Multiloculated hydrocephalus: a serious complication of cerebrospinal fluid shunt infection with gram negative organisms

A.B. Jamjoom, FRCS Ed (SN)

ABSTRACT

Objectives: To define the risk of multiloculated hydrocephalus (MLH) following gram negative organisms (GNO) shunt infection and to provide data for the first time about GNO shunt infection in Saudi Arabia.

Materials and methods: The author carried out a retrospective study of 11 consecutive cases of GNO shunt infection that were treated at King Khalid University Hospital (KKUH) from 1988-1994.

Results: There were 5 males and 6 females with a median age of 3 months. Hydrocephalus followed an intraventricular hemorrhage in 9 cases. The GNO shunt infection was diagnosed at a median of 30 days after the original shunting. Pseudomonas aeruginosa was the commonest organism. The median sterilization time of the cerebrospinal fluid (CSF) was 14 days of external ventricular drainage. Eight (73%) cases developed MLH which was diagnosed at a median of 2 months after the shunt infection. This was significantly higher than the 10% incidence of MLH following shunt infection by Staphylococcus epidermidis at KKUH (p=0.005). At a mean follow-up of 23 months, the mean shunt revision rate of our GNO shunt infected patients was 0.23/year. One patient died of multiple abscesses and 6 remain severely disabled.

Conclusions: Clinicians should be aware of the increased risk of MLH following GNO shunt infection particularly in patients with post hemorrhagic hydrocephalus. Every effort should be made to diagnose the shunt infection early and to treat it aggressively.

Saudi Medical Journal 1997; Vol. 18 (1) 54-58.

Keywords: Cerebrospinal fluid shunt infection, gram negative organisms, intraventricular hemorrhage, multiloculated hydrocephalus, ventriculoperitoneal shunt, ventriculoscopy.

Infection is a well recognized complication of cerebrospinal fluid (CSF) shunts that occurs in 5-10% of cases.1 Shunt infection is most frequently caused by Staphylococcus epidermidis which alone is responsible for 48% of these patients.1 Gram negative organisms (GNO) account for 19-22% of shunt infected cases.2 Considering the extensive literature on shunt infection, it is surprising that reports focusing on the specific problems associated with GNO shunt infections are very few.2,3 In particular, while the literature clearly documents the increased rate of morbidity and mortality associated with shunt infections due to GNO,3 their role in the etiology of multiloculated hydrocephalus has not been outlined. Multiloculated hydrocephalus (MLH), which is characterized by ventriculoperitoneal shunts or ventriculostomy within the ventricular system, is a difficult neurosurgical problem that carries a poor prognosis. In this study, the author undertook a retrospective review of all the cases of GNO shunt infection which were treated at King Khalid University Hospital (KKUH) over a 7-year period. The objectives of the study are to heighten the awareness of clinicians treating hydrocephalic patients of the increased risk of development of MLH following GNO shunt infection and to provide data for the first time about the clinical characteristics and outcome of patients with GNO shunt infection in Saudi Arabia.

Materials and methods The medical records and computerized tomography (CT) scans of all patients with CSF shunt infection due to GNO who were treated at KKUH between 1988-1994 were reviewed. The shunt infection was confirmed by a positive culture of GNO from the ventricular CSF. The infection was treated by prompt removal of the infected shunt, external ventricular drainage (EVD) and intravenous antimicrobial therapy based on the sensitivity of the infective pathogens. INSERT PHOTO 1 Ceftriaxone, amikacin and gentamycin were the...
most commonly used antibiotics. Intraventricular antibiotics were not used. The EVD was maintained with a change of drain, if the drain lasted more than 14 days, until the CSF culture was negative on three consecutive occasions. Following this a new ventriculoperitoneal (VP) shunt was inserted and the antibiotics were discontinued.

The following data was collected for each patient: age at time of diagnosis of hydrocephalus; sex; primary cause of hydrocephalus; duration between shunting and diagnosis of shunt infection; the infective organisms: CSF protein and white blood cell (WBC) at the time of diagnosis of the shunt infection; duration of EVD; development of MLH and how it was treated and late complications such as abscess formation and shunt malfunction. The shunt revision rate was calculated by dividing the total number of revisions by the total number of patients by the mean follow-up in years. The outcome was determined at follow-up and was considered “good” when the patient was developing normally or appeared to have a minor disability and “poor” when the patient had a serious psychomotor problem.

The diagnosis of MLH was made at follow-up and was based on the ventriculographic evidence of accumulation of dye in some dilated cavities but not others and the CT evidence of ventricular septations or asymmetrical dilatation of part or parts of the ventricular system and failure of that part to reduce in size despite evidence of adequate shunting elsewhere in the ventricular system (Fig. 1). The GNO shunt infection was considered to be the main cause of MLH when the CT scan of the patient, prior to the shunt infection, did not show evidence of multiloculation (Figs. 2a,b). Once the diagnosis of MLH was made another shunt or shunts were placed and in the last two patients endoscopic fenestration of intraventricular septations was attempted. The endoscopy was carried out via a frontal burr hole using a 5 mm rigid pediatric nephroscope and monopolar diathermy. In attempting to determine the significance of the incidence of MLH following GNO shunt infection, it was compared to the incidence of MLH in 21 patients with shunt infection caused by Staphylococcus epidermidis who were treated by us during the same period. The difference between the two groups was examined statistically using a chi-squared test.

**Results** Between 1988-1994, 560 CSF shunting procedures were performed and 37 patients were treated for CSF shunt infection at KKUH. Gram negative organisms caused shunt infection in 11 (30%) patients. Five of these cases had their original shunting procedure elsewhere. There were 5 males and 6 females with an age range of 8 weeks-3 years (median 3 months). Hydrocephalus was related to intraventricular hemorrhage (IVH) in 9 patients, aqueduct stenosis in 1 patient and Arnold Chiari malformation in 1 patient. The interval from shunting to the diagnosis of shunt infection was 8 days - 6 months (median 30 days). The infective organisms were Pseudomonas aeruginosa (3 cases); Klebsiella (2 cases); Enterobacter cloacae (2 cases); Seratia marcescens (2 cases); Escherichia coli (1 case) and Citrobacter diversus (1 case). The infection was related to a bowel perforation in one case. At the time of diagnosis of the shunt infection, the CSF protein ranged from 0.4-9.3 (mean 3.15) mg/l and the CSF WBC ranged from 15-50,000 (mean 250) cells/cu mm. The duration of EVD ranged from 9-28 (median 14) days. Eight (73%) cases developed MLH which was significantly higher than the 10% incidence of MLH following shunt infection by Staphylococcus epidermidis (p=0.004). The MLH was diagnosed from 1-8 (median 2) months after the shunt infection. There was no evidence of intraventricular multiloculation prior to the shunt infection on the computerized tomography (CT) scan in 5 cases and ultrasound in 3 cases. Because CT scans were not carried out routinely following the replacement of the shunt after control of the infection, an assessment of the early postoperative status of the ventricular system was not available. The MLH was therefore diagnosed at variable intervals after the shunt infection. The endoscopic fenestration of intraventricular septations while the existing VP shunt was in place, appears to be controlling the hydrocephalus at a follow-up of 6 months in one patient (Fig. 3). In another patient we were unable to fenestrate the rather thickened septations adequately and safety and the endoscopic procedure was abandoned. Seven patients were treated by the insertion of another VP shunt and one of these patients later underwent another shunting procedure for an encysted left frontal horn (Fig. 4).

At follow-up of 6-48 (mean 23) months, the mean shunt revision rate for our patients with GNO shunt infection was 0.23 revisions/year. One patient with Pseudomonas shunt infection died 4 months later as a result of the MLH progressing to multiple abscesses which failed to respond to repeated surgery and antimicrobial therapy. Of the survivors, 6 patients all of whom had a severe IVH, are exhibiting major psychomotor retardation and therefore their outcome is “poor”.

**Discussion** The rate of GNO causing shunt infection at our unit is unusually high (30%). This is related to referral bias. Five of the cases reported here had their original shunt surgery elsewhere. Our neurosurgical unit is a tertiary referral center that receives patients with unusual shunt infections or when the infection proves difficult to eradicate. The pathogenesis of GNO shunt infection is not clear. It is thought that the infection is the result of direct wound
Fig. 1 CT scan (no contrast) showing a dilated 4th ventricle and left temporal horn despite the presence of a VP shunt. The right temporal horn is well decompresed.

Fig. 2a CT scan (plain) at presentation prior to the GNO shunt infection showing a dilated lateral and 3rd ventricle with no evidence of loculation.

Fig. 2b CT scan (IV contrast) after the GNO shunt infection showing ependymal enhancement and ventricular compartmentalization.

Fig. 3 CT scan (plain) showing a multiloculated hydrocephalus which appears to be controlled by one shunt following endoscopic fenestration of septations.
contamination during surgery or caused by a retrograde infection from an asymptomatic perforation of the bowel. Perforation of the bowel was documented in only one of our cases. The mortality rate of our GNO shunt infected patients was 9% which is within the 0-39% mortality rate reported in the literature for this disease. Fifty-five per cent of our cases had a poor outcome while Stamos et al reported a poor outcome in only 11% of their GNO shunt infected patients. Several reasons may account for the difference between our results and those of Stamos et al. First, in our series there was a higher incidence of post hemorrhagic hydrocephalus (PHH) than in Stamos’ study (82% vs 13%). It is recognized that severe IVH is associated with an unfavorable outcome. Second, the interval between the shunting and the diagnosis of the GNO shunt infection in our study (median 30 days) was longer than the interval stated in Stamos’ series (median 10 days). This may suggest a possible delay in the presentation, referral, diagnosis and treatment of our patients which is likely to influence the outcome. Third, while there was no record of the incidence of MLH in Stamos’ study, a significant number of our patients (73%) developed MLH which is known to be associated with a poor prognosis. The high incidence of PHH and the delay in presentation and treatment of our patients may provide an explanation for the very high incidence of MLH amongst our GNO shunt infected patients. Fourth, in our study Pseudomonas, Klebsiella, Enterobacter and Serratia accounted for the majority of GNO shunt infections whereas E.coli was the most frequently isolated bacteria in the Stamos’ study. Fifth, even though we had commenced with the appropriate treatment as soon as the diagnosis was made, clearing the CSF of the GNO on three consecutive specimens required a median of 14 days of EVD. This is much higher than the mean sterilization time of 6.5 days or less reported by Stamos et al who also recommended the use of intravenous rather than intraventricular antibiotics. This would reflect possibly more severe infection in our cases which may account for poorer results.

It is well established that neonatal meningitis and IVH are the commonest causes of MLH. The role of CSF shunt infection in the etiology of MLH has been suggested but not clearly outlined. It is logical to assume that the ventriculitis associated with the shunt infection may result in ventricular septations and closure of foramen of Monroe, aqueduct and the outlets of the fourth ventricle thus producing MLH. Although it is difficult to prove that the GNO shunt infection was the sole etiological factor for the development of MLH in 73% of our patients, there is no doubt that it was a contributing factor in the origin or progression of the loculation. This is not surprising since in 55 of the 57 cases of MLH due to neonatal meningitis reviewed by Albanese et al, the infective agents were GNO.

The definite treatment of MLH is surgical. The operative approaches described included: multiple shunt placements, craniotomy and transcortical fenestration of intraventricular septations and ventriculostomy with manual and laser fenestration. Multiple shunt placements, which were employed in most of our cases, have been the traditional method of treating this condition. Our shunt revision rate (0.23/year) is low compared to the revision rate of 0.55-3.04/year in MLH reported in the literature. This is probably related to the short follow-up of our patients. The use of ventriculostomy to change the multicellular hydrocephalus to a unilocular hydrocephalus has become very popular. It must be appreciated, however, that the endoscopic procedure may not be successful and may have to be repeated and that multicellular may recur.

Conclusion Our observations suggest that a combination of GNO shunt infection and PHH are associated with an increased risk of MLH. This implies that every effort should be made to avoid shunt infection, particularly in patients with PHH and that once the infection is diagnosed as being due to a GNO, the patient should be managed aggressively with the appropriate intravenous antimicrobial therapy and EVD after removal of the infected shunt. These patients should be followed up carefully with serial CT scans to detect the multiloculation early.
Acknowledgment The author is grateful to Dr. A.H.A. Mohammed and Dr. A. Al Bouki for their help with the data collection and to Dr. Z.A.B. Jamjoom and Professor Naim Ur-Rahman for their permission to include their cases. The author is also grateful to the Microbiology Department and Radiology Department of KKUH for their services to the patients reported and to Ms Cora Rivera for her excellent secretarial assistance.

References


