The Management of Patients with Cerebrospinal Fluid Shunts

John D. Loeser, MD, Clifford J. Sells, MD, and David B. Shurtleff, MD
Seattle, Washington

This article outlines the principles of management of the patient with a cerebrospinal fluid shunt, emphasizing the clinical and laboratory methods of determining shunt malfunction or infection. Appropriate therapies for each complication are described.

Continuous patient evaluation and prompt treatment of complications are required to maximize the beneficial effects of a cerebrospinal fluid (CSF) shunting procedure for hydrocephalus. Not all patients can be closely followed by the neurosurgeon who inserted the original shunt; not all complications require the services of a neurosurgeon to initiate appropriate therapy. The purpose of this article is to outline a rational program for the assessment and management of a patient with a CSF shunt and to emphasize key factors in the determination of optimal continuing therapy.

From the Departments of Neurological Surgery and Pediatrics, University of Washington School of Medicine, Seattle, Washington. Drs. Loeser, Sells, and Shurtleff are Affiliates of the Child Development and Mental Retardation Center at the University of Washington. Requests for reprints should be addressed to Dr. John D. Loeser, Department of Neurological Surgery, RI-20, University of Washington, Seattle, WA 98195.

Evaluation of the Patient

The general scheme for the evaluation of a patient with a CSF shunt is illustrated in Figure 1. This flow diagram will serve as the framework for the discussion of the management of a patient with a CSF shunt. Obviously, any evaluation must commence with a careful history and pertinent physical examination. A wide variety of CSF shunt problems have arisen that could have been readily solved if the patient or the patient's parents could have described the type of shunt system implanted at another institution. As children with hydrocephalus live longer, it is becoming all too common to be unable to obtain the original operative records. The name of the surgeon or hospital may not be recalled, the hospital records and x-rays may have been destroyed, or delays in obtaining information may interfere with optimal care. It is strongly urged that copies of the operative report describing the shunt system be given not only to the referring physician but also to the parents. The type of shunt is an important portion of the history and will influence both the diagnostic workup and some important aspects of the therapy of shunt malfunction.

Most patients with shunts are children; the his-

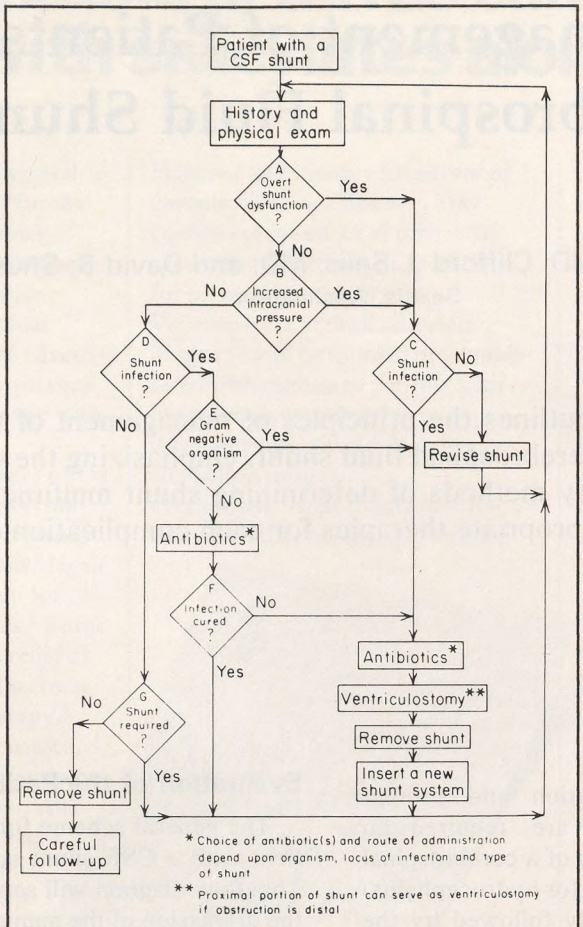

Figure 1. Flow diagram illustrating steps in the evaluation of a patient with a cerebrospinal fluid shunt.

tory must include growth and developmental landmarks so that recent head growth and performance can be placed in the perspective of past achievements. An interval history since the time of shunt insertion or last revision may reveal periods of shunt dysfunction or suggestions of infection.

The physical examination should include a general assessment of growth and development, behavior, and the various organ systems; but must focus upon size and shape of the head, skull sutures, scalp veins, fontanelles, and fundi. Cranial nerve, spinal cord, and long tract functions may reveal changes consistent with increased intracranial pressure. Signs of systemic infection, vascular or renal disease, or loculation of cerebrospinal fluid about the distal shunt must be sought; only after a firm data base is established by the history and physical examination can one prudently proceed to evaluation of the shunt system. Merely depressing the flushing device does not suffice to determine shunt adequacy. Confirmation of shunt function is established by special dynamic studies.[1,2]

The first determination to be made is the presence of "overt shunt dysfunction" (Figure 1, rhomboid A). This includes such problems as fluid collection around the shunt, palpable disconnection of shunt components, flushing device permanently depressed or incompressible, erosion of the shunt through the skin, or x-ray evidence of malposition or disconnection. If overt shunt dysfunction is observed, the shunt must be revised.

If overt shunt dysfunction is not established,

Table 1. Antibiotics of Choice in CSF Shunt Associated Infections

Organism	Antibiotic	Route of Administration	Dosage	Frequency
Staphylococcus	Methicillin	Intravenous Intraventricular	100 to 200 mg/kg/day 25 to 100 mg/day	q 4 to 6 hr qd
Staphylococcus susceptible to aqueous penicillin	Penicillin aqueous crystalline	Intravenous	25,000 to 300,000 units/kg/day	q 2 to 6 hr
B-Streptococcus or Streptococcus viridans	Penicillin* aqueous crystalline	Intravenous	25,000 to 300,000 units/kg/day	q 2 to 6 hr
E Coli or Klebsiella or Proteus	Gentamicin	Intramuscular Intraventricular	5 to 7 mg/kg/day 2 to 4 mg/day	q 8 hr qd
Pseudomonas	Gentamicin and/or Carbenicillin	Intramuscular Intraventricular Intravenous	5 to 7 mg/kg/day 2 to 4 mg/day 400 to 500 mg/kg/day	q 8 hr qd q 4 hr

*Neither aqueous penicillin nor carbenicillin should be administered intraventricularly or intrathecally; both are neurotoxic when injected directly into the central nervous system.

the physician must then ascertain from the history and physical examination and from tests of shunt function if the intracranial pressure is too high for the patient (Figure 1, rhomboid B). Elevated intracranial pressure is usually associated with rapid growth in skull circumference, bulging fontanelle, prominent scalp veins, and sutural diastasis. The child often manifests lethargy, nausea and vomiting, headache, retarded cognitive and motor development, and neurologic signs which may include sixth-nerve palsies, coma, and papilledema. The clinical impression can be confirmed by measuring shunt transmission pressures, cerebrospinal fluid flow, and ventricular pressure as has been described.[1,2] Symptomatic increased intracranial pressure mandates shunt revision.

When either overt shunt dysfunction or increased intracranial pressure are diagnosed, it is imperative to determine if there is bacteriologic contamination of the cerebrospinal fluid and the shunt (Figure 1, rhomboid C). This is done by sampling the cerebrospinal fluid from the shunt system or ventricles for culture and sensitivity, cells, glucose, and protein.[3] If there are no clinical signs of infection, the authors do not advocate delaying a necessary shunt revision to await the results of culture of the cerebrospinal fluid. Revising a shunt in the presence of CSF infection may result in continued and often more severe infection. When there are no clinical signs of infection, always sample the cerebrospinal fluid at the time of shunt revision. If a silent infection has been present, the

operative specimen will facilitate early, appropriate antibiotic therapy.

Therapy

Shunt infection and shunt dysfunction often occur together; an aggressive management program is required to promptly handle both problems. First, the patient must receive antibiotics appropriate for both the bacteria and the infection site. The route of administration is dependent upon the type of shunt and the locus of shunt obstruction. For example, drugs which do not penetrate the blood-brain barrier must be given directly into the ventricle. Table 1 lists the usual organisms associated with CSF shunt infection, and the dosage and route of administration of the commonly used antibiotics.[4-8] The table, however, is only a general guide. In all instances, cultures and sensitivity tests should be performed. Mean inhibitory concentrations of antibiotics in the ventricular fluid drawn prior to each intraventricular dose can be used to determine appropriate antibiotic dosage and frequency of administration. The antibiotic concentration in the cerebrospinal fluid immediately prior to the next dose should be at least two times the mean inhibitory concentration of the specific organism. The average shunted ventricular space is approximately 150 cc; individuals with enlarged ventricles should have doses of intraventricular medication increased proportionately. Clearance of antibiotics from the cerebrospinal fluid varies with each patient and should be evaluated carefully to avoid either ineffectively low doses or the accumulation of toxic amounts; in most patients daily administration is usually adequate. If a perforable cerebrospinal fluid device with access to the ventricular CSF space has not been used, a Rickham or Ommaya reservoir should be placed to avoid cerebral mantle destruction due to repeated ventricular taps. If the child has an unoperated myelo-meningocele, nonpenicillin agents may be introduced cautiously via the sac cerebrospinal fluid. Perforation of the sac should be through its margin and should transverse normal subcutaneous tissue, avoiding hemangiomatous and nervous tissue as well as the membranes over the dorsal surface of the sac.

Intraventricular antibiotic therapy should continue for 10 to 14 days. Systemic antibiotic therapy should continue for an additional three to four weeks.[5,6]

In addition to antibiotic therapy, the increased intracranial pressure must be immediately alleviated. When the shunt obstruction is distal, some shunt systems permit tapping the flushing device. With other types of shunt, ventriculostomy is required. The brain does not tolerate the combination of infection and increased pressure; both must be vigorously treated. Thirdly, after the CSF pressure has been normalized and adequate antibiotic levels established in blood, cerebrospinal fluid, and tissues, the infected, dysfunctional shunt must be removed. After three to seven days on external ventriculostomy drainage and antibiotics, an entirely new shunt system is inserted as far away from the old tract as possible. Antibiotics are continued for two to five weeks. One week after antibiotic cessation, appropriate cerebrospinal fluid and blood samples are taken to rule out continued infection. The patient must be carefully observed to ascertain that a suppressed infection does not recur.

When shunt function is apparently adequate and intracranial pressure is not elevated, the management is, of course, different. One must then consider the possibility of infection in a patient with a well-functioning shunt (Figure 1, rhomboid D). Usually, clinical assessment alone rules out infection; suspicious cases must be evaluated by sampling cerebrospinal fluid and blood. When gram-positive infection is documented, a three to six week course of the appropriate antibiotic is advocated. After a week without drugs, cerebrospinal fluid and blood are again cultured. If the infection persists, it will be necessary to replace the shunt to clear the infection (Figure 1, rhomboid F). When gram-negative infection is present (Figure 1, rhomboid E), the authors believe that total shunt replacement and coverage with the appropriate antibiotics are required.

Most patients will have neither shunt dysfunction nor infection. Their shunts will require no treatment at this evaluation interval. Periodically, the need for the shunt will be questioned (Figure 1, rhomboid G). Pressure and flow studies may seem to indicate that the shunt is not transmitting fluid.

Removal of this shunt is often a disaster; utmost caution is urged when contemplating shunt removal. If it is believed that the shunt can be safely removed, it is first prudent to ligate it in situ. After four to six weeks, if no problems have developed, the shunt may be removed. In fact, this is a very rare event in our institutions and this maneuver is not advocated.

The interval until the next evaluation is, of course, determined by the nature of the patient's problem. The authors routinely see the patient one month after shunt revision. If the shunt system is functioning well, one year is a standard time for a return visit to our clinic. More frequent assessment by the primary care physician may also be required.

Antibiotic Selection

The problem of selection of the appropriate antibiotic for a shunt-related infection cannot be overemphasized. In order to plan rational therapy, the following facts must be known:

1. Organism(s) causing infection
2. Antibiotic sensitivities of these organism(s)
3. Locus (or loci) of infection
   a. ventricular
   b. meningeal
   c. loculated intracranial
   d. within shunt
   e. within skin incisions
   f. intravascular
   g. intraperitoneal (or pleural, etc)
4. Distribution characteristics of antibiotics which could be used for each organism; also optimal routes of administration and dosages.
5. Type of shunt system.

The most common errors involve failure to give a drug which gets to the site of infection by the route of administration; ie, gentamicin must be placed directly into the ventricle to achieve adequate levels in most ventricular systems. This is readily accomplished with a nonvalved flushing device (such as the Foltz To-and-Fro Flusher), which is one reason the authors prefer this type of shunt system. If one cannot backfill the ventricles from the flusher, a ventriculostomy is required for intraventricular instillation. In contrast, chloramphenicol administered systemically achieves excellent levels in all CSF spaces.

Preliminary data have already suggested that computerized axial tomography will revolutionize the assessment of hydrocephalic patients. Ventricular size can be accurately determined at frequent intervals with minimal radiation and noninvasive techniques that do not increase risk of infection. Whereas clinical assessment will remain the primary determinant of shunt function, the strategies for detailed testing will change. The concept of management of a patient with a CSF shunt which is proposed here will be applicable even with this technological advance.

Acknowledgement

Supported in part by Training Grant #NS 05211 from the National Institute of Neurological and Communicative Disorders and Stroke, and Grant #C 112 from the National Foundation - March of Dimes.

References

1. Hayden PW, Rudd TG, Dizmang D, et al: Evaluation of surgically treated hydrocephalus by radionuclide clearance studies of the cerebrospinal fluid shunt. Develop Med Child Neurol (suppl) 32:72, 1975
2. Harbert JL, Haddad D, McCullough DC: Quantitation of CSF shunt flow. J Nucl Med 14:405, 1973
3. Myers MG, Schoenbaum SC: Shunt fluid aspiration: An adjunct in the diagnosis of cerebrospinal fluid shunt infection. Am J Dis Child 129:220, 1975
4. McLaurin RL: Treatment of infected ventricular shunts. Child's Brain 1:306, 1975
5. Shurtleff DB, Foltz EL, Weeks RD, et al: Therapy of staphylococcus epidermidis: Infections associated with cerebrospinal fluid shunts. Pediatrics 53:55, 1974
6. Sells CJ, Shurtleff DB, Loeser JD: Gram-negative cerebrospinal fluid shunt-associated infections. Pediatrics 59:613, 1977
7. Kramer PW, Griffith RS, Campbell RL: Antibiotic penetration of the brain. J Neurosurg 31:295, 1969
8. Schoenbaum SC, Gardner P, Shillito J: Infections of the cerebrospinal fluid shunts: Epidemiology, clinical manifestations, and therapy. J Infect Dis 131:54, 1975