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Cerebro-Spinal Fluid Shunt Revisions, Importance of the Symptoms and Shunt Structure

Beyin-Omurilik Şant Revizyonları, Semptomların ve Şantın Mekanik Yapısının Önemi

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ABSTRACT

AIM: CSF shunt failure is still a frequent problem in children. This prospective study was designed for focusing symptoms and reasons of shunt failure. We also especially focused on the mechanical reasons of shunt failure.

MATERIAL and METHODS: We focused on the causes of shunt failures, and the symptoms and signs in patients who were operated for shunt malfunction between January 1, 2001 and December 31, 2005 in the neurosurgery department. All examination and operative data were collected prospectively. Evaluation of these data was with the chi-square and Fisher exact tests.

RESULTS: After the evaluation of data on 111 patients and 153 revision procedures, the major symptoms in this group were vomiting (62.16%), somnolence (59.45%) and headache (48.64%). In the majority of the shunt revisions (115 operations, 75.2% of the all 153 procedures), one or more mechanical problems of the shunt systems were identified in surgery.

CONCLUSION: Shunt failures in children sometimes appear with very unusual symptoms. Also, probable structural problems of the shunt systems seem very important for shunt failure according to patient characteristics and etiology of the hydrocephalus. A systematic approach including CT, shunt series and abdominal ultrasound is needed to rule out shunt malfunction.

KEYWORDS: Hydrocephalus, Shunt systems, Symptom

ÖZ

AMAÇ: Beyin-omurilik şant disfonksiyonu, çocukluk çağında halen önemli bir sorundur. Bu prospektif çalışma şant disfonksiyonunun semptom ve nedenleri üzerine odaklanmıştır.

YÖNTEM ve GEREÇ: Bu çalışmada, Ocak 2001, Aralık 2005 tarihleri arasında şant disfonksiyonu nedeniyle opere edilen hastaların disfonksiyon nedenleri ve semptomları incelenmiştir. Tüm preoperatif ve operatif veriler prospektif olarak toplanmıştır. Bu veriler Fisher exact test ve ki-kare testleri kullanılarak değerlendirilmiştir.

BULGULAR: Yüzonbir hasta ve 153 cerrahi prosedürün değerlendirilmesi sonucunda majör semptomlar kusma (69 hasta, %62,16), uykuya meyil (66 hasta, %59,45) ve baş ağrısı (54 hasta, %48,64) olarak tespit edilmiştir.Sekiz hastada ender görülen birtakım semptomlara rastlanmıştır. Bunlar amnezi, spastik monoparazi, ani körlük, ataksi, bradikinezi, disfaji ve üriner inkontinansdır. Şant revizyonlarının çoğunda (115 operasyon, %75,2) bir veya daha fazla mekanik sorunla karşılaşılmıştır. Bu mekanik sorunların çoğu tıkanma şeklindedir. Ayrıca abdominal pseudokist 12 revizyonda (% 7,8) karşımıza çıkmıştır.

SONUÇ: Şant disfonksiyonları bazen çok alışılmadık semptomlarla ortaya çıkabilir. Ayrıca muhtemel şantla ilgili yapısal sorunlar, şant disfonksiyonu açısından hasta karakteristiklerinden daha önemli bir etken gibi görünmektedir. Bu nedenlerle şant disfonksiyonu şüphesinde tüm sistemi gözden geçirecek kranial bilgisayarlı tomografi, şant serisi direkt grafileri, batın ultrasonografisi gibi testlerin her hastaya uygulanması önemlidir.

ANAHTAR SÖZCÜKLER: Hidrosefali, Şant sistemleri, Semptomlar

INTRODUCTION

Cerebrospinal fluid (CSF) shunts are still the most common treatment option for hydrocephalus. However, shunt failure is still a frequent problem. Previous reports indicate that the 1-year failure rate of CSF shunts is approximately 25–40% (7, 11, 26, 35, 39 ,46).

MATERIAL and METHODS

This study includes patients who were operated for shunt malfunction between January 1, 2001 and December 31, 2005. All patients had a 3-piece shunt. The study focused on the causes of shunt failures, symptoms and radiological findings. Some patients presenting with mechanical shunt malfunction also developed shunt infection. Mean follow-up time was 32

months (between 12-60 months). Data about patients' age and gender, etiology of hydrocephalus, symptoms, date of last shunt placement, revision date, valve type, date of shunt failure, radiological findings and cause of shunt failure were prospectively collected and stored in a database software program (Microsoft © Access 2000), then analyzed using a commercial statistical software package (SPSS, version 13.0 for Windows; SPSS, Inc., Chicago, IL). The independent sample t -test, Chi-square and Fisher's exact tests were used for statistical analysis.

Etiology of failure

The causes of the failures were characterized as infections, valve malfunctions, proximal and distal catheter obstructions, disconnections, trapped ventricular systems, migration of shunt parts, abdominal pseudocysts, over-drainages, subdural hemorrhages and inappropriate implantation of the ventricular catheters.

Symptoms

Majority of the symptoms were somnolence, headache, vomiting, bulging fontanel, fever, seizure, macrocephalus and wound problems such as CSF leak, redness, wound dehiscence, ulceration and CSF collection.

There were also unusual patient complaints like lack of appetite, abdominal pain and irritability.

Causes of shunt malfunction

All patients with shunt malfunction underwent neurological and radiological examinations, computerized tomography (CT) scanning of the brain, abdominal ultrasonography and plain x-ray films of the whole shunt system. CSF was obtained from all patients during surgery. In every shunt revision operation, all parts of the shunt systems were investigated considering mechanical reasons for failures. And mechanical problems were determined as proximal and distal occlusions, migration of the shunt system, valve malfunction, disconnection, and shortened distal catheter of the shunt systems. On the other hand, non-mechanical problems were infection, abdominal pseudocyst, valve insufficient, overdrainage, subdural fluid collection and trapped fourth or lateral ventricles.

There was more than one mechanical problem in some cases. In addition, some patients had both mechanical and non-mechanical problems.

There was no other mechanical or non-mechanical problem for shunt failure in valve insufficient group. All the patients in this group had radiological hydrocephalus and/or symptom of the increased cranial pressure, and these patients were treated by changing the valve system with lower one.

RESULTS

Descriptive data

There were 111 patients (64 boys and 47 girls) with CSF shunt malfunction who were admitted to the hospital and underwent 153 shunt revision procedures. All patients had ventriculo-peritoneal CSF shunting systems. Only one revision surgery was required for 86 patients, and 25 patients had two or more revisions. Table I shows patient characteristics and surgical details of study population. Etiology of the hydrocephalus showed a wide variety. Spina bifida aperta (39 patients, 35.1%) and congenital hydrocephalus (18 patients, 16.21%) are the two frequent causes of hydrocephalus in this population. Table I also shows the summary of the descriptive data.

Symptoms

The mean age of study population was 67.57 months (min: 0, max: 348). The most frequent symptoms were vomiting (69 patients, 62.16%), somnolence (66 patients, 59.45%) and headache (54 patients, 48.64%).

No neurological symptoms were identified in 18 patients. However, shunt dysfunctions were recognized in 15 of these patients by incision site problems after the first neurosurgical operations were performed without any neurological symptoms. These operations were not only shunt procedures. Shunt failures were discovered in 5 patients following incision problems like CSF leakage or pseudo-meningocele on spina bifida aperta or tethered cord reconstruction surgery procedure sites. Another 10 shunt failures were determined in early stages of first shunting procedure. There were also surgical incision problems like CSF leakage around the valve system at cranial incision in these patients.

In other 3 patients, there was no neurological symptom or incision problem. Shunt failures were recognized by the examinations for non-neurological symptoms in these patients.

The only symptom in 4 patients was headache. All the other patients had two or more symptoms when they were admitted to the hospital. Symptoms and etiological factors are given in Table I.

Some symptoms were seen at different rates according to the age groups. Incision site problems were mostly seen in the 0-12 months age group, which was statistically significant (chi-square; $p<0.001$). In contrast, the number of patients with vomiting and somnolence were high in older ages. There was also statistically significant differences between age groups considering vomiting (chi-square; $p<0.002$) and somnolence (chi-square; $p<0.001$). Figure 2 shows these relations. Headache symptom was excluded in this figure because of the differences in the expression of this symptom between age groups.

There were also many unusual symptoms in the study group like lack of appetite in 50 patients, abdominal pain in 7 patients and irritability in 42 patients.

Very rare and unusual symptoms like amnesia, spastic mono-paresis, bilateral sudden total blindness, total ophthalmoplegia, ataxia, bradykinesis and urinary incontinence were seen in 7 patients but all of these patients had one or more neurological symptoms, as well.

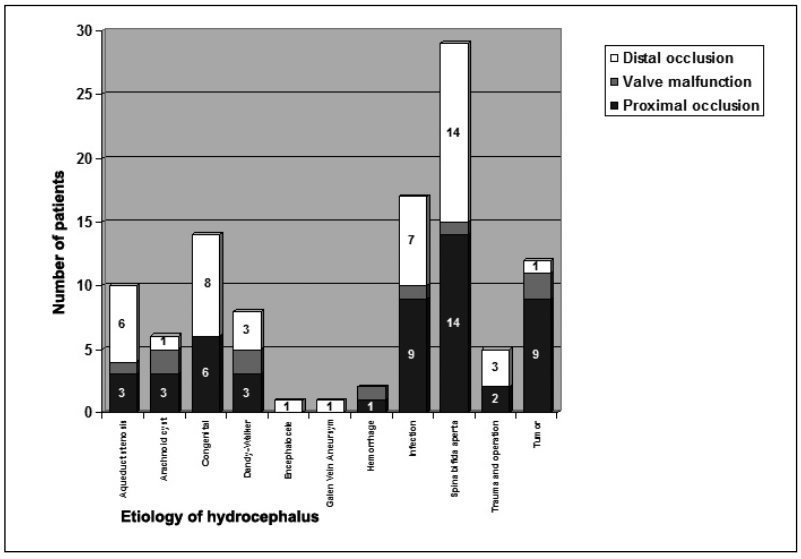

Figure 1: Relations between occlusions of the subparts of the shunt systems and etiology of hydrocephalus.

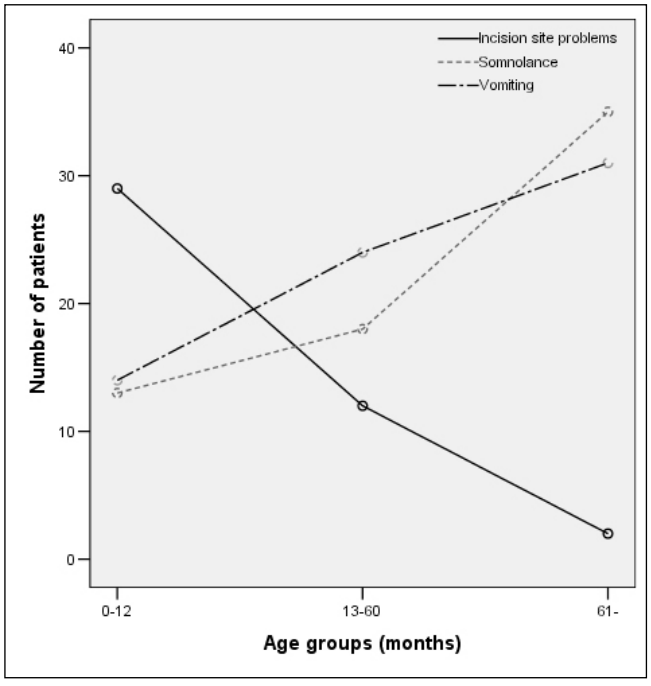

Figure 2: Some of the symptoms which showed different rates according to the age groups.

Reasons of Shunt Revisions

107 operations for mechanical (69.9% of the all 153 procedures) and 46 operations for non-mechanical (30.1% of the all 153 procedures) reasons were performed in the study group (Table II).

Mechanical and non-mechanical reasons appeared together in 3 patients.

Although occlusions of the proximal (in 50 revisions; 32.67%) and distal (in 45 revisions; 29.4%) end of the shunt system were most common findings in revision surgery, infection (in 36 revisions; 23.5%) and valve occlusions (in 10 revisions; 6.53%) were also noted frequently. Revision rate due to infection is significantly higher in the first 12 months of age (p=0.02; chi-square), and 19 of the 36 infection patients (52.7%) were seen in this group. Although 47.3% (9 of 19) of the patients under one year old underwent revision surgery one month after the first operation due to infection, this ratio was determined as 29.4% (5 of 17 patients) for older patients. Mean revision time due to infection was 69.15 day in the group of under one year old patients and 147.88 day for others.

On the other hand, mean revision time for mechanical and non-mechanical dysfunction subgroups were 1171.6 and 155.5 days respectively. There is a statistically difference between mean revision time of subgroups (independent sample t test, $p<0.001$). Mean shunt revision times according to the reasons of dysfunction are summarized in Figure 3.

Only 3 shunt revision surgeries (8.3%) were performed after one year due to infection.

Abdominal pseudo-cysts (APC) were found in 12 revisions, and all of them had CSF infections.

Mechanical shunt problems due to occlusion were categorized within 3 subgroups. These are proximal occlusions, distal occlusions and valve obstructions (malfunction). Distribution of the mechanical problems according to the etiological factors are given in Figure 1. In all etiological subgroups, the number of valve malfunctions was less than proximal and/or distal occlusions. Distal and proximal occlusions were equal in the myelomeningocele group which has the largest number of patients. Similarly, proximal and distal occlusion rates were very close to each other in the subgroups of infection, congenital hydrocephalus, Dandy-Walker malformation and trauma, and there was no statistically significant difference between them. In the tumor subgroup, the proximal occlusion rate was higher than for distal or valve occlusions, which was statistically significant (chi-square; $p<0.05$ for all).

Three patients from the infection group also had catheter or valve obstructions. These patients were excluded in the comparison of the groups.

DISCUSSION

Symptoms

Nulsen and Spitz (34) described the first working CSF shunt in 1952. Immediately after the invention, articles about rates and causes of shunt failures started to be published in the literature (2, 18, 22, 27, 35, 37, 43, 46, 49). Efficiency of CT scanning, magnetic resonance imaging, iodinated contrast

Table I: Patients' Characteristics; Symptoms and Number of Shunt Revisions According to the Age Groups. Very Rare and Unusual Symptoms are Not Added to the Table Because of the Limited Number of Patients

Characteristics	Age			Total
	0-12 months	13-60 months	61-	
Number of Patients (for 111 patients)	38	32	41	111
Male	22	20	22	64
Female	16	12	19	47
Number of Revisions (for 153 procedures)				
1	28	24	34	86
2	6	5	4	15
3	4	2	1	5**
4	1	1	3	3**
5	0	1	1	2
Neurological Signs (for 153 procedures)				
Somnolence	13	18	35	66
Headache	0	13	41	54
Vomiting	14	24	31	69
Stretched anterior fontanel	31	13	0	44
Irritability	14	20	8	42
Abdominal pain	1	1	5	7
Fever	9	6	5	20
Lack of appetite	19	20	11	50
Seizure	0	1	0	1
Other Signs (for 153 procedures)				
Macrocephalus	22	7	1	30
Cranial, abdominal or spinal surgical incision problems ±	29	13	4	46

** Some of the patients were operated on many times but were in different age groups.
± Incision problems include both early and late surgical site problems that are related to hydrocephalus.

studies, shunt taps, doppler ultrasonography and intracranial pressure monitoring for the diagnosis of patients with shunt malfunction were described in the past decades (5, 15, 23, 29, 31, 33, 41, 50, 51, 52). Few studies in the literature have focused on the symptoms and signs of shunt malfunction, and the data are collected from the patient history and physical examinations in most of them (4, 14, 24, 25, 48). Watkins and colleagues (53), Garton and colleagues (16) and Piatt (38, 39) evaluated the predictive value of clinical findings and pointed out one or two symptoms or signs.

In 2001, Garton and colleagues published a prospective controlled clinical study which was focused on the clinical symptoms of the patients with shunt failure(16). They determined that most valuable and statistically frequent symptoms are somnolence (or decreased level of consciousness), irritability, vomiting, headache and high body temperature, respectively. Similarly, our study group has the same symptom characteristics in almost the same sequence. Vomiting, somnolence, headache and irritability were the main symptoms in this study. However, 18 of the patients had no major neurological symptom listed above. These patients were recognized only with secondary signs like CSF collections after spinal surgeries or incidental radiological examinations. Also, three of the symptoms showed a linear relation with the age of the patients. While wound problems were naturally mostly seen in younger patients, vomiting and somnolence were determined in older patients (Figure 2). Lack of appetite, which is an easily distinguished symptom by parents, also was an important clinical symptom in all age groups (Table I).

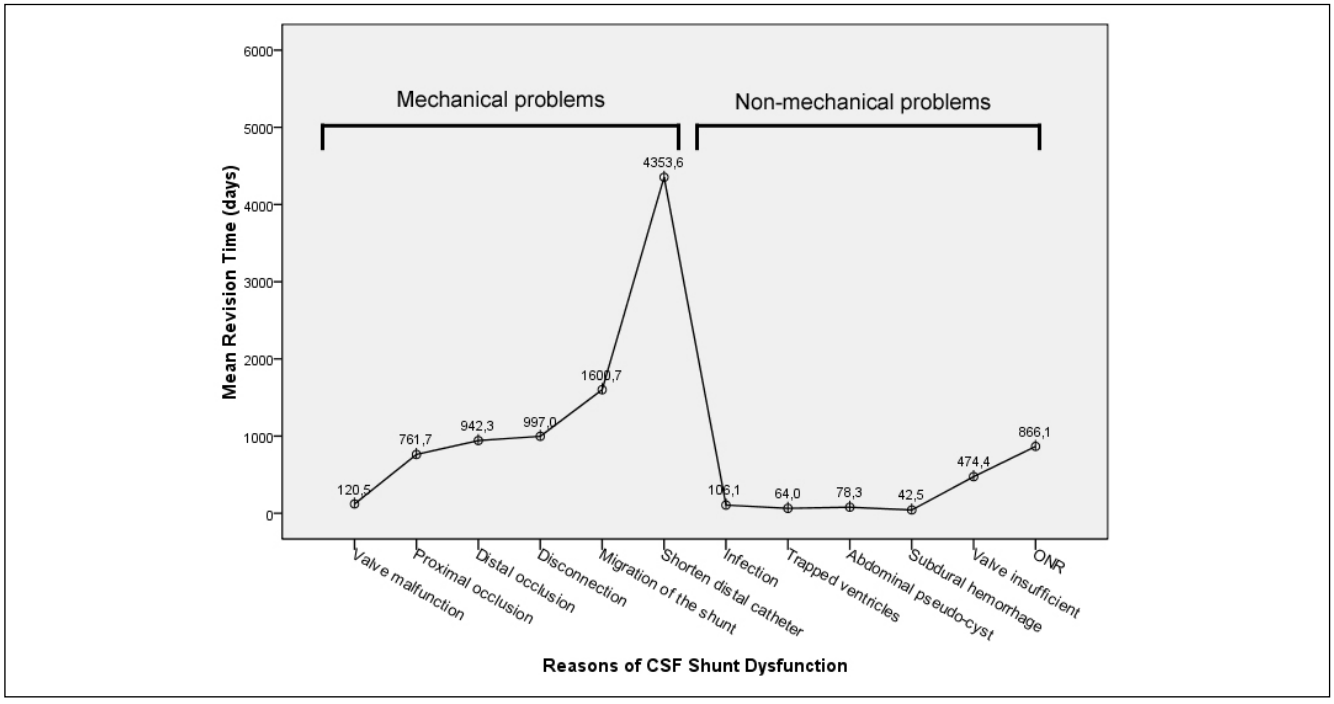

Figure 3: Mean revision times of the subgroups. Mechanical problems appear significantly later than non-mechanical problems.

Some unusual symptoms like amnesia, spastic mono-paresis, bilateral blindness, total ophthalmoplegia, ataxia, bradykinesis and urinary incontinence were also seen in seven patients. There are very few reports in the literature on the relations between shunt dysfunction and these symptoms (8, 16, 32). Blindness related to shunt dysfunction was first reported by Constatini et all in 1987 (8).

Reasons of Shunt Revisions

Although the treatment of hydrocephalus is still mostly dependent on the shunt systems, no numerical data regarding mechanical reasons of shunt revisions is found in the literature. In this study, the major reasons of shunt revisions were mechanical problems and they mostly occurred as occlusions of one or more part of the shunt systems (Table II).

The number of the shunt revisions caused by mechanical problems was 107, and 99 of them occurred because of the occlusion. More than one mechanical problem was found in 16 patients. Although the valve mechanisms have more complex and sensitive structure, proximal (ventricular) and distal (abdominal) catheters were frequently (more than 5 times) affected by the occlusion. Proximal and distal catheters showed similar occlusion rates in majority of all age and etiological groups (Table II, Figure 1). For shunt failure, structural characteristics of the shunt systems seem to be more important than the patient's characteristics like age and illness. Occlusion rates of the different shunt parts were not mainly related to the etiology of the hydrocephalus (Figure 1). However, proximal catheter occlusion was more frequent than for the distal catheter and valve obstruction only in the patients who underwent CSF shunting procedure after brain tumor excision, which could be caused by the elevated CSF protein or invasion of the proximal catheter with growing tumoral mass.

The second most frequent cause of shunt revision was determined as infection (30 shunt revisions). In the literature, shunt infection rate was reported to change between 3 and 15% and show a steady decrease after the first few months (10, 9, 18, 26, 28, 36, 40).

McGirt et al. examined 160 patients with CSF shunt revisions and reported that the percentage of shunt failures caused by infection decreased linearly after the operation. In their study, forty-five percent of shunt failures in the first month were attributed to infection, while only 6% of shunts failing after 2 years were attributed to infection (28). In this study, only 3 revision surgeries were made after one year of replacement. In addition, 52.7% of the patients with shunt infection were under one year old, and only 16.6% of them were older than 60 months.

Abdominal pseudocysts were found to be responsible for 12 shunt revisions in this study (7.8% of all revisions), and APC was accompanied by CSF infection in all patients. Although APCs are reported to occur at a rate between 0.33 and 6.8% (1, 3, 6, 9, 13, 19, 20, 21, 42, 45, 53) in literature, review of the studies shows that infection rates of the APCs change between 17 and 80% (12, 13, 17, 20, 21, 42, 44, 54) . Mobley et al. (2005) reported a review of literature and found the average infection rate of the APC as 42% (30). Most of the infections occurred in the first three months after operations. Similarly,

Table II: Patients' Characteristics; Etiology of Hydrocephalus and Reasons of the Shunt Revisions According to Age Groups

Characteristics	Age			Total
	0-12 months	13-60 months	61-	
Etiology of hydrocephalus (for 111 patients)				
Spina bifida aperta	19	11	9	39
Congenital	5	6	7	18
Aqueduct stenosis	4	4	4	12
Infection	1	5	5	11
Tumor	1	2	6	9
Dandy-Walker M.	2	2	2	6
Encephalocele	1	1	2	4
Arachnoid cyst	2	1	2	5
Trauma and operation	0	0	4	4
Hemorrhage	2	0	0	2
Galen Vein Aneurysm	1	0	0	1
Reasons of shunt revisions (for 153 procedures)				
Mechanical problems±	*Total 107 procedures, 69.9 % of all 153 procedures*			
Proximal occlusion	13	17	20	50
Distal occlusion	14	13	18	45
Valve malfunction	4	2	4	10
Shorten distal catheter	0	0	9	9
Disconnection	1	4	1	6
Total migration of the shunt	2	0	1	3
Non-mechanical problems±	*Total 46 procedures, 30.1 % of all 153 procedures*			
Infection	19	11	6	36
Abdominal Pseudo-cyst	4	5	3	12
Valve insufficient	0	3	4	7
Trapped fourth or lateral ventricle	2	2	0	4
Subdural hemorrhage	2	0	0	2
Overdrainage	0	0	1	1
Wrong localization of the ventricular catheter	1	0	0	1

±: Three patients had mechanical and non-mechanical problems together. Two of them had APC and distal-valve catheter occlusion, and one of them had trapped lateral ventricle and proximal catheter occlusion. All patients were added to the mechanical problems group because their symptoms depend on the occlusions.

abdominal pseudocysts and subdural hemorrhage showed a tendency to appear within the first 100 days. Although valve mechanisms were seems to be more resistant to the occlusion or malfunction than the catheter systems, valve occlusions (occurred on average 120.5 days after operation) appeared as the earliest mechanical problem. Similar to the occlusion rates, mean distal and proximal catheters occlusion times were very close to each other. Other mechanical problems which can be related to the increasing length of the patients like disconnection, shortening of the distal catheter or migration of the shunt systems appeared later than the occlusive problems.

This study shows that major symptoms of the shunt failures which show different disturbances among the age groups are vomiting, somnolence, headache and irritability. Also, shunt failures can cause unusual symptoms like lack of appetite, abdominal pain, amnesia, spastic mono-paresis, sudden total blindness or total ophthalmoplegia, ataxia, bradykinesis and urinary incontinence. For all of the symptomatic patients, a systematic approach including brain CT, shunt series (x-ray examinations) and abdominal ultrasound is needed to rule out the shunt malfunction.

Mechanical problems which depend on the shunt systems are the main cause of shunt failures in all age groups. Infection is the second most common reason of the shunt revisions, in especially the first year of the life. Unexpectedly, both of distal (abdominal) and proximal (ventricular) catheter systems show greater tendency for occlusion than valve systems. Studies regarding shunt revision times demonstrate that mechanical problems appear significantly later than non-mechanical problems.

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