



Hydrocephalus in children under the age of five from diagnosis to short-/medium-/long-term progression: a retrospective review of 142 children

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Abstract

The aim of this study is to evaluate the clinical history and prognosis of children with early-onset hydrocephalus. The retrospective study's inclusion criteria were hydrocephalus diagnosis before the age of 5 years, independent of aetiology, and birth details, January 1, 2000 to December 31, 2014. Overall, 142 children were entered into the study, divided into 11 aetiological groups: premature-birth post-intraventricular haemorrhage (16%), brain tumours (16%), spina bifida (15%), aqueductal stenosis (8%), post-meningitis (8%), post-haemorrhage (8%), Dandy–Walker malformation (6%), unknown origin (6%), arachnoid cyst (5%), miscellaneous obstruction (4%), and various causes (8%). In total, 23 patients died, primarily from the tumour group. Ventriculostomy, performed 42 times, was successful in 20 patients. Overall, 226 internal shunts were placed in 99 children. Infectious complications affected 19% of children after shunt placement and 51% after mechanical complications. Mean follow-up was 4 years 10 months, with 61% of children progressing fairly well, especially those with aqueductal stenosis, cysts, and unknown or diverse obstructive causes. Post-meningitis hydrocephalus displayed the poorest outcome. Isolated obstructive hydrocephalus exhibited better prognosis, with most obstructive aetiologies effectively treated via ventriculostomy. Children treated by shunt placement were more at risk of complications. Aetiologies with associated abnormalities and neurological sequelae had poorer outcomes.

Keywords Hydrocephalus · Paediatrics · Development · Prognosis

Abbreviations

CNS	Central nervous system
CSF	Cerebrospinal fluid
ETV	Endoscopic third ventriculostomy
PHH	Post-haemorrhage hydrocephalus
PIVHH	Post-intraventricular haemorrhage hydrocephalus
PMH	Post-meningitis hydrocephalus
VPS	Ventriculoperitoneal shunt
QOL	Quality of life

Introduction

Hydrocephalus is a common neurological event caused by several pathologies [1]. Based on its physiopathology, a structured aetiological classification can be made. Obstructive hydrocephalus arises from an obstruction in the ventricular system, with idiopathic aqueductal stenosis, X-linked hydrocephalus, brain tumours (especially infratentorial), or Dandy–Walker and Chiari malformations as potential aetiologies. Obstructive hydrocephalus appears to be a good indication for endoscopic third ventriculostomy (ETV). Communicating hydrocephalus is caused by deficient resorption of cerebrospinal fluid (CSF) at the arachnoid villi, as seen in post-haemorrhagic and post-infectious hydrocephalus. Given this setting, a ventriculoperitoneal shunt (VPS) often proves more effective than ETV. However, both phenomena can occur simultaneously, which renders it difficult to choose the most appropriate treatment. Finally, a rare cause of hydrocephalus is choroid plexus papilloma, diagnosed before the age of 2, associated with abnormally increased CSF production [1–4].

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Table 1 Population characteristics

Aetiology	N (%)	Death (%)	Mean age at diagnosis	Mean age at treatment	Mean follow-up time
PIVHH	23 (16)	5 (22)	10 d	1 mo 3 d	65 mo 9 d
Brain tumour	23 (16)	12 (52)	32 mo 6 d	32 mo 14 d	29 mo 9 d
Spina bifida	21 (15)	3 (14)	12 d	1 mo	87 mo 14 d
Aqueductal stenosis	12 (8)	0 (0)	2 mo 6 d	6 mo 6 d	41 mo 21 d
PMH	12 (8)	1 (8)	5 mo 6 d	5 mo 12 d	63 mo 6 d
PHH	11 (8)	0 (0)	13 mo	13 mo 6 d	73 mo 24 d
Dandy-Walker	9 (6)	1 (11)	2 mo 21 d	12 mo 24 d	55 mo 21 d
Unknown	8 (6)	0 (0)	11 mo 18 d	12 mo 24 d	50 mo 21 d
Arachnoid cyst	7 (5)	0 (0)	7 mo 21 d	9 mo 9 d	57 mo 21 d
Miscellaneous obstruction	5 (4)	0 (0)	4 mo 18 d	4 mo 18 d	35 mo
Various	11 (8)	1 (9)	4 mo 27 d	5 mo 12 d	60 mo
TOTAL	142 (100)	23 (16)	8 mo 14 d	10 mo 6 d	58 mo

N number, *mo* months, *d* days, *PIVHH* post-intraventricular haemorrhage hydrocephalus, *PMH* post-meningitis hydrocephalus, *PHH* post-haemorrhage hydrocephalus

Currently, hydrocephalus is treated using either ETV or VPS, yet complications can occur. These mostly involve infection (meningitis or peritonitis) or mechanical problems (ETV failure, shunt malfunction, obstruction, migration, etc.). Complications are more commonly seen with VPS than ETV. Intracranial haemorrhage is a serious ETV complication occurring during or immediately following the procedure [1–4].

Outcome depends on aetiology, associated malformations, delay until treatment, and occurrence of complications [1–3].

The main aim of this study was to evaluate: (1) the prognosis of children diagnosed with hydrocephalus of several aetiologies before the age of 5; (2) determinants of outcome like aetiology, age at diagnosis and treatment, treatment type, complications or sequelae, as well as associated neurological abnormalities.

Method

This clinical trial was conducted at the “Cliniques universitaires Saint-Luc” in Brussels, Belgium. The hospital archives were searched for cases using International Classification of Diseases (World Health Organization) codes 331.3, 331.4, 741.00, 741.03, and 742.3. The children were born January 1, 2000–December 31, 2014.

All cases involved hydrocephalus diagnosed before the age of 5, including all aetiologies. The exclusion criteria were absence of hydrocephalus (codification error), external and ex vacuo hydrocephalus, prenatal diagnosis with miscarriage or abortion, and incomplete files. For each patient, the following parameters were collected and recorded: age and symptoms at onset; age at treatment and treatment type;

occurrence of complications, neurological impairments or sequelae and their therapeutic sanctions; length of hospitalizations. All events occurring after December 31, 2014 were not considered.

Population

A total of 142 children were included in the trial and divided into 11 groups according to hydrocephalus aetiology: post-intraventricular haemorrhage in premature children (PIVHH, 16%), brain tumour (16%, of which 61% were infratentorial), spina bifida (15%, with confirmed Arnold-Chiari malformation in 86%), aqueductal stenosis (8%), post-meningitis hydrocephalus (PMH, 8%), post-haemorrhage hydrocephalus (PHH, 8%), Dandy–Walker malformation (6%), unknown origin (6%), and arachnoid cyst (5%). The remaining aetiologies were varied and separated into two groups: hydrocephalus of miscellaneous obstructive causes (e.g., Chiari I malformation, choroid cysts or foramen of Monro membranous obstruction, 4%) and other causes (e.g., craniosynostosis, achondroplasia, Hurler’s disease, occipital meningocele, as well as syndromic and antenatal hydrocephalus, 8%).

There were 84 boys (59%) and 58 girls (41%). Only three groups comprised predominantly boys, namely PIVHH, PHH, and miscellaneous obstructive causes.

In total, 23 children died (16%), predominantly (52%) in the brain tumour group. Two deaths were directly related to treatment, namely post-operative haemorrhage and acute VPS malfunction with brain herniation. The population characteristics have been presented in Table 1.

Table 2 Management of hydrocephalus

Aetiology	ETV (%)	ETV failure rate (%)	Time lapse to ETV failure [mean, (min – max)]	Shunts (VPS, VCS, CPS) (%)	Infectious complications (%)	Mechanical complications (%)
PIVHH	9	100	3 d	52	21	58
Brain tumour	48	55	42 d (3–197 d)	61	10	48
Spina bifida	10	100	6 d (0–14 d)	95	15	54
Aqueductal stenosis	67	12	14 d	50	10	60
PMH	33	50	12 d (7–14 d)	75	15	46
PHH	18	100	79 d (3–152 d)	64	25	38
Dandy-Walker	11	0	NA	89	39	61
Unknown	38	67	309 d (30–601 d)	75	0	78
Arachnoid cyst	29	100	162 d (136–183 d)	86	10	50
Miscellaneous obstruction	80	0	NA	40	50	0
Various	9	100	92 d	73	30	35
Total	28	52	71 d (0–601 d)	70	19	51

This table represents the percentage of patients who underwent ETV and occurrence of failure with the time until failure expressed in days. Additionally, the percentage of patients fitted with internal shunts is provided, along with the infectious and mechanical complication rates on those shunts

ETV endoscopic third ventriculostomy, PIVHH post-intraventricular haemorrhage hydrocephalus, PMH post-meningitis hydrocephalus, PHH post-haemorrhage hydrocephalus, VPS ventriculo-peritoneal shunt, VCS ventricular-cardiac shunt, CPS cysto-peritoneal shunt, d days, NA not applicable, min minimum, max maximum

Outcome

Type of education and clinical evaluation by a paediatric neurologist were primarily considered for outcome assessment, yet neurodevelopmental tests were only rarely available. Based on these findings, the patients were divided into four categories: green (normal development), yellow (mild developmental delay, less than 1 year, e.g., repeating a year at school), orange (moderate developmental delay, more than 1 year) and red (severe developmental delay; e.g., non-autonomous or institutionalised).

Results

Hydrocephalus diagnosis

Diagnosis was established on average at 13 months (mo) or younger (Table 1), mostly at birth or soon afterwards. In the event of PMH, there was approximately 1 mo between infection onset and occurrence of hydrocephalus. Only a few children with brain tumours or PHH, both acquired aetiologies, were diagnosed after the age of 2 years (yrs).

Management of hydrocephalus

Nine children (6%) did not require any treatment, versus five preterm children who died prior to treatment initiation.

The ages at diagnosis and treatment were similar between groups (Table 1). Surgical treatment was postponed in two children with aqueductal stenosis and one with Dandy–Walker syndrome (20, 21 and 90 mo, respectively), as their hydrocephalus was stable, with neurological status at diagnosis unaffected. This explains the higher mean age and broader age spread at treatment in these groups.

ETV was performed on 40 patients, twice for two of them, thereby representing the major procedure conducted in hydrocephalus of miscellaneous obstructive causes (80%) and aqueductal stenosis (67%). Internal shunts (mainly VPS, but also 10 cysto-peritoneal and two ventricular-cardiac shunts) were placed in 99 children. Internal shunt placement was the main surgical option for spina bifida (95%), Dandy–Walker (89%), arachnoid cysts (86%), PMH (75%), and hydrocephalus of unknown origin (75%) (Table 2). Later, both surgeries were required in some patients.

External ventricular shunts were initially implanted in 25 patients. For four, hydrocephalus stabilized following external shunt placement, while for others either shunt placement or ETV was performed.

Overall, 32% of patients receiving shunts needed only one. The mean number of shunts per patient was three, with

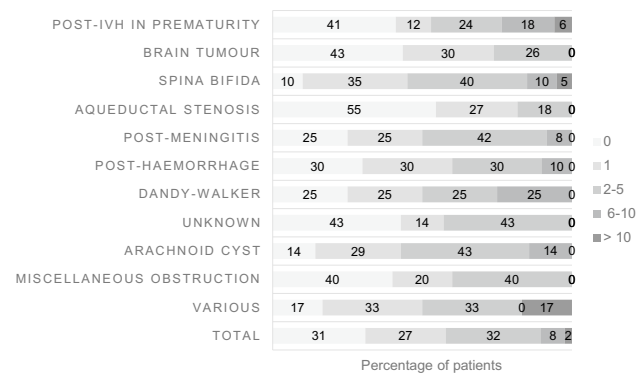


Fig. 1 Number of complications per patient This graph represents the number of complications occurring in each patient. For each aetiological group, indicated on the left, each bar coincides with the percentage of patients from the specific group presenting with a certain number of complications, as indicated by the shades of grey: the lightest to the darkest shade of grey corresponds to zero, one, two to five, six to ten and more than ten complications, respectively. *IVH* intraventricular haemorrhage

the highest average required for PIVHH, PHH and various aetiology groups.

Figure 1 illustrates the number of complications directly related to hydrocephalus, along with the corresponding treatments per patient. We noted that all groups except for spina bifida comprised over 50% of children exhibiting either no complication at all or only one. A high number of complications occurred in patients suffering from PIVHH, spina bifida, PMH, or PHH.

Complications occurred more frequently following shunt placement than ETV, namely 43 infectious complications (19% of 226 internal shunts placed) after an average time of 5.5 mo [range 3 days (d)–60 mo].¹ Meningitis occurred 34 times, peritonitis four times, and wound infection five times. Altogether, 115 mechanical complications (51%) were observed, following 12 mo on average (range 1 d–10 yrs 4 mo).²

ETV outcomes are summarized in Table 2. The overall ETV failure rate was 52%, after 2.5 mo on average (range 0–20 mo).³ ETV proved the most successful technique in patients with obstructive causes and aqueductal stenosis. The mean age of children undergoing functional ETV was 17.5 mo (range 1–62 mo).⁴ Of those with ETV failure, the mean age was 17 mo (range 0–57 mo).⁵ The failure rate was 41% for children aged 0–6 mo at surgery, 67% for 6–12 mo,

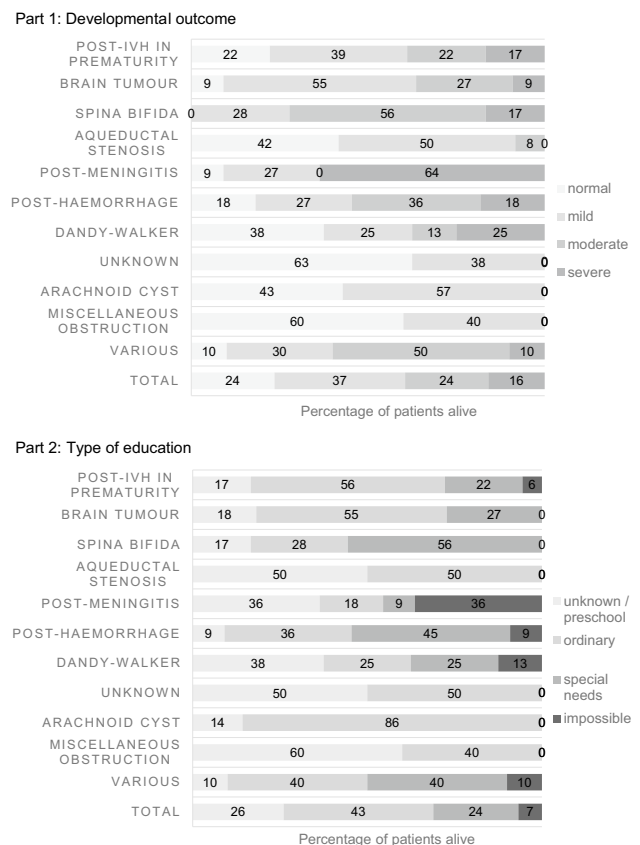


Fig. 2 Development and education Both graphs concern children that were still alive by the clinical study's end. The groups have been added on the left. The first graph shows the developmental outcome. The bar represents the percentage of patients alive in each group with normal development (the lightest shade of grey) versus mild (light grey), moderate (grey) or severe developmental delay (the darkest shade of grey). The second graph depicts the education type followed. The bar illustrates the percentage of patients alive in each group enrolled in an unknown type of schooling or being pre-schooled (the lightest shade of grey), attending ordinary education (light grey), benefiting from special needs education (grey) or being unschooled due to all education possibilities proving impossible (the darkest shade of grey). *IVH* intraventricular haemorrhage

60% for 1–2 yrs, and 55% for over 2 yrs. One child developed meningitis and another contracted a surgical wound infection following ETV (5% of 42 ETVs carried out).

Other complications, such as cerebral haemorrhage, subcutaneous CSF collection, bone defects, and skin necrosis (secondary to repeated interventions), were more common after shunt placement. Seven children developed extracerebral fluid collection (blood or CSF) post-ETV, though subdural shunts (external or peritoneal) were required in only two.

¹ (Minimum–maximum) in years (yrs) months (mo) and days (d).

² (Minimum–maximum) in years (yrs) months (mo) and days (d).

³ (Minimum–maximum) in years (yrs) months (mo) and days (d).

⁴ (Minimum–maximum) in years (yrs) months (mo) and days (d).

⁵ (Minimum–maximum) in years (yrs) months (mo) and days (d).

Outcome

Mean follow-up was 4 yrs 10 mo (Table 1). Figure 2 provides an overview of the children's development and education type. Of the children still alive, 24% developed normally (green), whereas development was mildly delayed in 37% (yellow), moderately in 24% (orange) and severely in 16% (red).

Children with aqueductal stenosis, arachnoid cyst, and hydrocephalus of either unknown or diverse obstructive causes experienced fair to good outcomes (green and yellow). Neurodevelopmental prognosis was intermediate for children with brain tumours, Dandy–Walker, and PIVHH, with 62% on average presenting none or only slight delay. Concerning children with spina bifida, PMH, PHH, or hydrocephalus of various aetiologies, development was often notably delayed (orange and red), with the highest ratio of severe delay (64%, with all children of this group displaying poor outcome) observed in PMH. Children with spina bifida were more or less severely burdened by lesions to the spinal cord, whereas the other children with poor outcomes and hydrocephalus of various aetiologies presented syndromes, craniosynostosis, Hurler's disease, achondroplasia, or sequelae of severe prematurity. Motor, neurosensory, and intellectual deficits were more common in these groups.

Epilepsy affected 28 patients (20%), though only transiently in five, primarily children with PMH (92%) or PHH (46%), i.e., aetiologies with severe brain sequelae.

Figure 2 provides an overview of development and education, with a correlation observed between outcome and education, namely between children attending ordinary school (43%) and those categorized as green and yellow, as well as between children requiring special education (24%, mainly for motor deficit) and those categorized as orange and red. Eight 'red' children (7%) could not attend any type of school, four of whom presented with PMH. A total of 26% were pre-schoolers or enrolled in an unknown scholarship.

Discussion

This study sought to investigate the outcome of patients with early-onset hydrocephalus. The first observation concerns the diversity of underlying hydrocephalus causes in young patients. This was associated with difficulties analysing the patient group as a whole. Early-onset hydrocephalus may be associated with other malformations and sequelae likely to negatively impact the outcome. In our cohort, patients with spina bifida, PMH, PHH, and hydrocephalus of various aetiologies presented poorer outcomes, with moderate or severe delay observed in 62%. Aqueductal stenosis, arachnoid cyst, and hydrocephalus of unknown origin and miscellaneous obstructive causes

were associated with better outcome in 97% of cases. Isolated hydrocephalus without any other clinical or radiological findings appears to be less common, with an incidence of 24% in our study; these patients' neurological status is more likely to improve following treatment.

Management of hydrocephalus

Our study provides an overview of the surgical procedures performed and their complications. We assessed the cases of 40 children receiving ETV and 99 receiving internal shunts. In some patients, ETV was performed for less evident aetiologies, with the ultimate objective to provide these children with a chance, however minute, of remaining shunt-free. Complications occurred more frequently after shunt placement (70% for all patients) than ETV (52% failure and 5% of infections). ETV and VPS were shown to be equally effective for managing hydrocephalus [6]. A prospective study comparing ETV (115 children) and shunt (43 children) as index surgery in aqueductal stenosis before 2 yrs of age reported ETV success rates of 68, 66, and 64% versus shunt success rates of 95, 83, and 79%, at 3, 12 and 36 mo, respectively [7]. With an adjusted hazard ratio, the failure risk appeared significantly higher for ETV within 3 mo post-surgery and in the event of prior shunting [7, 8]. The failure risk was also higher at younger age (especially before 6 mo) regardless of treatment type [7–9]. Overall, ETV remains a reasonable choice [7]. In our study, the ETV failure was 41% for children 0–6 mo of age at surgery, 67% for children 6–12 mo, 60% for children 1–2 yrs and 55% for those over 2 yrs old.

Several studies attest to the superiority of ETV combined with choroid plexus cauterization, primarily developed in Sub-Saharan Africa, given that shunt failure can be considered a death sentence in that region due to lack of health care facilities. This procedure is currently gaining increasing interest in developed countries [10–15].

Epilepsy

Epilepsy in hydrocephalus children appears to be related to associated brain abnormalities or encephalopathy sequelae, observed in up to 14% of cases in some studies [16]. Epilepsy affected 20% of our patients, principally concerning PMH and PHH, namely aetiologies with severe brain sequelae, as previously reported in the literature [17]. Conversely, in one published study, children with aqueductal obstruction, cysts, and encephaloceles presented the worse outcomes and highest epilepsy burden (33 and 29% vs. 17% for the entire population, respectively), though many suffered from associated brain malformations [18].

School and future

The future of children presenting with early-onset hydrocephalus should always be questioned, as predicting their outcome appears challenging. In our study, 61% of children progressed and developed fairly well, especially those with aqueductal stenosis, cyst, and unknown or miscellaneous obstructive causes. Among these, 43% attended normal schools and 24% received special education, whereas 7% were unable to attend any school. These results are in line with previously-published studies. Among these, 41–67% of children subsequently attended ordinary school and 33–50% received special education as necessary, whereas 7–9% were unable to enrol in schooling [16, 19, 20]. Overall, 20% required special education for motor deficits [10], compared to 18% of our own population. Paulsen et al. found that one-third of studied cases attended university versus 41% within the general population. However, spina bifida patients and adults with an intelligence quotient below the normal range were excluded from study participation [5]. In addition, 44% accomplished professional training, 45–56% were socially independent, whereas 20% required professional assistance [19, 20]. Any significant difference in outcome depending on aetiology could not be demonstrated [19]. It should, however, be noted that patients with hydrocephalus treatment in infancy displayed slightly lower quality life scores than the control group, whereas those that additionally suffered from epilepsy or cerebral palsy scored the lowest [5].

To conclude, isolated obstructive hydrocephalus, e.g., aqueductal stenosis, arachnoid or choroid cysts, and hydrocephalus of unknown origin, seems to be associated with more manageable outcomes. In brain tumours, the outcome is overshadowed by the tumour prognosis itself. Concerning spina bifida patients, their outcome seems to be related to hydrocephalus occurrence, being inversely related to the lesion level [21, 22]. Dandy-Walker syndrome can be associated with other central nervous system (CNS) malformations, which may negatively impact child development [23]. PIVHH and prematurity, as well as PHH and PMH, can cause severe brain injury via ischaemia and atrophy. Preterm child development is impaired by hydrocephalus in clear correlation to its severity (shunted vs. arrested vs. none) [24].

Thus, whilst outcomes appear affected by hydrocephalus aetiology, they are even more impacted by complications, associated CNS malformations, and secondary CNS lesions.

Early treatment of symptomatic hydrocephalus has significant impact on future evolution. Although published literature does not provide enough data to confirm a significant QOL difference between ETV and VPS patients [25], it must be emphasised that children treated by ETV exhibit fewer complications, while also less frequently needing surgical revision or further hospitalization. VPS placement in

these children is, however, not necessarily associated with comparatively more complications and poor outcome, as many patients implanted with a shunt experience a fairly good clinical progression. Pure obstructive hydrocephalus is more likely to be successfully treated by ETV. It is thus reasonable to attempt ETV in all possible cases of obstructive aetiologies, with the aim of providing these children with the chance to live without a shunt and the associated complication risks, despite ETV's lower success rate. With close follow-up, VPS can be performed as soon as ETV failure has been noticed. In the event of congenital aqueductal stenosis diagnosed prenatally, without any other brain abnormality, it appears judicious to reassure the future parents, given that ETV is most likely to be successful, with relatively good developmental prognosis.

Limitations

Though 142 patients were included in the trial, they were divided into 11 differently sized groups, some particularly small. As a result, statistical analysis and comparison among groups were rendered difficult. Also, due to the retrospective design of the study, not all information was available for all patients.

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Compliance with ethical standards

Conflict of interest The authors declare that they have no conflicts of interest.

Ethical standards The trial protocol was approved by the ethical committee of the university and the study complied with its recommendations. Owing to its retrospective nature.

Informed consent For this type of study, formal consent is not required.

References

1. Volpe JJ (2008) Neurology of the newborn, 5th edn. Elsevier Saunders, Philadelphia
2. Lyon G, Evrard P (2000) Hydrocéphalies, collections liquidiennes péricérébrales chroniques. Neuropédiatrie, 2nd edn. Elsevier Masson, Paris, pp 83–96
3. Johnston MV, Kinsman S (2004) Congenital anomalies of the central nervous system. In: Behrman RE, Kliegman RM, Jenson HB (eds) Nelson textbook of pediatrics, 17th edn. Elsevier Saunders, Philadelphia, pp 1983–1993
4. Pinton F, Ponsot G (2010) Hydrocéphalies. In: Chabrol B, Dulac O, Mancini J, Ponsot G (eds) Neurologie pédiatrique, 3rd edn. Médecine Sciences Flammarion, Paris, pp 162–172
5. Lindquist B, Fernell E, Persson EK, Uvebrant P (2014) Quality of life in adults treated in infancy for hydrocephalus. Childs

- Nerv Syst 30(8):1413–1418. <https://doi.org/10.1007/s00381-014-2425-4>
6. Limbrick DD, Baird LC, Klimo P, Riva-Cambrin J, Flannery AM (2014) Pediatric hydrocephalus: systematic literature review and evidence-based guidelines. Part 4: cerebrospinal fluid shunt or endoscopic third ventriculostomy for the treatment of hydrocephalus in children. *J Neurosurg Pediatr* 14(Suppl 1):30–34. <https://doi.org/10.3171/2014.7.peds14324>
 7. Kulkarni AV, Sgouros S, Constantin S (2016) International infant hydrocephalus study: initial results of a prospective, multicenter comparison of endoscopic third ventriculostomy (ETV) and shunt for infant hydrocephalus. *Childs Nerv Syst* 32(6):1039–1048. <https://doi.org/10.1007/s00381-016-3095-1>
 8. Lam S, Harris D, Rocque BG, Ham SA (2014) Pediatric endoscopic third ventriculostomy: a population-based study. *J Neurosurg Pediatr* 14(5):455–464. <https://doi.org/10.3171/2014.8.PEDS13680>
 9. Kadrian D, van Gelder J, Florida D, Jones R, Vonau M, Teo C, Stening W, Kwok B (2008) Long-term reliability of endoscopic third ventriculostomy. *Neurosurgery* 62(Suppl 2):614–621. <https://doi.org/10.1227/01.neu.0000316265.59596.8c>
 10. Kamalo P (2013) Exit ventriculoperitoneal shunt; enter endoscopic third ventriculostomy (ETV): contemporary views on hydrocephalus and their implications on management. *Malawi Med J* 25(3):78–82
 11. Kulkarni AV, Riva-Cambrin J, Browd SR, Drake JM, Holubkov R, Kestle JRW, Limbrick DD, Rozzelle CJ, Simon TD, Tamber MS, Wellons JC, Whitehead WE (2014) Endoscopic third ventriculostomy and choroid plexus cauterization in infants with hydrocephalus: a retrospective hydrocephalus clinical research network study. *J Neurosurg Pediatr* 14(3):224–229. <https://doi.org/10.3171/2014.6.PEDS13492>
 12. Mweshi MM, Amosun SL, Ngoma MS, Nkandu EM, Sichizya K, Chikoya L, Munthali J (2010) Endoscopic third ventriculostomy and choroid plexus cauterization in childhood hydrocephalus in Zambia. *Med J Zambia* 37(4):246–252
 13. Stone SS, Warf BC (2014) Combined endoscopic third ventriculostomy and choroid plexus cauterization as primary treatment for infant hydrocephalus: a prospective North American series. *J Neurosurg Pediatr* 14:439–446. <https://doi.org/10.3171/2014.7.PEDS14152>
 14. Warf BC (2005) Comparison of endoscopic third ventriculostomy alone and combined with choroid plexus cauterization in infants younger than 1 year of age: a prospective study in 550 African children. *J Neurosurg Pediatr* 103(6):475–481. <https://doi.org/10.3171/ped.2005.103.6.0475>
 15. Zandian A, Haffner M, Johnson J, Rozzelle CJ, Tubbs RS, Loukas M (2014) Endoscopic third ventriculostomy with/without choroid plexus cauterization for hydrocephalus due to hemorrhage, infection, Dandy-Walker malformation, and neural tube defect: a meta-analysis. *Childs Nerv Syst* 30(4):571–578. <https://doi.org/10.1007/s0038101323449>
 16. Platenkamp M, Hanlo PW, Fischer K, Gooskens RHJM (2007) Outcome in pediatric hydrocephalus: a comparison between previously used outcome measures and the hydrocephalus outcome questionnaire. *J Neurosurg* 107(1 Suppl):26–31. <https://doi.org/10.3171/PED-07/07/026>
 17. Klepper J, Büsse M, Straßburg HM, Sörensen N (1998) Epilepsy in shunt-treated hydrocephalus. *Dev Med Child Neurol* 40(11):731–736
 18. Tully HM, Ishak GE, Rue TC, Dempsey JC, Browd SR, Millen KJ, Doherty D, Dobyns WB (2016) Two hundred thirty-six children with developmental hydrocephalus: causes and clinical consequences. *J Child Neurol* 31(3):309–320. <https://doi.org/10.1177/0883073815592222>
 19. Preuss M, Kutscher A, Wachowiak R, Merkenschlager A, Bernhard MK, Reiss-Zimmermann M, Meixensberger J, Nestler U (2015) Adult long-term outcome of patients after congenital hydrocephalus shunt therapy. *Childs Nerv Syst* 31(1):49–56. <https://doi.org/10.1007/s00381-014-2571-8>
 20. Paulsen AH, Lundar T, Lindegaard KF (2015) Pediatric hydrocephalus: 40-year outcomes in 128 hydrocephalic patients treated with shunts during childhood. Assessment of surgical outcome, work participation, and health-related quality of life. *J Neurosurg Pediatr* 16(6):633–641. <https://doi.org/10.3171/2015.5.peds14532>
 21. Bellin MH, Dicianno BE, Levey E, Dosa N, Roux G, Marben K, Zabel TA (2011) Interrelationships of sex, level of lesion, and transition outcomes among young adults with myelomeningocele. *Dev Med Child Neurol* 53(7):647–652. <https://doi.org/10.1111/j.1469-8749.2011.03938.x>
 22. Verhoef M, Barf HA, Post MW, van Asbeck FW, Gooskens RH, Prevo AJ (2006) Functional independence among young adults with spina bifida, in relation to hydrocephalus and level of lesion. *Dev Med Child Neurol* 48(2):114–119. <https://doi.org/10.1017/S0012162206000259>
 23. Bolduc ME, Limperopoulos C (2009) Neurodevelopmental outcomes in children with cerebellar malformations: a systematic review. *Dev Med Child Neurol* 51(4):256–267. <https://doi.org/10.1111/j.1469-8749.2008.03224.x>
 24. Fletcher JM, Landry SH, Bohan TP, Davidson KC, Brookshire BL, Lachar D, Kramer LA, Francis DJ (1997) Effects of intraventricular hemorrhage and hydrocephalus on the long-term neurobehavioral development of preterm very-low-birthweight infants. *Dev Med Child Neurol* 39(9):596–606
 25. Kulkarni AV, Shams I, Cochrane DD, McNeely PD (2010) Quality of life after endoscopic third ventriculostomy and cerebrospinal fluid shunting: an adjusted multivariable analysis in a large cohort. *J Neurosurg Pediatr* 6(1):11–16. <https://doi.org/10.3171/2010.3.PEDS09358>