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

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OBJECT Treatment for hydrocephalus has not advanced appreciably since the advent of CSF shunts more than 50 years ago. The outcome for pediatric patients with hydrocephalus has been the object for several studies; however, much uncertainty remains regarding the very long term outcome for these patients. Shunting became the standard treatment for hydrocephalus in Norway during the 1960s, and the first cohorts from this era have now reached middle age. Therefore, the objective of this study was to review surgical outcome, mortality, social outcome, and health-related quality of life in middle-aged patients treated for hydrocephalus during childhood.

METHODS Data were collected in all patients, age 14 years or less, who required a CSF shunt during the years 1967–1970. Descriptive statistics were assessed regarding patient characteristics, surgical features, social functioning, and work participation. The time and cause of death, if applicable, were also determined. Kaplan-Meier survival estimates were used to determine the overall survival of patients. Information regarding self-perceived health and functional status was assessed using the 36-Item Short Form Health Survey (SF-36) and the Barthel Index score.

RESULTS A total of 128 patients were included in the study, with no patient lost to follow-up. Of the 128 patients in the study, 61 (47.6%) patients died during the 42–45 years of observation. The patients who died belonged to the tumor group (22 patients) and the myelomeningocele group (13 patients). The mortality rate was lowered to 39% if the patients with tumors were excluded. The overall mortality rates at 1, 2, 10, 20, and 40 years from time of initial shunt insertion were 16%, 24%, 31%, 40%, and 48% respectively. The incidence of shunt-related mortality was 8%. The majority of children graduated from a normal school (67%) or from a school specializing in education for physically handicapped children (20%). Self-perceived health was significantly poorer in 6 out of 8 domains assessed by SF-36 as compared with the background population. Functional status among the survivors varied greatly during the follow-up period, but the majority of patients were self-dependent. A total of 56% of the patients were socially independent, and 42% of the patients were employed.

CONCLUSIONS Approximately half of the patients are still alive. During the 42–45 year follow-up period, the mortality rate was 48%. Two deaths were due to acute shunt failure, and at least 8% of the deaths were shunt related (probable or late onset). The morbidity in middle-aged individuals treated for pediatric hydrocephalus is considerable. The late mortality rate was low, but not negligible. Twelve patients died during the last 2 decades, 1 of whom died because of acute shunt failure. Although the shunt revision rate was decreasing during the study period, many patients required shunt surgery during adulthood. Forty-one revisions in 21 patients were performed during the last decade. Thus, there is an obvious need for life-long follow-up in these patients.

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KEY WORDS pediatric hydrocephalus; very long term outcome; health-related quality of life
Only a few decades ago, there was no established effective treatment for the hydrocephalic condition. In 1962, Laurence and Coates reported an overall survival rate into adulthood of 20%.5 The Norwegian neurosurgeon Arne Torkildsen (1899–1968) was an important contributor to a new and effective treatment for hydrocephalus (HC) in 1937. The ventriculocisternostomy procedure was an internal shunting between 1 of the brain’s lateral ventricles and the cisterna magna, thereby bypassing a blockage in the cerebral aq- ueduct. However, it did not have any effect upon communicating HC. The modern shunting era began in the 1950s when ventriculoatrial (VA) shunts set a new standard in HC treatment.6,9 The first VA shunt was implanted in Norway in 1961, and VA shunts were the standard treatment procedure until 1979. Beginning in 1980, use of ventriculoperitoneal (VP) shunts became more common because of a few, but fatal, complications from VA shunting.7 Since then, VP shunts have been the standard treatment for patients with HC.

Shunting has dramatically improved the outlook of children with HC. The outcome for children treated for HC has been reported in several studies, but questions remain concerning the very long term outcome for these children.10,15 Self-perceived health and quality of life (QOL) data are important measures of patient outcome, although these parameters have not been commonly used in previous neurosurgical series for HC treatment. Leaving the protected life of childhood and entering the adult world requires independent decisions regarding studies, occupation, and partnership: have middle-aged adults with a history of treatment for pediatric HC achieved the same results as their peers? And how do they consider their own health?

The aims of this study were to determine the 40-year outcomes of children treated for HC. We investigated etiology, surgical morbidity, shunt dependency, and mortality in 128 hydrocephalic children who underwent shunt treatment between 1967 and 1970 (both years included). Furthermore, we assessed health-related QOL (HRQOL), functional outcome (Barthel Index score), and social outcome in this patient cohort.

Methods

Patients

We retrospectively analyzed a nonselected cohort consisting of 128 children, age 14 or less, who underwent first-time shunt insertion for HC in the Department of Neurosurgery, Rikshospitalet, in Oslo, Norway, during the period from 1967 to 1970—years prior to the introduction of CT scans in our hospital in 1977. HC diagnosis was determined during this period through clinical signs such as raised intracranial pressure and increased head circumference, as well as through the use of ventriculography, in which positive contrast media or air is introduced into the ventricular system after ventricular punctures. The patients were collected from surgical protocols of the relevant time period. The information obtained included patient sex, patient age at first shunt implantation, cause of HC, number and causes of shunt revisions, patient death, and cause of death. All types of CSF shunts were included. The type of shunt and shunt hardware were chosen according to the neurosurgeon’s preference at the time of the operation.

The follow-up assessments were ended on the 31st of December 2012, implying an observation time of 42 to 45 years among the patients who are still alive.

Patients who lacked clinical and radiographic follow-up since January 2000 were invited to the Neurosurgical Department to obtain cerebral CT or MR images and radiographs of the shunt system in order to assess shunt function.

Instruments

Assessment of HRQOL

The 36-Item Short Form Health Survey (SF-36) is a generic questionnaire as part of the Medical Outcomes Study that was used to assess HRQOL in 8 domains: physical functioning, social functioning, role limitations due to emotional problems, pain, mental health, vitality, and general health perception. All raw scores were linearly converted to a 0–100 scale, providing a sum score for each domain. In addition, individual domain scores were used to calculate the physical component summary and a mental component summary. Higher domain and summary scores indicate a higher level of functioning or health-related well-being.16

Assessment of Functional Status

The Barthel Index (BI) is a well-established and validated scale using 10 variables to measure performance in basic activities of daily living (BADL) primarily related to personal care and mobility. Scores range from 0 to 100, with a higher score denoting greater independence. The purpose of using this scale was to assess functional status and illustrate differences among subgroups within our cohort.

Statistical Analysis

Data were analyzed using SPSS statistical software. Mean scores ± standard deviation for the 8 SF-36 domains and the summary scores were calculated from the raw scores for the total group and for each of the subgroups using the SF-36 algorithm in SPSS.

Kaplan-Meier survival estimates were used to determine overall survival of patients. All statistical analysis was performed using SPSS 16.0.0 software (SPSS Inc.).

Ethical Approval

Ethical approval was obtained from the medical ethics committee of Norway, the Regional Committee of Medicine and Health (REK).

Results

Patient Characteristics

A total of 128 patients with childhood HC who underwent primary shunt surgery over a 4-year period were studied.

The male to female ratio was 1:17. The etiologies for HC are listed in Table 1. Figure 1 shows the number of patients receiving each type of shunt at initial implantation and at the end of the follow-up period.
The majority of the primary shunts were the VA type (94.5%), the standard treatment in the 1960s. The mean patient age at the time of the first shunt placement was 2 years (median 3.2 months); almost half of the patients underwent shunt implantation within the first 3 months of life, and 65% had shunts inserted by age 6 months. Given that the shunts were revised in the years following initial implantation and combined with an increasing preference for the use of VP shunts, the ratio has changed over the course of the follow-up period. At last follow-up, 45% of the shunts in the study patients are the VP type. Patients whose HC was associated with myelomeningocele (MMC), who accounted for 33% of the patients, received a shunt earlier in life than patients in the other diagnostic categories (p < 0.01).

**Shunt Revisions**

Figure 2 shows the number of shunt revisions performed in the study patients. Revisions were performed in 422 instances; 111 (26.3%) were elective (prophylactic) revisions for preserving the patency of the distal catheter as the patients were growing. Otherwise, a shunt system was revised only in the presence of clinical symptoms of shunt failure. The majority of the patients had between 1 and 5 procedures, with a mean revision number of 3.3. No revision of the shunt was performed in 27 patients (21%), of whom 4 patients (3.1%) are still alive at follow-up. Thirty-three patients (25.8%) have a shunt system that has not been revised within the last 30 years: 30 patients with VA shunts, 2 with VP shunts, and 1 with a subduroperitoneal shunt.

The annual shunt revision frequency decreased over time in the study population. More than 30% of the revisions were performed during the 1st year after initial shunt insertion, and 73% were conducted within the 1st decade. The mean annual revision rates (number of revisions performed each year compared with the number of patients at risk, i.e., still alive) during the 1st, 2nd, 3rd, and 4th decade after primary shunt insertion were 0.29, 0.05, 0.04, and 0.06, respectively. Twenty-one percent of all revision procedures were performed after age 18, and 9.7% were performed during the last decade.

Shunt independence and the need for shunt revision during the 40-year follow-up period are illustrated in Fig. 3.

**Shunt Dependency**

Forty-two years after initial shunt insertion, 59 of the 67 patients (88%) who are still alive have a prosthetic shunt system implanted. Endoscopic third ventriculosternostomy was successfully performed in 4 instances: 2 patients with MMC and 1 patient with a cerebral tumor had the shunt explanted after successful treatment with endoscopic third ventriculostomy (ETV) in the 2000s; 1 patient with HC secondary to meningitis was treated with ETV in 2011 when diagnosed with aqueduct stenosis and gradually increasing ventricular size. ETV treatment failed in 1 instance, and the patient required reimplantation of a VP shunt 3 years after endoscopic treatment because of recurrence of clinical symptoms indicating raised intracranial pressure.

In addition, 1 male patient with communicating HC...
was treated with fenestration of lamina terminalis after having explanted the shunt because of infection, and ETV was considered unfavorable because of small ventricular size.

Thus, successful treatment with internal CSF diversion in cases of shunt malfunction occurred in 5 out of 6 (83%) patients with ventriculomegaly.

In 3 patients (4.5%), their shunts were explanted during the observation period and they are considered to be shunt independent: 1 male with congenital communicating HC had the shunt explanted after getting meningitis 9 years after the initial shunt placement—he did not display clinical symptoms of shunt dependency in the follow-up period and had stable radiological findings; 1 female patient with MMC had the distal catheter removed and the remaining shunt system was ligated at the age of 11 because of symptoms of overdrainage; and 1 female patient with cerebellar astrocytoma, who underwent placement of a shunt prior to resection of the tumor, had the shunt removed a few years after the tumor extirpation.

In a number of patients, shunt independence has been discussed, but there is no objective proof to that effect, and they are not identified as such in this analysis.

Therefore, more than 40 years after initial shunt treatment, 64 out of 67 patients still have an artificial CSF diversion system; 5 patients have a functional stoma (ETV/lamina terminalis fenestration); and 59 have a prosthetic shunt system.

During the 4 decades studied, the diagnosis of spontaneously “arrested HC” has been used with differing frequencies. To determine the “shunt in function” status or persistent shunt dependency of the 67 patients still alive, our criteria were the following: 1) patients not having symptoms indicative of raised intracranial pressure, 2) radiological verification of stable size of the lateral ventricles and cortical width, and recognizable cortical relief not suspicious for raised intracranial pressure, and 3) radiological verification of shunt continuity with intraventricular placement of the proximal shunt, length of the distal shunt signifying a reasonable likelihood of intravascular placement of the distal shunt in VA shunt patients, and length of the distal shunt indicating intraperitoneal placement of the distal catheter in VP shunt patients.

The majority of the patients apparently have “well-drained” ventricles, a continuous shunt system, and do not show any clinical signs of raised intracranial pressure more than 4 decades after primary shunt insertion, which supports a functional shunt system. How this represents continuous shunt dependency cannot be clarified without using more invasive diagnostics.

**Mortality and Morbidity**

In total, 61 of the 128 (47.6%) patients died during the follow-up period. Thirty-one (24%) patients died during the first 2 years after initial shunt insertion, and 15 of these were patients with tumors. Thus, the 2-year mortality rate in nontumor patients was 12%. No deaths occurred within the 10-year period representing 24–34 years after initial surgical intervention, and 5 deaths occurred during the last 6 years of follow-up. Deaths associated with shunt treatment complications are listed in Table 2. Death due to acute shunt failure was verified in 2 patients. In addition, there were 2 patients without known underlying disease who suffered from sudden death in which shunt-related
...death must be considered the most likely cause. Furthermore, there were 3 patients who died a short time after the last shunt revision: 1 male patient with multiple malformations died at 12 years old, 3 days after the last shunt revision; 1 patient with HC of unknown origin died at 3 months old, 7 days after revision of his VA shunt; and 1 patient with subarachnoidal hemorrhage died 4 days after the last shunt revision.

Deaths due to acute or late onset of complications associated with VA shunt treatment were seen in a limited number of cases: an 8-month-old male MMC patient died from a pulmonary embolism; a male patient with secondary HC due to a low-grade intracranial tumor died after 20 years (at age 30) of cor pulmonale caused by long-time VA shunt treatment; and 2 MMC patients died at age 5 months because of intracranial infection.

Across the entire series, the shunt-related mortality rate was 7% before age 20 and at 8.6% by age 40.

Additionally, severe morbidity was observed in patients with a delayed diagnosis of shunt failure: 1 male patient with a pineal gland tumor and 1 female patient with HC due to intraventricular hemorrhage lost vision in both eyes after episodes of severely elevated intracranial hypertension; 1 male patient with noncommunicating HC due to neonatal intracranial infection had considerably reduced functioning after an acute episode of shunt failure when he was 28 years old, 26 years after his last shunt revision.

Surgery-associated complications that reduced functional outcome were seen in some patients: a 6-year-old male patient was hemiparalyzed after an elective craniotomy to remove detached ventricle catheters, and a well-functioning male patient with congenital HC developed subacute epidural hematoma 3 days after a minimal head trauma. Postoperatively, he had symptoms of a brainstem lesion and was in a coma for several weeks; in the following years he had significant sequelae before he died 15 years later.

Some intraoperative complications that are rarely seen today seem relevant to mention because of their potentially fatal consequences if not dealt with promptly: manipulation of an adherent atrial catheter caused cardiac tamponade in 2 patients, both of whom survived after a successful acute thoracotomy.

Shunt infection that resulted in temporary explantation of the shunt system, hospitalization, and intravenous antibiotics occurred 10 times in 9 patients (8% per patient, 2.4% per operation).

**Cause of Death in Patients Who Died More Than 20 Years After Shunt Insertion**

Twelve patients died during the last 20 years of follow-up. Seven out of 12 patients had MMC. The cause of death in these 7 patients was sepsis (secondary to decubitus infection; n = 1), cerebral herniation (Chiari crisis with respiratory arrest due to acute shunt failure; n = 1), death due to hemorrhagic gastritis (n = 2), accidental drowning (general seizure in the water; n = 1), and unknown causes (n = 2). One patient with congenital HC died in a traumatic accident. One tumor patient died from pneumonia. One patient with an intracranial tumor died from pulmonary hypertension caused by long-term VA shunting.

**Functional Outcome**

BI scores were available in 64 patients; overall, 61% had a BI score at or above 80, which means a good deal of independence in daily living. The mean BI score was 81, but the results revealed great differences in the level of independence, ranging from 5 to 100. Motor sequels were mainly found in the spina bifida and the intracranial hemorrhage groups. In total, 5% patients needed walking aids, and 30% needed a wheelchair. The most common sequel was impaired motor function, which was most often found in patients with MMC due to spinal deformity. Epilepsy was registered in 10 out of 67 (14%) patients at follow-up.

**Social Outcomes**

The educational achievements of these patients are illustrated in Fig. 4. Integration into the normal school system was possible for 67% of the children, but many of these students were 1–2 years behind their age group and required additional support. Twenty percent were attending schools adapted for physically handicapped children. These were mostly spina bifida patients with associated motor deficits who required special educational facilities. Only a minority (13%) of patients remained in a school for mentally handicapped children. As expected, meningitis, tumor, and intraventricular hemorrhage were the 3 conditions most associated with a poor educational outcome. In terms of work, 28 (42%) patients were currently employed (defined as working more than a 50% schedule); 21 (31%) were employed in the open labor market, and 7 (10%) were in sheltered workplaces. Thirty-nine (58%) patients were unemployed; of these, 31 (46%) were not employed because of chronic illness.

In terms of social integration (Fig. 5), the majority of the patients were socially independent (56%). The remaining patients were dependent on daily (33%) or weekly (11%) care. Five patients in the study cohort still live with their parents, 3 of whom require daily assistance. Two patients live in institutions. Eleven patients live in sheltered units, all requiring daily assistance.

Of the 64 patients with complete information, 17 (27%) patients reported having a spouse or a partner, 5 (8%) were...
divorced, and 42 (65%) were living without a partner. Seventeen (27%) patients have 1 or more children, and 27% of females in the study group reported a pregnancy.

**Self-Perceived Health**

The SF-36 was completed by 60 of the 67 patients (34 women and 26 men). The mean age of the survey responders was 44.7 years (median 41.7 years, range 40–56 years). The reference group included data from the Norwegian general population (age range 40–49 years). Compared with the reference group, the HC cohort reported poorer self-perceived health in 6 of the 8 surveyed domains (Table 3). There were some differences among the sexes. The male participants had significantly (p < 0.05) lower scores in 7 domains (physical functioning, role physical, general health, vitality, social functioning, role emotional, and mental health), and the females had significantly lower scores in 6 domains (physical functioning, role physical, bodily pain, general health, social functioning, and role emotional).

The mean SF-36 scores were assessed for significant differences (p < 0.05) in self-perceived health associated with the presence or absence of shunt infections, patient age at time of first shunt insertion (grouped as 0–3 months, 4–12 months, and >1 year).
TABLE 3. Summary of mean SF-36 scores and 95% CIs, stratified by sex and compared with the sex- and age-matched normative population

<table>
<thead>
<tr>
<th>Health Domains</th>
<th>Total HC Population (95% CI)</th>
<th>Male (95% CI)</th>
<th>Female (95% CI)</th>
</tr>
</thead>
<tbody>
<tr>
<td>No. of patients</td>
<td>60</td>
<td>26</td>
<td>216</td>
</tr>
<tr>
<td>Physical functioning</td>
<td>65.1 (55.5–74.7)†</td>
<td>70.8 (56.5–85.0)†</td>
<td>91.9 (90.0–93.5)</td>
</tr>
<tr>
<td>Role physical‡</td>
<td>62.5 (51.7–73.3)†</td>
<td>71.2 (54.0–88.3)†</td>
<td>86.4 (82.6–90.2)</td>
</tr>
<tr>
<td>Bodily pain</td>
<td>68.0 (61.3–74.6)</td>
<td>76.3 (66.4–86.2)</td>
<td>78.9 (75.5–82.3)</td>
</tr>
<tr>
<td>General health</td>
<td>69.5 (63.3–75.7)†</td>
<td>67.7 (56.0–79.3)†</td>
<td>79.3 (76.5–82.1)</td>
</tr>
<tr>
<td>Vitality</td>
<td>49.4 (43.7–55.1)†</td>
<td>53.9 (45.5–62.2)†</td>
<td>65.4 (62.5–68.3)</td>
</tr>
<tr>
<td>Social functioning</td>
<td>71.0 (64.1–78.0)†</td>
<td>70.7 (59.1–82.3)†</td>
<td>87.6 (84.8–90.4)</td>
</tr>
<tr>
<td>Role emotional§</td>
<td>70.6 (60.7–80.4)†</td>
<td>76.9 (62.1–91.8)†</td>
<td>89.2 (85.7–92.7)</td>
</tr>
<tr>
<td>Mental health</td>
<td>70.9 (66.3–75.4)†</td>
<td>69.1 (61.8–76.4)†</td>
<td>80.6 (78.5–82.7)</td>
</tr>
</tbody>
</table>

Norm = normative.
* The SF-36 normative data for the Norwegian population (age range 40–49 years), from Loge and Kaasa, 1998.
† Significant differences between the HC group and the normative population.
‡ Role limitation due to physical health problems.
§ Role limitation due to emotional health problems.

Who Were the Nonrespondents?

Three patients (4%) did not respond. We were unable to contact 2 patients with MMC despite several attempts by telephone and letter. One patient with MMC did not want to participate because of a lack of interest but reported having a busy work situation.

Discussion

Mortality

The present series demonstrates more than 50% overall survival after 42–45 years of follow-up in children treated for HC predominantly with VA shunts in the late 1960s. It is very difficult to compare death rates in various series because of different patient mixes, length of follow-up, and the era in which the study was conducted. Shurtleff et al. reported that 46% of shunt-treated hydrocephalic children died by age 15.13 Their patients were also treated with shunts in the 1960s. In the present study, we demonstrate a 15-year mortality rate of about 38% (Fig. 3). Amacher and Wellington reported a 21% death rate over a 5- to 12-year follow-up study.1

Our high mortality rate (39% in patients without tumors) must be considered in light of the era in which treatment was conducted, that is, before CT scans and without modern diagnostic tools. There was also at the time a lack of established routine controls for assessing persistent shunt function and recognizing shunt malfunctions.

Lower death rates are seen in more recent studies,4 as well as in our own study of children treated predominantly with VP shunts in the 1980s, nearly 20 years later than the present series.10

We have chosen to discuss the present data in comparison with our series from the 1980s, since the latter includes all children younger than 15 who underwent primary shunt surgery, before ETV was introduced as a routine procedure in our department. This implies a comparison of primary VA shunts from the era before introduction of CT (in 1977) with VP shunts from the 1980s, with more than 20 years of follow-up for both groups.

In the present series, the difference in overall patient survival over time is clear, as illustrated in Fig. 6. However, the cause of death is far from clear in many patients. The most striking difference is in early mortality during the first 5 years after initial shunt implantation. Substantial differences in the case-mix preclude meaningful statistical analysis. Moreover, routine controls to assess persistent shunt function were not common practice, and the critical signs of shunt malfunction were not established in the noncentralized welfare system at that time. When doubt existed regarding patient diagnosis and prognosis, shunting was sometimes seen as “the only relevant treatment,” although life expectancy for the child was considered limited.

Shunt-related deaths occurred in 11 patients (8.6%), and at least 2 of these were due to acute shunt failure. Anoth-
er 2 deaths were related to cardiovascular complications from VA shunting (acute pulmonary embolism and late-onset cor pulmonale after 20 years). Two life-threatening episodes of cardiac tamponade after manipulation of the cardiac catheter were successfully treated by acute thoracotomy. No further cardiovascular complications were observed during the last 25 years of observation, despite the fact that half of the patients still have a VA shunt.

Shunt Function

“Time to first failure” has been used as an indicator of primary shunt function in several studies. The report from Kulkarni and colleagues compares shunt treatment in recent years (2008–2012) with historic controls from the 1990s.3 They focus on time to first failure and found that treatment seems to have improved over time. Analysis is hampered by differences in patient selection, underlying cause of HC, and patient age.

Although the time to first revision can be interpreted from Fig. 3, this does not illustrate the time to first shunt failure. The regular use of elective elongations of the atrial catheter implies that most of the first revisions, as well as many of the subsequent ones during the first years of life, were implemented to avoid acute shunt failure and infections before such serious complications took place. It was well known that cranial migration of the distal catheter tip was especially pronounced during the 1st year of life.

In the present series, shunt revision data in patients from 3 to 42 years after initial treatment can be compared with later follow-up studies. Thus, the number of individuals who never underwent a shunt revision mostly consists of children who died early (not from shunt failure). Only 4% of the initial cohort are long-time survivors without revisions, and 3 out of 4 long-term survivors have experienced at least 1 episode of acute shunt failure.

Shunt Dependency

Apart from the 3 individuals (4%) considered to be shunt independent, the remaining 64 patients either have a prosthetic shunt system or have received a ventriculocisternostomy. Fifty-nine out of 64 patients have a prosthetic shunt system. The last 5 patients have been successfully converted to a functional ventriculocisternostomy.

The proportion of acquired shunt independence in this study is lower than in findings from a comparable study performed by the same authors with a half-as-long observation period,30 but the findings of the present study are comparable with other papers.

During the 4 decades studied, the diagnosis of arrested HC has been used with differing frequencies. According to today’s understanding of HC, many would claim that the diagnosis of arrested HC was a rather populist approach for neurosurgeons to handle the uncertainty of shunt function and shunt independence in adolescent patients. Some patients in this cohort were considered to have arrested HC during the late 1970s and 1980s, but many of these diagnoses have been proven wrong. Some patients may present with their first shunt malfunction very late in life, and our study confirms that many patients require reoperation when they are adults—some of them for the first time. One patient in our study was revised for first time after 30 years. This is a reminder that the long-term absence of shunt malfunction symptoms does not mean that the shunt is out of function.

It should be noted that there are 4 patients still alive at follow-up who have had no revision of their initial shunt.

Functional Outcome

Neurological sequels are common in HC patients.3,9 Most often this is due to the underlying cause of HC rather than the HC itself, although in some cases it results from the severity of HC and episodes of acute shunt malfunction. Eighty-nine percent of all patients who required walking assistance were patients with MMC. Motor sequelae were mainly found in the spina bifida group. Among the MMC patients, 66% were in need of a wheelchair or other walking assistance.

Social Outcome

With the increased life expectancy of patients with childhood HC after the introduction of shunt treatment in the 1960s, problems associated with adulthood are becoming more evident.11,12,16,17

Data from the Norwegian background population show that 81% males and 77% females in their 40s are living with a partner. The poor partnering (relationship) rate of 35% in our cohort is lower than expected. Similar results are found in MMC population studies.2,14

Even if cognitive impairment in the HC population has been reported in the literature, no intelligence test was used in this study. Some reports demonstrate an overestimation of subjective health in such patients. Participants in the study showed a wide range of cognitive function. It is recognized that patient perspective on their outcome is a highly relevant measure of health. SF-36 is a multidimensional tool that cuts across broad domains of physical, social-emotional, and cognitive health. More specific measures of each of these domains exist; neuropsychological testing is one of the preferred methods, which may be the subject for further studies.
Methodology

The collection of data in this present study was retrospective. Effort has been made to determine cause of death by reviewing the patients’ records and autopsy reports, but in some cases, the cause of death remains hidden, especially if it occurred in the early years. Therefore, our findings most likely underestimate the total number of shunt-related deaths.

No patients were lost to follow-up, but assessment of social and functional outcomes and HRQOL data was based on voluntary patient participation. Therefore, the 3 patients who did not cooperate in filling out the questionnaires may influence the overall results.

The fact that the questions concerning social outcome were not validated is another limitation of the study. There may have been self-selection in terms of willingness to submit information.

Finally, studying outcomes of middle-aged adults who were treated for childhood HC in the late 1960s amounts to evaluating standards of care more than 4 decades ago.

Conclusions

More than one-half of Norwegian patients who underwent first-time shunt insertion for HC in the Department of Neurosurgery, Rikshospitalet, in the 1960s survived into middle age, and the patients who are still alive are between 43 and 59 years of age. However, the 47.5% mortality rate is high, which is mainly because of severe comorbidity. Shunt-related deaths are not negligible, accounting for 8%. In our series, 96% of the patients still alive have a shunt system, and only 4.5% have become shunt independent. At follow-up, 75% of the 67 individuals have experienced at least 1 episode of acute shunt failure. Therefore, we conclude that the vast majority of patients are still shunt dependent. This supports the dictum, “Once a shunt–always a shunt.”

We conclude that HC has a substantial effect on the functionality in adulthood, but the results show considerable variation. Nearly half of the patients are outside the labor market because of chronic illness.

The risk of shunt malfunction is gradually declining over the years, but it persists through life. Therefore, adults treated for HC during childhood require life-long follow-up.

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Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions

Conception and design: all authors. Acquisition of data: Lundar, Paulsen. Analysis and interpretation of data: Lundar, Lindegaard. Drafting the article: all authors. Critically revising the article: all authors. Approved submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Lundar. Statistical analysis: Paulsen, Lindegaard.

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