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Short Communication

Some Recent Advances in our Understanding of Hydrocephalus and Further Developments in Shunt Technology with Improved Outcomes

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INTRODUCTION

During the past 20 years since this article was published there have been some fresh theories into the potential causes, classification and effects of hydrocephalus, with more clinical research now focused on methods to reduce the complications of CSF shunts and avoid shunts altogether whenever possible. Some studies have demonstrated encouraging improvements in long-term outcomes and quality of life in both children and adults with hydrocephalus.

However, in a systematic search for prospective multicenter randomized controlled trial studies done in pediatric hydrocephalus, Kestle et al. identified only 10 such studies between 1990 and 2019 (1). These studies included 3740 children from 189 centers and addressed questions related to the management of premature children with intraventricular hemorrhage, compared different shunt valves, assessed surgical techniques of shunt placement, compared endoscopic and shunt treatments and in one study compared the incidence of hydrocephalus after fetal or postnatal closure of myelomeningocele. Only two of these studies found a significant difference in the primary outcome, the other 8 found no difference between treatment arms. The positive results were found in the DRIFT (2) and MOMS (3) studies. Both of these RCT studies involved hydrocephalus in patients at the lowest end of the age spectrum: premature babies with post-hemorrhagic hydrocephalus (PHH) and babies with spina bifida and hydrocephalus operated in utero at 20-25 weeks gestation.

Early irrigation of the ventricles after premature-associated hemorrhage to remove damaging blood products and cytokines from the CSF, a technique labelled DRIFT (Drainage, Irrigation and Fibrinolytic Therapy), produced a significantly lower incidence of death or severe cognitive disability in the treatment group at 2 years and this benefit persisted at 10-year follow up (2,4). The technique involves local anesthetic insertion of ventricular catheters and continuous lavage with artificial CSF for 2-3 days. This is time consuming so a more rapid neuroendoscopic lavage technique has been developed to try to achieve similar improvements in outcome.

Similarly, fetal surgery for myelomeningocele has benefitted patients by reducing the need for hydrocephalus shunt placement compared with postnatal closure of spina bifida lesions(3). However, fetuses with severe ventriculomegaly do not enjoy this benefit (5).

Recent studies into CSF dynamics in hydrocephalus have resulted in a challenge to the old simple classification of hydrocephalus into obstructive and communicating types. In 2011 Rekate proposed a new classification of hydrocephalus which was reached by consensus after meetings of interested clinicians and basic scientists (6). In this classification there is a point of obstruction in all cases of hydrocephalus apart from the rare CSF-producing choroid plexus tumor (Papilloma/Carcinoma).

Obstruction between the spinal and cortical subarachnoid spaces (SSAS & CSAS) is rarely diagnosed but is probably the cause of many or most cases of normal pressure hydrocephalus (NPH). Blockage between the SSAS and CSAS results from either subarachnoid hemorrhage or infection. It frequently involves the area around the brainstem selectively, and this form of hydrocephalus can be amenable to endoscopic third ventriculostomy. Another potential point of obstruction is the venous sinuses which causes hydrocephalus in babies or in adults post-craniectomy. However it causes pseudotumor cerebri with a fixed-volume skull such as in adults without a history of craniectomy. If the skull is not of a fixed volume and the ICP is in communication with atmospheric pressure as in small babies the ICP cannot go above the atmospheric pressure, and the patient develops hydrocephalus.

A recent meta-analysis of the results of CSF shunt insertion in 2461 patients over 60 years old with a diagnosis of NPH showed an overall 75% rate of improvement in gait and mobility, together with a greater than 60% improvement in cognition and 55% improvement of continence (7). These initial gains decreased by 5% during the first 3 years of follow-up and adjustable valves were associated with a reduction in revisions (12% vs 32%) and subdural collections (9% vs 22%), compared with less expensive fixed pressure valves.

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Another recent large retrospective cohort study of the timing of treatment of post-traumatic hydrocephalus showed a clear benefit from early detection and shunting of patients who present with progressive low-pressure hydrocephalus after severe head injury, in terms of shortening their post-traumatic amnesic time during rehabilitation and a sustained improvement in their functional outcome (8).

Finally, a useful development in shunt technology has been the more widespread impregnation of shunt catheters with antibiotics which leach out from the catheter wall killing surrounding gram-positive bacteria during the first two weeks following shunt implantation. The efficacy of these catheters was tested recently in the BASICS trial which was a multi-centre, single-blind, randomised controlled trial. It included patients with hydrocephalus of any aetiology undergoing insertion of their first ventriculo-peritoneal shunt irrespective of age in neurosurgery centres in the UK and Ireland (9). Patients were randomly assigned to receive standard shunts, antibiotic-impregnated shunts or silver-impregnated shunts. The sample size was 3505 patients with a follow-up duration for 6-24 months. The shunt revision rate due to infection was significantly lower (2%) in the antibiotic-impregnated group compared to the other two groups (6% each). Although the benefit might seem modest the study does support the routine adoption of antibiotic catheters in patients who are having their first ventriculo-peritoneal shunt insertion and this practice will benefit patients with hydrocephalus by reducing risk and harm from shunt infection.

CONCLUSION

New theories of the CSF mechanisms underlying hydrocephalus, together with broader classifications of the different types of hydrocephalus have increased the potential for neurosurgeons to target treatments more effectively and hopefully improve long-term outcomes for patients of all ages. Shunt technology continues to develop in ever more sophisticated ways and this progress will help to reduce complication rates and improve outcomes in patients as they live longer with their implanted CSF shunts. A similar commentary on this article after a further 20 years might ideally focus on novel medical methods of precisely modulating CSF production from the choroid plexus, rather than just small incremental improvements in the mechanics and complications of CSF shunts.

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