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Review article

Contemporary management and surveillance strategy after shunt or endoscopic third ventriculostomy procedures for hydrocephalus

M. Burhan Janjua *, Caitlin E. Hoffman, Mark M. Souweidane

Department of Neurological Surgery, Weill Cornell Medical College, New York Presbyterian Hospital, 525 East, 68th Street, New York, NY 10065, United States

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ABSTRACT

The management of hydrocephalus can be challenging even in expert hands. Due to acute presentation, recurrence, accompanying complications, the need for urgent diagnosis; a robust management plan is an absolute necessity. We devised a novel time efficient surveillance strategy during emergency, and clinic follow up settings which has never been described in the literature. We searched all articles embracing management/surveillance protocol on pediatric hydrocephalus utilizing the terms "hydrocephalus follow up" or "surveillance protocol after hydrocephalus treatment". The authors present their own strategy based on vast experience in the hydrocephalus management at a single institution. The need for the diagnostic laboratory testing, age and presentation based radiological imaging, significance of neuro-opthalmological exam, and when to consider the emergent exploration have been discussed in detail. Moreover, a definitive triaging strategy has been described with the help of flow chart diagrams for clinicians, and the neurosurgeons in practice. The triage starts from detail history, physical exam, necessary labs, radiological imaging depending on the presentation, and the age of the child. A quick head CT scan helps after shunt surgery while, a FAST sequence MRI scan (fsMRI) is important in post ETV patients. The need for neuro-opthalmological exam, and the shunt series stays vital in asymptomatic patients during regular follow up.

1. Introduction

Hydrocephalus is a commonly encountered clinical diagnoses in neurosurgical practice across the age spectrum. This condition often arises as a result of a blockage in cerebrospinal fluid (CSF) flow within the intracranial compartments. The immediate diagnosis is primarily clinical, with or without the enlargement of cerebral ventricles usually associated with the clinical sequelae from an increase in intracranial pressure (ICP). Hydrocephalus is attributable to a myriad of causes ranging from congenital, perinatal insults, acquired causes of tumor, infection, traumatic and non traumatic hemorrhage, iatrogenic, and/or idiopathic causes, and rarely CSF overproduction. The overall incidence is estimated at 0.9–1.8/1000 births (reported range of 0.2–3.5/1000 births) [1]. The subsequent reported increase in incidence is thought to be due to increase survival of preterm infants.

The clinical manifestation depends on the etiology and the dynamics involved. Acute hydrocephalus is a life-threatening pathology which demands an urgent neurosurgical intervention. The acute increase in ICP can be catastrophic due to risk of transtentorial herniation, and/or herniation of the cerebellum into the foramen magnum. In contrast, slowly progressing hydrocephalus often manifests with the non-specific symptoms of headaches, nausea, vomiting, excessive lethargy, and sleepiness as well as neuro-ophthalmological complications of decrease in visual acuity are often attributed to increase in ICP [2]. Therefore, the presentation depends on the acuity of the disease ranging from decrease in psychomotor activity, altered sensorium, and progressive cerebral dysfunction. A detail history is prudent which often entails altered behavior as an early sign of pediatric hydrocephalus. A common neuro-opthalmological exam finding is papilledema, which is caused by optic nerve ischemia, traction, or transsynaptic neuronal degeneration. In some cases, papilledema can be the first manifestation because of close association of the ventricular system and visual pathway [3].

The acute/subacute presentation of noncompliant versus compliant hydrocephalus is still a challenging issue. Nevertheless, radiological findings can be clueless and/or misleading. However, the clinical examination stays vital in these cases. A detail neuro-opthalmological exam can also be helpful if, not limited by the optic atrophy. To avoid this near miss there should be a low threshold for an urgent exploration surgery to rule out and/or treat the primary pathology. The current modalities in practice for the

* Corresponding author.
E-mail address: burhan167@yahoo.com (M.B. Janjua).

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diagnosis and treatment of hydrocephalus are highly dependent on the realm of the clinical picture and radiological findings. Traditionally MRI of CT scan has been used as an imaging modality of choice depending on age and mode of presentation. The preferable treatment option is CSF diversion to the circulatory system or the peritoneum, or endoscopic third ventriculostomy.

Since complications and recurrence of hydrocephalus is not uncommon, there is a desperate need for a treatment algorithm entailing a quick diagnosis, and triaging strategy for these patients during emergency or follow up scheduled visit. Additionally, literature on long term surveillance is tenuous. An anecdotal evidence from day to day practice is highly supportive of this treatment strategy. Another important aspect of hydrocephalus treatment is an overwhelming parental concern for prompt diagnosis and treatment. A detail history of the child including birth history and a through neurological exam is vital. A careful examination of the skin along the distal tubing is also helpful to isolate the cause in shunted hydrocephalus patients. Therefore, a focused algorithm would be much more helpful for the patients as well as for the parents in these challenging circumstances.

The main objective of this study is to define and establish a treatment paradigm for all hydrocephalus patients after shunts or endoscopic third ventriculostomy procedures especially in an emergency and follow up clinic settings.

2. Methods and results

We conducted a literature review to find any previously published studies with a primary focus on the surveillance strategies or an established protocol laid for all patients who were treated for hydrocephalus. We used MEDLINE literature search utilizing the terms "hydrocephalus follow up" or "surveillance protocol after hydrocephalus treatment". These patients underwent ventriculoperitoneal shunt or an endoscopic third ventriculostomy procedure for treatment of hydrocephalus. For an immediate and/or during long term postoperative follow up period, no definitive study highlighting hydrocephalus surveillance strategy and/or a treatment protocol was identified. Numerous studies discussed the importance of currently in use imaging modalities and their efficacy after shunting or endoscopic third ventriculostomy procedures in treated cases of hydrocephalus.

The authors have tried to summarize their results with a treatment algorithm which include, the presentation, diagnosis, laboratory testing, imaging of choice, and the need for a through neuroopthalological exam. Furthermore, the treatment options have been separately discussed in emergency setting, during immediate postoperative, and long term follow up period after shunts or ETV procedures particularly, when presenting in extremis. These flow chart diagrams (Figs. 1–4) are self elaborative, and are equally helpful for a quick overview.

3. Discussion

A thorough understanding of cause and effect relationship of hydrocephalus to cerebral dysfunction is prudent in making treatment decisions. Unfortunately, this consideration is often lacking. Therefore, treatment decision must be made on prediction of risk involved. Whether shunts or neuroendoscopic CSF diversion, these treatment modalities are based on presentation, inherent complexity of case, and the expertise available on hand. Therefore, authors stress upon the understanding of the pathophysiology by studying the CSF hydrodynamics. Acute increase in ICP in the setting of blockage of CSF flow dictates the need of an urgent CSF diversion. There should be no hesitation to move on to a simple exploration as only a reliable investigation. Slowly progressive increase in ICP can be often elusive in the clinical picture. When CSF production exceeds absorption, ICP rises, but CSF production deceases as ICP rises to high levels, and a state of compensation ensues through transventricular absorption of CSF. This state of compensated hydrocephalus is observed in patients with a longstanding nonfunctional shunts and is not necessarily permanent due to parlous nature of this partially attainted balance [4]. Physicians must be alert and aware of an insidious subclinical progression with or without any preceding history of any traumatic event. A close communication is recommended to the patient or family regarding the expected outcome from treatment or continued observation. Moreover, physician's and patient/family's definition of treatment success must be correlated.

Apart from high pressure headaches, low pressure headaches are equally disabling. Slit ventricle syndrome (SVS) is such an entity with a triad of intermittent headaches accompanying findings of a slowly refilling shunt reservoir on physical examination with "slit" ventricles on radiographic examination [5–7]. These patient have a combination of intermittent affliction of intracranial hypo and hypertension, attributable to the siphoning of CSF through the ventricular shunt system resulting in coaptation of ventricular walls or slit ventricles. Baskin et al. described a treatment algorithm for these patients where an intracranial ICP fiberoptic electrode assisted monitoring is performed to observe the ICP for next 24–48 h. If asymptomatic, then shunt can be removed. However, an ETV can be performed in case of raised ICP. In case of failure of ETV, a new VP shunt is suggested. In the presence of persistence of disabling symptoms a pharmacologic treatment along with close neurosurgical follow up is recommended [8].

In the realm of hydrocephalus management, ventriculoperitoneal (VP) shunts are still considered as a first line treatment modality for hydrocephalus. This procedure is an effective long-term solution, common complications include an immediate shunt infection, proximal or distal catheter shunt malfunction and/or abdominal pseudocyst formation. Asymmetric or inappropriate ventricular size changes could also ensue after VP shunt, suggesting a need for regular evaluation of shunt function [9]. Additionally, an immediate subdural hygroma (25% of adults, 6% of children) can also be related to the shunt surgery [10]. Endoscopic third ventriculostomy (ETV), is now considered as an alternate and effective approach where third ventricle is cannulated with a rigid endoscope to create an opening in the floor of third ventricle in front of the mammillary bodies. This internal CSF diversion has become an important alternative to CSF shunting for children with hydrocephalus [11]. This procedure has some specific indications, improved outcome, and some limitations. The indications for an EVT procedure are still debatable in infantile periods with uncertainty in the predictors of late failure. Even with good postoperative outcome, late failure can occur, and may lead to rapid deterioration. Additionally, poor penetration through the membrane of liliequist during the initial procedure, anomalous anatomy, and/or altered CSF hydrodynamics can result in the closure of the an ETV stoma. The utility and safety of an ETV has been proven for obstructive hydrocephalus or triventricular hydrocephalus that occur secondary to aqueductal stenosis. For other cases, however, it is necessary to examine the anatomy, etiology, as well as the patient's age before considering this as a definitive treatment option. Canadian pediatric neurosurgery group developed a model to predict the probability of ETV success in treatment of hydrocephalus based on child's individual characteristics. They concluded age of the child was by far the strongest predictor. Infants less than 6 months of age had the lowest predicted ETV success, with progressively higher success observed as a child ages. The effect of a previous CSF shunt appeared to be less in magnitude than age in prediction of ETV success [12]. Although, there is no

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