

OCCASIONAL PAPER

From congenital to idiopathic adult hydrocephalus: a historical researchPaolo Missori,¹ Sergio Paolini² and Antonio Currà¹

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Reports of idiopathic adult hydrocephalus prior to the 1965 description of idiopathic normal pressure hydrocephalus by Salomón Hakim and Raymond Adams are lacking in the literature. Congenital paediatric and congenital adult hydrocephalus were considered to trace the early descriptions of idiopathic adult hydrocephalus. Through traditional or digital libraries and internet search engines, the historical sources dealing with idiopathic adult hydrocephalus were explored. The research shows that many distinguished physicians through the centuries have contributed to the discovery of idiopathic adult hydrocephalus. Early descriptions of the disorder were related to autopsy studies in the 18th and 19th centuries. From the second half of the 1800s, idiopathic adult hydrocephalus appeared to have been forgotten in the medical literature.

Keywords: cerebrospinal fluid; clinical neurology; dementia; history; hydrocephalus

Abbreviations: INPH = idiopathic normal pressure hydrocephalus

Introduction

Idiopathic normal pressure hydrocephalus (INPH), according to cerebrospinal fluid measurements, is a slow evolving neurological disease related to adult age, and its clinical picture was defined in the 1960s by Salomón Hakim and Raymond Adams (Hakim, 1964; Adams *et al.*, 1965; Hakim and Adams, 1965). At that time, the literature available on this subject was very scarce; Hakim and Adams found only three papers dealing with symptoms associated with adult hydrocephalus, and in only one of McHugh's cases did the symptoms and radiological studies fit with a diagnosis of INPH (Riddoch, 1936; Foltz *et al.*, 1956; McHugh, 1964).

Therefore, the Adams and Hakim description could be considered the first neurological account of INPH occurring in adult patients. However, it is surprising that experienced clinicians or pathologists could not detect such a disease for many centuries when the observational method is a key point of medical

evaluation. Research was carried out to find possible previous reports of idiopathic adult hydrocephalus already denominated as INPH. The unexpected results are reported here.

Materials and methods

Idiopathic adult hydrocephalus is considered to be a synonym of INPH. To determine the origin of idiopathic adult hydrocephalus (or INPH) better, the history of congenital or paediatric hydrocephalus was analysed. Through traditional or digital libraries and internet search engines, historical sources dealing with idiopathic adult hydrocephalus were considered, starting from the origin of congenital paediatric hydrocephalus in the archeological Egyptian collections, through the Greek period and the Middle Ages, and up to 20th century in English, French, German, Italian, Latin and Spanish. Early editions were preferred when available, otherwise subsequent book editions were used.

The various hydrocephalus denominations appearing throughout the centuries in the different languages (hydrocephalus, dropsy in the head, dropsy of the brain, watery head, water in the head, hydrocephale, eaux dans le cerveau, wassersucht, wasserkopf, bol-hoof, idrocefalo, idropisia della testa, hydrocephalo, hidrocefalo) were considered.

Idiopathic adult hydrocephalus is considered to affect patients over 50 years of age with a normal head circumference (52–58 cm) and symptoms ranging from absent to Hakim's triad. A pathological finding of cerebrospinal fluid exceeding 30 ml (the normal capacity of adult lateral and third ventricles) is considered suggestive of hydrocephalus. Whenever the amount of cerebrospinal fluid was reported in libras (pounds) or ounces, the value was determined by the following conversions: 1 libra = 12 ounces; 1 ounce = 30 ml or 30 cc.

Congenital paediatric, congenital adult, acute, or secondary chronic (post-infectious, post-trauma or post-haemorrhagic) hydrocephalus was differentiated from idiopathic adult hydrocephalus.

Results

The ancient period: the early findings of paediatric or adult congenital hydrocephalus (fifth century B.C.–800 A.D.)

Reports of paediatric hydrocephalus date back to very ancient times in the history of medicine. In ancient Egypt, the high level of medicine supports the hypothesis that the disease was known (Nunn, 1996). The medical papyri available to date do not report paediatric hydrocephalus, but some archeological surveys show the presence of possible paediatric or adult congenital hydrocephalic skulls in the Mediterranean region (Maslahat *et al.*, 1910; Armelagos, 1969). Among these finds, the discovery of Egyptian skulls compatible with children or youths affected by paediatric hydrocephalus (El Batrawi, 1935; Kuzawa and Armelagos, 1996; Tillier *et al.*, 2001) and large adult skulls with congenital hydrocephalus and plagiocephaly from cemeteries in the north of Egypt (Dery, 1913, 1915), without a doubt demonstrate the presence of this disease. In addition, a small statue of Pent-en-nessa in the Egyptian Museum in Cairo documents the condition (Pahor, 1992).

Medical knowledge was diffuse in the Mediterranean region. As a consequence, the Greek Hippocrates (fifth century B.C.) is credited as being the first physician to describe and treat a possible hydrocephalus (Davidoff, 1929; Lifshutz *et al.*, 2001). In '*De morbis*', Hippocrates provided a very short description of an ambiguous disease in which '*Ἦν ὕδωρ ἐπὶ τῷ ἐγκεφαλῷ...*': 'water over the head' was cured through selected food; but he was not referring to children and not using the term hydrocephalus (Hippocrate, 1526; Foesio, 1595). Surgery was also proposed: '*Deinde postquam cibis eum refeceris, demum secto iuxta sinciput capite, ad cerebrum usque perforato, et velut sectionem per terebram curato*': 'Then after administering food, finally incised the head next to the frontal region, penetrated up to the brain, and healed for trephine opening'. Although Hippocrates was probably merely draining the subdural or subarachnoid space (Lifshutz *et al.*, 2001), in some children with opened sutures and large

hydrocephalus, a much thinned brain can allow subcutaneous-subdural-transcortical tapping and drainage of the ventricular cerebrospinal fluid.

After Hippocrates, Celso (c. 14 B.C. to c. 37 A.D.) was the first to refer to Greek physicians describing '*ὕδροκέφαλον*' 'hydrocephalus' as collections of humor over the head for which a '*sinapi*' or chisel were the cure (Wedel, 1713). Following descriptions in the Greek medical literature of children affected with a visible and palpable fluid collection on the head, suggest that hydrocephalus was a well known clinical condition (Lascaratos *et al.*, 2004). Galen (129–216 A.D.) and Oribasius (325–395 A.D.), both from the Anatolic Pergamon city, educated at the great medical school of Alexandria in Egypt, and physicians to the Roman emperors; Aetius (502–575 A.D.) from the Anatolic Turkish Amida; and Paul of Aegina—a Greek island (c. 625–690 A.D.) detailed this disease, providing medical and surgical therapy. Notably, Galen, and later other physicians (Oribasius and Aetius), believed hydrocephalus to be a subcutaneous fluid collection on the head due to improper handling by the midwife during parturition. In these children, Galen distinguished between two extracranial (subcutaneous or subperiosteal) and two intracranial (extradural or subdural) forms (Galien, 1565; Kühn, 1827; Grunert, 2007). The assumption that hydrocephalus was an extra-axial accumulation rather than a ventricular system enlargement persisted in medical knowledge until the Renaissance and Andreas van Wessel's report of cerebrospinal fluid collecting in the ventricles. In contrast, Galen related the accumulation of humors (or 'water') in the cerebral ventricles to apoplexy in adult patients, a notion lasting up to the 18th century (Schutta and Howe, 2006; Schutta, 2009). A later theory from Nemesius of Emesa (late 4th century), in which the cerebral ventricle cavities were, to a variable extent, where fantasia (lateral ventricles), reason (third ventricle), and memory (fourth ventricle) resided, contributed to entangle the diagnosis of idiopathic adult hydrocephalus (Mundinus, 1482; Peyligk, 1499; Hundt, 1501; Nemesius, 1565; Garcia *et al.*, 1983) (Fig. 1).

The Middle Ages period: from Muslim medicine to early European reports of paediatric hydrocephalus (801–1400 A.D.)

In the Middle Ages, Islamic medicine developed the scientific method through the introduction of observation and systematic experimentation. This golden age of Islamic medicine inherited and strengthened, from the Greek and Roman literature, the surgical treatment of paediatric hydrocephalus (Avicenna, 1473; Albucasis, 1541, 1778; Rhazae, 1544; Alpagus, 1547; Rahimi, 2007). The Muslim physicians' writings served for centuries as teaching texts in European medical schools. The Muslim doctor Abu Bakr Muhammad al-Razi (850–925, Persian physician) is credited as composing the first writing on paediatric diseases (Rabdill, 1971; Modanlou, 2008). In the Latin editions of '*Liber medicinalis ad Almansorem*', Chapter 3 of '*egritudinibus puerorum*' deals with

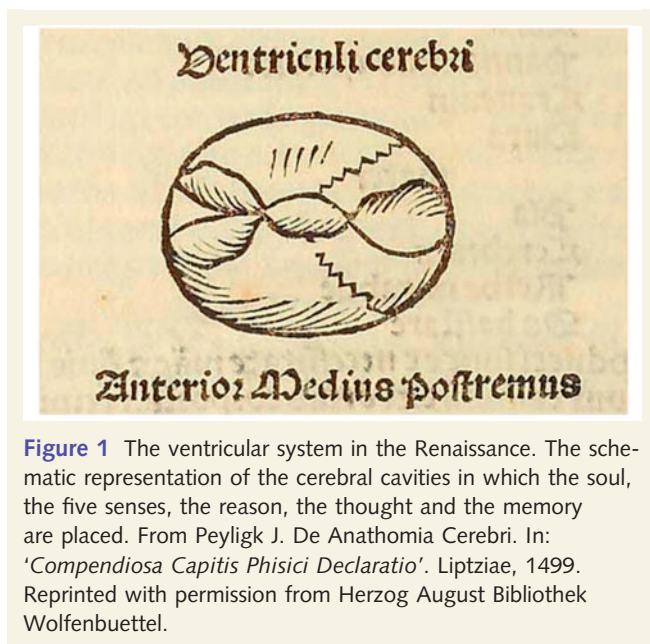


Figure 1 The ventricular system in the Renaissance. The schematic representation of the cerebral cavities in which the soul, the five senses, the reason, the thought and the memory are placed. From Peyligk J. *De Anathomia Cerebri*. In: *'Compendiosa Capitis Phisici Declaratio'*. Liptziae, 1499. Reprinted with permission from Herzog August Bibliothek Wolfenbuettel.

'*magnitudine capitis*' (Raazai, 1481; Rhazi, 1510; Rhazae, 1544). A child with progressive growth of the cranial bones is reported:

Et ego vidi puerum cuius caput adeo augmentabatur in longitudine et latitudine quod corpus eius susterre non poterat et non cessavit augmentari quousque mortuus est puer. Magnitudo autem capitis, aut erit ex grossa ventositate generata, in ossibus capitis, aut ex aggregatione aque inclusae in eo, quae non invenit viam exitus.

The pathological condition is caused by wind (*grossa ventositate generata*) or water enclosed without exit (*aggregatione aque inclusae in eo, quae non invenit viam*), exiting upon death. Medical treatment was subsequently suggested and there are no citations of adult patients affected by hydrocephalus. Yet, idiopathic adult hydrocephalus is not reported in any of these Islamic medical manuscripts.

During this period, semantic trouble occurs when, in the 12th century, the main Muslim translator is Gerard of Cremona. Oribasius and Aetius described paediatric hydrocephalus with the Greek term: *ὄγκος*: 'mass, tumour, or swelling'. Later, the Muslim literature accurately translated the term into *ورام* - *waram*: 'swelling'. Gerard of Cremona translated *ورام* - *waram* into *'apostema'*: abscess, inadvertently changing the original meaning (Pormann, 2004). Since that point, for many centuries, the medical authors included some diseases (i.e. tumours, abscesses, aneurysms, hernias) under the term *'apostemata'*, and among these was paediatric hydrocephalus.

At the end of the late Middle Ages, paediatric hydrocephalus was detailed in some of the early non-Muslim surgical treatises. In 1268, Guglielmo da Saliceto (1210–1277, Italian physician, author of the earliest record of human dissection) published his first surgical book *'Chirurgia'*, promoting new concepts in opposition to contemporary medical and surgical procedures. In a vulgar Latin language printed edition *'La ciroasca'*, many *'postema'* causes of

disease are reported; the first was *'laqua assunada in li cavi de li fantolini novelamente nadi'*, 'the water collected in newborn children's heads' (Guglielmi, 1486). The Hippocratic classification and Muslim surgical treatment was detailed, but Saliceto writes *'ma io al tempo de mo non ho viduto nissuno che ne scampi ne credo che alcuno ne possa scampare over evader'*: 'I have never seen anyone surviving and I believe no one can survive'. Nevertheless, Saliceto provides the original description in two young children; the first is simply observed in one Cremona's *'hospitalia'*, and the second has medical and surgical treatment. In both cases, a successful result was obtained.

The most appreciated of Saliceto's students, Lanfranco di Milano (1250–1306, Italian physician), followed his teacher's classification (*apostematibus*) and nomenclature in the book *'Chirurgia Magna'* edited in 1296, but he did not describe personal observations of children affected by hydrocephalus (Tabanelli, 1965).

The famous Guy de Chauliac (c.1300–1368, French physician) followed Saliceto's classification and put hydrocephalus in children as the first *'apostemata'*: 'the distinct diseases', naming it *'aque in capitibus puerorum'*: 'water in children's heads' (Guidonis, 1513).

The progressive increase in medical literature on paediatric diseases was enriched in 1484 by the book *'Libellus aegritudinum puerorum'* by Cornelius Roelans (1450–1525, Flemish physician) (Roelans, 1484). Roelans described *'de apostemate cerebri puerorum'* in children, in which the water was collected in the head—*'de aqua in capite congregata'*—and was therefore responsible for large heads, *'magnitudine capitis'*.

Pietro d'Argellata (died 1423, Italian physician), composed *'Chirurgia'* sometime between 1391 and 1423. In a 1499 edition, congenital hydrocephalus was included among *'de apostematibus capitis'* and defined as *'De aquositate quae repint in capitibus puerorum'*: 'The watery fluid which resides in children's heads' (Petri de, 1499). The therapy was primarily based on natural vegetal unguents, but whenever unsuccessful results were obtained, surgical incision was proposed to allow the water to exit freely: *'Si autem post hunc modum curari non poterit, quia ista non sufficient siant cauteria punctualia...sic aqua possit libere exire'*.

As in the previous Muslim literature, not one of these authors reports a case of idiopathic adult hydrocephalus.

The Renaissance (1401–1600): the early hypothesis of adult hydrocephalus

During the Renaissance, human anatomy diffused into the medical studies and universities through anatomical dissections, with normal and abnormal anatomy becoming distinguished and practiced by artists such as Leonardo da Vinci, Michelangelo Buonarroti, Raffaello Sanzio, Antonio Pollaiuolo and many others (Andrioli et al., 2004; Frati et al., 2006). Human anatomy was reborn with Antonio Benivieni (1443–1502, Italian physician), the first pioneer to practice the autopsy. Posthumously, in 1507, Benivieni's treatise was published on 15 autopsies to determine the cause of death and correlate some autopsy findings with prior symptoms (Benivieni, 1507). The autopsy studies allowed

previous ventricle representations to be discarded based on Nemesius of Emesa's localization theory (Fig. 1).

Giovanni da Vigo (1450–1525, Italian physician), in his 1514 '*Practica in Chirurgia*', placed '*de aquositate in capitibus puerorum*' among '*De apostematibus frigidis et calidis*', and the cure was identical to that of the physicians in the Middle Ages (Ioannes, 1515).

As with Benivieni, the first edition of a book dealing with 34 paediatric diseases by Leonello Vittori (1450–1520, Italian physician) was posthumously printed in 1544. The author detailed the water enlarging the sutures in one child who had been mishandled by the midwife or suckler by excessively squeezing the head (Victoriis, 1546). Both medical and surgical therapy was proposed.

Another paediatric publication in 1549, by Sebastian Oestreicher (Austrius Sebastianus, died 1550, German physician), refers to the previous description of paediatric hydrocephalus by Guglielmo da Saliceto and Cornelius Roelans without any new suggestions (Austrius, 1549).

Giovanni Filippo Ingrassia (1510–1580, Italian physician and anatomist) in 1553 considers the hydrocephalus a cold tumour and reports: '*...bimulum puerulum Panhormi inspeximus cuius et coronalis, et recta suturae in tantum dilatatae sunt, ut digiti spatium interfuerit...per earum apertarum suturarum spatium diaphanum quoddam conspiciebatur, non aliam substantiam, quam purissimam aquam repraesentans*': '*...in Palermo we have observed a child two years old in which the coronalis and sagittalis sutures were so enlarged, that a finger could be placed...through these openings an opaque space was detected, which called to mind very pure water*'.

João Rodrigues de Castelo Branco (Amato Lusitano, 1511–1568, Portuguese physician), an experienced cadaver dissector, wrote the '*Centuriae duae*' in 1554 and reported a questionable case of a 15-day-old child with a head tumour diagnosed as hydrocephalus (Lusitani, 1554).

Puer quindecim dies natus de repente in tumorem magnum in capite tactu mollem lapsus est, quem morbum Geaeci hydrocephalon appellant: cui occurrens hoc usi sumus medicamento, quo intra tres dies ex toto inflatio illa evanuit, sic enim medicamentum habet.

Hydrocephalus disease was not well known in medical practice. Amato Lusitano advised an ointment made with chamomile, wormwood and melilot, obtaining successful results in 3 days.

In 1543, Andreas van Wessel (Vesalius, 1514–1564, Flemish anatomist) brought to light the first edition of the '*De humani corporis fabrica*'. In a 1555 edition, Vesalius reported on a 2-year-old girl seen in Augsburg (Vesalii, 1555):

...Augustae Vindelicorum in bienni puella observavi, cuius caput in mensibus plus minus septem ita increverat, ut nullum viri caput unquam viderim, quod non molle illi cederet. Fuitque is affectus, quem veteres hydrocephalum vocarunt, ab aqua quae in capite asservatur, sensimque colligitur...huic puellae fuerit collectaverum in ipsius cerebri cavitate, adeoque in dextro sinistroque illius ventriculis: quorum cavitas amplitudine ita increverat, ipsum cerebrum ita extensum fuerat, ut

novem fere aquae libras, aut tres Augustanas vini mensuras (ita me ament Superi) continuerint.

'Her head increased in more or less seven months, so much I had never seen a man's head, since to him (the head) should not soften.' Notably, hydrocephalus was never linked to enlarged ventricles in ancient medicine. In Vesalius the term '*hydrocephalum*' is correctly adopted, for the first time the cerebrospinal fluid is described to collect within the ventricles of the brain, and the amount is measured as 9 libras (3240 cc).

During these years, the academic circles were filled with the anatomic dispute between Andreas van Wessel and his colleague Matteo Realdo Colombo (Renaldus Columbus, c.1516–1559, Italian anatomist and surgeon). In his '*de re anatomica*', Columbus described the brain of patients who died from a powerful apoplexia, '*magnam aquae copia, perspicuam quidem, sed glutinosam*': 'a great amount of water, undoubtedly clear but glutinous', but not idiopathic adult hydrocephalus (Columbi, 1559).

Job Fincel (Fincelius, c.1520–1582, German humanist and physician) started the literature genre of the wonder-book. In 1556, Fincelius reported many miraculous events precluding the end of the world (Fincelius, 1556; Barnes, 1988). Among wonders and some natural happenings, in 1547 Fincelius observed an infant whose head enormously increased from the 14th day of life up to the fifth month.

In 1558, Jérôme de Monteux (Hieronymus Montuus, 1518–1559, French physician) reported a fabulous case of hydrocephalus in a baker (Montuo, 1558):

...augescere caput ad tantam magnitudinem vidimus, ut mole bovis caput superaret, et totam cum naso faciem occultaret. lussimus primum baculum in ore immitti, ne deperdita respiratione morbo succumberet. Mox omni diversionum genere, cum tenuissima victus ratione servatus est, sensimque caput detumuit.

'We saw the head increase in so large a degree to overcome the head of a cow, and all the face was covered by the nose. We ordered first to put a stick in the mouth, to avoid death by breathing stoppage due to the disease. Soon with every possible change, (the man) was nourished with very slight meals, and afterwards the head decreased.'

In 1573, Jacques Dalechamps (1513–1588, French physician and botanist) published his book '*Chirurgie française*' (Dalechamps, 1573). He extensively described the Greek aetiopathogenetic theories of children's hydrocephalus, connecting the previous Arab experience and reporting the surgical therapy (Fig. 2), and he cited the case of Andreas van Wessel:

M. André Vessal anatomiste excellent de nostre aage, ou plus-tost restaurateur de l'Anatomie oubliee, corrompue et depravee, escrit avoir veu à Ausbourg une fille de deux ans malade de cest'affection, qui avoit la teste plus grosse que nul homme...l'eau estoit accumulee iusques au poix de neus livres, qui est une chose admirable et presque incroyable: et s'estoit amasee en la capacité des deux ventricules antérieurs du cerveau.

Jacques Dalechamps was amazed at the 9 libras of water in the anterior ventricles, but he did not mention adult patients affected by hydrocephalus.

In 1582, Ambroise Paré (1510–1590, French surgeon) referred to the van Wessel child affected by hydrocephalus and reported briefly on his surgical experience in four children (Paré, 1582).

In 1583, Geronimo Mercurialis (1530–1606, Italian physician and philologist) published the first known treatise about paediatric disease (Mercurialis, 1583). The author performed a very deep analytical review of paediatric hydrocephalus, cited all previous literature from Aristoteles to Rhazis, and stated that ‘...*Quamquam me non lateat etiam in adultioribus eum fieri*’: ‘...Although I have not seen any, (the hydrocephalus) can occur also in older (patients)’. The statement is not well defined but can be considered the first hypothesis regarding adult patients affected by hydrocephalus and the first connection with paediatric hydrocephalus.

During these years, the Latin translation from the Hippocratic Greek ‘*Ην ὕδωρ ἐπὶ τῷ ἐγκεφάλῳ...*’: ‘water **over** the brain’ was ‘*Aqua si in cerebro suborta fuerit...*’: ‘water **in** the brain’ (Cornarius, 1546; Mercurialis, 1588; Foesio, 1595). It is possible that Andreas van Wessel’s previous observation of cerebrospinal

fluid within the ventricles in congenital hydrocephalus supported this translation, opening a new surgical perspective, ventricular drainage of cerebrospinal fluid.

Gabriele Falloppio (Fallopius, 1523–1562, Italian anatomist and physician) was demoralized by the surgical results of paediatric hydrocephalus. Surgery is proposed as an extreme option for congenital hydrocephalus due to its high mortality: ‘...*quasi tutti morono coloro, che hanno l’acqua di sotto dal craneo, anzi che un solo fu da me aperto, e commutò la vita in morte*’ (Fallopio et al., 1637).

In 1597, Schenck von Grafenberg (1530–1598, German physician) collected observations of children with congenital hydrocephalus from Rhases, Albucasis, Avicenna, Lusitanus, Fallopius, Pareus and Fincelius (Schenckius, 1597, 1600). Andreas van Wessel’s child was described as ‘*hydrocephali nova species-res mira*’: ‘a new species of hydrocephalus—an extraordinary thing’ because the water was inside the cerebral ventricles.

The post-Renaissance or prelude period: 1601–1761

Radical changes came about in 1600 when numerous European scientists discovered and demonstrated new theories, theorems and laws. The congenital paediatric hydrocephalus appeared in many books all over Europe with the first images (Fig. 3). Danish (Bartholin, 1670), Dutch (Tulpii, 1652; Tulpius, 1670), German (Sennerto, 1633; Cummi, 1670; Wilhelm, 1682; Schröck, 1696), Italian (Cortesi, 1632; Fabricii, 1648), Scottish (Burnet, 1678) and Swedish (Lohrman, 1666) physicians describe children affected by this disease. Among these, Daniel Sennert (1572–1637, German physician) and Thomas Burnet (1635–1715, Scottish physician) in 1633 and 1678, respectively, wrote: ‘*Quidem hydrocephalo, qui tumor capitis totius aquosus est, quandoque etiam adulti corripiuntur...*’ (Sennerto, 1633) and

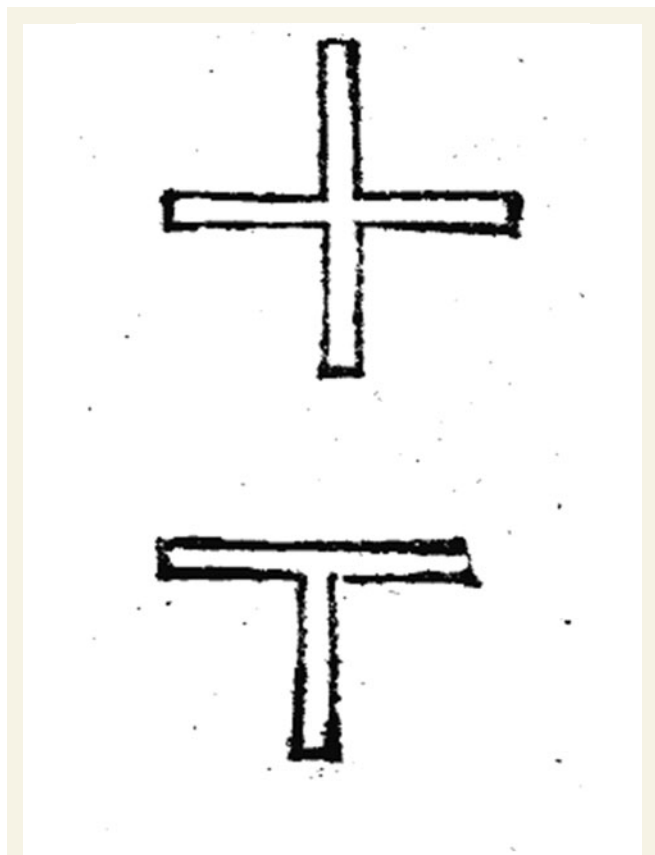


Figure 2 Jacques Dalechamps proposes the same cutaneous surgery and surgical incisions performed by the Greek and Muslim physicians. In children this procedure will be performed till the 19th century. To note the skull’s trephination is not reported by any of these physicians due to the anatomical gap allowing cerebrospinal fluid’s subcutaneous escape (Dalechamps, 1573).

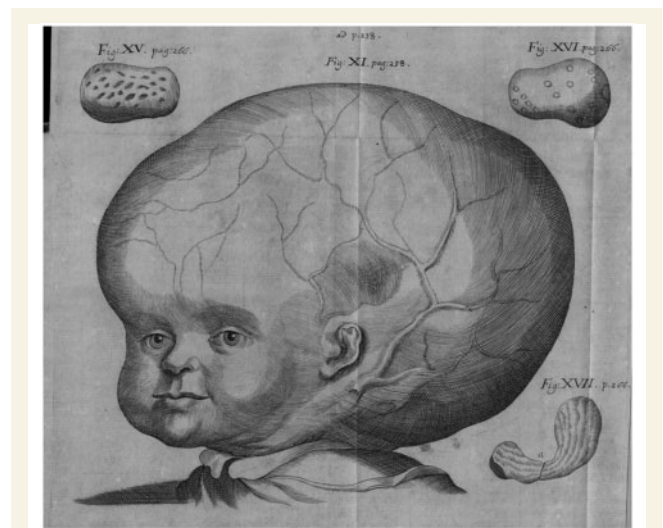


Figure 3 An early representation of paediatric hydrocephalus from Schröck Lucas (1646–1730, German physician and naturalist) contribute to diffuse the knowledge of the disease. The graceful appearance of the child lacks scientific representation. From *Biblioteca Nazionale Centrale di Roma*.

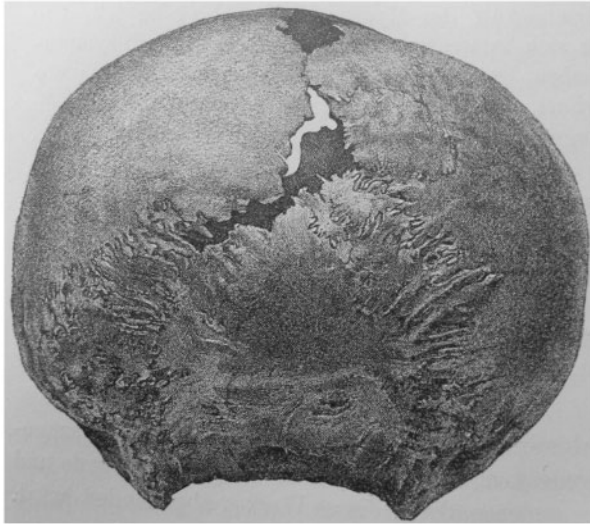


Figure 4 Congenital adult hydrocephalus. Posterior view of Borduni's skull (Schultze, 1901). The posterior sagittal and left lambdoid sutures are partially closed (private collection).

'*De Hydrocephalo. Infantibus malum hoc accidit frequentius quam adultis*' (Burnet, 1678). Hydrocephalus is not confined to children but can also occur in adults. Jacob Spon (1647–1685, French physician and archaeologist) and George Wheler (1650–1723, English botanist) write an account of the 1675–1676 trip to Italy, Greece and Asia Minor in the same year (Spon *et al.*, 1678). As a curiosity, Spon and Wheler reported what appears to be the first description of a hydrocephalic skull in an adult: '*j'aime mieux vous entretenir d'une tête prodigieusement grosse que l'on conserve au convent de l'Observance. C'etoit la tête d'un nommé Borduni fils d'un Notaire de Marseille*': 'I would rather speak of a prodigiously large head that is retained in the convent of the Observance. This was the head of a man appointed Borduni son of a notary in Marseille'. The man died at the age of 50 years in 1616, and the skull can be seen in the Salle d'ostéologie of the Muséum d'Histoire Naturelle de Marseille (Fig. 4).

During those years, Théophile Bonet (1620–1689, Swiss physician) published in French and Latin his extensive textbooks '*Sepulchretum Anatomicum*', counting over 3000 autopsies appearing between the end of the fifteenth and the first-half of the 17th century (Bonet, 1670, 1679). In the section on apoplexy of the '*Sepulchretum*', there are 22 cases with variable fluid in the head, but it is not clear whether it represents hydrocephalus or normal cerebrospinal fluid (Schutta and Howe, 2006). On the other hand, paediatric hydrocephalus is detailed in four children, but there is a lack of correlation with a large amount of serum found in the heads of adults (Bonet, 1670). The '*Sepulchretum Anatomicum*' inspired Morgagni's work that followed.

The assumption that hydrocephalus can happen in adult patients continued to be stated through the 18th century. In 1712, Veit Riedlin (1656–1725, German physician) (Fig. 5) detailed the first case of adult congenital hydrocephalus in the medical literature (Riedlin, 1712). Riedlin described the clinical history and anatomical findings of a 24-year-old man living in Augsburg from



Figure 5 Veit Riedlin, who in 1712 published the first medical report of adult congenital hydrocephalus in a 24-year-old man (private collection).

1678 to 1703: '*horrendae magnitudinis capite instructus*'. Cognitive function was not altered, '*... intellectu tamen semper sic gaudebat ...*', though the use of the hands was impaired after an apoplectic event, '*... nec manuum usu destitutum fuisse; at supervenisse tum apoplexiae quendam insultum, qui eum sic debilitavit, ut ex illo tempore artuum usu omnino fere destitutus vivat ...*'. Riedlin noted the closure of the sutures, '*tam arcte clausae conspiciebantur*', in contrast to younger patients who had an enlargement of the sutures, '*uti in aliis hoc morbo defunctis suturae minus firmiter cohaerentes quin hiantes repertae fuere*'.

Theodor Zwinger (1658–1724, Swiss physician, physicist and botanist) made undefined rare observations of hydrocephalus in adult patients in 1724: '*Dantur etiam observationes, licet raras, hydrocephali in adultis*' (Zwinger, 1724). Zwinger's observation is possibly a reference to Riedlin's report and other adult congenital hydrocephalus cases published afterwards (Ekmark, 1763; Michaelis, 1784; Gall, 1810; Elliotson, 1844; Vrolik, 1854; West, 1854). In all of these cases, the large heads of patients from infancy support a congenital origin of hydrocephalus, not allowing for a correct diagnosis of idiopathic adult hydrocephalus.

Early descriptions of idiopathic adult hydrocephalus (1761–1851)

Anatomy acquired the first modern pathologist in Giambattista Morgagni (1682–1771, Italian anatomist and pathologist), who

correlated living symptoms with post-mortem anatomical features. In his 1761 Latin masterpiece, Morgagni detailed 700 autopsy cases, among which some findings could support the early description of idiopathic adult's hydrocephalus (Morgagni, 1761). Morgagni distinguished '*de apoplexia serosa*' from '*de hydrocephalo*' as two distinct entities. Hydrocephalus was extensively reported in children from both his anatomical dissections and the observations of other authors. The opinion at that time was, in children, a dissolution of the brain by the water, but Morgagni stated that the mechanism can also occur rarely in adults ('*Cum igitur totius adutorum cerebri in mucum, nedum in aquam dissolutio sic rara sit*'—Liber I, Epist. XII, Case 14, p. 95). Morgagni differentiated between more (*multa*) and less (*pauca*) cerebrospinal fluid (*aqua*) in the ventricles. Although the limit is unclear, Morgagni determined the amount of cerebrospinal fluid in ounces (*uncia*) or pounds (*libra*) in some cases. In May 1727, Morgagni performed an autopsy on a 60-year-old male (Liber I, Epist. VIII, Case 27, p. 63). The clinical picture before death was characterized by impaired cognitive features. The major findings were blood collection in the lungs and a serous pericardial accumulation of 3 ounces. In the cerebral ventricles, Morgagni measured 3 ounces of cerebrospinal fluid (90 cc). In a 63-year-old man, the amount of water in the ventricles was 2 ounces (60 cc) (Liber I, Epist. IV, Case 2, p. 26). One more case was that of a 70-year-old male who died one month after a fall due to secondary complications. In the ventricles, Morgagni found 4 ounces of serous fluid (120 cc) (Liber IV, Epist. LI, Case 9, p. 286). In no case did Morgagni associate the copious cerebrospinal fluid in the ventricles with paediatric hydrocephalus.

In 1751, the first edition of the world encyclopaedia was published by Diderot and D'Alembert. Hydrocephalus disease was reported only in the 1782 edition, and the congenital origin was related to children (Diderot et al., 1782). Adult patients were not considered because '*dans les adultes les sutures serrées ne permettent pas la distension des os du crâne*': 'in the adults the sutures close not allowing distention of the bones of the skull'.

At the end of the 18th century, many journal accounts reported the death of Jonathan Swift (1667–1745, Irish author, journalist and prose satirist of '*Gulliver's Travels*'), dean of St. Patrick's Cathedral, and one of Dublin's foremost citizens. Swift became insane in his last years due to a progressive impairment of memory, language and cognitive functions over 3 years, leading to dementia. Death occurred at 78-years-old and autopsy 'found much water in the brain', 'to such an amount as to fill the basin and by their quantity to call forth expressions of astonishment from the medical gentlemen engaged in the examination' (Swift, 1763; Houston, 1835).

In 1802, William Heberden, Jr. posthumously published his father's (William Heberden, Sr., 1710–1801, English physician) book '*Commentarii de morborum historia et curatione*', the same book in which '*Pectoris dolor*' or *angina pectoris* is republished and 'Heberden's nodes' of osteoarthritis are described for the first time; and a contemporary English translation (Heberden, 1802a, b). In two sentences, the pathological description of an adult patient with copious cerebrospinal fluid in the ventricles is found:

Adultus quidam impatibili dolore capitis occupatus est, cupiditas cibi modo summa erat, modo nulla delirabat membra

distendebantur stupebat et brevi exinctus est. Ventriculi cerebri adeo turgebant ut scalpello adacto aqua in notabilem altitudinem exiliret.

'An adult was seized with intolerable pains of the head, sometimes had a voracious appetite, and sometimes none, became delirious, convulsed, stupid, and in a short time died. The ventricles of the brain were found so distended with water that, as soon as a puncture was made, the water flew out a considerable distance.'

In 1813, Matthew Baillie (1761–1823, Scottish physician and pathologist), writing in *Medical Transactions*, reported the case of a 56-year-old man who died after an 11-month history of motor aphasia and right hemiparesis (Baillie, 1813).

I had much more than a common curiosity to examine the brain after death...and the brain was examined in my presence on the 8th of January, 1806, viz. two days after he died.... The lateral ventricles of the brain however were found to contain rather more than six ounces of water (180 cc)... The result of this examination very much surprised me. Although symptoms of pressure upon the brain were very strongly marked, yet none had occurred during any part of the disease, which usually denote the accumulation of water in the ventricles.

In 1815, William Heberden, Jr (1767–1845, English physician) reported the clinical and pathological findings of a patient 'some years turned of fourscore. I had attended him occasionally, but not often, during the last ten years of his life' (Heberden, 1815). The main complaint was increasing deafness, and an acute stroke was the cause of death.

An able surgeon who conducted the examination' writes: '...The substance of the brain was healthy. The ventricles were greatly enlarged, and contained about eight ounces (240 cc) of transparent water...' Heberden was surprised 'that so large an accumulation of fluid (not less than three quarters of a pint), and consequently so great a compression, should ever be sustained with little or no manifest inconvenience. The reason of this may perhaps be suggested by the very gradual manner in which it may be supposed to have been collected, and in that wonderful power, with which every living body is endowed, of accommodating itself to new circumstances....'

In 1818, Leopold Anton Gölis (1765–1827, Austrian physician and pathologist) (Fig. 6), a paediatrician and dissector in the Institute for the Sick Children of the Poor in Vienna, described the clinical and autopsy findings of many children affected by hydrocephalus, opening 180 bodies that died of this disease. Due to Gölis' great experience with children, he described hydrocephalus in a 35-year-old man, case XXXVI, with 8 ounces (240 cc) of serum in the brain ventricles and refers to three old patients affected by hydrocephalus but lacking the typical paediatric external cranial vault alterations (Gölis, 1815, 1818) (Figs 7–10).

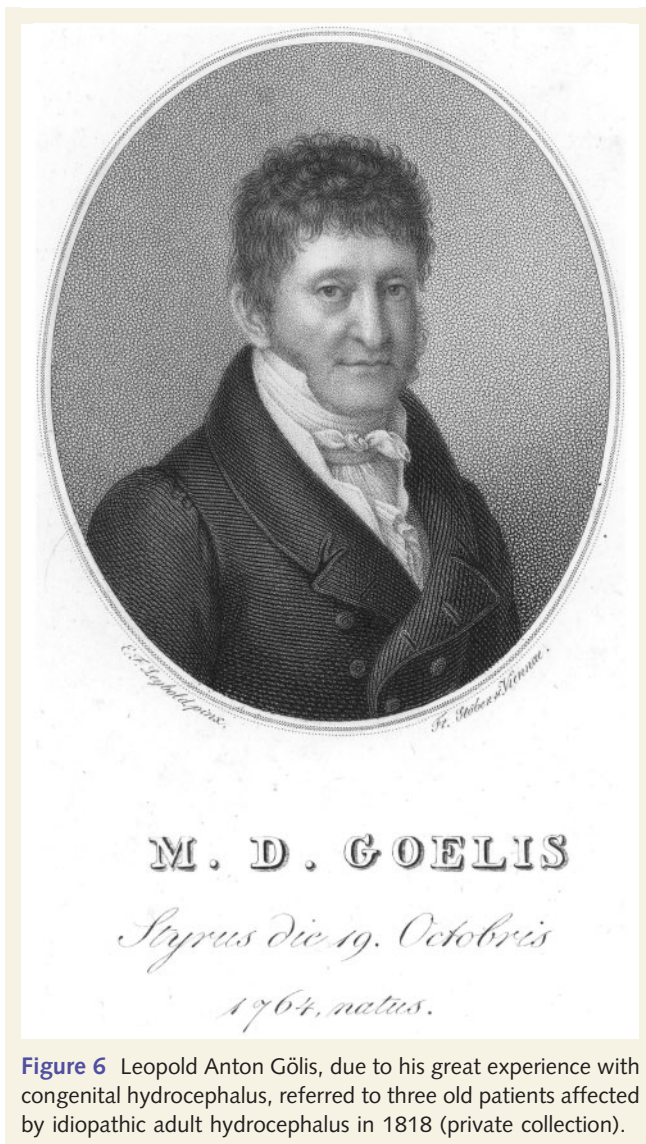


Figure 6 Leopold Anton Gölis, due to his great experience with congenital hydrocephalus, referred to three old patients affected by idiopathic adult hydrocephalus in 1818 (private collection).

Liegt nun aber gleich schon im Leben und in der Beschaffenheit des Kindes vorheräschend die Disposition zu diesem Leiden, so fehlt es doch nicht, wenn gleich an seltneren Beispielen von Jünglingen, Männern und Greisen, welche am chronischen Wasserkopfe zu leiden anfangen.

Ein f.f. Hoftrompeter starb in seinem 79sten Jahre, und ein Erzieher junger Fürsten im 71sten seines Alters an der chronischen innern Kopfwassersucht, und in einem noch vor wenig Jahren allgemein geschätzten und vielerfahrenen Arzte Wiens ist dieses Leiden schon deutlich entwickelt. Bei keinem dieser drei Kranken bemerkte ich an der äussern Form des Kopfes eine Veränderung, was Bonet, Zadutus, Fallopius, Rosenstein, Morgagni, Burserius, Storch, Riedler, Wichmann und Loder in ähnlichen Fällen behaupten.

Wenn im hohern Mannes und Greisenalter bei der vollendeten Vereinigung der oft völlig verschmolzenen Suturen das feste Knochengewölbe dem innern Drucke der stagnirenden sich häufenden Flüssigkeit einen unbezwingbaren Damm entgegensehrt, und deren Gewalt einzig gegen das mitten liegende

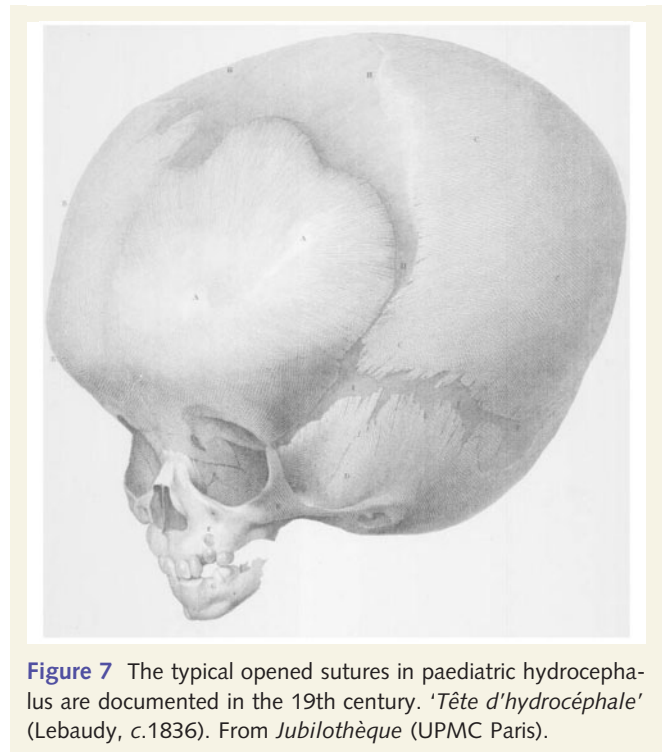


Figure 7 The typical opened sutures in paediatric hydrocephalus are documented in the 19th century. 'Tête d'hydrocéphale' (Lebaudy, c.1836). From *Jubilothèque* (UPMC Paris).

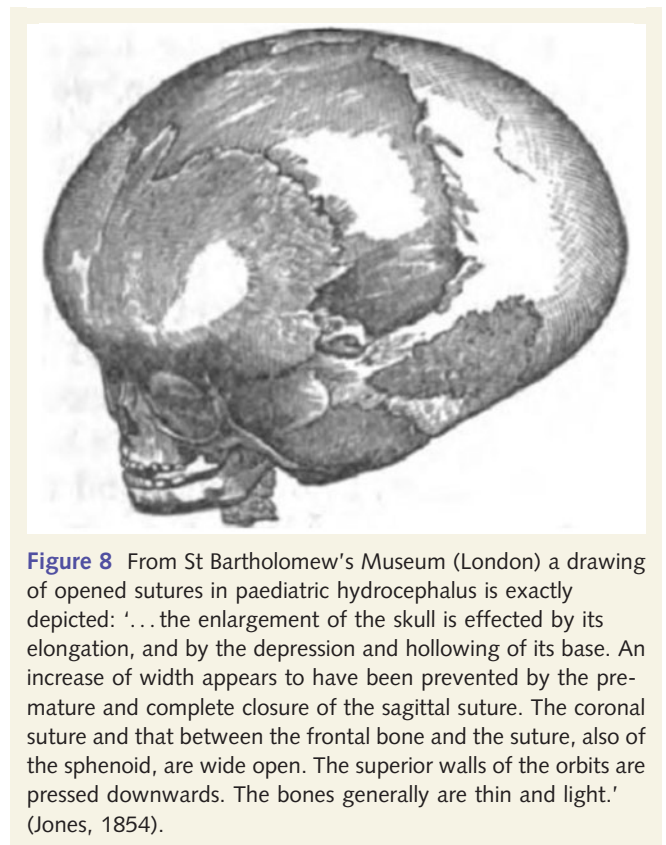


Figure 8 From St Bartholomew's Museum (London) a drawing of opened sutures in paediatric hydrocephalus is exactly depicted: '... the enlargement of the skull is effected by its elongation, and by the depression and hollowing of its base. An increase of width appears to have been prevented by the premature and complete closure of the sagittal suture. The coronal suture and that between the frontal bone and the suture, also of the sphenoid, are wide open. The superior walls of the orbits are pressed downwards. The bones generally are thin and light.' (Jones, 1854).

Gehirn hinwirkt so ist im kindlichen Alter die Kraft bemerkenswerth, mit der die Ränder der Schädelknochen auseinander gedrängt, und die Knochensubstanz selbst gedrückt und gedehnt wird.



Figure 9 The closure of the sutures may occur in children which survive hydrocephalus. 'Skull of an hydrocephalus child.' (Vimont, 1832).

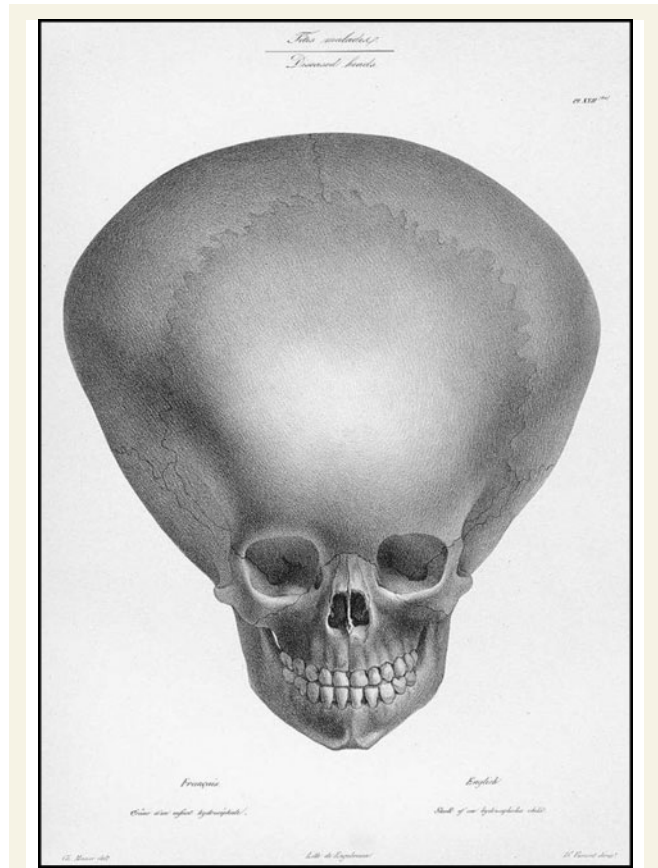


Figure 10 Closed sutures in paediatric hydrocephalus. 'The head of a child some 5 years of age... Every portion of the cranium was of stony hardness, and with no amount of pressure with the fingers could we produce the slightest indentation' (Blackman, 1854).

'Although the disposition for this disease is mainly present during life in children, there are, rarely, cases of youths, adults and old men who start to suffer chronic hydrocephalus.

A trumpet player died in his 79th year, and an educator of young princes in his 71st year, and an experienced medical doctor appreciated worldwide up to a short time ago developed this disease.

In nobody was there an external cranial vault alteration, like in similar cases of Bonet, Zadutus, Fallopius, Rosenstein, Morgagni, Burserius, Storch, Riedler, Wichmann and Loder.

In adult and old age, when skull sutures are closed, the internal pressure exerted by the stagnant fluid, progressively increasing, meets in the hard bone an impossible hamper; therefore, the pressure pushes with his strength against the brain. In young age, the high forces produce the expansion of the bone flaps and dislocation of the cranial vault.'

In 1819, Etienne Moulin (1795–1871, French physician) wrote the '*Traité de l'apoplexie, ou hémorrhagie cérébrale*' (Moulin, 1819). In old patients he noted:

Cette maladie est extrêmement commune. Elle a été méconnue jusqu'à ce jour; aucun ouvrage sur les hydrocéphales n'en fait

mention: une observation plus attentive et des circonstances aussi favorables que celles où l'auteur s'est trouvé n'auraient sans doute pas manqué de la faire reconnaître ...

Les vieillards décrépits et les sujets affaiblis sont ceux qu'elle atteint de préférence ...

La début de l'hydropisie est toujours très-let, et sa marche progressive; les signes de la compression cérébrale ne se dessinent que par degrés ...

L'état du malade s'aggrave à mesure que l'épanchement cérébral augment ...

La mort est la terminaison la plus fréquente de l'hydrocéphale chronique, surtout lorsqu'elle affecte des vieillards très décrépits.

'The disease is extremely common. It was unknown until this day; no book on hydrocephalus makes mention: a more careful observation and favourable conditions as those the author has found, would probably not fail to recognize ... The old decrepits and the weakened subjects are those reached preferably ... The beginning of the dropsy is still very slow, and progressive; the signs of brain compression develop by degrees ... The patient's condition worsens as the brain effusion increases ... Death is the most common

end of the chronic hydrocephalus, especially when it affects old people much deteriorated.'

In 1826, Friderico Dörner (Friedrich Dörner, German physician, dates unknown) discussed his medical thesis '*De hydrocephalo chronico senili*' at the University of Würzburg (Dörner, 1826). The author referred briefly to Morgagni, Gölis and Moulin as previous and related descriptions of hydrocephalus in adults. In his '*dissertatio*', the author described various symptoms that he believed were related to the disease. Among these symptoms, urinary and walking impairment was reported: '*In Hydrocephalo vesica urinaria et intestinum rectum paralyti afficiuntur*' and '*Hydrocephalici vacillant*'. Out of three '*exempla morbi*', two autoptic findings are compatible with a diagnosis of idiopathic adult hydrocephalus. In a 73-year-old man, the cerebral lateral ventricles were filled with clear water and all the posterior horns were enlarged; in the cranial cavity the amount of water was 6 ounces (180 cc): '*Ventriculi laterales sero limpido referti et cornibus inprimis posterioribus admodum divaricati*', '*Aqua in cranii cavo contenta unciarum sex pondus fere aequavit*'. In a 67-year-old man, the lateral ventricles were filled and enlarged with 5 ounces (150 cc) of clear water: '*Ventriculi omnes limpida aqua, pondere fere unciarum quinque expleti et dilatati*'.

In 1831 and 1836, Gabriel Andral (1797–1876, French physician) roughly described chronic hydrocephalus in adult patients (Andral, 1831, 1836):

Some writers have of late described under the name of chronic hydrocephalus of the old a disease characterized by the gradual weakness of the various cerebral functions in which as they assert there is nothing to be found on examination after death but a considerable accumulation of serum either in the subarachnoid cellular tissue of the external surface of the brain or in the ventricles.

Enfin exhalation graduelle et insensible constituant l'hydrocéphale chronique, maladie très différente chez l'enfant et le vieillard, liée chez le premier à un défaut de développement du cerveau, très rare chez l'adulte, assez fréquente chez le vieillard, et caractérisée par un affaiblissement graduel de l'intelligence et des mouvements, puis par le coma, respiration stertoreuse qui amène la mort. Toutes ces diverses formes réclament un traitement plus ou moins actif qui consiste spécialement dans les révulsifs.

'Finally, gradual and imperceptible exhalation produces chronic hydrocephalus, an illness very different in children and the elderly, the first linked to altered brain development, very rare in adults, fairly common in the elderly and characterized by gradual weakening of the intelligence and movement, followed by coma, which causes breathing impairment until death. All of these forms require a more or less active treatment, which especially consists of revulsive therapy.'

After these descriptions, idiopathic adult hydrocephalus appears to have been forgotten in the medical literature, although some progress in the surgical treatment of paediatric hydrocephalus was published from the end of the nineteenth through the 20th century.

In 1949's milestone book by Dorothy Russel (1895–1983, physician and pathologist) dealing with hydrocephalus, she stated that the adult form was 'the most obscure' (Russell, 1949). According to Russel, all reported hydrocephalus cases can be placed in the category of maldevelopment, gliosis of the aqueduct, inflammation, sinus thrombosis, or neoplasm. The use of the term 'idiopathic in connection with hydrocephalus' was not considered because a post-meningitic process should be regarded as the causative factor.

This position was followed by this outstanding description of the clinical symptoms and radiological features of idiopathic adult hydrocephalus by four French physicians in 1950 (Roger *et al.*, 1950):

'M.me L. . . , 68 ans. En 1948, elle commence à éprouver des difficultés pour marcher; elle avance à petits pas un peu saccadés trainant la jambe droite, et son entourage remarque un aspect anormal du regard. En novembre 1949, se produit une aggravation brutale. M.me L. doit s'aliter du jour au lendemain, sans ictus vrai, avec une hémiparésie bilatérale, des troubles passagers de la parole, une incontinence sphinctérienne et une accentuation considérable des troubles psychiques: indifférence affective, amnésie confusionnelle, grande apathie physique et intellectuelle. . . . La ventriculographie révèle une énorme dilatation des ventricules latéraux, du type carré, avec des cornes temporales moins distendues que les autres. . . . Il s'agit donc d'une hydrocéphalie communicante.

'M.me L. . . , 68 years old. In 1948, she began to experience difficulty walking, it has been slow and a bit jerky training the right leg, and her caretaker noticed an abnormal gaze. In November 1949, she has fast worsening. M.me L. was in bed the day after, without true stroke, with bilateral hemiparesis and transient disturbances of speech, sphincter incontinence and a considerable increase in mental disorders: affective indifference, amnesia, confusion, great physical and intellectual apathy. . . . The ventriculography reveals a huge dilatation of the lateral ventricles, square type, with temporal horns less enlarged than the others. . . . This is a communicating hydrocephalus.'

In 1964 Paul McHugh (1931–, English physician and psychiatrist) published some cases of 'occult hydrocephalus' (McHugh, 1964). In two asymptomatic patients (64- and 72-year-old) the post-mortem findings showed unexpected 'marked symmetrical dilatation' of the ventricular system. One other patient (52-year-old) presented a progressive difficulty in gait and cognitive functions. A ventriculogram demonstrated enlargement of the lateral and third ventricle, but after a ventriculo-cisternostomy the patient died due to infections.

Discussion

For many centuries, physicians followed the Hippocratic and Galenic doctrines of humors, and hydrocephalus was known and treated in children only as an extracranial or subdural fluid collection, which is clearly far from the current definition of hydrocephalus. At the end of the Middle Ages and the beginning of

Table 1 Early descriptions of patients affected by idiopathic adult's hydrocephalus

Author	Year	Number of patients	Age (years)
Giambattista Morgagni	1761	3	60, 63, 70
Matthew Baillie	1813	1	56
William Heberden, Jr	1815	1	> 80
Leopold Anton Gölis	1818	3	71, 79
Etienne Moulin	1819	–	–
Friedrich Dörner	1826	2	73, 67
Gabriel Andral	1833	–	–
Henri Roger	1950	1	68
Paul McHugh	1964	3	64, 72, 52

the Renaissance, the reports from Guglielmo da Saliceto (Guglielmi, 1486), the extension of surgical practice, the birth of paediatric science, the introduction of printed books, and the use of anatomic dissections changed the approach of the medical culture. In 1543, Andreas van Wessel was the first to describe hydrocephalus as a cerebrospinal fluid collection inside enlarged ventricles, completely changing the ancient concept of hydrocephalus (Vesalii, 1555; Grunert, 2007). Geronimo Mercurialis was the first to publish a paediatric book with a chapter on children's hydrocephalus and to realize a hypothesis of adult hydrocephalus (Mercurialis, 1588). This author presumably links the grotesque adult case of congenital hydrocephalus by Jérôme de Montoux (Montuo, 1558). Jacob Spon and George Wheler mentioned the first extraordinary case of adult congenital hydrocephalus (Spon, 1678). Veit Riedlin published the first medical report of adult congenital hydrocephalus in 1712, observed in his medical practice along with pathological findings after death.

The first observations of idiopathic adult hydrocephalus were related to the cadaver reports of Giambattista Morgagni (Morgagni, 1761) (Table 1). Interestingly, Morgagni measured the amount of water in the ventricles (procedure already performed in paediatric hydrocephalus) instead of determining the size of the wall ventricles. This methodological approach was followed in subsequent accounts from Baillie, Heberden Jr and Dörner. Both Heberden Jr. and, later, Dörner reported enlarged brain ventricles together with cerebrospinal fluid measurements for the first time. It is interesting to note that, only with the introduction of ventriculography by air from Dandy in 1918 'to produce a shadow in the radiogram' (Dandy, 1918) and Evan's ratio (Evans, 1942), that the diagnosis of hydrocephalus was made through ventricular size calculation.

The clinical picture of idiopathic adult hydrocephalus was very slowly delineated in the literature. The history of Jonathan Swift is suggestive of the clinical picture and autopsy findings of idiopathic adult hydrocephalus. Yet, the early descriptions of idiopathic adult hydrocephalus are pathological findings in asymptomatic patients (Morgagni, Baillie, Heberden Jr). Gölis, due to his experience with paediatric hydrocephalus, was the first physician who clearly associated hydrocephalus with adult patients, recognizing the possible cause of progressive neurological impairment. Moulin, Dörner and Andral gave us early outstanding descriptions of symptoms related to idiopathic adult hydrocephalus. The most detailed triad of

symptoms in idiopathic adult hydrocephalus was related by French neurologists in 1950; they summarized the clinical picture characterized by progressive walking, cognitive, and urinary impairment, which for the first time were confirmed with the radiological picture of enlarged ventricles. The Dorothy Russell's point of view was adopted in 1964 by McHugh. Indeed he put the adult congenital together with idiopathic adult hydrocephalus assuming a decompensation in adult life of a long-standing congenital hydrocephalus.

When Salomón Hakim and Raymond Adams described six adult patients affected by hydrocephalus (Hakim and Adams, 1965), only three were primary hydrocephalus patients (aged 52, 63 and 66 years), whereas the others were secondary to trauma or cyst. These authors conjoined the gait, urinary and cognitive impairment as a clinical peculiarity of patients affected by INPH. The very relevant result was the responsiveness of these patients to lumbar tap test and shunt surgery.

In conclusion, the discovery of idiopathic adult hydrocephalus followed a mosaic scheme in which many distinguished European physicians (anatomists, pathologists and paediatricians) contributed to putting the pieces in place. The neurological research by Salomón Hakim and Raymond Adams closed the main background of the disease, but opened some fuzzy sides of the pathogenesis that are still unresolved.

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