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Transitional care in pediatric neurosurgical patients

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ABSTRACT

The relatively young specialty field of pediatric neurosurgery addresses a number of surgical diseases that, while first encountered in children, may involve long-term post-operative sequelae that persist into adulthood. These diagnoses present a challenge for care providers as patients transition from pediatric to adult age groups. Brain tumors, shunted hydrocephalus, and myelomeningocele are three of the most common examples of this interesting category. The provision of coordinated transitional care to affected pediatric neurosurgical patients is made all the more difficult by the common comorbidity of developmental delay, affecting not only personal and social growth but also the character and composition of the care team. This article seeks to provide a background for some of the common pediatric neurosurgical diseases requiring a transitional care framework for survivors entering young adulthood, a summary of the adult surgical care challenges faced by these patients, and a rationale for different approaches to optimize care.

Introduction

Pediatric neurosurgeons treat a wide array of diseases from infancy to adulthood. Some, principally related to trauma care, require one or a few major interventions but may not require long-term management. Other conditions, such as congenital spinal deformities or central nervous system tumors, require not only complex initial interventions but also ongoing follow-up and monitoring for deleterious sequelae of operative intervention, subsequent oncologic therapy, or natural disease progression. As pediatric neurosurgery is a relatively new and very small field, there is a shortage of providers in the United States trained to provide such long-term care to patients.[1,2] In addition, due to advances in pediatric neuro-oncology and other specialty treatments, pediatric neurosurgical patients are experiencing progressively longer survivals and more frequently require neurosurgical care into adulthood. The mechanisms for provision of such treatment, and for reimbursement of coordination of care, have not yet been established. However, general models for provision of seamless transitional care have been proposed for patients entering young adulthood, and it is clear that a number of childhood neurosurgical diseases require such extended monitoring and treatment.

Shunted hydrocephalus

The improvement in early diagnosis, together with the introduction of valved shunt systems, new operative techniques, and supportive care of patients with hydrocephalus—especially in neonates with intraventricular hemorrhage of prematurity—has markedly improved long-term survival for children requiring cerebrospinal fluid (CSF) diversion. Along with survival, however, has come a greater appreciation for the long-term needs of patients with ventricular shunts, both in terms of ongoing neurosurgical interventions (shunt revisions) and in the diagnosis and treatment of long-term cognitive and motor deficits found in this group of patients.

The immediate and short-term complications of ventriculoperitoneal shunt placement—infection, overdrainage, mechanical obstruction, and disconnection—are well-documented[3] but only recently, researchers have reported outcomes after longer follow-up periods. Shunt survival (and thus need for revision) appears to be related to several factors, including age at first shunt placement, hydrocephalus etiology (tumor vs. congenital vs. other), time period to first revision, and others. Vinchon et al.[4] recently reported a series of 456 patients shunted in childhood with a median 23-year follow-up, in which 18% required at least one shunt revision after 20 years of age. Interestingly, 5% of the study population underwent a *first* revision after 20 years of age. Overall mortality at 20 years of age was 18%, with several additional

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survivors ultimately dying of shunt-related causes in their 20s. The authors produce evidence of the need for ongoing vigilance for shunt complications even decades after placement and found that only a small percentage of patients became truly shunt-independent. In addition, early optimism for the long-term patency of endoscopic third ventriculostomies has been tempered by reports of late and sudden failures requiring urgent re-operation and resulting in nearly universal mortalities.[5] These patients, just as those with ventricular shunts, thus are also in need of long-term follow-up.

On the other hand, a small but significant proportion of patients in this group may become shunt-independent. These patients are usually detected when their shunts fail and are yet asymptomatic and thus do not require revision.[6,7] It appears that patients with post-hemorrhagic hydrocephalus and hydrocephalus resulting from complete brain tumor resection have a higher likelihood of reaching shunt independence than patients with other hydrocephalus etiologies. Further work in this area is needed to help define appropriate clinical and imagining protocols for follow-up.

Importantly, measures of long-term functional and social outcomes are lacking in the literature, although a few reports have begun to provide initial information. In the report by Vinchon et al.,[4] only 18% of shunted patients were free of any functional sequelae in long-term follow-up. The remainder suffered from motor, cognitive, or behavioral deficits; epilepsy; vision loss; chronic pain; endocrine disorders; or obesity, with a frequency ranging from 9% to 48%. The worst outcomes were found in patients shunted from birth, and those treated in earlier decades of the study (1980s and 1990s). Other reports have indicated similar levels of long-term functional impairment. Given the potential severity and breadth of functional disorders detected, the need for long-term multidisciplinary follow-up is apparent.

Central nervous system tumors

Central nervous system (CNS) tumors are the most common solid tumors of childhood, with nearly 4500 patients newly diagnosed annually in the United States.[8] Common pediatric CNS tumors include astrocytoma (low- and high-grade gliomas), medulloblastoma, ependymoma, and other primitive neuroectodermal tumors. These lesions are increasingly treated in a multimodality fashion, by multidisciplinary teams.[9,10] Although cure rates have increased dramatically in the past two decades with advances in surgical technique and adjuvant chemotherapy and radiation therapy, both recurrence and late treatment effects present challenges to care providers as surviving patients enter young adulthood.

Treatment varies for each CNS tumor type and risk profile but often includes chemotherapeutic agents with potentially significant toxic side effects, and in some cases intensive radiation therapy, with its concomitant risks of local tissue damage, secondary malignancies, and acquired vascular abnormalities. Secondary effects include pituitary/hypothalamic insufficiency, hypofertility or infertility, growth and cognitive impairment, and spinal problems secondary to radiation therapy.[10–12] The degree to which such morbidities may require specific neurosurgical intervention in adult years is not known, but neurosurgeons are certain to encounter patients with other side effects and are thus positioned to help triage care in such cases.

Improved risk-stratification, targeted molecular therapy, and selective reduction in radiation therapy may limit or mitigate some of the secondary effects of cancer therapy, but long-term follow-up will always be required in patients undergoing treatment of pediatric CNS malignancies. In addition, as new, targeted therapies are introduced as alternatives to standard chemotherapy and radiation therapy, care providers will need to become familiar with a wholly new profile of treatment side effects, such as rash, fatigue, and pneumonitis in the case of epidermal growth factor inhibitors and hemorrhage, hypotension, and proteinuria in the case of vascular endothelial growth factor inhibitors.[10,12]

Much of the follow-up for CNS tumor survivors will be managed by neuro-oncologists and informed primary care physicians, but neurosurgeons will play a key role in providing education about long-term consequences of therapy and in neurosurgical interventions, including re-operation for tumor recurrence, management of CSF diversion, and, rarely, spine surgery for cervical instability acquired after radiation therapy.

Myelomeningocele

The clinical implications of myelomeningocele, or spina bifida aperta, have changed over the past two decades. At one end of the spectrum there is a decrease in incidence due to the recognition of the importance of antenatal folic acid supplementation in women, as well as enhanced technology that allows for earlier detection that may be followed by parental decisions to electively terminate the pregnancy. On the other is a rising prevalence, as patient survival has increased, due to greatly improved supportive management.[13] The value of a multidisciplinary approach is perhaps nowhere more apparent than in this condition, with coordinated care provided by urologists, orthopedists, neurosurgeons, physical and occupational therapists, specialized nurses, and pediatricians collaborating to provide care for these medically complex patients. Data quantifying precisely the efficacy and cost-effectiveness of such approaches are limited, but at least one study found that patients who lost access to a dedicated clinic practice suffered untoward adverse effects, including higher rates of renal failure and amputation.[14]

Shunt placement for hydrocephalus associated with myelomeningocele has generally been credited with improved early outcomes. However, long-term morbidity and mortality in patients who remain shunt-dependent may be paradoxically increased due to shunt-specific complications.[15] Other predictors of survival and improved mobility include the use of clean intermittent bladder catheterization and operative correction of secondary spinal cord tethering. In addition to a high frequency of orthopedic conditions (e.g., scoliosis, kyphosis, joint contracture, club foot, and hip subluxation), patients with myelomeningocele also are frequently found to have Chiari II malformations and syringomyelia, which have unique clinical manifestations in the myelomeningocele population.[16] These lesions require vigilance in both the initial assessment and long-term monitoring.

Two major longitudinal studies of myelomeningocele survivors—from Cambridge, England, and Chicago—suggested that cognitive outcomes were strongly related to maintenance of shunt function.[13,17] Several studies have attempted to measure the cost of disrupted care in patients with myelomeningocele, assessing potentially preventable complications, such as urinary tract sepsis, renal calculi, pressure sores, osteomyelitis, shunt malfunction, secondary spinal cord tethering, gait deterioration, and others.[18,19] Adults with myelomeningocele also have a high rate of severe obesity, which results in a distinct subset of long-term health and functional challenges.[20]

Other pediatric neurosurgical diseases

The list of diseases cared for by pediatric neurosurgeons is long, and a complete review of any process that may require long-term surgical intervention is beyond the scope of this article. Laws