2.1 General data

2.1.1 Patients and data collection
Data were collected on 400 patients undergoing primary shunt operations for hydrocephalus between December 1989 and January 1992. These patients were operated on in eight different Dutch neurosurgical centres (table 2.1.1).

When the surgical procedure had been completed, the surgeon filled out a patient form and sent it to the study coordinator. After the surgery there was a follow-up period. At the end of the follow-up period, the study coordinator collected the adjuvant data on each patient. The data were collected from the patient’s medical record and/or through contacting the patient’s neurologist or general practitioner.

Of the patients 222 were male, 178 female. We divided the study population in three age groups: 0-1 year of age = group A (71♂ and 54♀), 1-15 years of age = group B (21♂ and 18♀) and over 16 years of age = group C (130♂ and 106♀). The minimum follow-up period was two years and 175 days, the maximum follow-up period was five years and 65 days. The mean follow-up period was three years and 294 days. A shorter follow-up period was unavoidable in case of mortality during the follow-up period.

2.1.2 Etiology of hydrocephalus and most prevalent pre-operative symptoms
Overall, tumor is the most prevalent cause of hydrocephalus (104). Next are normal pressure hydrocephalus (59) and meningomyelocele (39) (table 2.1.2). But for each age group a more specific preponderance in etiology could be discerned. In group A meningomyelocele is the most important cause of hydrocephalus (37), followed by perinatal hemorrhage (31) and aqueductal syndrome (19). In group B the majority of cases is caused by tumor (24). Tumor is most prevalent in group C (75), but in this last group normal pressure hydrocephalus (59) as well as subarachnoid hemorrhage (44) are also frequent causes of hydrocephalus.

Symptoms at presentation were an enlarged head (104), headache (88), NPH symptoms and impaired consciousness (75).

2.1.3 Surgical procedure
2.1.3.1 Surgeon
The shunts were inserted by 46 neurosurgeons or neurosurgical residents. The number of
insertions per surgeon varied from 1 to 48. Twenty-two surgeons inserted less than five shunts. For statistical analysis we divided the surgeons in two groups: one group with five years or less of experience (17), the other with more than five years of experience (29).

### 2.1.3.2 Duration of operative procedure
The duration of the shunting procedure varied from 22 to 180 minutes with a mean of 65 minutes.

<table>
<thead>
<tr>
<th>centre</th>
<th>group A</th>
<th>group B</th>
<th>group C</th>
<th>total</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>40</td>
<td>11</td>
<td>66</td>
<td>117</td>
</tr>
<tr>
<td>2</td>
<td>15</td>
<td>4</td>
<td>44</td>
<td>63</td>
</tr>
<tr>
<td>3</td>
<td>3</td>
<td>3</td>
<td>16</td>
<td>22</td>
</tr>
<tr>
<td>4</td>
<td>2</td>
<td>0</td>
<td>13</td>
<td>15</td>
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<tr>
<td>5</td>
<td>14</td>
<td>7</td>
<td>7</td>
<td>28</td>
</tr>
<tr>
<td>6</td>
<td>28</td>
<td>6</td>
<td>44</td>
<td>78</td>
</tr>
<tr>
<td>7</td>
<td>12</td>
<td>6</td>
<td>23</td>
<td>41</td>
</tr>
<tr>
<td>8</td>
<td>11</td>
<td>2</td>
<td>23</td>
<td>36</td>
</tr>
<tr>
<td>total</td>
<td>125</td>
<td>39</td>
<td>236</td>
<td>400</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>etiology</th>
<th>A</th>
<th>B</th>
<th>C</th>
<th>total</th>
</tr>
</thead>
</table>
meningomyelocele                 | 37 | 1  | 1  | 39    |
aqueductal syndrome              | 19 | 2  | 11 | 32    |
tumor                           | 5  | 24 | 75 | 104   |
inflammation                     | 11 | 2  | 11 | 24    |
SAH\(^a\)                        | 1  | 0  | 44 | 45    |
NPH\(^b\)                        | 0  | 0  | 59 | 59    |
perinatal hemorrhage             | 31 | 2  | 0  | 33    |
miscellaneous                    | 3  | 1  | 3  | 7     |
unknown                         | 10 | 2  | 11 | 23    |
pseudotumor cerebri             | 0  | 0  | 2  | 2     |
Arnold-Chiari                   | 2  | 1  | 0  | 3     |
trauma                          | 2  | 3  | 5  | 10    |
intracran. hematoma             | 2  | 0  | 12 | 14    |
other                           | 2  | 1  | 2  | 5     |
total                           | 125| 39 | 236| 400   |

\(^a\)subarachnoid hemorrhage
\(^b\)normal pressure hydrocephalus
2.1.3.3 Drainage route

The frontal (158: 139 R, 7 L, 12 both) and parietal (156: 137 R, 19 L) drainage routes were equally used. The occipital route (86: 74 R, 8 L, 4 both) was less frequently used. In 348 patients a ventriculoperitoneal (VP) shunt was inserted, while 45 patients got a ventriculoatrial (VA) shunt. The El Shafei procedure was performed in six patients. In this procedure CSF is drained to the jugular vein in a retrograde way. In one patient the exact route of drainage remained unknown.

2.1.3.4 peri-operative antibiotics

334 patients (83.5%) were on antibiotics in the perioperative period. The antibiotics used were: floxapen (155), cotrimoxazol (96), cefuroxim (38), kefsol (27), cloxacillin (6), ceffacidal (3) and vancomycin (1). A combination of antibiotics was given in one case. In two cases the type of antibiotic remained unknown.

2.1.4 Shunt systems

2.1.4.1 Valves

Most frequently used was the Pudenz Schulte valve (101). The Holter (79), Orbis Sigma (63) Hakim (33), Accu-flo (30), Phoenix (26), Pudenz flushing valve (24) and Raimondi valve (20) were less frequently inserted. More than half (211) of these valves had 'medium' as a specification. The ‘low’ valve was inserted 89 times. The Orbis Sigma valve (flow limitation) was used in 63 cases. (Table 2.1.3 and 2.1.4).

2.1.4.2 Shunt components

In 90 cases a one-piece shunt was inserted. The other patients got a combination of several shunt components which are connected to get a complete shunt system. In five patients an anti siphon device (ASD) was inserted. A reservoir was part of the system in 178 patients. The mean number of shunt connections was 1.9, with a mean of 1.3 in the youngest group, 2.4 in the middle group and 2.2 in the oldest group.

The mean ventricular catheter length was 6.7 cm, varying from 6.1 cm in group A to 7.0 in group C. The mean abdominal catheter length was 29.7 cm: 33.2 cm in group A, 33.5 cm in group B and 26.5 cm in group C.

2.1.5 Pre-operative external ventricular drainage (EVD)

A hundred and eleven patients underwent EVD before internal shunt insertion. Reasons for this EVD were an increase in intracranial pressure (43), often in combination with hemorrhage (26), infection (11) and/or the need for pressure recording (11).

2.1.6 Cerebrospinal fluid analysis

Pre-operative ventricular CSF analysis (protein, leucocytes, erythrocytes) was done in 80 patients. All these patients had an EVD. In addition to this, 59 patients had their CSF analyzed during the operation. There was a certain overlap between patients in these groups,
so for statistical analysis we decided to include only the most recently obtained CSF values of protein, leucocytes and erythrocytes. For example, for a patient whose CSF was analyzed prior as well as during the shunt operation, we included the values obtained during the shunt operation.

Table 2.1.3 Valve types per age group

<table>
<thead>
<tr>
<th>valve type</th>
<th>group A</th>
<th>group B</th>
<th>group C</th>
<th>total</th>
</tr>
</thead>
<tbody>
<tr>
<td>PS medical</td>
<td>52</td>
<td>9</td>
<td>40</td>
<td>101</td>
</tr>
<tr>
<td>Holter</td>
<td>17</td>
<td>5</td>
<td>57</td>
<td>79</td>
</tr>
<tr>
<td>Phoenix</td>
<td>1</td>
<td>5</td>
<td>20</td>
<td>26</td>
</tr>
<tr>
<td>Pudenz flushing valve</td>
<td>6</td>
<td>3</td>
<td>15</td>
<td>24</td>
</tr>
<tr>
<td>Accu-flo</td>
<td>2</td>
<td>4</td>
<td>24</td>
<td>30</td>
</tr>
<tr>
<td>Orbis Sigma</td>
<td>28</td>
<td>3</td>
<td>32</td>
<td>63</td>
</tr>
<tr>
<td>Hakim</td>
<td>7</td>
<td>3</td>
<td>23</td>
<td>33</td>
</tr>
<tr>
<td>Raimondi</td>
<td>11</td>
<td>5</td>
<td>4</td>
<td>20</td>
</tr>
<tr>
<td>Sophy</td>
<td>0</td>
<td>0</td>
<td>7</td>
<td>7</td>
</tr>
<tr>
<td>Delta</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Hakim Medos (np)(^b)</td>
<td>1</td>
<td>1</td>
<td>7</td>
<td>9</td>
</tr>
<tr>
<td>Hakim Medos (p)(^c)</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>1</td>
</tr>
</tbody>
</table>

| total                         | 125     | 39      | 230     | 394\(^a\) |

\(^a\) An El Shafei procedure was performed in six cases. In this procedure no valve is used.
\(^b\) np=non-programmable
\(^c\) p=programmable

Table 2.1.4 Valve specification per age group

<table>
<thead>
<tr>
<th>valve</th>
<th>group A</th>
<th>group B</th>
<th>group C</th>
<th>total</th>
</tr>
</thead>
<tbody>
<tr>
<td>low pressure</td>
<td>65</td>
<td>7</td>
<td>17</td>
<td>89</td>
</tr>
<tr>
<td>medium pressure</td>
<td>30</td>
<td>25</td>
<td>156</td>
<td>211</td>
</tr>
<tr>
<td>high pressure</td>
<td>1</td>
<td>0</td>
<td>4</td>
<td>5</td>
</tr>
<tr>
<td>flow</td>
<td>28</td>
<td>3</td>
<td>32</td>
<td>63</td>
</tr>
<tr>
<td>adjustable</td>
<td>0</td>
<td>0</td>
<td>8</td>
<td>8</td>
</tr>
<tr>
<td>normal</td>
<td>0</td>
<td>3</td>
<td>10</td>
<td>13</td>
</tr>
<tr>
<td>low/medium</td>
<td>1</td>
<td>0</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>level II</td>
<td>0</td>
<td>1</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>unknown</td>
<td>0</td>
<td>0</td>
<td>2</td>
<td>2</td>
</tr>
</tbody>
</table>

| total            | 125     | 39      | 230     | 394\(^a\) |

\(^a\) An El Shafei procedure was performed in six cases. In this procedure no valve is used.
Results obtained through lumbar puncture were not included in the analysis because lumbar CSF may have a different composition compared to ventricular CSF.

2.1.7 Statistical analysis
For statistics we used the $\chi^2$-test and the Student’s t-test. In both tests, a p-value of 0.05 or less was considered significant. An analysis of variance was done once to look at the influence of several factors together on the outcome. With this test, significance was again attained with a p-value of 0.05 or less.

2.2 Results

2.2.1 Number of revisions and interval until revision
Of the 400 patients included in this study, 109 (27%) underwent a revision operation. Of these patients, 51 needed a second revision and 19 needed a third. Two patients underwent a seventh revision. Overall there were 200 revisions. Therefore the mean revision rate per patient was 50%.

Looking only at the first revision, the revision rate per patient is 27% (see above). In group A 55 patients (44%) needed a first revision. In group B a first revision was necessary for 7 patients (18%) and in group C 47 patients (20%) underwent a first revision. These differences in revision rates we found between the three age groups are significant ($\chi^2$; p<0.0001). The mean age of the whole group that underwent revision was 24 years, for the group without revision this was 36 years.

Between hospitals, there were no significant differences in the rate of first revisions per patient with, all age groups considered, a minimum of 6% to a maximum of 28%.

The mean number of days until the first revision was 230 (min. 2 days, max. 5 years). For group A this was 262 days (min. 2 days, max. 5 years), for group B 205 days (min. 30 days, max. 3 years) and for group C 195 days (min. 3 days, max. 3 years).

Figure 2.2.1 shows a shunt survival curve for all age groups together, excluding patients who died during the follow-up. After two and a half years of follow-up, almost 85% of the shunts are still functioning well. The highest number of malfunctioning shunts occurred in the first 4 months after insertion.

2.2.2 Causes of shunt dysfunction
The causes of shunt dysfunction were divided in five categories: proximal dysfunction, valve dysfunction, distal dysfunction, infection and ‘unknown’ (see table 2.2.5).

2.2.2.1 Proximal dysfunction
The most prevalent cause of shunt dysfunction was proximal catheter problems (89; 45%). These problems could either be obstruction (debris (55), plexus (15) or blood (1)) or malposition (18).

Proximal dysfunction within a month after primary insertion was caused by debris
obstructing the catheter or, by malposition. Obstruction later on was due to plexus ingrowth or, again, debris. In 2.2.6.2 the correlation between proximal shunt obstruction and ventricular catheter length will be analyzed further.

2.2.2.2 Valve dysfunction
In 35 (17.5%) cases it was the valve which caused shunt malfunction, but in 18 of these cases the exact valve problem remained unknown. Ten revisions were performed because of valve obstruction. In the other seven patients the valves were patent, but they had the wrong resistance: four valves had a resistance too low which caused overdrainage, three valves had a resistance too high causing underdrainage.

2.2.2.3 Distal dysfunction
Distal catheter dysfunction occurred less frequently (31; 15.5%). Usually there was a malposition (19) of the abdominal catheter. Obstruction was found in five cases, a cyst in one case and in six cases the cause of the distal dysfunction remained unknown.

2.2.2.4 Infection
2.2.2.4.1 Definition of infection  Infection was diagnosed according to the CSF shunt infection criteria based on clinical and laboratory findings as discussed by Walters et
Clinical manifestations such as fever alone or in combination with other symptoms of infection, peritoneal or meningeal signs of inflammation, wound infection or induration of the shunt tract, along with positive cultures of either CSF, blood, shunt device or any other combination of these, contributed to the diagnosis of infection.

2.2.2.4.2 Epidemiology of infection
Infection was the reason for revision in 22 cases (see table 2.2.6). The most frequently found organism causing infection was Staphylococcus (S.) epidermidis (50%), followed by S. aureus (23%). In 19 cases infection occurred within one and a half month after the primary shunt operation. The other three occurred later: respectively 3, 12 and 19 months postoperatively. In two cases the infection was caused by an organism already cultured from the CSF at the time of the operation.

The overall infection rate was 5.5% and in 3% of the cases an infection occurred after primary shunt insertion. For group A we found an infection rate of 6.0% for primary operations and revisions together. In group B this rate was 1.9% and in group C the infection
rate was 2.2%. Per hospital the infection rate per procedure varied from 1 to 6.7%.

Looking at the infection rate per etiology of hydrocephalus, we see considerable differences. Of all patients with a perinatal hemorrhage, 15.2% needed a revision because of infection. For patients suffering from hydrocephalus by an inflammatory process, 12.5% underwent revision and patients with meningomyelocele-related hydrocephalus needed revision for an infected shunt in 10.3% of the cases. However, patients with a tumor or normal pressure hydrocephalus only needed revision because of infection in 1.9 and 1.7% of cases respectively.

In patients who underwent EVD prior to primary shunt operation, the infection rate per procedure was 6.8%. This rate is significantly higher than the rate of the group as a whole ($\chi^2; p=0.0447$).

### Table 2.2.6 Infection

<table>
<thead>
<tr>
<th>Infecting Organism</th>
<th>Revision 1</th>
<th>Revision 2</th>
<th>Revision 4</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>S. epidermidis</td>
<td>5</td>
<td>5</td>
<td>1</td>
<td>11 (50%)</td>
</tr>
<tr>
<td>S. aureus</td>
<td>2</td>
<td>3</td>
<td></td>
<td>5 (23%)</td>
</tr>
<tr>
<td>Streptococcus</td>
<td>2</td>
<td></td>
<td>2</td>
<td>2 (9%)</td>
</tr>
<tr>
<td>E. coli</td>
<td>2</td>
<td></td>
<td>2</td>
<td>2 (9%)</td>
</tr>
<tr>
<td>Meningococcus</td>
<td>1</td>
<td></td>
<td>1</td>
<td>1 (4.5%)</td>
</tr>
<tr>
<td>Miscellaneous</td>
<td></td>
<td>1</td>
<td></td>
<td>1 (4.5%)</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>12</strong></td>
<td><strong>9</strong></td>
<td><strong>1</strong></td>
<td><strong>22 (100%)</strong></td>
</tr>
</tbody>
</table>

2.2.2.4.3 Treatment of infection In most cases infection was treated with antibiotics in combination with shunt removal, temporary EVD and eventual replacement of the shunt system. In one patient shunt removal and insertion of a new system at the other side was performed in the same procedure. This patient later developed another infection, caused by the same organism.

2.2.2.4.4 Peri-operative antibiotics and infection Twelve patients needed a first revision because of infection: ten out of 334 patients (3.0%) from the group that did got peri-operative antibiotics caught an infection, two out of 66 patients (3.0%) from the group that did not get peri-operative antibiotic prophylaxis became infected. Thus, the patients with an infection were equally divided between the group with antibiotics and the group without ($\chi^2; p=0.632$).

2.2.3 Revisions and etiology of the hydrocephalus A revision was necessary for 51% of patients with a meningomyelocele. The revision rates for patients with an aqueductal syndrome, a perinatal hemorrhage, an inflammatory process or a combination of etiologies were also high: resp. 44%, 39%, 33% and 43%. Of the patients with the most prevalent etiologies, patients with a tumor had the lowest revi-
sion rate (17%). These differences in revision rates per etiology are significant ($\chi^2$; $p=0.003$). See table 2.2.7. Although we did not see many revisions among NPH patients (22%), some of them did not improve clinically after primary shunting without having been revised (7 out of 59).

2.2.4 Surgical procedure and revision rate

2.2.4.1 Surgeon

The group of surgeons with five years or less of experience performed a total amount of 178 primary shunt operations. The more experienced group performed a total of 219 primary shunt operations. In three cases we have not been able to trace the identity of the surgeon. Of 109 first revisions, 51 were on patients who had their primary shunt surgery done by an ‘unexperienced’ surgeon and 58 were on patients operated on by an ‘experienced’ surgeon. Statistical analysis showed no significant difference in these numbers ($\chi^2$; $p=0.630$). The experience of the surgeon did not influence the duration of the operative procedure (t-test; $p=0.328$). In fact, the ‘unexperienced’ surgeons needed less time (63 minutes versus 66 minutes).

2.2.4.2 Duration of the operative procedure

The duration of the operative procedure was not significantly different for patients who later underwent revision (64 minutes) compared to those who did not (65 minutes), ($\chi^2$; $p=0.761$).

2.2.4.3 Drainage route

Of all frontal shunts, 46 out of 158 (29%) needed at least one revision. For parietal shunts this was about the same: 41 of 156 shunts (26%). Twenty-two of the 86 (26%) occipital shunts were revised. Thus, the location of the ventricular drain was of no significant importance ($\chi^2$; $p=0.976$).

Three out of six patients with an El Shafei shunt needed a first revision. One of them had a shunt infection, the second had a distal catheter obstruction with thrombus and the third had not improved clinically after shunting. In these three patients the El Shafei shunt was replaced with a VP shunt at revision. Eventually they did well having a VP shunt.

The revision rates for VP shunts and VA shunts were almost equal: resp. 27% and 24%.

2.2.5 Shunt system and first revision

2.2.5.1 Valves

Generally, there are no significant differences in revision rates per valve ($\chi^2$; $p=0.151$), as can be seen in table 2.2.8. But looking more closely at the figures, it is clear that the Phoenix (15%) as well as the Orbis Sigma valve (17%) had low revision rates and that the Raimondi valve (50%) had a rather high revision rate compared to the other valves. With the Raimondi valve (used in 20 patients), which is a distal catheter slit valve, two distal dysfunctions occurred (10%). With the other valves the rate of distal dysfunction was 12%.
### Table 2.2.7  Etiology and first revision in different age groups

<table>
<thead>
<tr>
<th>etiology</th>
<th>A</th>
<th>B</th>
<th>C</th>
<th>total&lt;sup&gt;c&lt;/sup&gt;</th>
<th>% infection</th>
</tr>
</thead>
<tbody>
<tr>
<td>meningomyelocele</td>
<td>37</td>
<td>1</td>
<td>1</td>
<td>39 (51%)</td>
<td>10</td>
</tr>
<tr>
<td>aqueductal syndrome</td>
<td>19</td>
<td>2</td>
<td>11</td>
<td>32 (43%)</td>
<td>6</td>
</tr>
<tr>
<td>tumor</td>
<td>5</td>
<td>24</td>
<td>75</td>
<td>104(17%)</td>
<td>2</td>
</tr>
<tr>
<td>inflammation</td>
<td>11</td>
<td>2</td>
<td>11</td>
<td>24 (33%)</td>
<td>13</td>
</tr>
<tr>
<td>SAH&lt;sup&gt;a&lt;/sup&gt;</td>
<td>1</td>
<td>0</td>
<td>44</td>
<td>45 (20%)</td>
<td>2</td>
</tr>
<tr>
<td>NPH&lt;sup&gt;b&lt;/sup&gt;</td>
<td>0</td>
<td>0</td>
<td>59</td>
<td>59 (22%)</td>
<td>2</td>
</tr>
<tr>
<td>perinatal hemorrhage</td>
<td>31</td>
<td>2</td>
<td>0</td>
<td>33 (39%)</td>
<td>15</td>
</tr>
<tr>
<td>miscellaneous</td>
<td>3</td>
<td>1</td>
<td>3</td>
<td>7 (43%)</td>
<td>--</td>
</tr>
<tr>
<td>unknown</td>
<td>10</td>
<td>2</td>
<td>11</td>
<td>23 (35%)</td>
<td>9</td>
</tr>
<tr>
<td>pseudotumor cerebri</td>
<td>0</td>
<td>0</td>
<td>2</td>
<td>2 (0%)</td>
<td>--</td>
</tr>
<tr>
<td>Arnold-Chiari</td>
<td>2</td>
<td>1</td>
<td>0</td>
<td>3 (0%)</td>
<td>--</td>
</tr>
<tr>
<td>trauma</td>
<td>2</td>
<td>3</td>
<td>5</td>
<td>10 (10%)</td>
<td>--</td>
</tr>
<tr>
<td>intracran. hematoma</td>
<td>0</td>
<td>0</td>
<td>12</td>
<td>14 (7%)</td>
<td>--</td>
</tr>
<tr>
<td>other</td>
<td>0</td>
<td>0</td>
<td>2</td>
<td>5 (10%)</td>
<td>--</td>
</tr>
<tr>
<td>total</td>
<td>125</td>
<td>39</td>
<td>230</td>
<td>400(27%)</td>
<td></td>
</tr>
</tbody>
</table>

<sup>a</sup> subarachnoid hemorrhage  
<sup>b</sup> normal pressure hydrocephalus  
<sup>c</sup> number of patients, (% first revision)

### Table 2.2.8  Valve type and first revision in different age groups

<table>
<thead>
<tr>
<th>valve type</th>
<th>A</th>
<th>B</th>
<th>C</th>
<th>total&lt;sup&gt;c&lt;/sup&gt;</th>
</tr>
</thead>
<tbody>
<tr>
<td>PS medical</td>
<td>52</td>
<td>9</td>
<td>40</td>
<td>101(27%)</td>
</tr>
<tr>
<td>Holter</td>
<td>17</td>
<td>5</td>
<td>57</td>
<td>79(29%)</td>
</tr>
<tr>
<td>Phoenix</td>
<td>1</td>
<td>5</td>
<td>20</td>
<td>26(15%)</td>
</tr>
<tr>
<td>Pudenz flushing valve</td>
<td>6</td>
<td>3</td>
<td>15</td>
<td>24(38%)</td>
</tr>
<tr>
<td>Accu-flo</td>
<td>2</td>
<td>4</td>
<td>24</td>
<td>30(20%)</td>
</tr>
<tr>
<td>Orbis Sigma</td>
<td>28</td>
<td>3</td>
<td>32</td>
<td>63(17%)</td>
</tr>
<tr>
<td>Hakim</td>
<td>7</td>
<td>3</td>
<td>23</td>
<td>33(33%)</td>
</tr>
<tr>
<td>Raimondi</td>
<td>11</td>
<td>5</td>
<td>4</td>
<td>20(50%)</td>
</tr>
<tr>
<td>Sophy</td>
<td>0</td>
<td>0</td>
<td>7</td>
<td>7 (14%)</td>
</tr>
<tr>
<td>Delta</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>1 (100%)</td>
</tr>
<tr>
<td>Hakim Medos (np)&lt;sup&gt;a&lt;/sup&gt;</td>
<td>1</td>
<td>1</td>
<td>7</td>
<td>9 (33%)</td>
</tr>
<tr>
<td>Hakim Medos (p)&lt;sup&gt;b&lt;/sup&gt;</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>1 (0%)</td>
</tr>
<tr>
<td>total</td>
<td>125</td>
<td>39</td>
<td>230</td>
<td>394(27%)</td>
</tr>
</tbody>
</table>

<sup>a</sup> non-programmable  
<sup>b</sup> programmable  
<sup>c</sup> number of patients, (% revision)
Because the mechanism of the Orbis Sigma valve (flow limitation) is based on a different principle than the mechanism of the other valves (differential pressure), we compared the Orbis Sigma valve alone with the other valves together for the occurrence of shunt dysfunction in each age group. Table 2.2.9 shows the mean number of revisions per subgroup.

An analysis of variance on the influence of the main factors age and valve type on shunt dysfunction showed a significant effect of these main factors together (analysis of variance; p<0.0001). There is also a significant effect of age alone, as well as valve type alone. The Orbis Sigma valve causes significantly less shunt dysfunction (analysis of variance; p=0.008) than the other valves. The youngest age group needs significantly more revisions (analysis of variance; p<0.0001), see also paragraph 2.2.1.

As shown in Table 2.2.9, the Orbis Sigma valve has positive effects mainly in age groups A and B. In group C the mean number of revisions for the Orbis Sigma valve and the other valve types is almost the same (respectively 0.19 and 0.20). In group C we paid special attention to NPH patients. In these patients, the Orbis Sigma valve did not cause significantly less shunt dysfunction than the other valve types, although the mean number of revisions was lower with the Orbis Sigma valve (0.17 versus 0.23; t-test, p=0.589).

For the group as a whole, valve specification (Table 2.2.10) was of significant importance for revision rate ($\chi^2$; p=0.003). Focusing on the three most frequently used valves, we found the following results: there was no significant difference between low pressure valves and medium pressure valves (t-test; p=0.483), but shunt systems in which the Orbis Sigma valve had been incorporated, necessitated significantly less revisions than shunt systems in which the low or medium pressure valve had been used (t-test; resp. p=0.001 and p=0.026).

2.2.5.2 Shunt components
The number of shunt connections is of no significant importance for the revision rate ($\chi^2$; p=0.749). The mean number of connections in the group that underwent revision was 2.0, in the other group it was 1.9.

The ventricular catheter length was significantly shorter ($\chi^2$; p=0.035) in the group that needed revision (6.4±1.7 cm) than in the group that did not (6.8±1.6 cm). Per age group, we looked at the relation between ventricular catheter length and the occurrence of obstruction of the proximal catheter. In group A the ventricular catheter length in the group with proximal obstruction was 5.2 cm, whereas the ventricular catheter length in the group without obstruction was 6.2 cm. This difference was almost significant (t-test; p=0.073). In group B the ventricular catheter length in the groups with and without revision was respectively 6.0 and 6.4 cm. This difference was not significant (t-test; p=0.642). The ventricular catheter length in group C for patients with and without obstruction was resp. 6.0 and 7.0 cm. The difference in length was almost significant (t-test; p=0.092). In all age groups together, the difference is significant: (t-test; p=0.003). There was no difference in the abdominal catheter length between the groups with and without revision ($\chi^2$; p=0.108). In the children under one year of age we paid extra atten-
tion to the correlation between revision rate and abdominal catheter length. But again, there was no significant difference between the groups with and without revision.

### 2.2.6 CSF composition and revision

For protein, leucocytes and erythrocytes we first compared patients who had had EVD prior to shunt operation with those who had not (fig. 2.2.2). Protein and leucocyte concentration were significantly higher in the EVD group (t-test; resp. p=0.01 and p=0.02). The mean erythrocyte concentration was higher in the group with EVD (2018 vs. 1817/μl) but this was not significant (t-test; p=0.159).

Again drawing a distinction between patients who had had and who had not had EVD prior to primary shunt insertion, we then looked at possible differences in CSF composition in those who did and those who did not undergo a first revision. The leucocyte counts in EVD patients with revision were significantly higher than in those without revision (t-test; p=0.002). There was no difference for protein or erythrocyte counts (t-test; resp. p=0.901 and p=0.214), see fig. 2.2.3. In the patients who had not had EVD, no difference was found in any of the CSF parameters: erythrocyte counts (t-test; p=0.837), leucocyte

<table>
<thead>
<tr>
<th>Table 2.2.9</th>
<th>Mean number of revisions per subgroup</th>
</tr>
</thead>
<tbody>
<tr>
<td>age group</td>
<td>Orbis Sigma valve</td>
</tr>
<tr>
<td>A</td>
<td>0.18</td>
</tr>
<tr>
<td>B</td>
<td>0.00</td>
</tr>
<tr>
<td>C</td>
<td>0.19</td>
</tr>
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</table>

<table>
<thead>
<tr>
<th>Table 2.2.10</th>
<th>Valve specification and first revision in different age groups</th>
</tr>
</thead>
<tbody>
<tr>
<td>valve</td>
<td>A</td>
</tr>
<tr>
<td></td>
<td></td>
</tr>
<tr>
<td>low pressure</td>
<td>65</td>
</tr>
<tr>
<td>medium pressure</td>
<td>30</td>
</tr>
<tr>
<td>high pressure</td>
<td>1</td>
</tr>
<tr>
<td>flow&lt;sup&gt;b&lt;/sup&gt;</td>
<td>28</td>
</tr>
<tr>
<td>adjustable</td>
<td>0</td>
</tr>
<tr>
<td>normal</td>
<td>0</td>
</tr>
<tr>
<td>low/medium</td>
<td>1</td>
</tr>
<tr>
<td>level II</td>
<td>0</td>
</tr>
<tr>
<td>unknown</td>
<td>0</td>
</tr>
<tr>
<td>total</td>
<td>125</td>
</tr>
</tbody>
</table>

<sup>a</sup> number of patients, (% revision)

<sup>b</sup> flow=Orbis Sigma valve
counts (t-test; p=0.082) and protein concentration (t-test; p=0.460). See fig. 2.2.4.

To look very closely at the influence of erythrocyte concentration on revision rate, we compared patients with the lowest erythrocyte levels (<500/μl) with those with the highest erythrocyte levels (>3000/μl). Again, no difference in revision rate could be detected (t-test; p=0.353)

2.2.7 Morbidity
Postoperative morbidity was defined as ‘added morbidity’ after operation. Nine patients (2.3%) showed morbidity after their primary shunt operation. In one patient a treatment other than the shunt insertion itself was the cause of this morbidity. This patient had a venous leg thrombosis. In another patient a pre-existing tumor caused morbidity. Symptoms resulting from a subdural hematoma occurred in two cases. Symptoms after postoperative hemorrhage also occurred twice. Postoperative seizures, transient hemiplegia and transient CSF hypotension were also mentioned in single cases as morbidity symptoms.

Eventually the morbidity was persistent in only four cases (1%) after primary shunt insertion: one as a result of a subdural hematoma, one because of tumor, one because of postoperative hemorrhage and in one case seizures were the cause of morbidity.

In five cases a postoperative radiologic diagnosis of subdural hygroma or hematoma was made, without any clinical signs.

2.2.8 Mortality
In the follow-up period, 68 out of 400 patients died: ten of them were in group A, nine were in group B and the other 49 were in group C. The most important cause of death was a pre-existing tumor (26); pulmonary (9) and cardial (7) problems were also important.

The mortality of two patients was shunt related. One appeared to have a subdural hematoma, the other died of a shunt infection (aspergillus) which caused obstruction of the shunt system.

After revision another nine patients died. Again, tumor was the most frequent cause of death: three patients. Another three patients died of unknown causes and two succumbed after pulmonary problems. Only one death could be explained by shunt dysfunction. The proximal catheter was found to be obstructed by debris.

The overall shunt related mortality was 0.5%.
Figure 2.2.2  CSF composition in each patient group (mean; standard deviation)

Figure 2.2.3  CSF composition in patients with and without revision. All patients with EVD (mean; standard deviation).
2.3 Discussion

Since the implantation of the first valves in the early fifties, progress has been made in the development of more suitable biomaterials and new valve-systems have become available. The physiology and hydrodynamics of the cerebrospinal fluid circulation were better understood and new diagnostic methods became available. Also, great improvements were made in pre- and postoperative care\textsuperscript{101}. However, postoperative complications in shunt surgery remain a major concern. The majority of these complications are shunt obstructions; either proximal, distal or valve-related. In the literature, shunt infections are also mentioned as an important cause of many shunt problems.

This study analyzed 400 patients who underwent a total of 600 CSF shunt operations. These patients were collected from eight Dutch neurosurgical centres and were followed postoperatively for at least two and a half years. We wrote requests for participation in the study to all major Dutch neurosurgical centers and eight hospitals were willing to participate. Because the incidence of shunted hydrocephalus patients is about 650\textsuperscript{127}, we considered the 400 patients collected during two years in this study (± one third of the whole Dutch population) representative for all Dutch neurosurgical hydrocephalus patients.

In this part the data described in part 2.1 and 2.2 will be further analyzed and discussed in relation to data from the recent literature.

\begin{figure}[h]
\centering
\includegraphics[width=\textwidth]{figure224.png}
\caption{CSF composition in patients with and without revision. All patients without EVD (mean; standard deviation)}
\end{figure}

2.3 Discussion

Since the implantation of the first valves in the early fifties, progress has been made in the development of more suitable biomaterials and new valve-systems have become available. The physiology and hydrodynamics of the cerebrospinal fluid circulation were better understood and new diagnostic methods became available. Also, great improvements were made in pre- and postoperative care\textsuperscript{101}. However, postoperative complications in shunt surgery remain a major concern. The majority of these complications are shunt obstructions; either proximal, distal or valve-related. In the literature, shunt infections are also mentioned as an important cause of many shunt problems.

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In this part the data described in part 2.1 and 2.2 will be further analyzed and discussed in relation to data from the recent literature.
2.3.1 Shunt revision rate per patient
The study-population consisted of 400 patients who underwent 200 revision operations during the follow-up period. Therefore the mean revision rate per patient was 50%. The rate of first revisions per patient was 27%. There were no major differences in revision rates between Dutch hospitals.

In 1994 Lund-Johansen et al. studied a population of adult patients only and found 66% revisions per patient (follow up time varied from 1 to 9 years). In 1994 Kast et al. reported 78% revisions per patient in a slightly younger population (2 months to 26 years old) than our study-population. A study performed in the children’s hospitals in Toronto and Paris showed an even higher revision rate of 120%.

The overall revision rate per patient we found is rather low compared to the other studies. This difference cannot be explained by differences in age or etiology of the patient population. Our Dutch population consisted of children as well as adults. Adults apparently need less revisions than infants (see 2.3.5). Even with this mixed population we observed less revisions than Lund-Johansen et al. reported in an adult population. A possible explanation for the low revision rate in the study could be that during the last decades, improvements in overall medical management of shunt patients have probably enabled Dutch neurosurgeons to reach a relatively high standard in CSF shunt surgery.

2.3.2 Causes of shunt dysfunction
2.3.2.1 Proximal dysfunction
The most frequent cause of shunt malfunction was a dysfunction of the ventricular catheter. This was found in 45% of the revisions. Dysfunction in the first postoperative month was either because of a catheter malposition or an obstruction by debris. Dysfunction more than six months after the first operation was more commonly caused by obstruction through plexus ingrowth or, again, debris.

In a 1993 review, percentages of 39 to 47 were reported; later studies found revision rates in the same range. These studies also report that ventricular dysfunction shortly after the operation is usually caused by debris. Ingrowth of choroid plexus was a frequent cause of proximal dysfunction later on.

2.3.2.2 Valve dysfunction
In the 200 revisions encountered in this study, valve obstruction was the cause of malfunctioning in ten cases (5%). In only 7 cases (3.5%), problems were caused by over or underdrainage in the presence of an otherwise patent valve. One of the patients died of a subdural hematoma probably caused by overdrainage.

In the literature, over or underdrainage is the cause of shunt dysfunction in 7 to 12% of cases. According to Sainte-Rose, overdrainage may be directly or indirectly related to more than 40% of the shunt failures. He not only mentioned subdural hematomas, craniosenosis, slit ventricles and orthostatic hypotension as complications due to overdrainage, but also shunt obstruction. Regarding valve obstruction, Sainte-Rose et al.
reported on two series of patients, one treated with shunts incorporating the Orbis Sigma valve, the other with shunts incorporating standard differential pressure valves. They found a valve obstruction rate of 18.3% for the Orbis Sigma valve, compared to 9.7% for the standard differential pressure valve (see also 2.3.6.1). In spite of this high obstruction rate for the Orbis Sigma valve, the overall mechanical complication rate was actually lower for the Orbis Sigma valve compared to differential pressure valves. A valve obstruction rate of 1% over a 2-year period was found by Kast et al.\(^71\).

In our study, and also in previous studies, the actual rate of valve problems seems rather low. The catheter is probably more important than the valve in determining the rate of shunt dysfunction. However, Sainte-Rose’s statement that the indirect complications (shunt obstruction) of, for example, overdrainage are considerable, cannot be rejected by our study in which we only included direct (obvious) complications.

In paragraph 2.3.6.1 the different valve types will be discussed further.

2.3.2.3 Distal dysfunction
Our study shows a distal dysfunction rate of 16%. Most of these cases (9.5%) consisted of a malposition of the abdominal catheter, usually caused by initial placement of the catheter in the preperitoneal fat layer instead of the abdominal cavity or by migration of the distal catheter. We did not see migrations to unexpected locations like the colon, the thoracic cavity, etcetera.

Previous studies showed that 14 to 33% of shunt malfunction was caused by distal dysfunction\(^41,71,88,98,110\). In the literature, migration of the distal catheter to unexpected locations such as the colon\(^118\), the thoracic cavity\(^30\), the scrotum\(^73\), the anus\(^92\), the bladder\(^89\), the pulmonary artery\(^93\) and the stomach\(^66\) have been described.

2.3.2.4 Infection
2.3.2.4.1 Infection and epidemiology  In 22 cases, infection was the reason for a revision operation. The overall infection rate was 5.5% and in 3% of the cases an infection occurred after primary shunt insertion. Half of the infections in our study were caused by Staphylococcus epidermidis, followed by S. aureus in 22% of the infection cases. An important finding was that the infection rate in patients who underwent EVD before shunting was significantly higher than the overall rate. But even after EVD the infection rate remained relatively low (6.8%). This slight increase in infection rate is indeed relevant in a study like this, with a large population of 400 patients. Most cases of infection occurred shortly after operation: 19 of our patients showed signs and symptoms of infection within one and a half month after operation. Two patients in our study had a shunt infection caused by an organism which could later be cultured from CSF obtained during the previous shunt operation. Fifteen out of 22 cases (68%) were found in the youngest age group.

In the literature, infection rates of 3 to 29% are reported with a mean of 5 to 8\(^%\)\(^27,41,63,71,88,98,128\). Most infections are caused by S. epidermidis, followed by S. aureus\(^27,42,63,128\). The reason for S. epidermidis to become such a frequent cause of shunt
infection, is its property to form a mucoid glycocalyx which protects itself against antimicrobial drugs and host immune responses\textsuperscript{27}. The glycocalyx also induces increased adhesion of the bacteria to shunt material\textsuperscript{42}. Furthermore, host leucocyte immune function is impaired in the presence of shunt material\textsuperscript{27}. Although higher infection rates after EVD have been described before\textsuperscript{94}, other studies\textsuperscript{42} showed no difference in infection rates for patients with previous EVD compared to patients without. Most studies show that infections usually occur within 2 months after shunt insertion\textsuperscript{42,44,71}. Because of this short interval it is assumed that in these cases contamination took place during shunt operation\textsuperscript{44,58}. The literature on age and shunt insertion and infection shows that, in general, infection rates are higher in younger children\textsuperscript{27,29,41,42,98,128}. Sometimes the underlying etiology of the hydrocephalus is mentioned as being important for the susceptibility to infection: in case of a perinatal hemorrhage, the chance of an infection is high\textsuperscript{98}. A possible explanation for this high rate might be that perinatal hemorrhage most frequently occurs in preterm infants. In these preterm infants the immune system is still immature\textsuperscript{27}. Thus, age is probably more important than etiology. The high susceptibility to infection in younger children has been explained in several ways. As mentioned above, the immune system in babies, especially preterm babies, is still immature. Probably even more important is the bacterial skinflora. In babies there is a quantitative as well as a qualitative difference in bacterial skin flora compared to adults. Pople et al.\textsuperscript{99} showed that the higher bacterial density in the skin of newborn was a risk factor for infection. Furthermore, strains of coagulase negative staphylococci with a high degree of bacterial adherence are more often found in newborn than in older children\textsuperscript{99}. Another explanation proposed by Duhaime et al.\textsuperscript{44} was the higher temperature and humidity in the operating room during pediatric neurosurgery.

The data we found on infection are in concordance with reports from the literature. But it is clear that the Dutch infection rates are rather low compared to the rates found in other studies. A possible explanation could be that Dutch standards of asepsis during surgery are high. Also, in the last decade materials and skills have improved, which also might contribute to low infection rates

2.3.2.4.2 Treatment of infection All cases of infection except one in our Dutch study were treated according to the ‘gold standard’ as described by Gardner et al.\textsuperscript{58}. This consists of: removal of the shunt, EVD if necessary, administration of antibiotics and replacement by a new shunt system only after recovery. This treatment was sufficient in all cases. The one patient who was not treated according to this standard developed another infection caused by the same organism later on (see 2.2.2.4.3).

In a retrospective study on 38 patients with a shunt infection Morisette et al.\textsuperscript{94} found a cure rate of 94\% after treatment with intravenous antibiotics and shunt replacement, either with or without EVD. For other modalities of treatment like medical support, intravenous antibiotics alone and intravenous antibiotics with shunt revision (no replacement!) the cure rates were respectively 0\%, 25\% and 0\%. All patients who only received a supportive treatment died. Those who received antibiotics together with a shunt revision
always developed a recurrent infection. Morisette et al. also concluded that careful selection of antibiotics in combination with shunt removal (with eventual replacement), either with or without EVD is the proper way to treat shunt infections. Walters et al.\textsuperscript{129} supported this view: they reported that the best treatment for shunt infection consisted of antibiotics, shunt removal and delayed shunt replacement. Replacement during the same operation in which the infected system was removed gave results that were worse. They noted that EVD treatment of infections resulted in a higher mortality and morbidity but they could explain this by the severity of the underlying problem. In the literature it has been described\textsuperscript{1}, that in the instance of a shunt infection caused by an organism which is found in meningitis in the 'normal' population there is no absolute indication for shunt removal: this conclusion was based on successful treatments of H. Influenza, meningococcus and gonococcus infections with antibiotics alone. However, if the CSF had not become sterile within 72 hours, the shunt system was still removed. An alternative way of treating shunt infections, a third ventriculostomy\textsuperscript{68}, has been suggested by Jones et al. They reported four successfully treated cases of recurrent infections by third ventriculostomy and intravenous and/or intraventricular antibiotics.

Comparing the literature and our own experience in the treatment of shunt infection, the ‘gold’ standard appears to be the most suitable method to treat shunt infection. The cure rate is high and complications of infection are low. Other ways of treatment\textsuperscript{27,68,94} are possible in some cases, but they are not necessarily better. The increasing popularity of third ventriculostomy might change this situation.

2.3.2.4.3 Prevention of infection Our study showed that peri-operative antibiotics had no significant effect on infection rates. The relative number of infections in patients with or without infection was the same. Other preventive measures like planning the operation early in the morning and other suggestions done by Choux\textsuperscript{29} were not part of the procedure in a systematical way in the participating centres and their impact could not be studied hence.

From a metanalysis\textsuperscript{63} of studies of the past few years it can be concluded that antibiotic prophylaxis is only useful when the baseline infection rate is higher than 5%. Choux\textsuperscript{29} by following a strict operative protocol could reduce the infection rate from 7.8 to 0.3%. Some of the precautions constituted planning the operations early in the morning, reducing the duration of the operating to a minimum and unpacking the shunt material immediately before implantation. This study also agreed that the surgeon’s concern with the infection problem already influences the incidence of infection. Pople et al.\textsuperscript{99} mentioned such precautions as washing the patient with two separate chlorhexidine shampoos 2 to 24 hours pre-operatively, isolation of wound edges with antiseptic drapes and the changing of gloves just before the actual implantation of shunt material. They also stated the rate of infection will already decline by greater alertness of the surgeon. Another study\textsuperscript{44} concluded that airborne bacteria from personnel present in the operating room are an important cause of shunt infections, even more important than endogenous bacteria. This study has several suggestions to reduce this surgeon-related contamination, varying from
washing or covering the surgeon’s face and increasing the distance between surgeon and
wound, to installing a transparent mechanical barrier between surgeon and patient.
Another precaution mentioned in the literature is impregnating the shunt catheter with
antibiotics. A recent report\textsuperscript{18} showed that catheters impregnated with antibiotics (rifampin/clindamycin) protected against infection in the immediate postoperative period. In a
study\textsuperscript{112} performed by Schierholz et al. in 1997, a shunt impregnated with broad-spectrum antibiotics was tested. This study also showed that shunt impregnation with antibiotics could reduce the incidence and amount of bacterial colonization on the surface of shunt catheters.

With the Dutch low infection rates, radical methods of prevention may not be necessary. If we do want to reduce the infection rate even more, measures for prevention should be aimed at the youngest patients who run the highest risk of infection. The impregnation method\textsuperscript{18,112} as well as a strict peri-operative protocol might be useful in reducing infection rates in high risk patients.

2.3.3 Revisions in the various age groups
Our results clearly show a relation between age and the need for a first revision operation. In the group under one year of age the first revision rate was 44\%, versus 20\% in older age groups. Looking at the occurrence of obstruction, which is the most prevalent cause of dysfunction in the three age groups, a relatively high rate of obstruction is found in the youngest group.

Previous studies have also reported age as an important factor determining shunt dysfunction\textsuperscript{27,29,31,41,42,71,98,99}. Another hypothesis was put forward by Liptak\textsuperscript{78}. An erect position changes the ventricular pressure and the cerebrospinal fluid flow through the ventriculoperitoneal shunt system. The increase in movement and time spent in the an erect position by children over one year of age may prevent obstruction of catheters and valves by means of the increase in CSF drainage.

Although Liptak’s hypothesis is interesting, it is probably not the only explanation for the relatively high rate of dysfunction in the youngest group. The etiology of the hydrocephalus may be a more important factor\textsuperscript{41,98}, and shall be dealt with separately (see 2.3.4).

2.3.4 Revisions related to underlying etiology of hydrocephalus
We found a significant influence of the etiology of hydrocephalus on shunt revision: there was a predisposition to shunt dysfunction in meningomyelocele, aqueductsyndrome, infection and perinatal hemorrhage.

These findings are concomitant with the data from the literature showing the etiology of hydrocephalus to be of great influence on the later occurrence of shunt dysfunction\textsuperscript{41,98}. In a 14-year retrospective study\textsuperscript{98} of shunt operations a clear, almost significant, trend was seen regarding etiology and the need for a revision operation. Posthemorrhagic hydrocephalus and hydrocephalus caused by meningomyelocele in particular have been reported as being relevant to a high rate of shunt obstructions\textsuperscript{41,98}. In posthemorrhagic hydrocephalus there might be sedimentation of blood cells on the shunt valve. This sedi-
mentation can impair valve closure or cause obstruction\textsuperscript{26}. Another possibility is that platelets mediate blood cell adhesion to the shunt material\textsuperscript{26}. In meningomyelocele as well as aqueductal syndrome and infection, a weak physical condition might lead to a higher frequency of shunt dysfunction overall\textsuperscript{76,91}.

In spite of the many different explanations that have been given, the exact reason why etiology is so important in shunt dysfunction remains unknown.

\section*{2.3.5 Operative procedure and incidence of shunt revision}

\subsection*{2.3.5.1 Surgeon}
In our Dutch study there was no relation between the experience of the surgeon and the incidence of revisions. Also, there was no difference in the duration of the surgical procedure between ‘experienced’ and ‘unexperienced’ surgeons.

In the literature, there are divergent views regarding the experience of the neurosurgeon\textsuperscript{41,79,98}. According to Piatt et al.\textsuperscript{98}, the experience of the surgeon was of no significant importance for the rate of revisions. Di Rocco et al.\textsuperscript{41} could not detect any difference in the percentage of shunt dysfunction after operations performed by residents compared to those performed by staff surgeons either. But a Norwegian study\textsuperscript{79} showed that inexperienced surgeons performed significantly more inadequate operations, leading to a higher revision rate.

\subsection*{2.3.5.2 Duration of the surgical procedure}
As reported by others\textsuperscript{27,41,79,98} this study showed that the time spent in the operating room did not affect the revision rate.

\subsection*{2.3.5.3 Drainage route}
There was no difference in revision rates between frontal and parietal shunts. Neither could we detect a difference in revision rates between VP and VA shunts.

But in a study in which they analyzed the functioning duration of frontal and parietal shunts in a population of 114 children, Albright et al.\textsuperscript{2} found that frontal shunts functioned for a significantly longer period than parietal shunts, even if in both groups ventricular catheters were implanted in a correct position. But according to Bierbrauer et al.\textsuperscript{19}, parieto-occipital shunts survived longer than frontal shunts. In accordance with our study, Piatt et al.\textsuperscript{98} found no difference between frontal and parietal shunts.

In the literature it is widely known that the number of dysfunctions of ventriculoperitoneal and ventriculooatrial shunts is roughly the same\textsuperscript{27,88}.

\section*{2.3.6 Shunt system versus revision}

\subsection*{2.3.6.1 Valves}
Our study indicates a high rate of dysfunction for a distal slit valve (Raimondi valve; 50\%). Significantly less revisions were seen in shunts with the Orbis Sigma valve (17\%). In this study we found a high revision rate of 44\% for the low pressure valve; the difference in revision rates for low and medium pressure valves (23\%) was not significant. It
was significant for the Orbis Sigma valve (17%) compared to the differential pressure valves, as discussed before in 2.2.5.1.

Recent studies reported an increased risk for distal catheter obstruction with the use of distal slit valves in the peritoneal catheter\textsuperscript{31,109,110}. These slit valves apparently constituted a canal into which the omentum could grow and cause obstruction at the distal end of the catheter. Some studies\textsuperscript{17,110} reported a lower incidence of revisions for the Orbis Sigma valve compared to differential pressure valves\textsuperscript{11,110}. Sainte-Rose et al.\textsuperscript{110} showed that shunts with Orbis Sigma valves survived longer than shunts with differential pressure valves. A higher number and larger proportion of proximal obstructions occurred with the standard differential pressure valves. Also, the proportion of slit ventricles was significantly lower using the Orbis Sigma valve. They did find, however, a higher rate of valve obstruction with the Orbis Sigma valve than with the differential pressure valves (18.3 versus 9.7%). The high obstruction rate could be explained by the valve architecture: the Orbis Sigma valve is a high resistance system with a narrow orifice that might be easily occluded. Aschoff et al.\textsuperscript{9} reported that, although there are some encouraging results with the Orbis Sigma valve in pediatric patients, the results in adult patients are disappointing. They stated that the considerable decrease in overdrainage problems with the Orbis Sigma valve may be outweighed by the increased rate of shunt insufficiency with this valve. In another report\textsuperscript{12} Aschoff et al. also mentioned the high chance of valve blockade with the Orbis Sigma valve.

Not much has been reported about the designation of the valve (low, medium or high pressure or flow-limiting) with respect to the revision rate. But it is widely known that low pressure valves are generally used in the youngest age group, which is characterized by a different etiology of hydrocephalus and other age-related problems than the other groups. The etiologies themselves tend to cause more revisions in the youngest group. Also, it has been reported before\textsuperscript{98} that valve dysfunction is more prevalent in children under two years of age. In a recent study, Boon et al.\textsuperscript{20} reported a better outcome for NPH patients who were treated with low-pressure valves compared to NPH patients treated with medium-pressure valves. Most outcome parameters (gait, disability, reduction in ventricular size) showed trends in favor of the low-pressure valve, but only the dementia scale was significant. This outcome scale does not involve the occurrence of shunt complications; it is meant to be an indicator of the quality of life. We did not include any measurements of the quality of life in our study.

The advice given before\textsuperscript{31,109,110} to use open-ended distal catheters without valves seems to be justified by our results. However, the high rate of dysfunction with the Raimondi valve (which is a distal slit valve) cannot be explained by a more frequent occurrence of distal shunt obstruction with this valve. In fact, in our material the rate of distal dysfunction is actually lower with the Raimondi valve than with the other valves. The difference is mainly caused by a relatively high frequency of obstruction by debris. We observed that Orbis Sigma valves not only function better in pediatric patients than differential pressure valves, but also that these valves function just as well as the differential pressure valves in adult patients. Especially in NPH patients both valve types func-
tioned equally well. This is in contrast with previous reports\textsuperscript{11}, which stated that the Orbis Sigma valve would only give satisfactory results in children. A possible explanation for the good results in NPH patients might be that in these patients the CSF flow through the shunt is lower than the flow in the other patients with other etiologies\textsuperscript{69}. In 1987, Kadowaki et al.\textsuperscript{69} showed that the maximum CSF flow in one NPH patient was 0.12 ml/min, whereas the maximum flow in patients with other etiologies was often higher. The limited drainage capacity of the Orbis Sigma valve is probably just sufficient for these patients. The fact that our results show the same success rate for both valve types in older patients as well is more difficult to explain. Apparently, even in these patients, the low flow through the Orbis Sigma valve is enough to get a satisfactory clinical result. In contrast to previous findings\textsuperscript{11,110} we did not see a high rate of valve obstruction with the Orbis Sigma valve.

2.3.6.2 Shunt components

Our results show that the number of shunt connections was of no significant importance for the occurrence of a shunt revision. A short ventricular catheter did constitute a risk factor for a later shunt revision. The risk was mainly determined by the higher chances of ventricular catheter obstruction with a shorter ventricular catheter. The length of the abdominal catheter was not important for the incidence of revisions. This was also applied to the group under one year of age.

From the literature, opposite views are known concerning the influence of the number of shunt components on the frequency of shunt revisions\textsuperscript{41,79,98,109}. Piatt et al. reported that complex shunts (multi-component) had survived significantly shorter than simple shunts\textsuperscript{98} (single-component) and Di Rocco et al.\textsuperscript{41} stated that shunts in which surgeons had added an extra component ran a higher risk of failure. Sainte-Rose\textsuperscript{109} also warned that the number of shunt connections was important for the chance of dysfunction. On the other hand, a Norwegian study\textsuperscript{79} was published which showed no difference in the number of shunt connections in patients who needed a revision compared to patients who did not. According to several studies, the length of the abdominal catheter has no influence on the rate of distal shunt dysfunction\textsuperscript{109,110,116}. One study found a shorter abdominal catheter to be related to a higher rate of revisions\textsuperscript{110}. This was explained by the need for lengthening the catheter.

The fact that we did not find any influence of the number of shunt connections on revision rate might show that with the currently used materials the number of shunt connections is not as important as it used to be. Another explanation is that surgeons try to limit the number of connections because of previous reports of higher revision rates with multi-component shunts. The mean number of connections in this Dutch study was only 1.9, which is rather low.

A possible explanation for the higher revision rate with shorter ventricular catheters may be the shrinkage of the ventricles after implantation of a functioning shunt. When the excess of cerebrospinal fluid has been drained, the ventricles shrink and as a result the ventricular catheter may retract into the brain parenchyma. The shorter the ventricular
catheter, the greater the chance of ‘pseudo-retraction’.

The view held by some surgeons that in younger children the abdominal catheter shouldn’t be too long is rejected by our study. We couldn’t detect any influence of the length of the abdominal catheter on revision rates. The conclusion may be that an abdominal catheter length of up to 60 cm does not increase the risk of complications in younger children.

2.3.7 Cerebrospinal fluid composition versus revision operations
Because it has been assumed for years that CSF concentrations of protein and cells were important factors in shunt malfunctioning, we paid special attention to the relation between these concentrations and revision rates.

Our study showed no correlation between CSF protein concentration and the occurrence of shunt dysfunction. It should be remarked that the maximal protein concentration was 3.5 g/l. We also didn’t find any significant difference in erythrocyte concentrations between patients who needed a revision operation and patients who did not. Shunts were inserted in a single patient with a cell count above 20,000 cells/μl without the need for a revision later on. Since the beginning of shunt surgery it has been assumed that a high CSF protein may induce shunt dysfunction because a high concentration of protein would cause a high viscosity, which is proportional to the resistance a liquid will encounter during flow. Current literature however, expresses a different view. High protein concentrations hardly affect viscosity. In fact, a high protein concentration is shown to lower the surface tension of a fluid which would facilitate valve opening. It has been described in vivo that a CSF erythrocyte count of more than 1000 per μl would increase the chance of shunt dysfunction (and revision). Sainte Rose et al already mentioned that a shunt operation in which a ventricular catheter was placed in a ventricle full of blood clots was doomed to failure.

The lack of influence of protein concentration on shunt dysfunction we found corresponds with results from other studies. In erythrocyte counts, however, the reports from the literature and our own results differ. We do not have a satisfactory explanation for this contrast between literature and our own clinical results. Because our study only included a few patients with extremely high erythrocyte counts (90% of the patients had counts below 5000 cells/μl), there is a possibility that we have observed too few patients with (extremely) high counts to show a correlation between these high counts and an increased shunt failure rate.

2.3.8 Morbidity
As mentioned before we defined morbidity as “added morbidity” after the operation. This definition makes the morbidity found in our Dutch study very low. After primary shunt operations the morbidity rate was 2.3%. After revision the rate was comparable: 1.5%. In only four cases the morbidity resulted in a permanent disability. Morbidity in these cases was caused by a subdural hematoma, postoperative seizures and postoperative hemorrhage. In one case the morbidity was caused by a pre-existing tumor.
In the literature a higher percentage is reported\textsuperscript{27}, but this is probably due to the definition used. Previous studies included morbidity caused by the pre-existing etiology of the hydrocephalus. In our study we would then observe a considerable morbidity rate of patients with a tumor especially. Generally, it may be concluded that a CSF shunt procedure harbours little risks in terms of morbidity.

### 2.3.9 Mortality

After primary shunt operation 68 out of 400 patients died, which implies a mortality rate of 17\%. Shunt-related mortality alone shows a different picture. Only two patients died of a shunt complication (0.5\%) after primary insertion. One of them had an infection, the other died after a subdural hematoma, probably caused by overdrainage. The majority of deaths were caused by progressive tumor growth. Another nine patients died after revision. Only one of those deaths was caused by a shunt complication. The proximal catheter appeared to be obstructed by debris.

Older studies\textsuperscript{27} report very high shunt-related mortality rates. Some studies found a rate of 60\%. Currently, a mortality rate of about 12\% is usually found\textsuperscript{27}. Another current study\textsuperscript{109} showed a shunt-related mortality rate of 1\%. It has been reported\textsuperscript{27} though, that in general more people die of the disease causing hydrocephalus, than of the shunt treatment.

The mortality rate we found, shunt-related and non-shunt-related together (17\%), looked substantially higher than the rates reported in the literature\textsuperscript{27}. The difference can be explained reasonably by the population studied: in most former studies only patients with a non-neoplastic cause of the hydrocephalus were included. When we exclude neoplastic cause in our study, we find a mortality rate of 11\% (shunt-related plus non-shunt related) which is similar to previous studies. As written above, older studies\textsuperscript{27} mentioned very high shunt-related mortality rates compared to our 0.5\%. This difference is hard to explain. Probably several factors act together. In other studies, infections were an important cause of death. We found a low infection rate. Also, in our study only 45 patients received a ventriculo atrial shunt. A probable cause of death in this type of shunt is pulmonary embolism. We did not observe this complication, whereas the older studies\textsuperscript{27} reported it as a major cause of death.

Overall, it can be concluded that in Dutch hospitals very few patients die of shunt-related causes.
2.4 Conclusions

2.4.1 Summary of previous conclusions
From this study it is clear that the revision rate after shunt operation in Dutch neurosurgical centers is low compared to other studies. Proximal dysfunction is the most frequent reason for shunt revision. Valves cause shunt problems in a minority of cases: only 35 cases (17.5%) overall. Similar for distal shunt problems. If distal problems occur at all, they are usually caused by an initial malplacement of the abdominal catheter in the peritoneal fat layer, or migration later on.

The infection rate after primary shunt insertion in the Netherlands is rather low (overall 5.5% and 3% after primary insertions). External ventricular drainage increases the chance of infection. Young age is a risk factor for infection. If infection occurs, it will mostly be in the first postoperative months. In Dutch hospitals, CSF shunt infection is treated according to the ‘gold standard’ by Gardner et al. This method of treatment is very efficient. Complications of infection are rare and the cure rate is high.

Overall, age is the most important determinant of shunt dysfunction. This can be explained by the occurrence of risk-increasing etiologies in the youngest age group, the weaker general condition of, especially, preterm infants and also the immaturity of the immune system of these infants.

The experience of the surgeon does not influence the time spent in the operating room or the need for a later shunt revision. There is no preferential drainage route for CSF shunts: VA and VP shunts show equal rates of complications, and there is no difference in complications between frontal, parietal and occipital drainage routes.

In the youngest age groups, the Orbis Sigma valve causes less complications than differential pressure valves. In the adult group, there is no difference in the number of complications between the Orbis Sigma valve and the differential pressure valves. In shunt systems incorporating the Raimondi valve, the revision rate is high. This cannot be explained by an increased rate of distal dysfunction.

A short ventricular catheter predisposes to shunt obstruction. A possible mechanism for this might be ‘pseudoretraction’. After the excess of cerebrospinal fluid has been drained to the abdominal cavity, the ventricles shrink and the ventricular catheter may retract into the brain parenchyma. A long abdominal catheter poses no problems in any of the age groups. There is no need to refrain from using long abdominal catheters in infants under one year of age. In fact, putting in a longer abdominal catheter might prevent a later revision to lengthen the shunt.

Neither moderately elevated CSF protein nor a high erythrocyte count caused additional problems in CSF shunting in our study. Shunts functioned successfully in patients with a CSF protein concentration as high as 3.5 g/l or, exceptionally, CSF with erythrocyte counts above 20,000 cells/µl.

Morbidity, defined as ‘added’ morbidity after shunting, and shunt-related mortality are very low in this study. A reasonable conclusion here can be that CSF shunt operations in the Netherlands harbour little adverse consequences for the patient.
2.5 Future directions

Although Dutch CSF shunt surgery has low morbidity and mortality rates, there are still many complications. Recommendations for the future resulting from this study could be the following:

1. As the Orbis Sigma valve has been shown to cause significantly less shunt dysfunction in young patients than differential pressure valves, we would recommend to incorporate this valve in all CSF shunt systems in children under 15 years of age.

2. Our results justify a prospective randomized trial to compare the Orbis Sigma valve with differential pressure valves in a selected group of adult patients. These patients should be patients with an expected low shunt flow rate, for example NPH patients.

3. To further decrease the infection rate, preventive measures should be aimed specially at the youngest age groups, in which the infection rate is highest. In these high-risk patients, shunt impregnation with antibiotics as well as a strict peri-operative protocol might be useful.

4. In 1998 Rekate\textsuperscript{106} stated: ‘The best shunt is no shunt’. Further use and improvement of endoscopic techniques such as the use of laser\textsuperscript{125} may decrease the need for shunts in the future.

5. Our study did not include the quality of life in the evaluation of shunt functioning. As Aschoff et al.\textsuperscript{15} mentioned, the patient’s quality of life is more important than the number of shunt revisions. Patients requiring several shunt revisions, may actually have an excellent quality of life. We therefore recommend future studies in which the quality of life should be evaluated.