ORIGINAL ARTICLE

Experience of ventriculo-peritoneal shunt insertion in late presenting congenital hydrocephalus

Zahid Hussain, Abdul Ghaffar, Shahzad Ahmed Qasmi, Junaid Mushtaq

Abstract

Objective: To evaluate the outcomes of insertion of ventriculo-peritoneal shunts in paediatric neglected hydrocephalus.

Methods: The quasi-experimental study was conducted at the Combined Military Hospital, Rawalpindi, from January 2012 to June 2014, and comprised infants of both genders who presented late with congenital hydrocephalus, having fronto-occipital circumference more than 98 percentile of matched age group. Pre-operative assessment was based upon detailed history, clinical examination, laboratory investigations and computed tomography scan of head. After insertion of ventriculo-peritoneal shunt, patients were followed up for outcomes. Data was analysed using SPSS 17.

Results: Of the 30 infants, 12(40%) were girls and 18(60%) were boys. Overall mean age was 7.73±1.41 months (range: 5-10 months). Mean fronto-occipital circumference was 54.30±3.08. Cerebrospinal fluid infection was documented in 12(40%). Abdominal wound complications were observed in 7(23.3%) infants. Ventriculo-peritoneal shunt was removed in 18(60%) and eventually replaced in 18(60%). In-hospital mortality on account of complications was encountered in 13(43.3%). Correlation of fronto-occipital circumference to mortality was significant (p=0.001).

Conclusion: To shunt or not to shunt remains a dilemma for poor-risk infants, but timely reporting of infants with hydrocephalus, proper case selection may improve the outcome of surgical intervention.

Keywords: Ventriculo-peritoneal shunt, VPS, Hydrocephalus, Neglected, Fronto-occipital circumference, FOC. (JPMA 66: 430; 2016)

Introduction

Hydrocephalus is one of the surgically treatable paediatric problem.[1] Cerebrospinal fluid (CSF) shunting is the most common way of managing hydrocephalus and ventriculo-peritoneal shunt (VPS) is the mainstay of CSF shunting though having high rate of complications ranging from 1% to 40% with the mean shunt survival being about 5 years.[2] The rate of complication is very high in neglected cases of hydrocephalus. Despite the significant advances in shunt technology, most of the complications are related to shunt hardware and infections.[3,4]

Neglected hydrocephalus can be defined as marked enlargement of head with thinned out scalp, pressure sores on bony prominences, with established neurological deficit as visual loss, marked ventricular dilatation with less than 2cm cortical mantle associated with absence / agenesis of the cerebellar vermis.

The idea of surgical intervention in neglected cases was related to availability of on-demand provision of free medical treatment, moral reasons to ease the problems of family with handling of the disabled and actively ill child. Various factors contributing to delay in neurosurgical advice were religious taboos, local quackery and lack of education.

The current study was planned to review the outcome of surgical intervention in neglected hydrocephalus and to assess the various risk factors having adverse effect on outcome.

Patients and Methods

The quasi-experimental study was conducted at the Neurosurgery Department of the Combined Military Hospital (CMH), Rawalpindi, from January 2012 to June 2014, and comprised infants of either gender who presented late with congenital hydrocephalus, fronto-occipital circumference (FOC) significantly higher than 98 percentile of matched age group, having signs of marked hydrocephalus such as tense fontanelle, irritability, perinaud's phenomenon and failure to thrive.

After getting written informed consent from respective parents, comorbid conditions of spina bifida, systemic anomalies and chest infections were recorded. Infants of normal or small FOC with hydrocephalus, secondary craniosynostosis, hydrocephalus ex-vacuo, post-

Combined Military Hospital, Rawalpindi Cantonment, Pakistan.
Correspondence: Zahid Hussain. Email: zahidsurgeon@gmail.com

meningitic hydrocephalus, active sepsis, multiple organ congenital anomalies, coagulopathies and hydrocephalus associated with tumours were excluded.

Pre-op assessment included detailed history, clinical examination, including FOC, fontanelle tension, scalp condition, pressure sores and systemic examination. Pressure sores were graded from 1 to 4; grade 1 was non-blanchable erythema of intact skin, grade 2 was partial thickness skin loss involving epidermis, dermis or both, grade 3 was full thickness skin loss but bone and muscle were not exposed, and grade 4 was full thickness tissue loss with exposed bone. Laboratory investigations, like complete blood count (CBC), coagulation profile, X-ray chest and computed tomography (CT) scan of head were carried out.

Patients were prepared, draped and the usual site of insertion of ventricular catheter was at keen's point unless scalp sores dictated deviation. Various types of shunts were used according to urgency, availability and priority of the scalp condition ranging from Pudenz burr hole valve tolow-profile delta valve and Chhabra valve.

Post-operatively, patients were followed at 10 days, fortnightly, monthly and 3 monthly for outcomes like clinical condition, abdominal distension, re-admission, duration of hospital stay, CSF infection, subdural collection, shunt malfunction, cranial wound complications, abdominal wound complications, shunt removal, shunt revision and in-hospital mortality.

Data was analysed in terms of frequencies, percentages and mean ± standard deviation (SD).Descriptive data was analysed for association using univariate analysis.

Results

Of the 30 infants operated upon, 12(40%) were girls and 18(60%) were boys. Overall mean age was7.73±1.41 months (range: 5-10 months). Mean FOC was 54.30±3.08cm. Four (13.3%) patients had had prior neurosurgical advice, but no documented CSF infection. Haemoglobin (Hb) ranged from 6.4 to 8.4 g/dl. Grade 1-3 pressure sores on bony prominences of parieto-occipital areas were seen in 19(63.3%). Procedure of VPS was tolerated by all 30 patients and made uneventful recovery from anaesthesia and were discharged on 2nd post-op day. Readmission was frequent and all 30 patients were re-admitted in post-op period ranging from 10-90 days. Re-admission hospital stay ranged from 2 to 30 days (Table-1).

Abdominal distension occurred in 8(26.6%) patients due to large amount of CSF shift to abdominal cavity and were managed conservatively. Subdural fluid collection was recorded in 4(13.3%) and 2(50%)of them required burr hole intervention. Cranial wound erythema/pressure necrosis of wound was seen in 9(30%) patients (Table-2).

Shunt malfunction developed in 12(40%) patients over 3 months and the same number was found to have CSF infection. Abdominal wound complications were observed in 7(23.3%) patients ranging from stitch abscess to wound dehiscence. VPS was removed for infection/malfunction in 18(60%) cases and eventually replaced in the same number of patients after CSF cell count dropped to 10 cells/mm^3. Post-operative complications led to mortality in 13(43.3%) patients. Significant association was found of larger pre-op FOC, post-op shunt removal, post-op CSF infection and shunt

Table-1: Demographics.

Variable	%/ Mean	Significance
Gender		
Male	60% (n=18)	P=0.384
Female	40% (n=12)	
Age in months		
Mean age	7.73 ± 1.41	P=0.151
Prior neurosurgical advice		
Yes	13.3% (n=4)	P=0.084
No	86.7% (n=26)	
Pre-op pressure sores		
Yes	63.3% (n=19)	P=0.067
No	36.7% (n=11)	
FOC in cm		
Mean	54.30 ± 3.08	P=0.001
Haemoglobin		
Mean	7.04 ± 0.58	P=0.106
Re-admission day		
Mean	41.83 ± 18.98	P=0.046
Duration of Hospital stay		
Mean	12.40 ± 8.36	P=0.630

FOC: Fronto-occipital circumference.

Table-2: Complications.

Variable	Present	Significance
Abdominal distension	26.7% (n=8)	P=0.709
Subdural collection	13.3% (n=4)	P=0.782
Cranial wound complication	30% (n=9)	P=0.394
CSF infection	40% (n=12)	P=0.036
Shunt Malfunction	40% (n=12)	P=0.885
Abdominal wound complication	23.3% (n=7)	P=0.417
Shunt removal	60% (n=18)	P=0.015
Shunt Revision	60% (n=18)	P=0.003
In-Hospital Mortality	43.3% (n=13)	P=0.036

CSF: Cerebrospinal fluid.

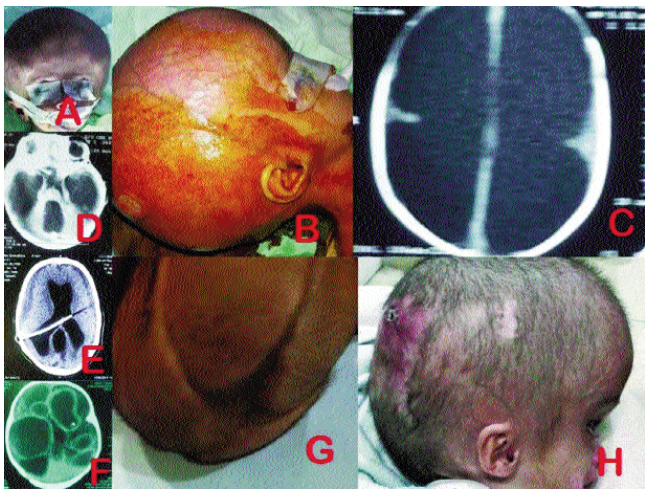

Figure: Scans and complications of Neglected Hydrocephalus. (A) Pre-op enlarged head with dilated veins and bulging fontanelle. (B) Pre-op pressure sore on bony prominence. (C) Computed tomography (CT)scan of head showing maximal hydrocephalus and thin cortical mantle. (D) CT Head showing gross cerebral atrophy with hydrocephalus. (E) Post-op CT Head showing shunt catheter in ventricles and bilateral chronic sub-dural haematomas (F) CT scan appearance of post-shunt infection ventriculitis. (G) Post-op sunken head and deformity. (H) Post-op cranial wound margin pressure sore.

revision to mortality ($p<0.05$ each).

Discussion

Despite innovations like endoscopic procedures, novel shunt designs and operative environment, Pudenzburr hole valve VPS placement remains the mainstay of paediatric hydrocephalus management in our setup. Worldwide and nationally, complication rates range between 25% and 40% within one year of insertion.[5] In our study population, the overall complication rates of shunting (mean follow-up of 6 months) were higher than usual, prompting us to investigate the prognostic factors related to poor outcome in neglected hydrocephalus.

Surgical intervention in these poor-risk patients come with reasons in our setup. First of all, parents have a big challenge in handling of such large head, co-morbid conditions of paraplegia, sphincter problems and actively ill, malnourished children who remain hospitalised most of the time on one account or other and have been guided by practitioners to pursue active treatment of neglected hydrocephalus. We also observed that once these patients got advice of VPS anywhere in the country, the parents pursued surgical intervention at our setup where free-of-cost treatment is being provided. Lack of education, quackery, socio-economic factors, religious taboos and living too far have largely contributed to late presentation. Before embarking on surgical intervention, detailed pre-op evaluation, including clinical assessment and investigations, should be carried out to minimise complications.

We analysed various factors for association with mortality using univariate analysis (Tables-1 & 2). Gender did not have any association with mortality. Age of infants ranged from 5-10 months but there was no significant impact on mortality ($p=0.151$). But young age has been reported as a risk factor for VPS complications, and in particular infectious complications.[6,7]

Pre-op FOC ranged from 49-60 cm and larger FOC had significant association with mortality ($p=0.001$), corresponding to report by Gather et al. that patients with FOC more than 60cm had only 10% chance of good outcome.[6] Fontanelle characteristics, head circumference at birth, and head growth velocity have been found to be good predictors for the need of shunt placement as quoted by Phillips BC et al.[8] Pre-op Hb ranged from 6.4 to 8.4 g/dl, mean 7.04g/dl which was optimised before surgery but we found no significant association of low Hb with complications and mortality ($p=0.106$). Pressure sores on bony prominences of parieto-occipital areas were present in 63.3% ranging from grade 1-3, which were thought to contribute to shunt/CSF infection but were found insignificant ($p=0.067$). All patients tolerated well the VPS procedure and made uneventful recovery from anaesthesia.

Follow-up visits have been a matter of non-compliance in our country and despite encouragement and facilitation we lost 13.3% of the patients to follow-up which may have resulted in a significant sampling bias and moreover they may be presumed to be either dead or had a good outcome. We found that most of the patients presented for follow-up only when they had developed some complication. Re-admission was frequent in post-op period with mean of 41.83 days correlating with shunt complications and onset of infection.[7] Re-admissions had significant association with mortality ($p=0.046$) but duration was not related to mortality. Re-admissions, intensive care, managing infections and VPS replacements in these patients are a costly affair and pose significant financial burden on strained economy of the country.

Subdural fluid collection was found on follow-up scans in 13.3% but only 6.6% required burr holes intervention. Cranial wound erythema/ pressure necrosis complications were seen in 30% which sometimes heralded CSF leak and put these patients at a very high risk of shunt infection.[9]

Shunt malfunction was observed in 40%, but in our group most of them happened before 3 months. The results vary

worldwide where the overall 30-day and 1-year failure rates for new shunts were 12.9% and28.8%, respectively, as reported by Al-tamim et al.[10] However, Radmanesh F et al. reported rates of shunt infection and shunt malfunction of 16.5% and 39.4%, respectively.[11] Parental response on shunt malfunction was studied and results showed, positive predictive value (PPV) of 41%, negative predictive value (NPV) of 79%, sensitivity of 83%, specificity of 34%, and accuracy of 52% for shunt failure.[12]

Post-operatively, CSF infection was documented in 40% and had significant association with mortality (p=0.036). While many factors like malnutrition, pressure sores, anaemia, comorbid conditions, uro-sepsis have contributed to shunt infections, but Kulkarni AV et al. found three variables associated with an increased incidence of shunt infection which can be prevented are, postoperative CSF leak, alternatives to placement of a CSF shunt in premature infants, and minimal manual contact with the shunt system.[13] Shunt infection usually occurs within a few months of shunt surgery and is associated with substantial risks of morbidity, including increased risk of seizure disorder and decreased intellectual performance.[14-16] Strategies to reduce this burden have been innovative like antibiotic impregnated ventricular catheter systems.[17] Hayashi T et al. first reported that irrigation alone, without topical antibiotics, contributed to the prophylaxis of shunt infection.[18] Antimicrobial suture has been used with some benefit by Rozzelle CJ et al.[19]

Abdominal complications were observed in 23.3% (not necessarily associated with peritonitis) and mostly heralded infective complications and route of entry for CSF infection. Some authors have published that leaving the distal end of a shunt catheter in place in a patient with acute peritonitis is a reasonably safe choice in specific patients, provided the source of infection is aggressively treated with systemic antibiotics and local debridement when necessary.[20]

VP shunt was removed due to infection or malfunction in 60% cases, and evidence favour that in cases with VP shunt infection it is essential to remove VP shunt and start systemic antibiotics.[21]

VP shunt was revised in 60% which had significant association with the in-hospital mortality. Shunt revision in our series was statistically higher than most series of early picked hydrocephalus operations.[22] Stone JJ reported that on average VPS patients had 2.66 revisions.[23] Various programmable shunts are available in the market to cater to low expansile force of maximal hydrocephalus patients, but no advantage was found in using a shunt system that is prohibitively expensive.[24]

Mortality on account of complications occurred in 43.3%. There was significant association with larger FOC, shunt removal, CSF infection and shunt revisions.

The limitations of our study included small sample size, 13.3% were lost to follow-up, and lack of long-term follow up. A prospective randomised controlled trial with extended follow-up is needed in the country to fully investigate the constraints and positive outcome predictors of VPS in paediatric hydrocephalus.

Conclusion

CSF shunting is the mainstay of treating hydrocephalus. Prognostic factors such as large FOC, pressure sores, infections and shunt revisions had direct impact on mortality rate. To resolve the dilemma of having VPS for neglected hydrocephalus, the clinical signs of disease progression have to be considered in association with CT scan findings of ventriculo-megaly for favourable outcome.

References

1. Kestle JRW, Garton HJL, Drake JM. Treatment of hydrocephaluswith shunts. In: Albright AL, Pollack IF, Adelson PD, eds. Principles and Practice of PediatricNeurosurgery. New York: Thieme, 1999; pp 75-90.
2. Crnich CJ, Safdar N, Maki DG. Infections associated withimplanted medical devices.In: Finch RG, Greenwood D, NorrbySR, eds. Antibiotic and Chemotherapy: Anti-Infective Agents and Their Use in Therapy.8th ed. London: Churchill Livingstone, 2003; pp 575-618.
3. Khattak A, HaiderA.Treatment of congenital Hydrocephalus. J Postgrad Med Inst. 2003; 17:11-3.
4. Khan I, Burhan M, Nadeem M. A reappraisal of contributing factors leading to shunt infection. J Ayub Med Coll Abottabad. 2009; 21:66-8.
5. Ali M, Aman R, Khan Z, Khan K.M, Siddique M, Khanzada K, et al.Frequency of causes of ventriculo-peritoneal shunt failure in hydrocephalus patients. J Postgrad Med Inst. 2011; 25:368-72.
6. Gathura E, Poenaru D, Bransford R, Albright AL.Outcomes of ventriculoperitoneal shunt insertion inSub-Saharan Africa. J Neurosurg Pediatr. 2010; 6:329-35.
7. Enger PO, Svendsen F, Wester K, CSF shunt infections in children:experiences from a population-based study. ActaNeurochir. 2003; 145:243-8.
8. Phillips BC, Gelsomino M, Pownall AL, Ocal E, Spencer HJ, O'Brien MS. Predictors of the need for cerebrospinal fluid diversion inpatients with myelomeningocele. J Neurosurg: Pediatrics. 2014; 4: 3470-4.
9. Jeelani NU, Kulkarni AV, Desilva P, Thompson DN, Hayward RD. Postoperative cerebrospinal fluid wound leakage s a predictor of shunt infection: a prospective analysis of205 cases. J NeurosurgPediatrics. 2009; 4:166-9.
10. Al-Tamimi YZ, Sinha P, Chumas PD, CrimminsD,Drake J, Kestle J, et al. Ventriculoperitoneal Shunt 30-Day Failure Rate:A Retrospective International Cohort Study. Neurosurgery. 2014; 74:29-34.
11. Radmanesh F, Nejat F, El Khashab M, Ghodsi SM, Ardebili HE. Shunt complications in children with myelomeningocele: effect of timing of shunt placement. J NeurosurgPediatrics.2009; 3:516-20.
12. Naftel RP, Tubergen E, Shannon CN, Gran KA, Vance EH, Oakes WJ,

et al. Parental recognition of shunt failure: a prospective single-institution study. J NeurosurgPediatrics.2012; 9:363-71.

13. Kulkarni AV, Drake JM, lamberti-pasculli M. Cerebrospinal fluid shunt infection: a prospective study ofrisk factors. J Neurosurg. 2001; 94:195-201.
14. Kontny U, Höfling B, Gutjahr P, Voth D, Schwarz M, Schmitt HJ. CSF shunt infections in children. Infection. 1993; 21:89-92.
15. Davis SE, Levy ML, McComb JG, Masri-Lavine L. Does age or other factors influence the incidence of ventriculoperitoneal shunt infections?Pediatr Neurosurg. 1999; 30:253-7.
16. Kulkarni AV, Riva -Cambrin J, ButlerJ,Browd SR, Drake JM, Holubkov R, et al. Outcomes of CSF shunting in children: comparison of Hydrocephalus Clinical Research Network cohort with historical controls. J Neurosurg Pediatr. 2013; 12:334-8.
17. Sciubba DM, Stuart M, Mcgirt MJ, Woodworth GF, Samdani A, Carson B, et al. Effect of antibiotic-impregnated shunt catheters in decreasing the incidence of shunt infection in the treatment of hydrocephalus. J NeurosurgPediatrics.2005; 103:131-6.
18. Hayashi T,Shirane R, Yokosawa M, Kimiwada T, TominagaT.Efficacy of intraoperative irrigation with saline forpreventing shunt infection. J NeurosurgPediatrics.2010; 6:273-6.
19. Rozzelle CJ, Leonardo J, Li V. Antimicrobial suture wound closure for cerebrospinal fluidshunt surgery: a prospective, double-blinded, randomizedcontrolled trial. J NeurosurgPediatrics.2008; 2:111-17.
20. Dalfino J, Adamo MA, Gandhi RH, Boulos AS, Waldman JB. Conservative management of ventriculoperitoneal shunts in the setting of abdominal and pelvic infections. J NeurosurgPediatrics.2012; 9:69-72.
21. Siraj M, Iltaf S, Saeed R. Management of ventriculo-peritoneal shunt infections. J Surg Pak. 2011; 16:114-7.
22. Notarianni C, Vannemreddy P, Caldito G, Bollam P, Wylen E, Willis B, et al. Congenital hydrocephalus and ventriculoperitoneal shunts: influence of etiology and programmable shunts on revisions. J Neurosurg Pediatrics. 2009; 4: 547-52.
23. Stone JJ, Walker CT, Jacobson M, Phillips V, Silberstein HJ. Revision rate of pediatricventriculoperitoneal shunts after 15years. J NeurosurgPediatrics. 2013; 11:15-9.
24. Warf BC. Comparison of 1-year outcomes for the Chhabraand Codman-Hakim Micro Precision shunt systems in Uganda: a prospective study in 195 children. J Neurosurg. 2005; 102:358-62.