ORIGINAL ARTICLE

Dandy-Walker Malformation: A Clinical and Surgical Outcome Analysis

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ABSTRACT

Objective: To determine the clinical presentations, complications and mortality in patients with Dandy-Walker Malformation (DWM) after surgery i.e., shunt with y-connector.

Study Design: Case series.

Place and Duration of Study: Neurosurgery Ward, JPMC, Karachi, from January 2009 to December 2013.

Methodology: Cases of DWM, with associated hydrocephalus, further confirmed on CT scan of brain, were admitted through OPD. Those who were previously operated, those with other associated co-morbid or anomalies were excluded from this study. Combined drainage of the ventricular system and posterior fossa cyst, via dual shunt i.e. cystoperitoneal and ventriculoperitoneal shunt with y-connector was performed in all patients. Complications and mortality after surgical intervention in these patients were noted upto one month after surgery. The data analysis for descriptive statistics was done on SPSS version 20.

Results: In this study of 70 patients, majority of the patients were female aged between 1 - 2 years. Hydrocephalus was the predominant symptom as being present in all patients, followed by cerebellar signs in 60 (86%), and other in 5 (7.14%) patients. Complications of surgery were infection and shunt fracture dislocation in 7 (10%) each, malpositioning and shunt blockage in 6 (8.5%) each within one month of surgery, intracranial haemorrhage in 5 (7.14%) patients. Only one patient (1.42%) expired after surgical intervention.

Conclusion: In DWM, the commonest presentation is that of hydrocephalus. Shunt malfunction and infection are the commonest complication after shunting. Dual shunt with y-connector has the lowest mortality when compared with other methods for treatment of shunt with y-connector.

Key Words: *Dandy-Walker malformation. Y-connector. Hydrocephalus. Shunt infection. Shunt malposition. Shunt block. Shunt fracture-dislocation. Ventriceal-peritoneal shunt. Cystoperitoneal shunt.*

INTRODUCTION

Dandy-Walker Malformation (DWM), first described by Dandy and Blackfan, is a congenital disorder that involves the cerebellum and the fourth ventricle. It is characterized by a triad of enlarged posterior fossa with elevation of the tentorium/torcula, cystic dilatation of the fourth ventricle and cerebellar vermis agenesis. A large number of concomitant problems including those of brain and other than brain may be present, but DW is recognized by the above three features. Hydrocephalus may be defined as a complication rather than part of this syndrome. In addition to complex pathogenesis with possible genetic and environmental antigenic etiologies, placental and umbilical cord abnormality may also be related to its development.[1] Clinical presentation was most often heralded by symptoms and signs of hydrocephalus with focal neurological findings being a less prominent feature.[2]

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Regarding treatment, there is still controversy over the optimum surgical management. Initially Surgery of the posterior fossa with membrane excision was the preferred method of treatment. Unfortunately, there was a high rate of complications, and many of the patients treated in this way still needed a shunting system.[3] Other options include ventriculo-peritoneal shunting, combined cyst and ventricular shunting, shunting with y-connector, endoscopic third ventriculostomy,[4] Ultra-sound-guided puncture via the lateral and III ventricles,[5] and ventricular fenestration. Therefore, although, surgical treatment remains controversial, prognosis varies greatly according to the severity of syndrome and associated co-morbidities.

In Pakistan, there is diversity of treatment options utilized at different centres. Therefore, this study aimed to determine the complications and mortality associated with shunt with y-connector at the study centre.

METHODOLOGY

In this case series, 70 consecutive cases of DWM were included who were treated in neurosurgery ward, Jinnah Postgraduate Medical Centre, Karachi, from January 2009 to December 20013. These patients with signs and symptoms suggestive of DWM with associated hydrocephalus and further confirmed on CT scan brain

(plain) were admitted through OPD. Those who were previously operated other associated co-morbid or anomalies were excluded from this study. Combined drainage of the ventricular system and posterior fossa cyst, via dual shunt i.e. cystoperitoneal (CP) and ventriculoperitoneal (VP) shunt with y-connector was performed in all patients. Complications and mortality after surgical intervention in these patients were noted upto one month after surgery.

The data was collected in a proforma and analyzed on SPSS version 20. Frequencies and percentages were presented for categorical variables while mean and standard deviation were presented for numerical variables.

RESULTS

This study included a total 70 patients. Majority of the patients were female and aged 2 years ± 1 SD. Hydrocephalus was the predominant symptom present in all patients, followed by cerebellar signs in 60 (86%) and other signs in 5 (7.14%) patients. The latter include delay in reaching milestones and abnormal gait (one patient) each, and psychiatric disorders (in 3 patients). Complications which were observed were shunt fracturedislocation, and infection in 7 (10%) each malpositioning, and shunt blockage in 6 (8.5%) patients each within one month of surgery and intracranial haemorrhage in 5 (7.14%). Only one (1.42%) patient expired after surgical intervention (Table I).

Table I: Complications after placement of shunt with y-connector (n=70).

Malpositioning	Shunt: fracture/ dislocation	Intracranial haemorrhage	Infection	Shunt blockage	Mortality
6 (8.57%)	7 (10%)	5 (7.14%)	7 (10%)	6 (8.5%)	1 (1.42%)

DISCUSSION

DWM is a rare disease. In addition to complex pathogenesis with possible genetic and environmental antigenic etiologies, placental and umbilical cord abnormality may also be related to its development. Hydrocephalus is a resulting complication, rather than part of triad that constitute this syndrome, and the predominant presenting symptom.[6] This study found hydrocephalus in all the patients. Other focal neurological symptoms due to cerebellar vermis agenesis are usually ignored by the patients as reflected in this study. However, these patients may also present with other non-CNS deformities.[7,8]

Cerebellum is believed to play a role in motor control, motor learning, and even cognition such as development of language and other cognitive skills. Thus, the effect of this disorder on intellectual development is variable, with some children having normal cognition and others never achieving normal intellectual development.[9] More severe intellectual impairment has been observed in patients with agenesis of the corpus callosum.[10] This study showed that in small group of patients reasons for seeking medical advise were psychogenic changes, irritability, delay in reaching milestones, and abnormal gait. These symptoms are usually ignored by the parents until these hampered the daily activities or studies.

Widely diverging opinions on the optimal therapy for intracranial cerebrospinal fluid cysts, mainly arachnoid cysts and the DWM, exist. Membrane excision was initially proposed by Dandy for the treatment of DWM. The results using this form of therapy have been poor initially, with a high failure rate. This therapy was based on an assumption that the blocked outlets of the fourth ventricle were responsible for expansion of the fourth ventricle and supratentorial hydrocephalus.[11] This method, has fallen into disrepute because of a high mortality rate of 10% and a failure rate of 70%.[12] Udvarhelyi and Epstein still advocate this treatment by way of cyst wall excision in children aged over 3 years.[13] This was then replaced by shunting due to high morbidity and mortality associated with excision.[14] Transtentorial herniation of cyst or enlargement has been noted following placement of ventricular shunt alone, which is associated with high mortality. Therefore, Kumar *et al*. advocated shunting both of cyst and ventricles.[15] The present experience with this procedure showed a very low mortality rate (1.42%).

Malfunction and infection were the commonest complications associated with use of shunt systems. Other complications included headaches and focal cranial nerve deficits following infection caused by a cystoperitoneal shunt due to tethering of the brainstem.[10] Chronic cerebral herniation and central brain herniation have also been noticed as a complication.[16] A single-catheter shunting system effectively drained the supratentorial and infratentorial compartments has been found to be associated with reduced complexity and potential risk associated with the combined shunting systems required by so many with DWM-related hydrocephalus.[17] The present results matched with other studies in a way that shunt malfunction and infection being the predominant complications in these patients with dual shunt with y-connector leading to a single channel for drainage. Endoscopic third ventriculostomy may be considered an acceptable alternative, especially in older children, with the aim to reduce the shunt-related problems.[18]

Endoscopic methods of transaqueductal placement of a single-catheter cyst-ventriculoperitoneal shunt has also been noted to be with good results. The introduction of endoscopic and stereotactic techniques has expanded the available treatment possibilities for posterior fossa cysts.[19] Successful endoscopic third ventriculostomy resulted in a slight decrease in ventricle size and varying degrees of reduction in cyst size.[20] The risk of failure increases with intracerebral infection likely because of obliteration of cerebrospinal fluid pathways. Central

brain herniation is noted in these patients.[14,21] It is also noted that failure rates are reported to be particularly high in the first year of age in patients of third ventriculostomy.[22,23]

Dual shunt with y-connector has been shown to be associated with low mortality when compared with other methods when mortality as high as 18% is seen.[24] A reduction in ventricle size noted on postoperative images occurred more frequently in patients with a VP shunt, whereas a reduction in cyst size was more appreciable in patients with a CP shunt. Use of single draining channel leads to less complications in terms of infection and shunt related system if dual channel is used for drainage. This study thus showed that double shunting in the form of y-connector should be a better options in patients with DWM.

CONCLUSION

DWM is a syndrome which although characterized by a triad, commonly present with signs and symptoms of hydrocephalus taken as a complication of this syndrome rather than part of the triad. Symptoms due to hydrocephalus are the most common presentation for referral to hospitals. Malfunction and shunt infection are recognized as the commonest complication after shunting. Dual shunt with y-connector has the lowest mortality when compared with other methods for treatment of shunt with y-connector. Thus, it can be considered as a good option in such patients with DWM.

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