, the outcome was always death. Following along with the same theme, another trocar-type system was developed in 1753 by Claude Nicholas Le Cat (1700–1768) and described in the *Philosophical Transactions of the Royal Society* (*London*). Mr. Le Cat included an illustration of a child with severe hydrocephalus (LeCat 1753). In his paper, Le Cat illustrates his system for CSF drainage. Le Cat, like Galvani, noted that his outcomes were dismal and that the mortality rate eventually reached 100%, but nevertheless he felt his system was a useful surgical intervention for treating hydrocephalus (Figs. 21 and 22).

In searching for surgical treatments of hydrocephalus, we reviewed the writings of Pierre Dionis (1645–1718), a prominent eighteenth-century French surgeon and anatomist who practiced in Paris. Dionis was appointed the first surgeon to Queen Maria Theresa. Later Louis XIV established a demonstratorship in operative surgery at the Jardin in Paris and appointed Dionis as the first holder of that position. His surgical textbook entitled "A Course of Chirurgical Operations Demonstrated in the Royal Gardens at Paris" became a popular and widely used eighteenth-century surgical handbook eventually being translated into several languages (Dionis 1733). In an English edition of his surgical handbook, Dionis discussed his views on hydrocephalus:

> The Etymology of the Hydrocephalum proceeds from *Hydro* Water, and *Cephale* which signifies a Head; so that tis a sort of *Dropsie*, with which the Head is so full of Water; that 'tis perfectly inundated. He went on to describe the two classic forms of hydrocephalus following the ancient classification of "external" and "internal" **(note** text, spelling and syntax has been copied exactly as printed). (Dionis 1740)



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XL. *A new* Trocart *for the Puncture in the* Hydrocephalus, *and for other Evacuations, which are neceſſary to be made at different Times; by M.* le Cat, *F. R. S. Tranſlated from the* French *by* Tho. Stack, *M. D. F. R. S.*

Read Oct. 31. 1751.

ON the 15 of October, 1744, Peter Michel, an infant of three months and a half old, ſon of a weaver, of the ſuburb of St. Sever of Rouen, was brought to me, having his head, for five weeks paſt only, as big as it appears in Fig. 1. All the ſutures of the ſcull were conſiderably ſeparated, aſunder; the exterior veins of the head very much ſwoln, and the eyes turned downward. This infant was pretty plump, and had had no diſtemper before this accident; but from the time it appear'd, he became very froward, far from being dull or lethargic, as ſome authors ſay.

A hydrocephalus of ſo enormous a ſize, and ſo ſpeedily formed, appear'd to me incurable by medicines in ſo young an infant; and entertaining no greater hopes from the operation, I exhorted the parents to patience. They came again to me, and earneſtly intreated me, ſaying, that their child could not poſſibly hold out long againſt a diſtemper, which gain'd ground ſo very faſt. They took the event on themſelves, and by force of intreaties made me reſolve on the operation.

I ſuſpected, that the cauſe of the deaths (and ſudden too for the moſt part) of thoſe, who had been

L l 2 punctured

Fig. 21 Text of the trocar system for treating hydrocephalus – by LeCat (1753) (Author's personal collection)

Dionis continued to embrace a common theme dating from antiquity, namely, that a common cause of hydrocephalus was due to head molding by the midwives shortly after birth, a practice which he denounced. For external hydrocephalus, Dionis suggests the surgeon make several cuts between the skin and the pericranium to relieve the fluid accumulated (subgaleal shunting) – but having said that, he declares that he has never seen a case of "external" hydrocephalus. As to "internal" hydrocephalus, he believes that this is a disease that is "incurable." Nevertheless, Dionis wrote that for treatment of this type, he preferred to use the trepan, the only treatment available. Included in his surgical handbook is an illustration of the instruments he used for trepanning. The surgical view of treatment of internal hydrocephalus here reveals an ancient treatment dating back to the Greeks – so sadly we see little if any progress over the previous 2000 years (Figs. 23 and 24).

Alexander Monro *secundus* (1733–1817) is remembered for his remarkable works on anatomy and his studies on hydrocephalus. Not appreciated by many readers today were his surgical efforts directed toward the treatment of hydrocephalic children. Monro was also an advocate for surgical drainage of CSF through a trocar but again following the earlier Hippocratic dogma that this was only to be attempted in cases of "external hydrocephalus." The outcomes continued to invariably fatal mostly due to sepsis and meningitis (Figs. 25, 26, and 27) (Monro 1797).

Nineteenth-Century Incunabula Period for Surgical Treatment of Hydrocephalus

With the introduction of asepsis techniques in the Listerian era (ca. 1870s), the use of the trocar was again resurrected as a surgical treatment for hydrocephalus. A popular advocate employing a trocar was a French surgeon Antoine Chipault (1866–1920). In a rather bizarre form of treatment, Chipault's technique (1894) required the trocar to be placed into the ventricle via the nose (Chipault 1894–1895). Unfortunately Chipault did not report his outcomes, so long-term follow-ups are not available. One may assume the results were not good considering some of the anatomical structures that lay in the pathway of that trocar. Following a different theme, Langenbeck and Hahn (Langenbeck and Hahn 1969) introduced a technique in which the trocar was passed through the orbital roof into the ventricle, a surgical approach later popularized in performing the frontal lobotomy. Despite these being "improved techniques," the reported mortality was still close to 100% (Figs. 28, 29, 30, and 31).

William W. Keen (1837–1932) was a prominent Philadelphia surgeon, early pioneer brain surgeon and professor of surgery at Jefferson Medical College. Keen became an early and

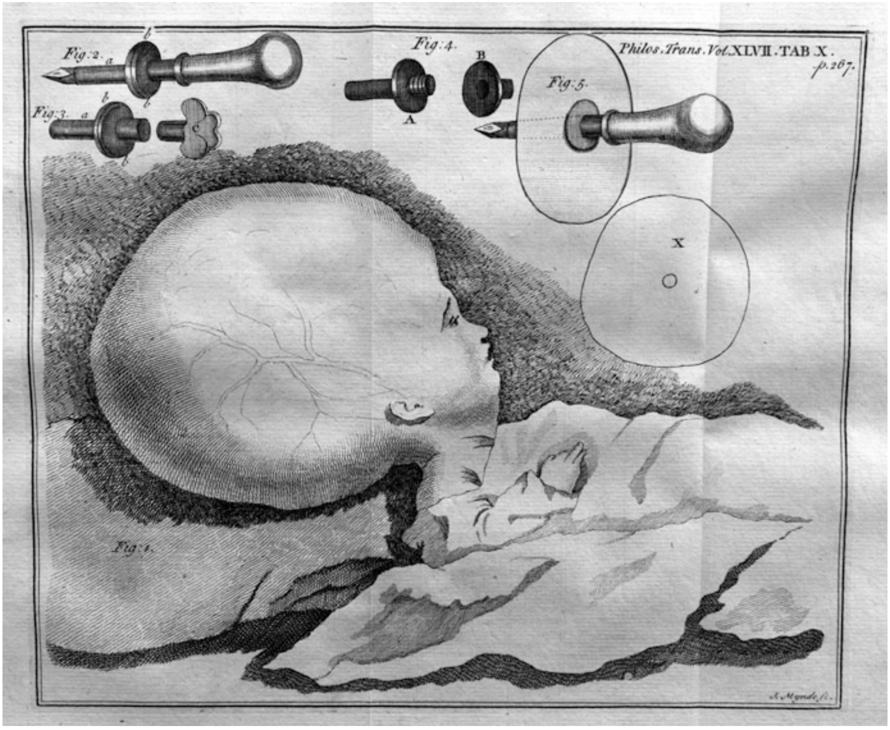

Fig. 22 Text of the trocar system for treating hydrocephalus – by LeCat (1753) (Author's personal collection)

vocal advocate of Listerian antisepsis. With now the ability to reduce the high morbidity due to infection, Keen developed a cannula for draining the lateral ventricles for hydrocephalus in 1888. Unfortunately the technique was useful only in the short term and not suitable for permanent diversion of CSF. In addition Keen found that the sudden collapse of the ventricles proved its own morbidity (Figs. 31, 32, and 33) (Keen 1888).

Toward the end of the nineteenth century and coming into the first part of the twentieth century, a number of new surgical approaches for hydrocephalus were advocated. In 1891 Heinrich Quincke (1842–1922) introduced the lumbar puncture as a treatment for reducing CSF fluid. He was to eventfully conclude that the results were temporary at best. Quincke quickly realized that CSF was being constantly produced so that fluid removed was rapidly replaced in a matter of hours (Figs. 34 and 35) (Quincke 1891, 1901; Haymaker and Schiller 1970). Quincke came to develop the lumbar spinal puncture while looking for the least harmful and simple way to treat hydrocephalus. He believed that part of the disease process was that the increasing pressure in hydrocephalus compressed the pacchionian granulations and blocked CSF absorption. His postulation was that by removing CSF, one could break the vicious cycle of overproduction and underresorption. He quickly recognized that the procedure was not only safe but could be used for both therapeutic and diagnostic purposes. Quincke would add an interesting modification in his technique. By moving the spinal needle back and forth in the soft tissue space between the skin and the dura, he could develop a fistula track for CSF drainage and diversion. Quincke felt this fistula tract provided for a slower and more steady absorption of CSF.

Surgeons were becoming more familiar with the concept of overproduction/blockage of CSF and developing surgical techniques to move CSF to another anatomical space. Among the first to address this concept was H. Ferguson (1853–1912) in 1898. Ferguson designed a surgery in which he connected the lumbar thecal sac

Fig. 23 Illustration from Dionis' textbook of surgical techniques showing a demonstration (Dionis 1733) (Author's personal collection)

to the peritoneal cavity by a wire passed through a vertebral body (Ferguson 1898). Caring this concept a step further, B. Heile in 1908 devised a technique whereby he anastomosed the lumbar thecal sac to the greater omentum with a vein graft (Heile 1908, 1925a). Kanusch again proposed a technique in 1908 for draining CSF into a dissected subaponeurotic space of the scalp. This technique of subgaleal shunting is still being used (Kanusch 1908). Johann von Mikulicz (1850–1905), a German surgeon, published a report in 1893 on a system he designed to "slowly" move CSF from the ventricles via a glass wool mesh to just under an osteoplastic flap. The wool allowed the CSF to travel from the ventricle to epidural space; he reported a "cure" in this case. Mikulicz would further modified system by designing a gold funnel with a disk. The funnel was placed through the disk through the skull and into the ventricle – this would become the first subgaleal-subarachnoid-ventricle shunt with diversion of CSF from the ventricle to the subgaleal space – a closed system (Aschoff et al. 1999). A further modification was designed by Sutherland and Cheyne. The authors published two successful cases in which the surgical technique involved forming a fine weave from 16 fine strands of 2-inch long catgut. This woven catgut was placed in the ventricle on one side with the other end placed in the subdural space. In both cases, they reported excellent results with collapse of the ventricles and overriding sutures (Sutherland and Cheyne 1898).

The concept of a "valved" shunt with unidirectional flow possible was introduced in 1907 by a Leipzig surgeon Edwin Payr (1871–1946). Payr harvested a saphenous vein graft, and taking advantage of luminal flow that is unidirectional, he placed the graft into the lateral ventricle and anastomosed it to the sagittal sinus. While clearly innovative, he was only able to report an average 4-month survival rate. At autopsy he found the lumens of the grafts remained patent with no reflux of the blood back into the ventricles (Payr 1908, 1911).

Early twentieth-century surgeons proposed draining CSF from the ventricles to other surgical cavities as an additional form of treatment for hydrocephalus (McClure 1909). In 1925 B. Heile published his technique for the lumbar ureteral shunt (uretero-subarachnoid anastomosis) as a diversion procedure (Heile 1925b). Unfortunately this technique required taking one kidney. This technique was further modified by Donald Matson (1913–1969) in 1949 (Matson 1949). Matson placed polyethylene tubes in the lumbar subarachnoid space and "tunneled" them into the peritoneal cavity. While a lumboperitoneal shunt is not commonly used for the treatment of hydrocephalus now, this technique is still popularly used in the treatment of pseudotumor cerebri. By the mid-1950s, the ventriculoatrial shunt was becoming the mainstay for the treatment of hydrocephalus. In 1954 ventriculo-pleural shunt was described by Joseph Ransohoff (1915–2001) and colleagues in six patients. The results of this