The clinical spectrum of shunt nephritis

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Abstract

Background. Shunt nephritis is an immune-complex-mediated glomerulonephritis (GN) associated with chronically infected ventriculoatrial shunts inserted for treatment of hydrocephalus.

Methods. Six patients aged 5–22 years with shunt nephritis are reported who have been observed between 1971 and 1994. The clinical course and long-term outcome are analysed in relation to the time of diagnosis and renal histopathology.

Results. The time of diagnosis of shunt nephritis ranged from 0.3 to 4.5 years after the last shunt operation. Diagnosis was delayed up to 1.5 years after the first clinical manifestations. All patients had signs of infection, i.e. recurrent fever, hepatosplenomegaly, anaemia, and cerebral symptoms. Renal manifestations consisted of haematuria (macroscopic in 3 patients), proteinuria (heavy in 5), renal insufficiency (4) and hypertension (2). Decreased C3 levels, cryoglobulins, and antinuclear factors were frequent. Cultures of blood and cerebrospinal fluid provided growth mainly of S. epidermidis. Renal biopsy revealed endocapillary GN (1), membranoproliferative GN (1) and endocapillary/extracapillary GN with crescents (2). All patients received antibiotics i.v. Complete recovery was observed in three of four patients in whom the shunt was totally removed, supported by transient external drainage of cerebrospinal fluid, and followed by placement of a ventriculoperitoneal shunt. One child with delayed diagnosis, presenting with a serum creatinine of 3.2 mg/dl, hypertension, and severe scarring on renal biopsy, rapidly progressed to irreversible ESRD within 5 months. Two patients without removal of the shunt died subsequently from sepsis.

Conclusions. The renal outcome of shunt nephritis is good if early diagnosis and treatment is provided including i.v. antibiotics and total removal of the infected shunt. The possible progression to ESRD requires frequent nephrological monitoring of patients with ventriculoatrial shunts.

Key words: end-stage renal disease; dialysis; infection; shunt nephritis; ventriculoatrial shunts.

Introduction

Shunt nephritis commonly occurs in patients with infected ventriculoatrial (VA) shunts inserted for palliation of hydrocephalus of congenital or acquired origin. It was first described by Black et al. in 1965 [1]. To our knowledge about 148 patients with this condition have been reported [2–34]. Although a review of the literature suggests that the frequency of the condition has been declining over the last decade, some recent reports demonstrate its continuing clinical importance. The series of patients presented here illustrates the wide clinical spectrum of this disorder. This report emphasizes the possible progression of shunt nephritis to end-stage renal disease (ESRD) and evaluates strategies to prevent this development.

Subjects and methods

Between September 1971 and February 1994, six patients (5 children, 1 adult) were admitted to the Departments of Paediatrics and Internal Medicine of the University of Heidelberg with a clinical picture consistent with shunt-associated glomerulonephritis (GN). Table 1 summarizes the past history, laboratory findings, bacteriological studies, treatment and outcome of these patients. All patients had implants of VA shunts due to congenital or acquired hydrocephalus. The clinical course of case no. 5 is described in detail because of its unusually rapid progression to irreversible ESRD.

Results

In our six patients the time of diagnosis of shunt nephritis was 0.3–4.5 (median 2.1) years after the latest shunt operation. The time from onset of clinical symptoms suggestive of shunt nephritis until diagnosis ranged from 2 to 16 (median 5) months (Table 1). All patients presented with recurrent fever, hepatosplenomegaly, anaemia, and cerebral manifestations,
i.e. vomiting, nausea, behavioural changes, or seizures. In addition, patient no. 6 showed skin purpura. GN was suggested by haematuria (3 macroscopic, 3 microscopic) and proteinuria which was heavy in five patients. No patient was nephrotic. Hypertension was noted in two patients and serum creatinine levels (SCR) were elevated in four. Sedimentation rate was increased in all patients. Decreased C3, cryoglobulins, and antinuclear factors were found if tested. Cultures of blood or CSF were positive in five and three patients respectively. Renal biopsy revealed endocapillary GN and membranoproliferative GN each in one case and endocapillary/extracapillary GN with crescents in two. Immunofluorescence studies in two patients showed granular deposits of IgM, IgG, and complement components (see Table 1).

All patients were treated with intravenous antibiotics. In four patients (nos. 2, 4, 5, 6) where the VA shunt was totally removed, no infection recurred; three of these appeared to have recovered completely after 2, 8 and 23 years. In nos. 2 and 4 a new VP and in no. 5 a new VA shunt was reinserted after transient external CSF drainage for 1–7 weeks. In patient no. 6 no replacement was required. Patient no. 5 developed ESRD despite replacement of the infected shunt (see Case report). The two children with only partial (no. 1) or no (no. 3) removal of the VA shunt had continuous infection, and died; in no. 1 supplied with bilateral VA shunts only one valve of the shunt could be removed, the other being left in place; in no. 3 the parents refused surgical treatment because the child was severely handicapped, i.e. blind, mentally retarded, and suffering from convulsions.

It is noteworthy that three patients (nos. 1, 2, 5) were initially treated with oral antibiotics for a presumed diagnosis of urinary tract infection. In the only adult patient (no. 6) the skin lesions first suggested the erroneous diagnosis of Schönlein–Henoch purpura; treatment by methylprednisolone pulses failed to improve renal function.

Case report (patient no. 5)

This girl, born on January 1984, presented at birth with a lumbosacral myelomeningocele, hydrocephalus, and neurogenic bladder. During the neonatal period her myelomeningocele was repaired and a VA shunt

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**Table 1. Clinical findings in six patients with shunt nephritis at the time of diagnosis and their clinical outcomes**

<table>
<thead>
<tr>
<th>Patient no.</th>
<th>Time of diagnosis</th>
<th>Age at diagnosis (year), sex</th>
<th>Cause of hydrocephalus</th>
<th>Time since first and latest shunt operation (year)</th>
<th>Delay in diagnosis (mo.)</th>
<th>Blood pressure (mmHg)</th>
<th>Haemoglobin (g/dl)</th>
<th>ESR (mm 1st/2nd h.)</th>
<th>SCR (mg/dl)</th>
<th>Haematuria</th>
<th>Proteinuria (g/day)</th>
<th>C3* (mg/dl)</th>
<th>Antinuclear antibodies</th>
<th>Cryoglobulins</th>
<th>Blood culture</th>
<th>Renal biopsy:</th>
<th>Time from diagnosis to last observation (yr)</th>
<th>Outcome at last observation</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Sept. 1971</td>
<td>9.9 F</td>
<td>MMC</td>
<td>9.8/15</td>
<td>5</td>
<td>105/75</td>
<td>9.6</td>
<td>68/120</td>
<td>0.9</td>
<td>M</td>
<td>1.6</td>
<td>ND</td>
<td>+</td>
<td>ND</td>
<td>Alk. faecalis</td>
<td>LM</td>
<td>0.5</td>
<td>ongoing infection, died</td>
</tr>
<tr>
<td>2</td>
<td>Apr. 1972</td>
<td>5.5 F</td>
<td>MMC</td>
<td>5.4/4.5</td>
<td>7</td>
<td>100/60</td>
<td>8.7</td>
<td>51/64</td>
<td>1.1</td>
<td>M</td>
<td>1.9</td>
<td>ND</td>
<td>+</td>
<td>ND</td>
<td>Staph. epid.</td>
<td>IF</td>
<td>0.6</td>
<td>ongoing infection, died</td>
</tr>
<tr>
<td>3</td>
<td>Oct. 1977</td>
<td>11.3 F</td>
<td>Arachnoidal cyst</td>
<td>11.1/2.6</td>
<td>16</td>
<td>125/80</td>
<td>7.3</td>
<td>30/85</td>
<td>1.0</td>
<td>M</td>
<td>1.4</td>
<td>28</td>
<td>+</td>
<td>+</td>
<td>Staph. epid.</td>
<td></td>
<td>22.8</td>
<td>SCR: 0.7 mg/dl urine: nil</td>
</tr>
<tr>
<td>4</td>
<td>Mar. 1988</td>
<td>12.3 F</td>
<td>Astrocytoma</td>
<td>4.3/0.3</td>
<td>2</td>
<td>145/80</td>
<td>6.0</td>
<td>26/61</td>
<td>1.7</td>
<td>m</td>
<td>0.6</td>
<td>42</td>
<td>+</td>
<td>+</td>
<td>Prop. Acnes</td>
<td></td>
<td>0.6</td>
<td>SCR: 0.7 mg/dl urine: nil BP 130/80</td>
</tr>
<tr>
<td>5</td>
<td>Aug. 1993</td>
<td>9.5 F</td>
<td>MMC</td>
<td>9.5/4.0</td>
<td>5</td>
<td>165/95</td>
<td>m</td>
<td>82/116</td>
<td>3.2</td>
<td>m</td>
<td></td>
<td>48</td>
<td>+</td>
<td>+</td>
<td>Sterile</td>
<td></td>
<td>7.7</td>
<td>SCR: 0.7 mg/dl urine: nil BP 125/70</td>
</tr>
<tr>
<td>6</td>
<td>Jan. 1994</td>
<td>22.1 M</td>
<td>Post traumatic</td>
<td>0.8/0.5</td>
<td>2</td>
<td>110/70</td>
<td>m</td>
<td>73/106</td>
<td>2.5</td>
<td>m</td>
<td></td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>Staph. epid.</td>
<td></td>
<td>2.5</td>
<td>SCR: 1.1 mg/dl urine: nil BP 135/80</td>
</tr>
</tbody>
</table>

**Abbreviations:** MMC, myelomeningocele; ESR, erythrocyte sedimentation rate; M, macroscopic; m, microscopic; GN, glomerulonephritis; MPGN, membranoproliferative GN; EEEN, endo-/extracapillary GN; PSR, partial shunt removal; CSR, complete shunt removal; ECSFD, external cerebrospinal fluid drainage; VPS, ventriculoperitoneal shunt; VAS, ventriculoaortal shunt; BP, blood pressure; SCR, serum creatinine; ND, not done; +, present; * normal range 55–120.
was placed. Subsequently recurrent urinary tract infections combined with fever occurred. Since a voiding cystourethrogram performed at the age of 3 months showed high-grade vesicoureteral reflux on the right side, reimplantation of the corresponding ureter was performed and daily intermittent catheterization of the bladder was started. During the further course several shunt revisions were performed, the latest in July 1989 being due to traumatic shunt disconnection.

From April 1993 the girl had recurrent episodes of fever, often associated with assumed urinary tract infections. The fever responded well to antibiotics. In August 1993 she was admitted to the children’s hospital of Karlsruhe because of fever up to 40 °C, nausea, vomiting, muscle weakness, shortness of breath, and behavioural changes. Physical examination showed hepatosplenomegaly, oedema, and hypertension (165/95 mmHg). Urinanalysis revealed erythrocytes (1600/µl), leukocytes (40/µl), and proteinuria (2.9 g/day); urine culture remained sterile. SCR was 3.2 mg/dl, potassium 7.4 mmol/l, phosphate 3.4 mmol/l, total protein 7.1 g/l, C-reactive protein 4.5 mg/dl (upper normal limit 1 mg/dl), haemoglobin 5.0 g/dl, blood bicarbonate 16 mmol/l. Two blood cultures remained sterile. Serum complement levels C3 and C4 were decreased to 48 (normal range 55–120) mg/dl and 4 (normal range 20–50) mg/dl respectively; antinuclear antibody titres were elevated to 1/50 (normal limit 1/25) and cryoglobulins were detectable. Renal ultrasound revealed enlarged hyperechogenic kidneys. On the evening of admission the girl presented a grand mals convolution. A cranial CT scan showed no signs of intracranial haemorrhage or increased pressure.

Treatment consisted of intravenous antibiotics (vancomycin, rifampicin and flucloxacillin), sodium bicarbonate and blood transfusions. Blood pressure was controlled with nifedipine, captopril and frusemide. The VA shunt was not removed because the parents refused it. The further course was complicated by continuing fever, malaise, anorexia, hypertension up to 190/140 mmHg, and increased SCR up to 6.8 mg/dl.

In September 1993 the girl was transferred to the University Children’s Hospital of Freiburg for removal of the shunt. Culture of the catheter tip showed Staph. epidermidis. Antibiotic treatment was continued with vancomycin i.v. During the next 3 days fever decreased, the clinical condition stabilized, SCR decreased, and serum levels of complement and C-reactive protein became normal. After 2 weeks of external CSF drainage a new VA shunt was implanted; after a further 2 weeks this shunt was revised because of malfunction with papillary oedema. Hypertension was controlled by clonidine, nifedipine, captopril, frusemide, and prazosin. After discharge from the hospital in October 1993, SCR increased steadily from 2.7 mg/dl to 3.7 mg/dl in January 1994. No clinical or biochemical signs of recurrent shunt infection were noted.

In February 1994 the girl was transferred to University Children’s Hospital of Heidelberg. In the presence of a fair clinical condition SCR had risen to 6.0 mg/dl and inulin clearance was 8 ml/min/1.73 m². Renal biopsy revealed endocapillary and extracapillary proliferative GN, with fibrocellular or fibrous crescents in three of 10 glomeruli (Figure 1); the other seven glomeruli were totally or almost totally sclerosed, and widespread tubular and interstitial fibrosis was present. After creation of a arteriovenous fistula, intermittent haemodialysis treatment was started on 28 March 1994. At present the patient is well, blood pressure is easily controlled by enalapril, 1.25 mg/day. Urine output ranges between 200 and 400 ml/day. The patient is thought to be candidate for cadaver kidney transplantation.

Discussion

Our series of young patients with shunt nephritis reveals a wide spectrum of outcomes, although the initial clinical presentation was similar. All patients showed signs of infected VA shunts as described above in detail. The high frequency of cerebral manifestations in our patients is well documented in the literature [6,10,28,29]; it may be attributed to increased intracranial pressure, hypertension, or other mechanisms. Haematuria, proteinuria, hypertension, and high SCR levels were often misinterpreted. Urinary tract infection was a frequent erroneous diagnosis, supported by the presence of a neurogenic bladder. Skin lesions were taken as signs of Schönlein–Henoch purpura. Therefore the diagnosis was often delayed.

In the following we present a short review of the literature on shunt nephritis, which contains the data of 72 cases published in an earlier review by Arze et al. [6] and of another 76 patients reported subsequently [2–5,7–34] (Table 2). The median intervals from the last shunt operation and from first clinical manifestation to the diagnosis of shunt nephritis were similar in both observation periods and ranged widely. Compared to the earlier review by Arze et al. we noted a higher incidence of hypertension, anaemia, renal
failure, haematuria, proteinuria, and nephrotic syndrome in the series reported more recently. The predominant finding on renal biopsy was MPGN. Immunofluorescence revealed granular immunoglobulin and complement deposits in all biopsy specimens examined, with a predominance of IgM (84%), IgG (66%) and C3 complement (94%). Electron-microscopy was reported in only 32 cases, usually presenting an increase of mesangial matrix and cells.

The true glomerular basement membrane was not thickened, but electron-dense deposits were observed in the subendothelial and mesangial areas [6,15,24]. In many patients cryoglobulins and antinuclear antibodies were detected. The serological and immunofluorescence findings point to an activation of the classical complement pathway in this type of immune-complex nephritis. If the outcome of the patients reported by Arze et al. [6] is compared to that of patients published subsequently, a higher incidence of persistent renal failure is noted, whilst mortality has decreased.

The initial event in shunt infection is colonization of the surface of the atrial part of a VA shunt, most commonly by a coagulase-negative staphylococcal strain. All our patients had a positive blood and/or CSF culture. According to the literature cultures were positive in a growing proportion of patients (Table 2). S. epidermidis accounts for about 75% of all shunt infections. Other responsible micro-organisms were S. aureus, E. coli, Corynebacterium bovis, Corynebacterium xerosis, Pseudomonas aerugonos, Pseudomonas cepacia, diphterois, Listeria monocytogenes, Bacillus subtilis, Bacillus cereus, Serratia, Propionibacterium acnes, Peptococcus, Mycobacterium gordonae, Micrococcus, Moraxella nonliquefaciens, Moraxella bovis, Acinetobacter iowo/DC3, alpha-haemolytic streptococcus spp. and the fungus Fusarium [3,6,17,20,30,33,35]. Negative blood or CSF cultures do not exclude shunt nephritis, especially after previous antibiotic therapy. Decreased serum levels of complement, detection of cryoglobulins or autoantibodies are helpful to support the diagnosis of shunt nephritis.

If the infection occurs within several months after the initial shunt insertion it must be assumed that it is due to direct contamination of the VA shunt during operation. For infections occurring later other forms of contaminations have to be considered, e.g. by minor injuries to the skin or by surgery which may induce S. epidermidis assisted by the high affinity for hydrophobic materials used for VA catheters [36]. Most coagulase-negative staphylococci produce an exopolysaccharide protective biofilm (slime) that surrounds the strain. All our patients had a positive blood and/or CSF culture. According to the literature cultures were positive in a growing proportion of patients (Table 2). S. epidermidis accounts for about 75% of all shunt infections. Other responsible micro-organisms were S. aureus, E. coli, Corynebacterium bovis, Corynebacterium xerosis, Pseudomonas aerugonos, Pseudomonas cepacia, diphterois, Listeria monocytogenes, Bacillus subtilis, Bacillus cereus, Serratia, Propionibacterium acnes, Peptococcus, Mycobacterium gordonae, Micrococcus, Moraxella nonliquefaciens, Moraxella bovis, Acinetobacter iowo/DC3, alpha-haemolytic streptococcus spp. and the fungus Fusarium [3,6,17,20,30,33,35]. Negative blood or CSF cultures do not exclude shunt nephritis, especially after previous antibiotic therapy. Decreased serum levels of complement, detection of cryoglobulins or autoantibodies are helpful to support the diagnosis of shunt nephritis.

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Table 2. Clinical findings in 148 patients with shunt nephritis at the time of diagnosis and their clinical outcomes

<table>
<thead>
<tr>
<th>Summarized by Arze et al. 1983 (n=72)</th>
<th>References [2–5] and [7–34] (n=76)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Children/adults</strong></td>
<td></td>
</tr>
<tr>
<td>Time from last shunt operation to diagnosis of shunt nephritis</td>
<td>4.4 yr.*</td>
</tr>
<tr>
<td>Interval from first clinical manifestation to diagnosis of shunt nephritis</td>
<td>(4 wk–14 yr)</td>
</tr>
<tr>
<td>Hypertension</td>
<td>7/72 (10%)</td>
</tr>
<tr>
<td>Anaemia</td>
<td>61/72 (85%)</td>
</tr>
<tr>
<td>Renal failure</td>
<td>33/72 (46%)</td>
</tr>
<tr>
<td>Haematuria</td>
<td>63/72 (88%)</td>
</tr>
<tr>
<td>Proteinuria</td>
<td>46/72 (64%)</td>
</tr>
<tr>
<td>Nephrotic syndrome</td>
<td>20/72 (28%)</td>
</tr>
<tr>
<td>C3 levels low</td>
<td>51/60 (85%)</td>
</tr>
<tr>
<td><strong>Staph. epidermidis</strong></td>
<td></td>
</tr>
<tr>
<td>blood culture</td>
<td>39/57 (70%)</td>
</tr>
<tr>
<td>CSF culture</td>
<td>26/36 (43%)</td>
</tr>
<tr>
<td>Renal biopsy</td>
<td>(n=47)</td>
</tr>
<tr>
<td>MPGN</td>
<td>27 (57%)</td>
</tr>
<tr>
<td>DMP</td>
<td>15 (32%)</td>
</tr>
<tr>
<td>EEGN</td>
<td>9 (15%)</td>
</tr>
<tr>
<td>others</td>
<td>5 (11%)</td>
</tr>
<tr>
<td><strong>Outcome</strong></td>
<td></td>
</tr>
<tr>
<td>complete recovery</td>
<td>38 (53%)</td>
</tr>
<tr>
<td>persistent urine abnormalities</td>
<td>16 (22%)</td>
</tr>
<tr>
<td>renal failure (ESRD)</td>
<td>4 (6%)</td>
</tr>
<tr>
<td>death</td>
<td>14 (19%)</td>
</tr>
<tr>
<td><strong>Hypertension</strong></td>
<td>32/50 (64%)</td>
</tr>
<tr>
<td><strong>Anaemia</strong></td>
<td>47/47 (100%)</td>
</tr>
<tr>
<td><strong>Renal failure</strong></td>
<td>43/70 (61%)</td>
</tr>
<tr>
<td><strong>Haematuria</strong></td>
<td>62/62 (100%)</td>
</tr>
<tr>
<td><strong>Proteinuria</strong></td>
<td>71/71 (100%)</td>
</tr>
<tr>
<td><strong>Nephrotic syndrome</strong></td>
<td>20/47 (43%)</td>
</tr>
<tr>
<td><strong>C3 levels low</strong></td>
<td>44/47 (94%)</td>
</tr>
<tr>
<td><strong>Staph. epidermidis</strong></td>
<td>54/63 (86%)</td>
</tr>
<tr>
<td><strong>blood culture</strong></td>
<td>20/25 (80%)</td>
</tr>
<tr>
<td><strong>CSF culture</strong></td>
<td>(n=60)</td>
</tr>
<tr>
<td><strong>Renal biopsy</strong></td>
<td>37 (62%)</td>
</tr>
<tr>
<td><strong>MPGN</strong></td>
<td>8 (13%)</td>
</tr>
<tr>
<td><strong>DMP</strong></td>
<td>9 (15%)</td>
</tr>
<tr>
<td><strong>EEGN</strong></td>
<td>6 (10%)</td>
</tr>
<tr>
<td><strong>others</strong></td>
<td>(n=67)</td>
</tr>
<tr>
<td><strong>complete recovery</strong></td>
<td>36 (54%)</td>
</tr>
<tr>
<td><strong>persistent urine abnormalities</strong></td>
<td>12 (18%)</td>
</tr>
<tr>
<td><strong>renal failure (ESRD)</strong></td>
<td>13 (5) (19%)</td>
</tr>
<tr>
<td><strong>death</strong></td>
<td>6 (9%)</td>
</tr>
</tbody>
</table>

Data are given as median and range; * mean

**Abbreviations:** GN, glomerulonephritis; MPGN, membranoproliferative GN; EEGN, endo-/extracapillary GN; DMP, diffuse mesangial proliferation; CSF, cerebrospinal fluid.
immunoglobulin and circulating immune complexes which are attracted by the kidney. Bacterial antigens have in fact been demonstrated in the glomeruli of patients with shunt nephritis [37,38], but despite the high frequency of shunt infections after insertion of VA shunts of about 12% [39], only 0.7–2.3% of the patients actually develop shunt nephritis [14,40, 41]. This suggests that a favourable immunological environment must be present for the development of shunt nephritis.

Usually the prognosis of renal function is good in shunt nephritis provided that the VA shunt is removed within a few weeks after the first signs of infection. Resumption of renal function has been reported even in cases initially requiring dialysis treatment for up to 2 months [27]. In three of our cases, early shunt removal followed by temporary external CSF drainage with or without subsequent implantation of a VP shunt was followed by complete recovery for up to 23 years. On the other hand our case report demonstrates that even a short delay in the removal of the VA shunt may precipitate a rapidly deteriorating course, which in our patient was accelerated by uncontrollable hypertension. Irreversible ESRD has been reported in six other children and one adult with shunt nephritis [4,9,10,31,42]. In three of these patients renal biopsy revealed endocapillary/extracapillary GN, with more than 70% crescents [9,10] and in two extensive glomerular sclerosis and tubular atrophy [4,42]. In all reported patients with ESRD the diagnosis was delayed for 0.2 up to 5 (median 0.9) years. Five of the seven patients with known ESRD were started on dialysis treatment (all since 1983), and two subsequently underwent successful renal transplantation; the other two patients died on conservative treatment. Up to now no recurrence of shunt nephritis in the transplanted kidney was reported.

Treatment of shunt nephritis requires at first immediate removal of the infected shunt [22,43]. A temporary external drainage of CSF should be performed for about 1–2 weeks before a new shunt is reinserted. By intraventricular instillation of antibiotics the chance for reinfection is reduced. Antibiotics (e.g. vancomycin, rifampicin) should be given intravenously for at least 10 days, but vigorous antibiotic therapy without shunt removal is rarely effective in resolving the shunt infection [21,35]. Following removal of an infected shunt, a VP rather than a VA shunt should be inserted. In view of the severe complications associated with VA shunts, e.g. shunt nephritis, bacterial endocarditis, sepsis, recurrent pulmonary embolism and cor pulmonale, the general use of VP shunts is preferred in most neurosurgical units today [39,43]. However, if patients progress to ESRD and if peritoneal dialysis is considered, the insertion of a VA shunt appears to be preferable in view of the danger of birefractive infection but corresponding experience is missing.

New strategies to prevent shunt infections include antibiotic prophylaxis during insertion of CSF shunts or in periods of possible bacteraemia, packs soaked in antiseptic agents to isolate wound edges, regular glove-changing before handling the shunt, and alternative biomaterials for VA shunts [36,44]. The value of these strategies to prevent shunt nephritis needs to be demonstrated. We believe that the general use of VP shunts is the most important option to reduce the incidence of this disease. However, in four cases shunt nephritis has also been reported after insertion of a VP shunt [37,45–47]. Therefore any patient with a shunt who develops fever or other signs of infection should be suspected of having shunt nephritis, and diagnostic procedures should be undertaken immediately to allow a definite diagnosis and immediate treatment.

Acknowledgements. We thank Prof. M. Brandis and Prof. R. Korinthenberg, Departments. of Paediatrics and Paediatric Neurology, University of Freiburg for information on patient no. 5, and to PD Dr. C. Benninger and the staff of the Divisions of Paediatric Nephrology and Neurology, University Children’s Hospital of Heidelberg, for the management of the patients.

References


18. Tott H, Redl J, Beregi E. Shunt nephritis with crescent forma-

19. Martinez-Vea A, Darnell A. Fever and vasculitis purpura in a
41-year-old woman with a ventriculo-atrial shunt for normotens-
ive hydrocephalus (clinical conference). Med Clin (Barc) 1988;
90: 786–793 (in Spanish)

20. Narchi H, Taylor R, Azmy AF, Murphy AV, Beattie TJ. Shunt 

caused by ventriculo-atrial shunt: possible cure without reopera-


23. Ter Borg EJ, van Ruijswijk KH, Kallenberg CG. Transient 
arthritis with positive tests for rheumatoid factors as presenting 

24. Fukada Y, Ohtomo Y, Kaneko K, Yabuta K. Pathologic and 
laboratory dynamics following the removal of the shunt in shunt 

25. Legrain V, Fontan D, Pujol JF, Guillard JM, Taieb A. Cutaneous 
vasculitis disclosing shunt nephritis. Ann Dermatol Venerol 
1993; 120: 788–789 (in French)

Uwamoto S, Suzuki K, Fukugawa M, Okuda T. Shunt nephritis: 
efficacy of an antibiotic trial for clinical diagnosis. Intern Med 
1993; 32: 291–294

27. Samtleben W, Bauriedel G, Bosch T, Goetz C, Klare B, Gurland 
HJ. Renal complications of infected ventriculocisternal shunts. 
Artif Organs 1993; 17: 695–701

28. Bayston R, Rodgers J. Role of serological tests in the diagnosis 
of immune complex disease in infection of ventriculocisternal shunts for 

29. Fijen CAP, Kuiper EJ, Tja HG, Daha MR, Dankert J. Complement 

nephritis associated with propionibacterium acnes. Infection 

31. Samtleben W, Bosch T, Bauriedel G et al. Medical complications 

32. Lee HS, Cha SH, Cho BS, Yang MH. A case of shunt nephritis. 

1995; 88: 911–918

1996; 85: 882–883

35. Frame PT, McLaurin RL. Treatment of CSF shunt infections 
with intrashunt plus oral antibiotic therapy. J Neurosurg 
1984; 60: 354–360

36. Pople IK, Bayston R, Hayward RD. Infection of cerebrospinal 
fluid shunts in infants: a study of etiological factors. J Neurosurg 
1992; 77: 29–36

37. Dobrin RS, Day NK, Que PG et al. The role of complement, 
immunoglobulin and bacterial antigen in coagulase-negative 

38. Groeneveld ABJ, Nommensen FE, Mullink H, Ooms ECM, 
Bode WA. Shunt nephritis associated with Propionibacterium 
acnes with demonstration of the antigen in the glomeruli. 
Nephron 1982; 32: 365–369

Ruperto Carola University of Heidelberg, 1994; 305–308

40. Schoenbaum SC, Gardner P, Shillitio J. Infections of cerebrospi-
nal fluid shunts: epidemiology, clinical manifestations and 

41. Bayston R, Swinden J. The aetiology and prevention of shunt 

42. McKenzie SA, Hayden K. Two cases of ‘shunt nephritis’. 
Pediatrics 1974; 54: 806–808

43. Blount JP, Campell JA, Haines SJ. Complications in ventricular 
4: 633–656

44. Bayston R, Grove N, Siegel J, Lawellin D, Basham S. Prevention of hydrocephalus shunt colonisation in vitro by 
impregnation with antimicrobials. J Neurol Neurosurg Psychiat 
1989; 52: 605–609

45. Patriarca PA, Laueru BA. Venticuloperitoneal shunt associated 
infection due to Haemophilus influenzae. Pediatrics 1980; 65:
1007–1009


47. Okoro BA, Ohaegbulam. Experience with ventriculo peritoneal 
shunts at the University of Nigeria Teaching Hospital, Enugu. 

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