Defining Excellence in Pediatric Neurosurgery

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Our children are the most important people in most of our lives. Is there any joy like the joy at hearing a child say, ''Daddy, I love you?'' Is there any grief like the grief at the death of one's child? Given the importance of our children, we want their care—especially their neurosurgical care—to be excellent. And the parents of our pediatric patients want the same excellent care for their children.

The fundamental principle of excellent pediatric neurosurgery is to care for our pediatric patients as if they were our own children. It is that simple. And it is that difficult. The principle has a corollary: If we cannot do a particular operation well enough to be comfortable doing it on our own child (were we to operate on our own, which we obviously would not), we should not do it on someone else's child but refer that child to someone we would want to operate on our child. If my child's magnetic resonance (MR) scan demonstrated a partially cystic, partially solid craniopharyngioma, I would not want a neurosurgeon who had not operated on a child with a craniopharyngioma in the past 3 years to do the operation. Neither should we do it on someone else's child.

Do you *do* excellent pediatric neurosurgery? In a teenage boy with shunted hydrocephalus and severe nocturnal headaches whose scan demonstrates small ventricles that have not changed in the past 2 years, do you diagnose migraine headaches because the scan is unchanged, do you assume that it is the shunt and ''explore'' it, or do you insert a monitor to measure his intracranial pressures? In a 16-year-old girl with vertex headaches, anxiety, and an incidental 1.5-cm Chiari malformation, do you assume that the headaches are caused by the Chiari malformation and decompress it, or do you consult a pediatric neurologist?

If the question, ''Do you do excellent pediatric neurosurgery?'' is asked of pediatric neurosurgeons, I suspect the majority of us would answer ''yes'' fairly quickly, whether in reality we are excellent or not. We usually do not know our outcomes—our percentage of shunt infections or of cerebrospinal fluid leaks—but we assume that we are excellent. I suspect that if the question is asked to ''general neurosurgeons,'' ''adult neurosurgeons'' whose practice is perhaps 10% pediatrics, the answers would be more hesitant and the percentage lower, particularly because most neurosurgeons believe that there is a correlation between surgical volume and outcome.

Do you *want to do* excellent pediatric neurosurgery? Excellence may be more difficult in pediatric neurosurgery than in other neurosurgical subspecialties because we treat such a spectrum of disorders: congenital, tumors, vascular, spine, and trauma. If I am honest about this, I think it unlikely that pediatric neurosurgeons can give excellent care to every child who comes to us. We may be able to give excellent care to children with tethered spinal cords but not such excellent care to children with unstable cervical spine fractures.

As a general rule, the rarer a child's condition is, the greater the likelihood is that we will not be able to treat the child with excellence. A 16-year-old child with hydrocephalus and basilar invagination secondary to the Hadju-Cheney syndrome came to me with headaches from hydrocephalus and slowly progressive quadriparesis from severe basilar invagination (Figure 1). I treated her hydrocephalus with a shunt but did not think I could give excellent care to her basilar invagination and referred her to a colleague with significant experience with these complex disorders. Maybe that is part of excellent care—knowing and accepting our limitations and referring to someone better. I realize it is hard for some neurosurgeons to publically acknowledge that there might be someone better.

CLINICAL EXCELLENCE IN PEDIATRIC NEUROSURGERY

If we *want to do* excellent pediatric neurosurgery, it will affect 5 areas of our clinical practice: relationships, judgment, diagnosis, treatment, and outcomes. Excellence in pediatric neurosurgery does *not* involve the speed with which we can insert a shunt.

Relationships With Children and Their Families

Although some may disagree with this, I believe that the foundation of clinical excellence in pediatric neurosurgery is the relationships with children and their parents. Those relationships—relationships with children who are suffering because of pain or disability, relationships with obnoxious parents who demand a scan because they are convinced their

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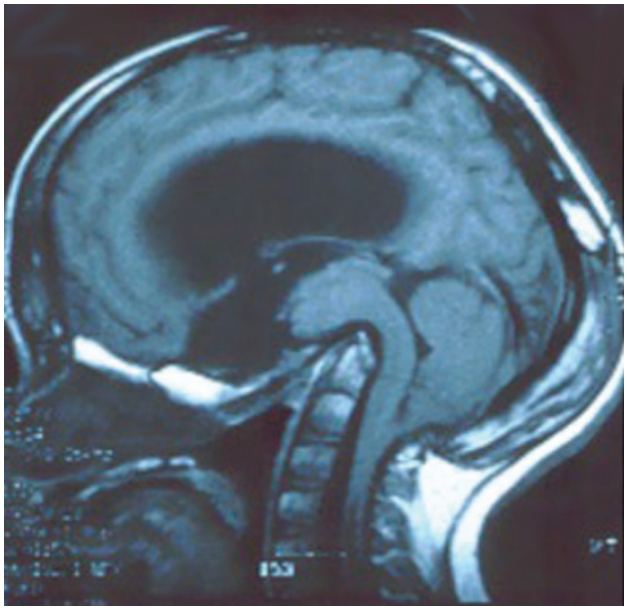

FIGURE 1. Hydrocephalus and severe basilar invagination in a teenager with Hadju-Cheney syndrome.

child's shunt is malfunctioning, and relationships with parents in agony, hearing the prognosis of their child's diffuse brainstem glioma—are one reason why some neurosurgeons choose not to do pediatric neurosurgery.

Excellence in relationships with some parents requires that we let them know that we value their disabled child, just as they value him or her; their child has value because the child is loved, even if he or she cannot be ''productive.'' Excellence requires compassion, having a relationship with the parents as a person rather than as a technician, grieving with them when their teenager has a terrible brain injury from a bicycle accident. It means being to them a physician who does neurosurgery rather than a neurosurgical technician. As my wife said, ''You can be an expert neurosurgeon…with the bedside manner of a fish.''

For me, the hardest parents to have excellent relationships with are those who (like neurosurgeons) are positively certain that they are right about their child's condition and the treatment the child needs. It goes without saying, but needs to be said, that no matter how compassionate and experienced we are, we will not have excellent relationships with all parents.

Relationships with the children themselves require wisdom. How do you tell a 10-year-old boy with a medulloblastoma that he will need radiation and chemotherapy? Partly by telling him the truth in such a way that he will understand that his parents and you will be there with him through it. Although the pain of children with neurosurgical disorders is one of the major reasons some neurosurgeons avoid pediatric neurosurgery, most of us who do pediatric neurosurgery consider the work to be a blessing—a gift from God—and the relationships to be priceless. When is the last time a patient drew you a picture after a spine fusion that you put on your refrigerator?

Excellence of Judgment

The second of the 5 attributes is excellence of judgment. In pediatric neurosurgery, our judgment needs to consider the outcome of the operation not only in the near future but also over the child's lifetime, eg, in a 3-year-old with occipitocervical instability, whether to do the fusion from the occiput down to C2 or C3 or C4. We have to consider the quality of life…for the child's lifetime.

This may be the attribute to which experience contributes the most. When I was in England on sabbatical several years ago, I heard someone say that neurosurgery is learned in 3 decades: In the first decade after residency, one learns how to operate; in the second decade, one learns when to operate; and in the third decade, one learns that hardest aspect of judgment, when not to operate.

Judgment *to* operate is a critical aspect of excellence. Fred Epstein had the judgment to go against conventional wisdom that said that intramedullary spinal cord tumors were inoperable, and his judgment to operate changed the treatment of children and adults with those tumors around the world. Last March, a 13-year-old boy was evaluated with a posterior III ventricular tumor, hydrocephalus, and a normal neurological exam (Figure 2). After agonizing about the various treatment options, I decided to do an endoscopic third ventriculostomy to treat his hydrocephalus and to remove the tumor through a transcallosal, interforniceal approach. The tumor turned out to be a mature teratoma with a small component of germinoma. If the tumor had been biopsied stereotactically, the focus of germinoma may have been missed, and his treatment would have been suboptimal.

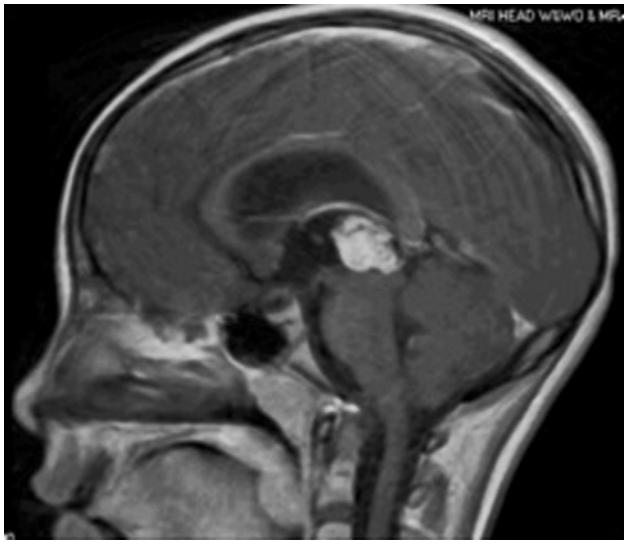

FIGURE 2. Posterior third ventricular mixed germ cell tumor.

Judgment not to operate is probably the hardest. A 9-year-old girl with a 3-year history of slowly progressive hemiparesis was evaluated in Kenya last January. Her MR image from Nairobi seemed to show a dorsally exophyic cervicomedullary astrocytoma, a generally resectable tumor. Our hospital agreed to donate her care, and she came to the University of Wisconsin for the operation, but her MR scan here demonstrated a diffuse tumor within the pons, medulla, and upper cervical cord. She had come halfway around the world to have the tumor removed, her care was being donated, the operation was scheduled, and yet it seemed there was no way an operation could make her better. I made the difficult decision not to operate, and she returned to Kenya to get radiotherapy there.

Excellence in Diagnosis

The third attribute, excellence in diagnosis, may be more difficult in children than in adults. So often, children provide either no history or only a limited one, and they may be less able, unable, or unwilling to cooperate for a detailed neurological exam. Therefore, we are often tempted to operate because their scans are abnormal: They have a Chiari I malformation, so we assume it to be the cause of their headaches; they have a conus that ends at the bottom of L2, so we assume it to be the cause of their bedwetting; they have a shunt, so we assume it to be the cause of their headaches.

Making the correct diagnosis in children with headaches and shunted hydrocephalus is perhaps the most difficult and most vexing of our diagnostic challenges, particularly in children who also have migraine headaches. In the past 2 years, I have seen 2 teenaged girls who each had >20 shunt operations at other hospitals within the previous 2 years. Every time they developed a bad headache, their ventricular catheter was replaced or a new shunt was inserted on the opposite side. Those children did not need a shunt revision; they needed the correct diagnosis. One girl's scans demonstrated slit ventricles; her shunt tap revealed high intracranial pressure; and an isotope shuntogram demonstrated shunt patency. She was effectively treated with a cranial expansion. The other girl had intracranial pressure monitoring for 48 hours that demonstrated no correlation with her headaches, which were 7 to 9 on a severity scale of 10 whether her intracranial pressure monitoring was −4 or 15 mm Hg. She had chronic daily headaches of childhood.

Excellence in Our Treatment

Fourth, excellence in our treatment, ie, our operations, goes a long way toward obtaining our ultimate goal, excellent outcomes. Excellent treatment necessitates knowing the latest data—whether gross total resections correlate with outcome in ependymomas (yes), whether D5 1/4 saline is an appropriate fluid after serious head injury (no), or whether endoscopic shunt insertions are associated with significantly improved shunt function (no).[1-5] I doubt there are any significant differences in excellent operations for pediatric neurosurgical disorders compared with adult vascular disorders or adult spine disorders. All are characterized by preoperative planning, intraoperative carefulness and gentleness, and the decisive factor, intraoperative judgment. No matter how carefully we plan the operation, how gently we handle the tissues, if we decide during a craniotomy for tumor that particular tissue is tumor when in fact it is normal, the child often suffers.

The treatment of hydrocephalus with shunts includes not just the operation to insert a shunt but the choice of which shunt to insert, where to insert it, and whether to use adjuncts such as ultrasound or Stealth to position the ventricular catheter. As far as which shunt to insert is concerned, 2 of the top 5 shunts inserted in the United States are programmable shunts, despite the fact that 2 randomized, prospective studies have shown that neither of the 2 commercially available programmable shunts functions any better than nonprogrammable shunts.[8,9]

As far as whether the entry site is frontal or posterior, no high-quality studies have answered that important question; a long-term retrospective study and a short-term prospective study came to opposite conclusions.[10,11] I recently polled 7 of the most experienced pediatric neurosurgeons in the United States and Canada about their shunt practice. When asked whether they inserted shunts frontally or posteriorly, the answers (in addition to my own) were exactly divided.

Of the 8 neurosurgeon responses, 4 replied that they used adjuncts to guide ventricular catheter insertion and 4 replied that they did not. Without adjuncts, our shunt outcomes are not, and will not be, excellent. In recent multicenter trials, the 5-year shunt function rate was approximately 30%, but the outcome of catheters positioned away from choroid plexus was significantly better.[3,9] To do excellent shunts, we have to position the tip of the ventricular catheter away from choroid plexus, and adjuncts are needed to do that.

Excellent Outcomes

Lastly, there are 2 aspects of excellent outcomes, technical outcomes and personal outcomes. Technical outcomes include factors that can be benchmarked such as infection rates, cerebrospinal fluid leak rates, and catheter disconnection rates. In my opinion, excellent outcomes in treating hydrocephalus would require shunt function of 75% at 5 years and infection rates of <5%, with the same success rate for hydrocephalus treated with endoscopic third ventriculostomies, ≥75% at 5 years.

It is a human trait to believe that our outcomes are better than they really are, so we need data about the common procedures we do. We published outcomes at the Children's Hospital of Pittsburgh of 5 common pediatric neurosurgical operations: initial shunt insertion, first shunt revision, craniotomy for brain tumor, correction of sagittal synostosis, and

release of tethered spinal cords.[12] We found a 65% 5-year function rate of initial shunts, a low mortality rate (1%) and morbidity rate (10%) after craniotomy for brain tumor, and a low frequency of transfusions for sagittal synostosis operations (20%).

Personal outcomes cannot be benchmarked. They are demonstrated partly by the cards and letters we receive from grateful parents, but probably more so by our relationship with families when our care of their child does not turn out well—when untethering a lipomeningocele results in paraparesis or tumor removal results in permanent hemiplegia.

In conclusion, we want excellence for our children who need neurosurgical care, and the parents of the children we care for want it for their children. Excellence in pediatric neurosurgery requires a relentless passion to give that, and only that, to the children we care for. We all fall short of the goal at times, but the children deserve nothing else.

Disclosure

The author has no personal financial or institutional interest in any of the drugs, materials, or devices described in this article.

REFERENCES

1. Adelson PD, Bratton SL, Carney NA, et al. Guidelines for the acute medical management of severe traumatic brain injury in infants, childen, and adolescents. *Pediatr Crit Care Med*. 2003;4(3)(Suppl):S60-S64.
2. Albright AL, Sposto R, Holmes E, et al. Correlation of neurosurgical subspecialization with outcomes of children with malignant brain tumors. *Neurosurgery*. 2000;47(4):879-885.
3. Kestle JR, Drake JM, Cochrane DD, et al. Lack of benefit of endoscopic ventriculoperitoneal shunt insertion: a multicenter randomized trial. *J Neurosurg*. 2003;98(2):284-290.
4. Lawton MT, Du R. Effect of the neurosurgeon's surgical experience on outcomes from intraoperative aneurysmal rupture. *Neurosurgery*. 2005; 57(1):9-15.
5. Smith ER, Butler WE, Barker FG II. Craniotomy for resection of pediatric brain tumors in the United States, 1988 to 2000: effects of provider caseloads and progressive centralization and specialization of care. *Neurosurgery*. 2004;54(3):553-565.
6. Kestle JR, Walker ML; Strata Investigators. A multicenter prospective cohort study of the Strata valve for the management of hydrocephalus in pediatric patients. *J Neurosurg*. 2005;102(2)(Suppl):141-145.
7. Pollack IF, Albright AL, Adelson PD. A randomized, controlled study of a programmable shunt valve versus a conventional valve for patients with hydrocephalus. *Neurosurgery*. 1999;45(6):1399-1411.
8. Boop FA. Long-term outcome in patients harboring intracranial ependymoma. *J Neurosurg*. 2005;103(1):4-5.
9. Kestle JR, Drake J, Milner R, et al. Long-term follow-up data from the shunt design trial. *Pediatr Neurosurg*. 2000;33(5):230-236.
10. Albright AL, Haines SJ, Taylor FH. Function of parietal and frontal shunts in childhood hydrocephalus. *J Neurosurg*. 1988;69(6): 883-886.
11. Bierbrauer KS, Storrs BB, McLone DG, Tomita T, Dauser R. A prospective, randomized study of shunt function and infections as a function of shunt placement. *Pediatr Neurosurg*. 1990-1991;16(6): 287-291.
12. Albright AL, Pollack IF, Adelson PD, Solot J. Outcomes analysis in pediatric neurosurgery. *Neurosurgery*. 1999;45(1):101-106.