The past, present, and future of pediatric neurosurgery

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It is uncomfortable to be given an honor you feel is undeserved. It is embarrassing to accept an honor you feel is undeserved. I stand before you with both emotions, but also with gratitude for the honor, knowing I stand on the shoulders of so many others who have led our subspecialty in the past two generations of pediatric neurorsurgeons, including Donald Darrow Matson.

Matson was born in 1913, the son of an army officer. He graduated from Cornell in 1935 and entered Harvard Medical School in 1939. While a medical student at Harvard, he published a paper on chronic pulmonary sepsis in the New England Journal of Medicine. After graduating from medical school, he undertook a 29-month surgical internship at the Brigham and Children’s Hospitals and then a fellowship in neurosurgery with Franc Ingraham until 1943. During that time, the pair wrote the classic paper, “Subdural Hematomas in Infancy,” which was published in the Journal of Pediatrics. Matson served in the army in Europe during World War II for 3 years, returned to Boston to join Dr. Ingraham, and worked with him as a research fellow in neurosurgery. He gained experience in hydrocephalus, craniosynostosis, and nerve regeneration as well as the use of polyurethane tubing, a precursor to shunts. He then served as a neurosurgery resident at Duke University Hospital, the Children’s Hospital in Boston, and Peter Bent Brigham hospital and was a fellow in neurosurgery at the Lahey Clinic. He rejoined Dr. Ingraham in 1948, and the two were partners for years. In 1954, they published the first pediatric neurosurgery textbook, Neurosurgery of Infancy and Childhood. When Ingraham retired in 1964, Matson succeeded him. He was a leader in several neurosurgical societies and president of the Harvey Cushing Society (1968–1969). He died in 1969 at the age of 55 during the height of his career, of an illness thought initially to be periarteritis nodosa but later determined to be Creutzfeldt–Jakob disease. In a memorial article published in the Journal of Neurosurgery, he was described as “a devoted husband and father, a warm and gratifying friend, a superb surgeon, a competent scientific investigator, a literary man in the field of medicine, an articulate spokesman for neurosurgery, and a teacher of men.” Thus, I do feel uncomfortable standing here.

I want to begin my lecture by giving thanks—thanks professionally to Don Reigel, who developed the pediatric neurosurgery department at the Children’s Hospital of Pittsburgh and introduced me in 1974 to the joy of caring for children with neurosurgical disorders; to Peter Jannetta, who offered me the position as chief of pediatric neurosurgery in 1991 and allowed me to develop it into the program I wanted it to be; and to my partners, Ian Pollack and David Adelson, who have been all I could have asked for in collegiality, collaboration, productivity, and the joy of working together. Personally, I thank my daughter Julie, my son Todd, and my wife Elizabeth for their love and support, for being willing to give up much of me so that I could do this work. Lastly, spiritually, I thank God for the privilege of being a pediatric neurosurgeon. In religious terms, it is a blessing—a gift from God—to be able to do this work.

Abbreviations used in this paper: ASPN = American Society of Pediatric Neurosurgery; CSF = cerebrospinal fluid; CT = computed tomography; DBS = deep brain stimulation; ICP = intracranial pressure; MR = magnetic resonance; RCT = randomized clinical trial; TBI = traumatic brain injury.
This afternoon, I would like to ponder three questions: where we were, where we are, and where we should go? I want to consider where we were in 1978 when I finished my neurosurgery residency, where we are now, 25 years later, and where we should be 25 years from now. We will consider three generations of pediatric neurosurgery, defining a generation as 25 years: the generation before 1978, the generation between 1979 and 2004, and the succeeding generation between 2005 and 2029.

Where Were We?

Pediatric neurosurgery was in its infancy in the generation that ended in 1978. At the start of that generation in 1953, there may have been only two pediatric neurosurgeons, Frank Ingraham and Donald Matson. By the end of the generation there were approximately 28 full-time pediatric neurosurgeons, contributors to the edition of *Pediatric Neurosurgery/Surgery of the Developing Nervous System*. The Pediatric Section of the American Association of Neurological Surgeons had been founded and included 75 members.

Pediatric neurosurgery fellowships were developed during that generation. In 1953, there was a so-called fellowship at the Boston Children’s Hospital, in which fellows functioned “like supernumerary residents,” to use Robin Humphrey’s term. By 1978 there were about five sustained fellowships—in Boston, Toronto, Philadelphia, Chicago, and Los Angeles.

Technologically, diagnostic tests evolved during that period, from ventriculography to angiography to CT scanning, which was introduced in the mid-1970s. The first shunt had been invented by Nulsen and Spitz in 1951.16 By 1978, shunts had become state-of-the-art systems such as the Raimondi Uni-shunt, which was wire reinforced with a distal slit valve. Because it had no barium in it, it never became calcified or brittle, but because of the wire reinforcement, it perforated the abdominal wall and all of the hollow abdominal viscera at one time or another. The operating microscope was introduced in the mid-1960s and binocular microscopes were in use by 1978.

At pediatric neurosurgery meetings authors presented case reports, retrospective case series, and an occasional clinical or laboratory investigation. In 1978 members at the pediatric section meeting discussed topics such as CSF antibiotic levels in treating shunt-related infections, the value of CT scanning in diagnosing arachnoid cysts, and the quality of survival in patients with cerebellar astrocytomas. Virtually every paper presented was either a retrospective case series or a technical note. Parenthetically, only four of the 10 papers presented during that meeting were subsequently published—one of the few characteristics of pediatric neurosurgery in 1978 that is still true.

The vast majority of pediatric neurosurgery manuscripts published during that generation were similar to the presentations themselves: either single case reports or retrospective case series; however, this form of increasing medical knowledge was probably appropriate during that era.

I believe that one highlight of that generation occurred in its final year, with the founding of the ASPN. The organization was founded by 16 neurosurgeons who established standards for pediatric neurosurgeons. They were committed to “advancing the development of pediatric neurosurgery through laboratory and clinical research,” impressive foresight as to how we should advance our field. In 1985, I declined an invitation to join the ASPN. I thought it was too cliquish, too expensive, and too weak academically; papers presented at the annual meetings and published subsequently in *Concepts in Pediatric Neurosurgery* were not peer reviewed. There was some truth to each of those criticisms, but they did not undermine the fundamental thrust of the organization: to define pediatric neurosurgeons and to contribute to the development of pediatric neurosurgery by clinical and laboratory research.

Such was the climate in 1978 when I began practice in pediatric neurosurgery; there were approximately 28 full-time pediatric neurosurgeons, five fellowships, clinical series involved retrospective data, first-generation CT scanners had been introduced, and the political organization was in its infancy.

Where Are We?

Between 1979 and 2004, the number of full-time pediatric neurosurgeons increased from 28 to approximately 135. The field grew from five unaccredited fellowships to 22 accredited fellowships in 2004 (more than the number of applicants for fellowships). Politically, pediatric section membership increased from 75 to 316. This second generation has been characterized by the maturation and expansion of the ASPN to include most of the active pediatric neurosurgeons in the US and Canada. The American Board of Pediatric Neurosurgery was developed to provide formal credentials for pediatric neurosurgeons. Accredited pediatric neurosurgery fellowships were developed. By 2003 we witnessed the decision to require reaccreditation in pediatric neurosurgery as well as the development of the means by which to enforce this process; in so doing, we became the first neurosurgical subspecialty to require reaccreditation.

Technological changes have been vast, most being unimaginable at the start of the decade. We went from CT scanning to MR imaging to functional MR imaging to MR spectroscopy; from binocular microscopes to ceiling-mounted, stereotactically coordinated microscopes with binocular vision for two observers and the capacity to obtain digital photographs and videos. Shunt technology evolved from the Raimondi Uni-shunt to antibiotic-impregnated, programmable shunt systems with built-in antishunt devices. Another development, the ultrasonic surgical aspirator, has allowed the removal of tumors in the basal ganglia and spinal cord. Endoscopes were developed, permitting the widespread treatment of hydrocephalus with endoscopic third ventriculostomy. Arteriovenous malformations are now often obliterated using stereotactic radiosurgery, avoiding craniotomies. Baclofen pumps, vagal nerve stimulators, and DBS were developed. We expanded our clinical practice to include disorders rarely treated in 1978—brachial plexus injuries, spasticity, dystonia, and intractable seizures.

The quality of presentations at national meetings improved. In 1982, I gave a lecture at the pediatric section meeting in Toronto entitled, “How to Prepare and Present a Medical Lecture.” I used approximately 300 slides that were shown from two racks of projectors, with three projectors in each rack and each rack focused on a separate...
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screen. We progressed from slide shows—done in diazo dyes if we wanted to avoid black-on-white background, slides that commonly contained information printed too small to decipher—to clear, computer-generated, multi-colored slides and now digital video. The proportion of oral presentations at national meetings that are subsequently published, however, remains 50% or less.

The academic highlight of this generation was probably the development of RCTs, in which clinical research allowed several questions to be definitively answered. I do not know when the first such trial was conducted. Our group undertook one such trial in 1991 involving intrathecal baclofen, but there were probably others before that.

Since then, pediatric RCTs have been performed for brain tumors, shunts, selective dorsal rhizotomies, and therapeutic muscle stimulation after rhizotomies. The work in RCTs has been led by John Kestle and Jim Drake.

Now, at the end of the generation, where are we, clinically? Let me summarize where we are in terms of three disorders—hydrocephalus, head trauma, and brain tumors—and then describe in more detail our status in relation to the problems I have focused on in the past 15 years, spasticity and movement disorders.

In hydrocephalus, we know from prospective randomized multicenter trials the following: 1) shunt function rates are approximately 40% at 5 years; 2) the mean infection rate is 8%; 3) there is no evident difference in function in 5 years between any of the commercially available shunts (including programmable devices); 4) insertion of the ventricular catheter with endoscopic guidance does not improve long-term shunt function; 5) placement of the catheter tip away from the choroid plexus significantly decreases the duration of shunt function.

We do not, however, know how to place shunt tips into the frontal horns of the lateral ventricles precisely and regularly. Nor do we know which children will respond to endoscopic third ventriculostomies.

In children who have sustained a TBI, little more is known today than in 1978. In the past 25 years, not one multicenter RCT has been completed in pediatric head injury. We believe, based on data derived from less rigorously designed pediatric studies collated in the pediatric trauma guidelines that hyperventilation, corticosteroid agents, and propofol are harmful; that ICP may be abnormally low; and we have evidence that cranial decompression may be helpful in cases of severe head injury.

Other unresolved issues include the optimal serum osmolality, glucose, or CO₂, optimal position of the head of the bed, optimal ICP in children after TBI (although the expert opinion indicates that it is < 20 mm Hg), or the effects of hypothermia, which have been purported to be beneficial in three single-institution studies; nor do we know the differences in responses of infants/toddlers/older children to our therapies. Do infants respond to manitol the same as 1-year-old children?

We have learned far more about brain tumors despite their rarity because of several prospective multicenter RCTs that have been conducted by national pediatric neurooncology groups. We know, for instance, that in low-grade astrocytomas the extent of resection correlates strongly with outcome and that chemotherapy is the initial treatment of choice for midline tumors that cannot be resected. Whole-brain irradiation is no longer used. Even conventional external-beam irradiation with four opposing fields is being replaced—because of its adverse effects on cognition—by conformal radiotherapy in children younger than 10 years of age.

We know that high-grade gliomas should be maximally resected because the extent of resection correlates with improved outcome and that patients should then undergo radiation therapy (56–60 Gy) and chemotherapy.

Furthermore, we have evidence that after therapy, the 5-year survival is 23% for anaplastic astrocytoma and 12% for glioblastoma multiforme.

We know that medulloblastomas in children should be maximally resected without entering the brainstem and this should be followed by irradiation and chemotherapy; after these therapies, long-term survival is greater than 70% if the patient is older than 3 years of age and the tumor has not spread from its initial site. In 1979 the standard dose in craniospinal irradiation was 3600 cGy for medulloblastomas, whereas in 2003 it had been reduced to 2400 cGy; the lower radiation dose is justified by the supplemental multidrug chemotherapy regimen. Amazingly, in cases of high risk medulloblastomas (disseminated tumors or those with bulky residual tissue), the survival rate has improved from less than 20% to approximately 60% (Tarbell NJ, personal communication, 2004).

We know that in patients with ependymomas, the extent of resection is the single most important factor related to survival. We do not, however, know any effective treatment for diffuse brainstem gliomas or recurrent posterior fossa ependymomas. Effective treatments for these lesions are far more likely to evolve from adjuvant therapy, even gene therapy, than from improved surgical procedures, which appear to have been optimized.

In 1979 treatment for spasticity and movement disorders, the area in which I have been most interested, involved Bishoff myelotomies and little else for the other pediatric movement disorders. Because of this, pediatric neurosurgeons did not focus their interest on these disabilities. Now we have good treatments for spasticity, whether it is associated with cerebral palsy, trauma, or familial spastic paraparesis, or whether spasticity is focal or generalized. Yet, pediatric neurosurgeons still have little interest in these children.

We treat focal spasticity with intramuscular injections of botulinum toxins, which have been shown to improve spasticity significantly in the upper and lower extremities, range of motion, and function in the injected muscle. For children with spastic diplegia, we can treat them with selective dorsal rhizotomies, improving their gait. In three RCTs, the authors have shown that rhizotomies significantly decrease spasticity in the legs and improve function as measured using a Gross Motor Function Measure.

Children with upper- and lower-extremity spasticity can be treated with intrathecal baclofen. In 1991, we reported a randomized double-blind clinical trial in which we found that bolus doses of intrathecal baclofen significantly reduced lower-extremity spasticity. Investigators in subsequent multicenter prospective (but not randomized or blinded) clinical trials have shown that continuous intrathecal baclofen infusion improves upper- and lower-extremity spasticity.

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We do not, however, understand the pathophysiology of
dystonia, athetosis, or chorea, nor do we know the extent
to which DBS improves secondary dystonia. Additionally
we are unable to classify or grade athetosis, or determine
the optimal good treatment for it—whether stereotactic or
by infusion of medications.

Thus, in the present generation, there are approximate-
ly 135 pediatric neurosurgeons, 22 accredited fellowships,
better-quality presentations at meetings, enormously bet-
ter technology, and better-quality clinical (and laboratory)
research.

Where Should We Go?

For the generation of you who are commencing pedi-
atrie neurosurgery, I have a question, a request, and a few
suggestions about potential areas to investigate.

First, the question: should we not focus on the disorders
that affect the most children? We write article after article
and spend millions of dollars researching brain tumors.
They are dramatic. They are serious. They are—despite
our perception—quite rare; they affect only three/100,000
children, approximately 2200 in the US each year. Yet ap-
proximately 10% of pediatric neurosurgeons today focus
primarily on brain tumors. We write about encephaloceles,
which affect one/20,000 children. Even hydrocephalus,
the traditional disease of pediatric neurosurgery, affects
about one/1000 children and is not close to being the most
common disorder. Cerebral palsy, which receives little at-
tention, affects twice as many children (two of every 1000
children or ~10,000 new cases every year). Traumatic brain
injury affects 191/100,000 children (~150,000–160,000/
year), with 5 to 10% of the injuries being moderate and 10
to 15% of the injuries being severe. Annually 7000 chil-
dren die of injuries to the brain and spinal cord. There is
more death and disability associated with TBI than all
other pediatric-related causes combined, 18-fold more
deaths than are caused by brain tumors.6 Epilepsy affects
two/100 children, with medically intractable seizures
occurring in 10 to 20% of them.7 We need to refocus not
on the view under our microscope but on a view outside
our present field of vision. I ask again, should we not
focus on the disorders that affect the most children?

Second, the request: will you consider going abroad for
a short time to do and teach pediatric neurosurgery in a
Third World country? Fifty percent of many Third World
country’s populations are younger than 18 years of age.
There are enormous needs for equipment and enormous
opportunities to teach, operate, and learn. You will prob-
ably be more of a physician there than anywhere since you
left medical school, and you may never again feel as worth-
while. You can take instruments that will remain there,
result in twisting and abnormal postures. They respond
minimally to oral medications. In prospective nonran-
domized nonblinded trials, we have shown that intrathecal
baclofen improves severe dystonia in nearly 90% of chil-

Drug Administration. Based on CSF flow, it is likely that
intraventricular infusions, compared with intrathecal infu-
sions, would achieve higher concentrations over the cor-
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into the parenchyma—for example, baclofen into the in-
ternal globus pallidus for dystonia. We can explore DBS
of other sites, such as the subthalamic nucleus for dysto-
nia. We should evaluate the use of motor cortical stimula-
tion, implanting epidural electrode strips over the motor
strip to alleviate hyperkinetic movement disorders that
involve only one extremity. Additionally for children with
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while. You can take instruments that will remain there,
functioning long after you are gone. You can take your
child and have an experience neither of you will ever for-
get. You will have a multiplier effect: the techniques you
teach will be used to treat countless patients after you
leave, and you can become a resource, so that the local
neurosurgeon can go to his local internet café, send you
images of his latest conundrum, and get your advice from
half a world away.

Third, I would like to suggest potential areas in which
to investigate the common disorders. In epilepsy, we can
explore the use of stereotactic radiosurgery for focal epi-
leptogenic lesions, whether in the mesial temporal lobe or
elsewhere. For children with focal lesions located in areas
that cannot be ablated without causing unacceptable mor-
bidity, we can work with companies to develop on-de-
and sensors to activate cortical stimulation from elec-
trodes surgically placed over the epileptogenic cortex. For
the treatment of children with generalized seizures, we
should investigate infusions of medications into the sub-
arachnoid space via implanted pumps. It is highly likely
that such infusions will result in higher concentrations of
the drugs in the CSF and brain than are achieved after oral
administration. Work could be done in the laboratory to
identify stem cell or gene transfer techniques to produce
cells that could be injected into epileptogenic foci to release
inhibitory neurotransmitters such as γ-aminobutyric acid.

In the treatment of trauma, there is so much to be done.
We must establish multicenter RCTs to answer fundamen-
tal questions about CO2, optimal levels of glucose and ICP,
the prevention of secondary injury, and the role of decom-
pressive craniotomies, as well as the effects of the different
therapies on children of different ages. After the acute head
injury has been treated, work is needed to evaluate neu-
rostimulation of alertness and the implantation of sensors to
pick up microvoltages from thoughts generated in cortical
tissue, transmitting them via electrodes to nerves or mus-
cles in paralytic limbs (techniques that would be applicable
in spina bifida as well).

In hydrocephalus, we need to develop technology that
will allow us to simply and reliably place shunt catheters
where we intend them to go. Another need is technology
that will allow noninvasive measurement of ICP. We might
consider placing shunts from the ventricle upward into the
subarachnoid space, where CSF would be absorbed. We
should find out how much CSF is absorbed along the spinal
dura mater.

Lastly, for the management of children with movement
disorders, we can study intraventricular infusion of med-
ication. I have submitted an “investigational new drug”
application for intraventricular baclofen to the Food and
Drug Administration. Based on CSF flow, it is likely that
intraventricular infusions, compared with intrathecal infu-
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experience an improved development if their occult hydrocephalus were treated in a timely manner.

Based on these remarks, you know that I foresee that pediatric neurosurgery at the end of your generation will be profoundly different from now, involving far more technology, with implantation of devices to measure ICP, regulate CSF flow in shunts, stimulate cerebral cortex and parietic muscles, and with greater involvement in neurorehabilitation.

In closing, thank you for the undeserved opportunity, but valued privilege, to present the Matson Lecture. From Matson to today’s practitioners, pediatric neurosurgeons share a common awareness—that children are the most valuable people in the lives of most people. Nothing causes the grief that the death of a child does, and few disasters cause the grief of a permanently brain damaged child. Pediatric neurosurgeons know that children are inherently worthwhile, whether they are bright or retarded, agile or disabled. They have worth ultimately because they are loved and valued by God. Because children have such worth and because we recognize it although most neurosurgeons do not, it is incumbent on you—the next generation of pediatric neurosurgeons—to do good quality clinical research. In contrast to most of the publications of my generation, data from multicenter multidisciplinary RCTs should become the norm. The children deserve them.

References


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