Posterior fossa syndrome

MARK M. SOUWEIDANE, M.D.
Weill-Cornell Medical College, New York, New York

Since the recognition 30 years ago that surgery on children with posterior fossa tumors can result in postoperative mutism, the features surrounding this behavioral and functional “syndrome” have gained much attention, but sadly, disproportionately less elucidation. The clinical spectrum is varied and one needs to look no further than the skillful description provided 20 years prior by Dr. Robin Humphreys to identify the afflicted child.

The child awoke immediately after operation and conversed appropriately. The next day he became increasingly irritable and refused to talk. He layed curled on his side, followed commands, refused to open his eyes. At 6 weeks postoperative he was alert, understood most conversation but did not engage in verbal speech. By 4 months he was able to verbalize with only ‘yes’, ‘no’ and ‘hi’. By 6 months he was capable of carrying on a conversation.

The reported incidence varies widely, but in this most recent publication by a highly skilled team of neuro- oncology specialists, the incidence of posterior fossa syndrome (PFS) reaches nearly 40%. Perplexing is why the frequency of PFS continues to increase during an era in which we have witnessed substantial reductions in overall surgical morbidity. The most likely explanation for this divergence is the expanding definitional criteria and wider recognition of this neurological and neurobehavioral syndrome. Although one may be tempted to challenge this most recent report as inflating the true incidence of PFS due to more relaxed inclusion criterion, the experience of this highly regarded group should serve as cautionary note: removing tumors from the cerebellum in developing children is far from risk free.

What has not yet come to light is how one predicts which patients are at risk for developing PFS and how to prevent this phenomenon. This current report correlates MR-based cerebellar atrophy at 1 year after surgery with PFS. Review of an MR image 1 year after surgery contributes little help regarding prevention. From the surgeon’s perspective, avoiding the syndrome is problematic. What is currently known is that surgical intervention for tumors that infiltrate the vermis, brainstem, deep cerebellar nuclei, cerebellar outflow tracts, or some combination of these structures is contributory. Because the median and paramedian cerebellum have been repeatedly cited as related to PFS, alternative surgical trajectories have been advocated, most notably the telovelar or cerebellomedullary fissure approach. As Dr. Wells and coauthors note, this surgical approach does not fully protect the patient from PFS nor has the approach been systemically compared with a transvermian approach. Further, because most medulloblastomas arise in the cerebellar midline some vermian injury is likely, if not through the approach than via the tumor resection. Some reduced risk may come with further clarification of the anatomical milieu of posterior fossa brain tumors just as neurosurgeons and neuroradiologists have done with supratentorial tumors.

Another option to be considered in PFS avoidance is to limit tumor resection in an effort to bypass injury of adjoining tissue. In this scenario one could envision using a tailored surgical approach (such as limited removal) in high-risk patients. This would be a legitimate option if those patients deemed to be at substantial risk could be readily identified and if subtotal tumor removal did not compromise their outcome. As stated by the authors in this current report and substantiated by most other published works, risk criteria for PFS defined on preoperative MR imaging have yet to be clarified. Additionally, repeated prospective cooperative group studies have supported the fact that significant residual tumor is a negative prognostic indicator. Thus, limited tumor removal does not currently appear to be a justifiable option unless the tumor is known to invade the brainstem and cerebellar peduncles.

The authors of this report suggest that some intraoperative adjuncts may contribute to reducing the incidence of PFS. Electrophysiological monitoring appears unlikely to affect the postoperative incidence of this syndrome because the exact anatomical basis remains elusive and no appreciable method exists for monitoring the dentato-rubo-thalamic tracts. Additionally, the avoidance of the Cavitron ultrasonic aspirator or self-retaining retractors remains only conjectural and anecdotal at present.

Another point worthy of consideration is whether a preventive mechanism is crucial. Tumor removal in other anatomical regions (such as the supplementary motor cortex and ventricular trigone) is associated with predictable neurological morbidity. In these situations the expected injuries are considered acceptable in the name of achieving a cure, a sentiment that is, in part, due to variable rates of recovery. Should we similarly accept that the risk of PFS as an unavoidable byproduct of medulloblastoma removal? Although we can be confident that the majority of these children will return to a baseline level of func-
The authors of the paper, “Postoperative cerebellar mutism following treatment of medulloblastoma: neuroradiographic features and origin,” published in this month’s Journal of Neurosurgery: Pediatrics would like to thank Dr. Souweidane for his insightful comments and certainly agree with many of his comments. The authors would also like to thank the Journal of Neurosurgery: Pediatrics for allowing us to reply and partially editorialize on some of the aspects of the published paper.

The senior author of the manuscript (R.J.P.) has had the opportunity to care for children with medulloblastomas for over the past quarter century. Three major realizations concerning the management of medulloblastoma have become apparent over this time period and deserve comment to put this paper and Dr. Souweidane’s comments in perspective. The first realization is that in the 1970s, with radiation therapy alone, only one-half or less of children with medulloblastoma were long-term survivors of this disease.Fortunately, with multimodality therapy, including improvements in neurosurgery resulting in the majority of patients with nondisseminated tumors undergoing total or near-total resections, survival rates have significantly improved to the point that in some series 80% or more of patients can be expected to survive for 5 years, the majority cured of their disease.

The second realization was the sobering understanding that the price paid for such survival rates is extremely high, especially in the very young child, as long-term neurocognitive sequelae are common. Studies initially emanating from Europe and then later from the US clearly demonstrated the overall poor level of intellectual functioning of these children and led to a reappraisal of the causes for such outcomes. Whole brain radiotherapy, used as part of prophylactic craniospinal radiotherapy, has been incriminated as one of the most important factors resulting in decreased overall intelligence and neurocognitive function over time, and this recognition has led to the stepwise reduction in the dose of craniospinal radiation therapy, and most recently the volume of posterior fossa boost radiotherapy, in attempts to at least reduce the degree of cognitive decline.

The third devastating concern, which became increasingly apparent in the 1990s, is that many children with medulloblastoma developed posterior fossa mutism, which although often was poorly defined, affects a sizable proportion of patients, whether it be 25 or 40%, and possibly over one-half of those initially affected will have permanent long-term sequelae including balance difficulties, speech impairments, and likely cognitive challenges.

Although it is impossible to prove the “new” occurrence of this syndrome in retrospect, the senior author of the paper did carefully evaluate children through the 1980s and rarely saw this syndrome, despite evaluating over 100 patients with medulloblastoma who underwent operations at his primary institution and receiving consultations from all over the country. This discrepancy raised the issue of why this syndrome was being observed; concerns were even more heightened when the Children’s Oncology Group, in a nationwide prospective study, found that institutions across North America self-identified this syndrome in nearly 25% of patients, using as Dr. Souweidane states somewhat subjective criteria. It also became apparent that this syndrome did not just occur randomly. In evaluating severely affected patients, it was noted that neuroimaging...