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Twenty-year outcome in young adults with childhood hydrocephalus: assessment of surgical outcome, work participation, and health-related quality of life

Clinical article

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Object. Shunting of CSF is one of the most commonly performed operations in the pediatric neurosurgeon's repertoire. The 1st decade after initial shunt insertion has been addressed in several previous reports. The goals of the authors' study, therefore, were to determine 20-year outcomes in young adults with childhood hydrocephalus and to assess their health-related quality of life (HRQOL).

Methods. Patients younger than 15 years of age, in whom a first-time shunt insertion was performed for hydrocephalus in the calendar years 1985–1988, were included in a retrospective study on surgical morbidity, mortality rates, academic achievement, and/or work participation. Information concerning perceived health and functional status was assessed using the 36-Item Short Form Health Survey (SF-36) and Barthel Index, which were completed by patients still alive by September 1, 2009.

Results. Overall, 138 patients participated, no patient being lost to follow-up. For the 20-year period, the overall mortality rate was 21.7%. The mortality rate was not significantly higher in the 1st decade after initial shunt insertion than in the 2nd decade ($p = 0.10$). Ten percent of the patients surviving still live with their primary shunt in place, whereas 81% required at least one revision, and among these individual the mean number of revisions was 4.2 (median 3, range 0–26). There was a significantly higher revision rate during the 1st decade after initial shunt insertion compared with the 2nd decade ($p = 0.027$). The majority of patients live lives comparable with those of their peers. At follow-up, 56% were employed in open-market jobs or were still students, 23% had sheltered employment, and 21% were unemployed. The HRQOL was slightly lower in the hydrocephalic cohort than in the normative population. A significant difference was found in 2 of 8 SF-36 domains—Physical Functioning and General Health.

Conclusions. During the 20-year follow-up period, 81% of the patients required at least one revision of the CSF shunt. The mortality rate was high: 24 patients died in the 1st decade and 6 died in the 2nd decade after implantation of the initial shunt. In total, 4 deaths (2.9%) were due to shunt failure. Shunt placement to treat childhood hydrocephalus has a substantial effect on social functioning in later life, although perceived health was positively found to be better than expected in young adults with hydrocephalus. *(DOI: 10.3171/2010.9.PEDS09548)*

Key Words • **hydrocephalus** • **20-year outcome** • **quality of life** • **pediatric neurosurgery**

Hydrocephalus is a pathological condition caused by abnormalities of production and/or absorption of CSF in the brain. This condition is particularly common in infants and children. The natural history of untreated congenital hydrocephalus is progressive cognitive decline and an early death, usually before the 3rd decade of life.[4,14] Hydrocephalus may be primary or secondary. Common causes of secondary hydrocephalus in children include brain tumor, intracranial hemorrhage, trauma, meningitis, and developmental anomalies of the brain.

Placement of a shunt for CSF diversion has been the first option to treat hydrocephalus. Complications such as shunt malfunction and shunt infection are relatively common, and shunt failure leading to death does occur in a number of cases.

Ten-year outcome in children with hydrocephalus has been reported in a few studies.[3,7,9] During the 1st decade after shunt placement, mortality and shunt revision rates have been demonstrated to be high. The impact of hy-

Abbreviations used in this paper: ADL = activities of daily living; ETV = endoscopic third ventriculostomy; HRQOL = health-related quality of life; MCS = Mental Component Summary; MMC = myelomeningocele; PCS = Physical Component Summary; SF-36 = 36-Item Short Form Health Survey; VA = ventriculoatrial; VP = ventriculoperitoneal.

drocephalus, however, should be considered not merely in terms of surgical morbidity and mortality. Less is known about longer-term outcome in patients with childhood hydrocephalus, particularly the achievements and self-perceived health in young adults with hydrocephalus. To better understand the impact of hydrocephalus over a greater time frame, we endeavored to determine the 20-year outcome in children with hydrocephalus. We examined case records and census data on diagnosis, shunt complications, shunt dependency, and mortality rates in a nonselected population. We also addressed scholastic achievement, work participation, functional outcome, and HRQOL.

Methods

We retrospectively analyzed a nonselected cohort consisting of 138 children age 14 years or younger who underwent first-time shunt insertion to treat hydrocephalus between 1985 and 1988 in the Department of Neurosurgery, The National Hospital, Oslo, Norway. The cases were collected from surgical protocols of the relevant time period. All types of CSF shunts were included. The case record data included sex, age at first shunt implantation, cause of hydrocephalus, number and causes of shunt revisions, death, and cause of death. For patients having been followed in other neurosurgical units, information was also obtained from these relevant departments.

Scholastic outcome was simplified into normal versus special schooling, and employment attendance into open, sheltered, or no work. Mortality rates and survival of the initial shunt was assessed using Kaplan-Meier analysis. Group comparisons employed t-tests, with significance accepted at the 5% level.

Instruments

Assessment of HRQOL. The generic questionnaire SF-36 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.[24] The survey comprises questions with yes/no answers and response choices scored on a 3- to 6-point category scale. For each question raw scores were coded, recalibrated in 10 items, summed and transformed to the eight 0–100 scales (0 = poorest possible health state; 100 = best possible health state) according to the SF-36 algorithms.[17] In addition, scores were summarized in 2 validated main scores: a PCS and a MCS.[23] Higher domain and summary scores indicate higher level of functioning or health-related well-being.

Assessment of Functional Status. The Barthel Index is a well-established and validated scale that uses 10 variables to measure performance in basic ADL[19] primarily related to personal care and mobility. Scores range from 0 to 100, with a higher score denoting greater independence. The purpose of using the Barthel Index was to assess functional status and illustrate differences among subgroups within our cohort.

Background Information Scheme. The Background Information Scheme gathers information on marital status, living arrangements, attained educational level, and employment.

Perceived health, or HRQOL, was assessed in patients alive on September 1, 2009. We mailed the questionnaires to the patients' home address after having contacted them by phone. For some individuals, the SF-36 was not considered feasible. Patients who, as judged by treating physician or a family member, would not understand the measures in the questionnaire or were unable to express independent view on their own health, were not asked to complete the SF-36.

Informed consent was obtained from all study participants; consent for incapacitated participants was obtained from the person authorized to give this consent.

The SF-36 and Barthel Index were chosen in the absence of disease-specific measures to estimate health status in adults with hydrocephalus. The Hydrocephalus Outcome Questionnaire is an existing disease-specific measure, but it is only to be used in children aged between 5 and 18 years.[13] Patients in this study ranged in age from 21 to 36 years.

The SF-36 is a valid generic measure that is not specific to sex, disease, or treatment group.[2] Comorbidity is a common feature in hydrocephalic patients, and the advantage in using SF-36 is its applicability in patients with more than one condition. Also playing a part in the decision to use this instrument was the availability of Norwegian normative SF-36 data, allowing for comparisons with the Norwegian background population.[16,17]

Statistical Analysis

Data were analyzed using SPSS (version 16.0). Mean scores and SD of the eight SF-36 domains and the two summary scores were calculated from the raw scores for the total group and each of the subgroups using the SF-36 algorithm in SPSS. Bonferroni correction was not been implemented in calculations (see Tables 4 and 5), but the results, when corrected for, can be read in the discussion section.

Ethical Approval

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

Results

Shunt History

In the 138 patients with childhood hydrocephalus, there was a statistically significant predominance of males (90 [65.2%] of 138; $p < 0.01$). The etiology of the hydrocephalic conditions are summarized in Table 1. Aside from hemorrhage (in 19% of the cases), other common underlying causes included MMC (17%) and neoplasm (17%). The different causes were equally distributed among the sexes.

The mean age at first shunt implantation was 23 months (median 6 months). In almost half (49%) of the patients, the shunt was placed during their first 6 months of life; in 86 (62%) the shunt was placed in the 1st year.

TABLE 1: Cause of hydrocephalus in 138 patients who underwent shunt treatment during childhood, including number of deaths 20 years after initial shunt insertion

Cause of Hydrocephalus	No. of Patients		
	Alive	Dead	Total (%)
hemorrhage	23	3	26 (19)
neoplasm	11	12	23 (17)
spina bifida (MMC)	17	6	23 (17)
intracranial CSF cysts	10	0	10 (7)
other malformations	11	4	15 (11)
aqueductal stenosis	8	1	9 (6)
postinfectious hydrocephalus	7	1	8 (6)
posttraumatic hydrocephalus	3	1	4 (3)
unknown	18	2	20 (14)
total	108	30	138 (100)

In the remaining 52 cases (38%) the patients were in the range of 1–14 years when the shunt was first inserted. The far most common first-time treatment for the hydrocephalus was VP shunt surgery (90% of cases); this was followed by VA shunt surgery (3.6% of cases). Of the remaining cases there were 4 subduroperitoneal shunts, 3 ventriculostomies (ad modum Torkildsen), 1 cisternoatrial shunt, and 1 cisternoperitoneal shunt. No patient underwent primary third ventriculostomy.

Twenty-four patients required no shunt revision, 10 of whom were alive at follow-up. The longevity chart for the primarily inserted shunt illustrates patients without shunt revision from a 20-year perspective (Fig. 1). Within 2 years of initial shunt insertion, more than half of the patients needed to undergo shunt revision. Overall, 81% of the patients needed secondary or follow-up surgery. These 114 patients had a total of 479 revisions of the shunt (mean 4.2, median 3.0, range 0–26). Elective elongation of the distal shunt catheter accounted for 28 of the revisions. Otherwise, a shunt system was revised only in the presence of clinical symptoms of shunt dysfunction, which in most cases was supported by radiological findings indicating shunt failure. Table 2 presents a summary of revisions stratified according to age of the child at the primary shunt surgery. Patients in whom the shunt was implanted during the first 6 months of life had a significantly higher number of later revisions ($p < 0.01$). Three hundred fifty-six (74%) of all revisions were performed during the first 10 years after the first shunt insertion, and 123 revisions (26%) were performed from 10 to 24 years after the primary procedure. The annual shunt revision frequency decreased over time; the mean annual revision rates during the 1st and 2nd decades after primary shunt insertion were 0.29 and 0.11, respectively. The annual distribution of deaths, shunt independence, and the need of shunt revision during the 20-year follow-up period are illustrated in Fig. 2.

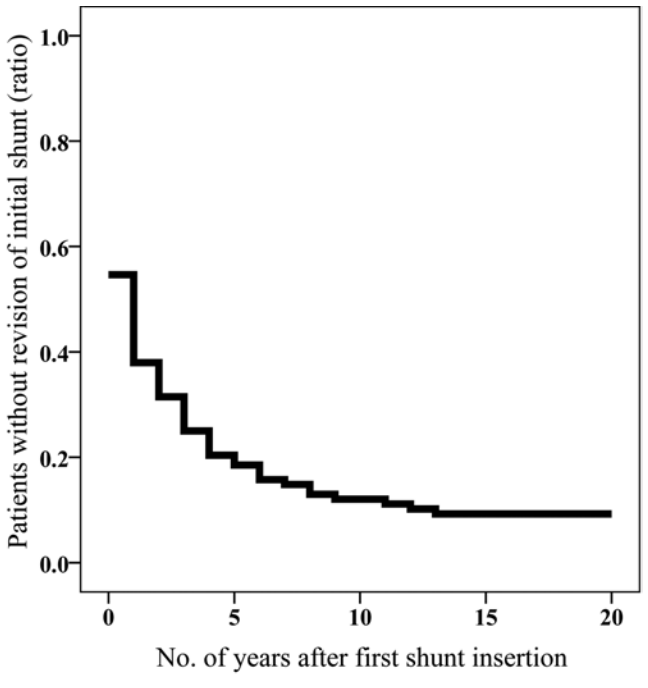

Fig. 1. Kaplan-Meier curve of patients who did not undergo revision of the initial shunt.

Shunt Dependency

At the time of this writing, 96 (89%) of the 108 surviving patients still retained a shunt. Of these, 78 (81%) patients have VP shunts. In the 12 (12.5%) in whom a primary VA shunt was placed, conversion to a VP shunt was performed in 11 cases during the follow-up period; just one patient has retained a VA shunt for 23 years.

Fifteen patients (10.9%) have had their shunt explanted during the follow-up period, and 12 remain shunt independent at present. The mean longevity of the shunt in these patients was 7.83 years (median 9.42 years, range 1 month–15.17 years). In 6 cases routine control radiological images revealed disconnected or displaced shunts; in 2 cases the shunt was found to be dysfunctional due to blockage during elective elongation surgery; 1 shunt was explanted due to infection; and 1 patient exhibited long-lasting symptoms, indicating overdrainage, and was therefore removed. In 3 patients signs of hypertension recurred and reimplantation of a shunt followed. Two of the patients, with MMC and aqueductal stenosis, respectively, were treated with ETV after shunt malfunction, but ETV failed to improve the symptoms and a shunt was reinserted.

TABLE 2: Number of shunt revisions in 138 children with hydrocephalus in a 20-year follow-up period shunt insertion

Age Group*	No. of Patients (%)	No. of Revisions				Total No. of Revisions
		0	1	2–4	>5	
<6 mos	68 (49)	6	9	26	27	304†
6 mo–1 yr	18 (13)	3	3	8	4	47
>1 yr	52 (38)	15	9	20	8	128

* Child's age at which the primary shunt was initially inserted.

† Significant difference of number of shunt revisions for this age group compared with that in the remaining patients, measured with independent t-test ($p < 0.05$).

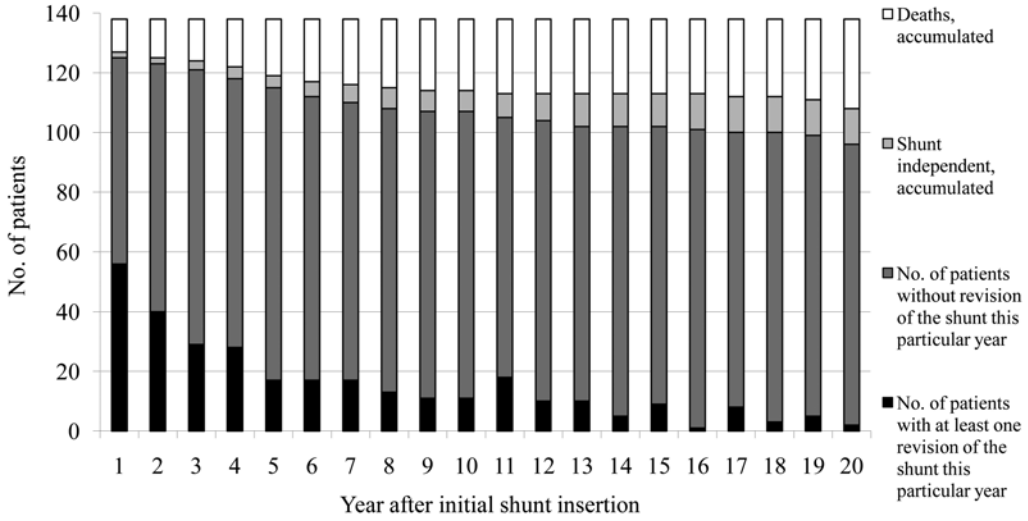

Fig. 2. Twenty-year annual distribution of data obtained in 138 shunt-treated children with hydrocephalus, showing accumulated deaths, acquired shunt independence, and the need of shunt revisions each year.

Mortality

Of the 138 patients evaluated, 30 (22%) died during the 20-year study period, and 24 died during the 1st decade after initial shunt insertion. Thirteen percent of patients died of nontumor-related causes. Analyses revealed survival rates of 93.5, 87.7, 82.6, and 78.3 at 1, 5, 10, and 20 years, respectively (from initial shunt insertion to death); the survival rate is illustrated in Fig. 3. Assessment of the cause of death among the aforementioned patients showed that 12 died of brain tumor, 11 of whom died within 9 years of the primary shunt procedure. One died of malignant schwannoma of the upper mediastinum. One died of a progressive metabolic disease affecting the nervous system (Hurler syndrome). Eight of the children had severe congenital syndromes or severe malformations for which life expectancy was considered limited. Two patients harbored a suboptimally functioning shunt for which further corrections were refused because other severe malformations were deemed incompatible with prolonged survival. In 2 patients who died, the cause of death was not likely shunt failure, although the precise cause remains uncertain.

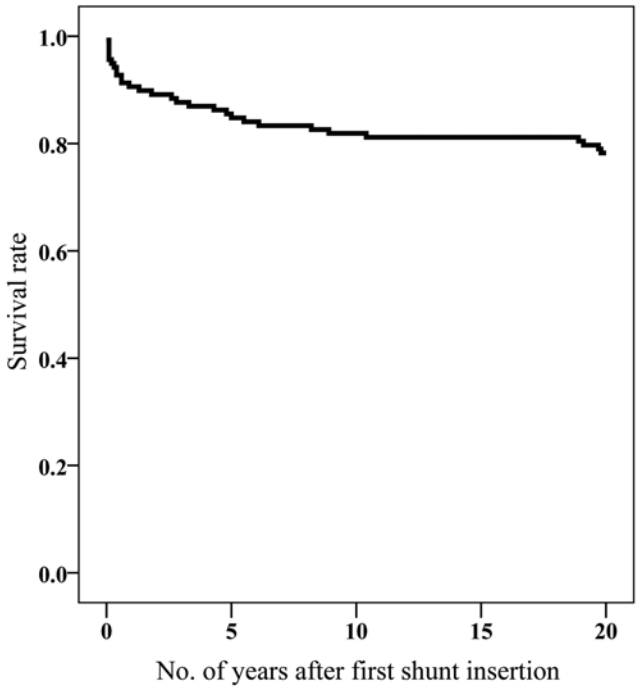

Fig. 3. Kaplan-Meier curve of survival rate in 138 children with hydrocephalus in whom a shunt was inserted during childhood.

Four patients (2.9%) most likely died of shunt failure. Two of the patients died during the 1st decade, whereas the other 2 died in the 2nd decade. One patient died of cerebral anoxia due to a blockage in the proximal catheter. In 1 patient there was a disconnection between the valve and the peritoneal catheter, and this patient died before being brought to neurosurgical attention. One patient with MMC died of herniation of the brainstem caused by prolonged intracranial hypertension despite several surgical attempts at CSF drainage. The last patient was found to be apneic the night before a planned shunt revision, and autopsy revealed cerebral anoxia caused by a blocked shunt.

Employment, Education, Living Arrangements, and Material Status

Among the 77 individuals who responded/could respond to the SF-36, 22 (23%) were students at the time they responded (Table 3). With respect to work, 53 reported to be currently employed; 31 (33%) were employed in the open labor market and 22 (23%) were in sheltered workplaces. In the working group the highest completed educational level was regular primary school (4 patients); 77% continued their school career in regular secondary schools and 10% had completed a higher educational level. Twenty individuals (21%) were not in contact with the labor market due to chronic illness.

Eleven patients reported living in sheltered units offering daily nursing assistance, and 9 were living in sheltered apartments with nursing assistance at least once a week; 32 patients lived with their families and 35 lived

TABLE 3: Education, work participation, and living arrangements in 95 patients

Education, Work, & Living	SF-36		
	Respondents	Nonrespondents*	All Respondents (%)
no. of patients	77	18	95
education level			
primary elementary school completed			
regular	74	1	75 (79)
special	3	17	20 (21)
secondary elementary school completed			
regular	72		72 (76)
special	1	7	8 (8)
higher educational level			
completed	5		5 (5)
ongoing	22		22 (23)
employment			
open employment	31		31 (33)
sheltered employment	22		22 (23)
no employment	2	18	20 (21)
residence			
independently	56		56 (59)
sheltered or in care	21	18	39 (41)
partner			
yes	21		21 (22)
no	56	18	74 (78)

independently. Consequently, 59% reported being fully independent in daily living and 41% needed some sort of assistance from family members if living at home or from nursing staff if living in sheltered living arrangements. Regarding material status, 22% reported having a spouse or a partner, and 78% not having a spouse or a partner.

Perceived Health of the Population Compared With Norwegian Background Population

The SF-36, Barthel Index, and Background Information Scheme were completed by 77 patients. Sixty-eight (88%) of the participants completed the questionnaires alone and 9 (12%) had assistance from a caregiver. Assistance was required mainly due to handwriting problems; 2 patients needed assistance due to visual impairments. The mean age of those who responded (51 males, 26 females) was 24.5 years (median 23.17 years, range 21–33 years).

Compared with the reference group, the young adults with hydrocephalus reported a poorer perceived health in 2 of the 8 SF-36 domains (Table 4). Male participants had significantly ($p < 0.05$) lower scores in 2 SF-36 domains (Physical Functioning and General Health) and females in 3 SF-36 domains (Physical Functioning, General Health, and Role Limitations due to physical health problems).

The mean SF-36 scores were investigated for each of the etiological subgroups and the results revealed significant difference in some of the domains illustrated in Table 5. Physical Functioning and PCS scores were poorer in the spina bifida group, and Role Limitation due to physical functioning and PCS scores were lower in the neoplasm group. The other malformations group scored lower for mental health and MCS scores. As to Physical Functioning, patients with hydrocephalus of unknown origin scored better than other respondents. No significant difference in perceived health was found in relation to the number of shunt revisions (grouped as 0, 1–4 or > 4 revisions).

Abilities in 18 Patients in Whom SF-36 was not a Feasible Measure of Perceived Health

In 18 patients considered to have limited mental resources as a result of the hydrocephalic condition itself or other diseases that reduced cognition and mental health, the SF-36 was not considered to be a useful measure of perceived health. This group was asked to complete only the Barthel Index and Background Information Scheme. One patient was deaf and mute. Limitations regarding physical functioning were also common in this group. Barthel Index ADL scores were significant below those of the patients who could respond to the SF-36 ($p < 0.01$; Table 6). Apart from a slightly higher frequency of shunt revisions (mean 5), neither etiology of hydrocephalus nor age at first shunt implantation (mean 13 months, median 5 months) differed significantly in this group compared with the other patients.

Who Were the Nonrespondents?

A total of 13 patients (12%) were nonrespondents. Lack of interest and time to complete the questionnaires were given as main reasons for not partaking in the sur-

TABLE 4: Summary of mean SF-36 scores and 95% CIs, stratified by sex, compared with a sex- and age-matched normative population without disabilities*

Health Domains	Total HC Population (95% CI)	Male (95% CI)		Female (95% CI)	
		HC Population	Norm Population†	HC Population	Norm Population†
no. of patients	77	51	233	26	270
Physical Functioning	80.6 (74.8–87.3)‡	86.3 (79.8–92.8)‡	94.7 (93.1–96.3)	69.4 (57.6–81.2)‡	94.0 (92.7–95.3)
Role Physical¶	76.3 (66.2–83.8)	84.3 (76.0–92.3)	88.5 (85.3–91.7)	60.6 (45.2–76.0)‡	85.2 (81.8–88.6)
Bodily Pain	80.3 (74.2–85.8)	83.7 (77.5–89.9)	83.4 (80.7–86.1)	73.5 (61.7–85.3)	79.7 (77.0–82.4)
General Health	75.5 (71.1–80.9)‡	77.1 (71.6–82.6)‡	83.0 (80.9–85.1)	72.2 (62.2–82.2)‡	82.5 (80.4–84.6)
Vitality	56.8 (52.8–61.2)	59.3 (54.7–63.9)	61.1 (58.9–63.3)	51.7 (43.2–60.2)	56.6 (54.2–59.0)
Social Functioning	84.9 (80.3–93.7)	86.5 (81.8–91.2)	88.0 (85.4–90.6)	81.7 (72.7–90.7)	85.3 (83.0–87.6)
Role Emotional§	86.6 (80.3–93.7)	89.5 (82.5–96.5)	84.4 (80.6–88.2)	80.8 (66.7–94.9)	78.9 (75.0–82.8)
Mental Health	75.8 (72.0–80.0)	77.0 (73.0–81.0)	77.9 (75.9–79.9)	73.4 (63.9–82.9)	76.4 (74.6–78.2)

* HC = hydrocephalus; Norm = normative.
† The SF-36 normative data are for the Norwegian population (age range 19–29 years), from Loge and Kaasa.
‡ Significant differences between participants with hydrocephalus and normative population.
¶ Role limitation due to physical health problems.
§ Role limitation due to emotional health problems.

veys. Nonresponse to the questionnaires reduced the effective sample size in our study, and the perceived health status and the disability of the nonrespondents remain broadly unknown. Compliance in QOL surveys has been reported to be lower in individuals with reduced physical functioning and chronic diseases;[12] therefore, we consider a response rate of 88% to be satisfactory in young individuals with shunt placed for hydrocephalus during childhood.

Discussion

Long-term outcome following shunt placement in patients with hydrocephalus has been evaluated in numerous studies since the 1960s. To date, no authors have provided a study regarding outcomes in a period exceeding 10–15 years. The present study was performed to determine surgical morbidity and mortality rates as well as social functioning and HRQOL during a period exceeding 20 years after first shunt implantation in an nonselected cohort of patients with hydrocephalus.

The male/female ratio in the studied cohort (1.9:1.0) is slightly higher than in other studied populations.[1,3,14] Our cohort, however, is an nonselected group of all children (< 15 years of age) who underwent a first CSF shunt

TABLE 5: Summary of mean SF-36 scores for each etiological subgroups compared with the remaining etiological subgroups as a whole*

SF-36 Health Domains†	Cause of Hydrocephalus								
	Hemorrhage	Neoplasm	Spina Bifida	Cysts	Aqueductal Stenosis	Other Malformations	CNS Infection	Postraumatic	Unknown
no. of patients	17	7	14	9	6	5	4	2	13
Physical Functioning			55.4 ± 31.3‡						97.7 ± 3.9¶
Role Physical§		42.9 ± 47.2‡							
Bodily Pain									
General Health									
Vitality									
Social Functioning									
Role Emotional**									
Mental Health						59.2 ± 34.8‡			
PCS		42.9 ± 13.3‡	45.1 ± 9.6‡						
MCS						42.0 ± 11.5‡			

* Mean scores are shown when significant differences were found with an independent t-test.
† The range of SF-36 domain scores is 0 (worst possible status) to 100 (best possible status).
‡ Significantly lower score for this etiological subgroup compared with the remaining, measured with an independent t-test ($p < 0.05$).
¶ Significantly higher score for this etiological subgroup compared with the remaining, measured with an independent t-test ($p < 0.05$).
§ Role limitation due to physical health problems.
** Role limitations due to emotional problems.

TABLE 6: Barthel Index of ADL in 95 patients with hydrocephalus

Variable	Barthel Index of ADL	
	SF-36 Respondents	SF-36 Nonrespondents
no. of patients	77	18
mean ± SD	94 ± 12.7	36 ± 13.7
median	100	37.5
minimum	45	15
maximum	100	55

placement through the calendar years 1985–1988 and were born in a geographically defined area. It is unlikely that our sample represents a more or less severely affected group of individuals with childhood hydrocephalus, and our findings can therefore be regarded as representative for children with hydrocephalus in general.

Overall Mortality Rate and Shunt-Related Deaths

The 20-year mortality rate of 22% (13% not related to tumors) is comparable with the rate in other reports, although our study differed in follow-up and patient selection, sometimes making it difficult to compare different series. In our relatively small sample size there is a clear difference in mortality in the 2 decades studied, although no significant difference was found ($p = 0.10$); 24 of 30 deaths occurred during in the 1st decade after initial shunt insertion. In another nonselected cohort of shunt-treated individuals, the 10-year mortality rate was reported to be 12.4%.[21] Lumenta and Skotarczak[18] reported a mortality rate of 13.7% in a long-term follow-up study (median 17 years, range 5–26 years) of congenital hydrocephalus in which peri- and intraventricular hemorrhage, tumor, and infection were excluded as causes. A 10-year mortality rate of 11% was reported by Casey et al.[3] when deaths due to tumors were excluded.

In our series there were 4 deaths (2.9%) that could be reliably attributable to shunt malfunction. Shunt-related deaths in other hydrocephalic populations have demonstrated rates of 2%–5%, which corroborates our findings.[3,9,11] These cases emphasize the potential dangers of a blocked shunt when not recognized promptly and dealt with in an expeditious manner. One patient did have diffuse symptoms for weeks before relation to shunt dysfunction was discovered. Therefore, providing guidelines regarding shunt failure symptoms to patients, family, and caregivers is still an important part of a neurosurgeon's work and should be emphasized. Additionally, surveillance scans should be used to identify cases of "diffuse" symptoms in patients with potential shunt failure; this could contribute to the detection of shunt dysfunction earlier to avoid secondary insults caused by prolonged intracranial hypertension, which in worst case scenarios can be fatal.

Shunt Revision and the Need of Follow-Up

Four (81%) of 5 patients in our study needed at least one shunt revision during the 20-year follow-up period. The annual number of revisions and the annual number of patients requiring revision surgery were significantly higher in the 1st than 2nd decade. There are a limited number of studies addressing functional outcome beyond 10 years after initial shunt placement. Lumenta and Skotarczak[18] reported a shunt revision rate of 71.2% in their cohort of individuals with hydrocephalus. Other comparable studies have revealed revision rates between 55% and 82% during a 10-year period.[3,20] Reducing the number of shunt revisions will always remain the desirable goal. Clearly, one of the best ways of managing shunt problems is avoiding them in the first place. Pediatric shunt surgery should be performed by pediatric neurosurgeons skilled in shunt hardware and techniques, and with an experience in deliberating the technical problems of shunt dependency and shunt revisions.

How often shunt-treated children and young adults should undergo follow-up evaluation is another topic for discussion. We recommend several visits during the 1st year, every year for children age 1–3 years, and every 2nd year in children between ages 4 and 16 years. Thereafter (after growth is completed), control examinations should be performed every 4–5 years. A control examination should include clinical consultation with emphasis on symptoms of possible shunt dysfunction supplied by MR or CT imaging of the brain. Plain radiographs of the shunt should also be acquired to assess shunt continuity, distal catheter position, and length.

Shunt Dependency

Twenty years after the initial shunt insertion 96 (89%) of the 108 patients still have a shunt implanted. We consequently have to assume that most of these individuals remain dependent an adequate shunt function. Twelve patients (9%) have been shunt independent during the observation period; in most of these cases the shunts were coincidently discovered to be dysfunctional and were explanted, and we can only speculate as to why some patients can live a shunt-free life after being shunt dependent in earlier life. Obviously there are different degrees of shunt dependence, and this group may have been less shunt dependent from the onset. In our cohort there was a higher frequency of shunt independence compared with another study in which shunt were removed in 3.2% of the patients after up to 15 years of follow-up.[10] Chronic intracranial hypertension, episodic or continuous, in previously shunt-treated children with apparent arrested hydrocephalus has been demonstrated in several reports;[25] therefore, intracranial pressure monitoring and a psychometric evaluation may be valuable tools in the continuing assessment of such patients.

This cohort of hydrocephalic children originates from the mid 1980s. For a long period after the introduction of shunt systems, ETV was performed only occasionally, but it has again become more common in recent years, in parallel with the development of advanced neuroendoscopic techniques.[5,6] Since the 1990s ETV has been used as our first option in selected cases. Although the vast majority of young adults with childhood hydrocephalus continue to be shunt dependent 20 years after first shunt insertion, there are reasons to believe that ETV can contribute to make a greater proportion of hydrocephalic patients shunt free in the future. The dictum "once a shunt, always a shunt" may no longer be as valid as in earlier decades.[8]

Work Participation and Social Functioning

We report that 23% of our population is not part of labor market because of chronic illness and severe disabilities. Twenty-three percent were still students and there are reasons to believe that in the future they will be part of the work force. Fifty-four percent of the patents were employed and fewer than half of these did have sheltered employment. We have reason to believe that the Norwegian Welfare System does have an impact on work participant rate in our cohort. Individuals with chronic disabilities receive governmental financial support and people with difficulty attending the open employment market are offered financial subsidies and sheltered work places to the highest possible extent. Besides financial independence, having a job is an important factor in social integration and self-esteem. In countries where social services are not as well established, it is possible that some of those in sheltered employment had been forced into the open employment market and that the most socially deprived individuals would be economically dependent of others.

A higher portion attending "normal" schools compared with those in other studies, where 60%–69% have been reported,[3,9] may partly be explained by national political tradition; all children should be treated equally and therefore integrated into regular schools. Potential risk of this integration policy may be that a somewhat larger proportion fall through the cracks at an earlier point in life and thus do not reach their potential outcome as young adults. To minimize disability and enhance participation in later life, it is important to assess and understand aspects of cognitive and behavioral problems in early childhood and the need for supportive schooling.

Approximately 40% remain dependent with respect to living arrangements, similar to 40% reported in another study of young adults with childhood hydrocephalus.[7] Despite being in their early 20s up to about age 35 years, the majority of the patients (78%) in the group were single. The young population (mean age 24.5 years) and the fact that behavioral problems[15] and physical handicaps are more common in hydrocephalic individuals may have been decisive here.

Perceived Health

This study showed that for 2 of the 8 SF-36 domains, the perceived health in young adults with hydrocephalus was poorer than that of a sex- and age-matched population. The difference was most pronounced for the Physical Functioning domain, covering walking, self-care ability, and strenuous activities, and the General Health domain. The marginal statistical significance for general health in both males and females were obliterated when Bonferroni correction was performed. The MCS (Vitality, Mental Health, Role Problems due to emotional problems and social functioning) and the Bodily Pain scores did not differ significantly from those of the Norwegian background population.

Table 5 illustrates tendencies of differences among the etiological subgroups. Hydrocephalic patients with spina bifida reported significantly poorer perceived scores for the Physical Functioning domain even when Bonferroni correction was performed. This finding confirms earlier observations.[22] Patients with hydrocephalus of unknown causes scored better on all domains, although a significantly higher score was obtained only in the Physical Functioning domain.

No significant difference in perceived health was found in relation to the number of shunt revisions. The authors of previous works correlating the number of shunt revisions and outcome have reported conflicting results. This present study corroborates those findings, with no significant differences according to the number of shunt revisions. Although many of patients with hydrocephalus live with a chronic condition where the prevalence of physical handicaps and other comorbidities by far exceeds that of the reference population, their perceived HRQOL may, viewing the cohort as a whole, appear more positive than one might expect.

Study Limitations

This study's limitation is that the chosen measure of perceived health, the SF-36 surey, was not feasible in all patients. A bias caused by the group in which SF-36 was considered to be unfeasible may have introduced an overestimation of HRQOL. Similar limitations do, however, apply to assessments of HRQOL when using questionnaires in general. The assessment of HRQOL in individuals unable to express themselves in words and terms understandable to the majority of us remains an important challenge. The population's lack of ability to communicate makes it difficult for us to say much about the individuals' experienced QOL, but this should not be interpreted in the worst light. Furthermore, neuropsychological testing may be a relevant alternative to the SF-36 survey for measuring perceived health and may be potential subject for future studies.

Conclusions

The results show that revision frequency was significantly higher in the 1st decade than the 2nd decade after initial shunt insertion and that the shunt revision rate was significantly higher in patients younger than 6 months of age when the first shunt was placed. After 20 years of follow-up, 10% of the patients still live with primarily inserted shunt hardware.

We conclude that the mortality rate in this cohort was high. Of the 30 deaths in total, 80% of the patients died during 1st decade and 20% in the 2nd. Most of the deaths were related to the primary cause underlying the hydrocephalic state in the first place. Nevertheless, and despite an almost omnipresent availability of CT scanning as well as modern shunt techniques, 4 children (2.9%) did die of shunt failure.

We found that the majority of hydrocephalic children live lives comparable with those of other young adults in our society. A substantial proportion of the patients nonetheless live a life dependent on a varying degree of assistance, and 1 of 5 is permanently outside the employment market. It is reasonable to ask for potential improvements, but a poor final outcome is also obviously related to irreversible primary brain damage, not only to damage

caused by the hydrocephalic state per se. Prevention has a role, consisting in prenatal diagnosis and counseling, as does aggressive medical treatment of the diseases known to cause hydrocephalus.

Despite the fact that hydrocephalus has several and severe effects on life, we think that HRQOL in young adults with childhood hydrocephalus seems to be less disheartening than anticipated.

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 to the study and manuscript preparation include the following. Conception and design: all authors. Acquisition of data: Lundar, Paulsen. Analysis and interpretation of data: Paulsen. Drafting the article: Paulsen. Critically revising the article: all authors. Reviewed final version of the manuscript and approved it for submission: all authors. Statistical analysis: Paulsen. Administrative/technical/material support: Lundar. Study supervision: Lundar, Lindegaard.

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