

Flannery

The 31st Annual Meeting of the
AANS/CNS Section on

PEDIATRIC

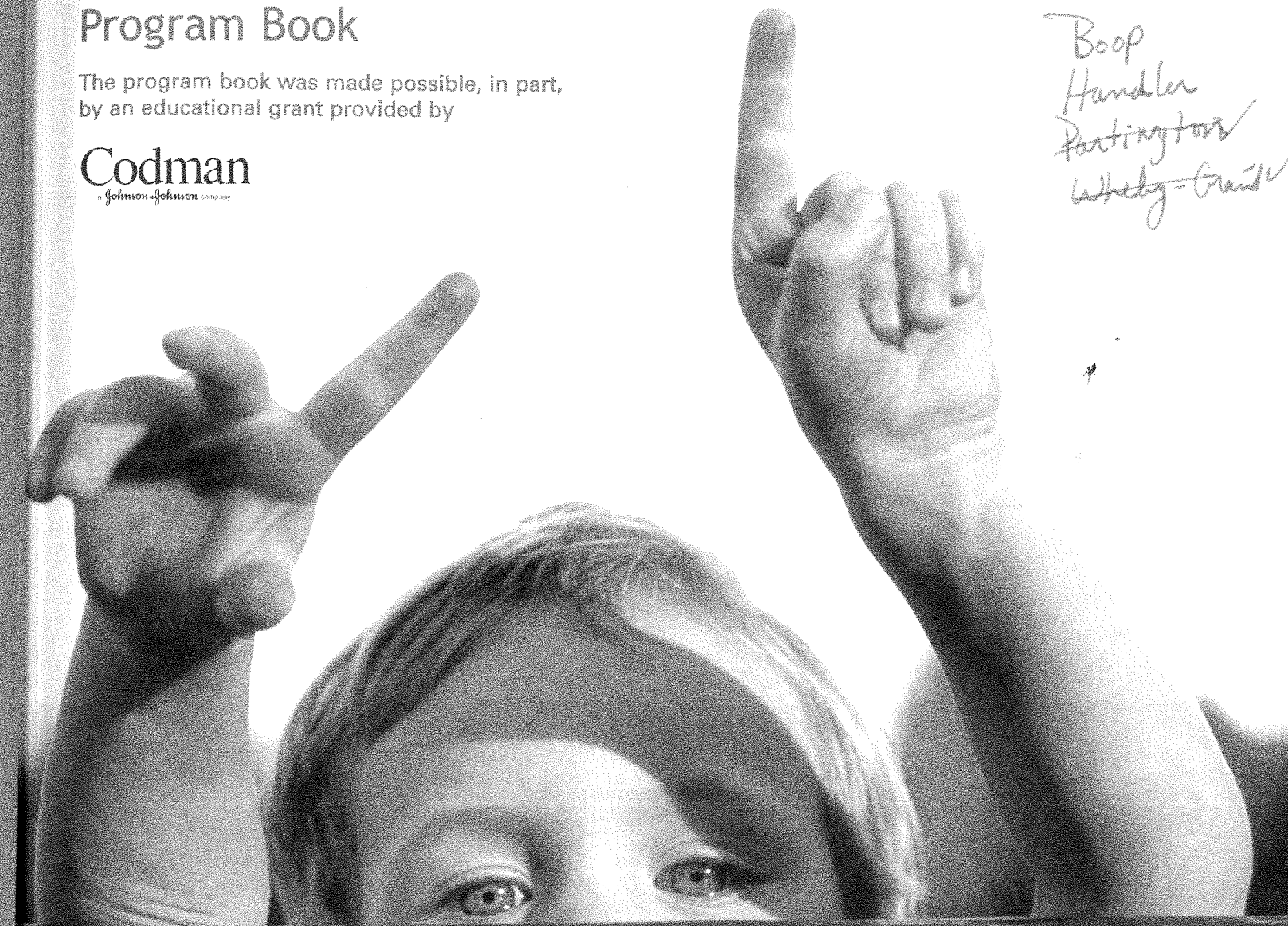
Neurological Surgery

Program Book

The program book was made possible, in part,
by an educational grant provided by



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Whelby-Grand*



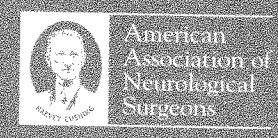
*My Shaver
Respectful
Papa
Lydia*

AANS/CNS Section on Pediatric Neurological Surgery

5550 Meadowbrook Drive • Rolling Meadows, IL 60008-3852

Phone: (847) 378-0500 • Fax: (847) 378-0600

www.neurosurgery.org/pediatric



December 4-7, 2002

Hyatt Regency Scottsdale
at Gainey Ranch

Scottsdale, AZ

Jointly Sponsored by
the American Association of Neurological Surgeons



American
Association of
Neurological
Surgeons



AANS/CNS Section on Pediatric Neurological Surgery

31st Annual Meeting
December 4-7, 2002
Scottsdale, AZ

Continuing Medical Education Credit

This activity has been planned and implemented in accordance with the Essentials and Standards of the Accreditation Council for Continuing Medical Education (ACCME) through the joint sponsorship of the American Association of Neurological Surgeons and the AANS/CNS Section on Pediatric Neurological Surgery. The American Association of Neurological Surgeons is accredited by the Accreditation Council for Continuing Medical Education (ACCME) to provide continuing medical education for physicians.

The American Association of Neurological Surgeons designates this educational activity for a maximum of 16 hours in category 1, toward the AMA Physician's Recognition Award, with an additional 4.75 hours for the Pre-meeting Coding Course. Each physician should claim only those hours that he or she actually spends in the educational activity.

Disclaimer

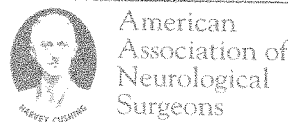
All drugs and medical devices used in the United States are administered in accordance with Food and Drug Administration (FDA) regulations. These regulations vary depending on the risks associated with the drug or medical devices compared to products already on the market, and the quality and scope of the clinical data available.

Some drugs and medical devices demonstrated or described on the print publications of the AANS/CNS Section on Pediatric Neurological Surgery have a FDA clearance for use for specific purposes or for use only in restricted research settings. The FDA has stated that it is the responsibility of the physician to determine the FDA status of each drug or device he or she wishes to use in compliance with applicable law.

Annual Meeting Sites

1972	Cincinnati
1973	Columbus
1974	Los Angeles
1975	Philadelphia
1976	Toronto
1977	Cleveland
1978	Philadelphia
1979	New York
1980	New York
1981	Dallas
1982	San Francisco
1983	Toronto
1984	Salt Lake City
1985	Houston
1986	Pittsburgh
1987	Chicago
1988	Scottsdale
1989	Washington, D.C.
1990	San Diego and Pebble Beach
1991	Boston
1992	Vancouver, BC
1993	San Antonio
1994	St. Louis
1995	Pasadena
1996	Charleston
1997	New Orleans
1998	Indianapolis
1999	Atlanta
2000	San Diego
2001	New York
2002	Scottsdale
2003	To Be Announced

Continuing Medical Education Credit.....	Cover
Disclaimer.....	Cover
Annual Meeting Sites	1
Pediatric Section Chairmen	2
Officers of the AANS/ CNS Section on Pediatric Neurological Surgery	2
Ad Hoc Committees	3
2002 Raimondi Lecturer	4
Raimondi Lecturers	5
Matson Memorial Lecturers	5
Kenneth Shulman Award Recipients	6
Traveling Fellowship Awards	7
Hydrocephalus Association Award Recipients	7
Program Schedule	8-13
Floor Plan of the Exhibit Hall	14
Exhibitor Listing	15-16
Acknowledgements.....	17
Disclosure Information.....	18-20
Scientific Program Oral Abstracts	21-49
Scientific Program Poster Index	50-51
Scientific Program Poster Abstracts	52-66
Notes	66-67
2002 Membership Roster	68-76



Jointly sponsored by the American Association of Neurological Surgeons



Officers and Standing Committees

Pediatric Section Chairmen

1972-73	Robert L. McLaurin
1973-74	M. Peter Sayers
1974-75	Frank Anderson
1975-76	Kenneth Shulman
1976-77	E. Bruce Hendrick
1977-78	Frank Nulsen
1978-79	Luis Schut
1979-81	Fred J. Epstein
1981-83	Joan L. Venes
1983-85	Harold J. Hoffman
1985-87	William R. Cheek
1987-89	David G. McLone
1989-91	Donald H. Reigel
1991-93	R. Michael Scott
1993-95	Arthur Marlin
1995-97	Harold L. Rekate
1997-99	Marion L. Walker
1999-2000	John P. Laurent
2001-2003	Thomas G. Luerssen

Officers

Chair	Thomas G. Luerssen, MD (2001)
Secretary	I. Richmond Abbott III, MD (2000)
Treasurer	Jeffrey H. Wisoff, MD (2001)
Membership Chair	Sarah J. Gaskill, MD (2002)

Executive Committee

Members At Large (two-year terms)	Andrew D. Parent, MD (2001)
	Frederick A. Boop, MD (2001)
	Alan R. Cohen, MD (2002)
	Ann-Christine Duhaime, MD (2002)

Standing Committees

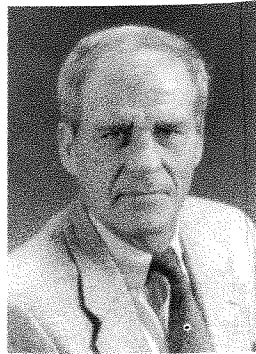
Nominating Committee	John P. Laurent, MD
	Harold L. Rekate, MD
	Marion L. Walker, MD
Rules and Regulations Committee	Chair: Cheryl A. Muszynski, MD (2002)
Membership Committee	Chair: Sarah J. Gaskill, MD (2002)
Program and Continuing Education Committee	Chair: Ann Marie Flannery, MD (2001)
	Vice-Chair: Joseph R. Madsen, MD (2002)
	Ex Officios: Harold L. Rekate, MD
	Thomas G. Luerssen, MD
Annual Meeting Chair	Harold L. Rekate, MD
Future Annual Meeting Chairmen	2003: To Be Announced
	2004: Mitchel S. Berger, MD
	2005: W. Jerry Oakes, MD
	2006: Michael H. Handler, MD

Ad Hoc Committees

Traveling Fellowship Committee	Chair: R. Michael Scott, MD
Lifetime Achievement Award	Chair: Robin P. Humphreys, MD
NEUROSURGERY//ON-CALL® Web site	Chair: Douglas L. Brockmeyer, MD
Publications Committee	Chair: Sarah J. Gaskill, MD
Liaison to the American Academy of Pediatrics	Joseph H. Piatt, Jr., MD (1997)
Liaison to the Joint Council of State Neurosurgical Societies	Michael Heafner, MD (2000)
Representative to AANS Executive Committee	Thomas G. Luerssen, MD
Representative to CNS Executive	Richard Ellenbogen, MD
Representative to the Quality Assurance Committee	Chair: Paul A. Grabb, MD (1999)
Representative to the Washington Committee	Marion L. Walker, MD (1999)
Representative to the Neurological Surgery Political Action Committee	Michael Medlock, MD

Volker K.H. Sonntag, MD

Volker K.H. Sonntag, MD, a native of Graudenz, Germany, received his Bachelor of Arts degree in Chemistry from Arizona State University. His studies then took him from University of Arizona Medical School to Tufts New England Medical Center Hospital, Youngstown Ohio and back again to Arizona, ultimately to practice neurosurgery in 1978. In 1983 he joined the Barrow Neurological Institute (BNI), where he has since remained. At the BNI, Dr. Sonntag has served on many boards and programs, including Vice-Chairman of the Neurological Surgery Division and Director of the Residency Program.



Dr. Sonntag has received many awards for his contributions to the teaching and mentoring of young neurosurgeons such as the Mentor of the Millennium award given to him by his residents in 2000. He has also received many honors for his community service, research, and outstanding achievements, including the Meritorias Service Award from the Joint Section on Disorders of the Spine and Peripheral Nerves in 1999. The North American Spine Society established the Volker K.H. Sonntag Fund for Research in 2001 in his honor.

Through laboratory and clinical research, Dr. Sonntag has devoted his career to improving the understanding of spinal disorders, especially cervical and upper cervical spine disorders. His academic output has been prolific. He has written more than 60 chapters for important neurosurgical texts and more than 200 articles for refereed and nonrefereed journals. As well, he has been invited as a Visiting Professor to more than 40 institutions.

In addition to his academic, teaching, and clinical obligations, Dr. Sonntag has been an active member and leader in many professional neurosurgical and spine societies throughout his career, holding more than 20 professional memberships including AANS, CNS, Neuro-Society of America, and the Society of Neurosurgeons. Since 1982, he has been a member of the American Association of Neurological Surgeons (AANS), and presently serves as Vice President of AANS. Above all, Dr. Sonntag is devoted to his family, his wife Lynne (of 28 years), and their three children.

Raimondi Lecturers

1978	E. Bruce Hendrick	1991	Judah Folkman
1979	Paul C. Bucy	1992	Olof Flodmark
1980	Floyd Gilles	1993	Maurice Albin
1981	Panel Discussion	1994	Blaise F.D. Bourgeois
1982	Panel Discussion	1995	Robert H. Pudenz
1983	Derek Harwood-Nash	1996	Samuel S. Flint
1984	Anthony E. Gallo, Jr.	1997	M. Michael Cohen, Jr.
1985	Frank Nulsen	1998	Robert A. Zimmerman
1986	William F. Meacham	1999	David B. Schurtleff
1987	Dale Johnson	2000	Steve Berman
1988	Joseph J. Volpe	2001	Alejandro Berenstein
1989	Martin Eichelberger	2002	Volker K.H. Sonntag
1990	George R. Leopold		

Matson Memorial Lecturers

1987	John Shillito	1995	John Holter
1988	E. Bruce Hendrick	1996	
1989	Martin P. Sayers	1997	Maurice Choux
1990	Roger Guillemin	1998	Lisa Shut
1991	Robert L. McLaurin	1999	Gary C. Schoenwolf
1992	Joseph Murray	2000	Postponed due to illness
1993	Eben Alexander, Jr.	2001	Donald H. Reigel
1994	Joseph Ranschoff	2002	David McLone

Award Recipients

- 1983 KIM MANWARING Neonatal Post-Hemorrhagic Ventriculomegaly: Management with Pulsed Lumbar Cisternostomy
- 1984 ARNO FRIED A Laboratory Model of Shunt-Dependent Hydrocephalus
- 1985 ANN-CHRISTINE DUHAIME The Shaken Baby Syndrome
- 1986 ROBERT E. BREEZE CSF Formation in Acute Ventriculitis
- 1987 MARC R. DELBIGIO Shunt-Induced Reversal of Periventricular Pathology in Experimental Hydrocephalus
- 1988 SCOTT FALCI Rear Seat-Lap Belts. Are They Really "Safe" for Children?
- 1989 JAMES M. HERMAN Tethered Cord as a Cause of Scoliosis in Children with a Myelomeningocele
- 1990 CHRISTOPHER D. HEFFNER Basilar Pons Attracts Its Cortical Innervation by Chemotropic Induction of Collateral Branch Formation
- 1991 P. DAVID ADELSON Reorganization of the Cortical-Tectal Pathway Following Neonatal Cerebral Hemispherectomy in Cats
- 1992 DAVID FRIM Effects of Biologically Delivered Neurotrophins in Animal Models of Neural Degeneration
- 1993 MONICA C. WEHBY Metabolic Demonstration of Retained CNS Function in the Rabbit Model of Infantile Hydrocephalus
- 1994 ELLEN SHAVER Experimental Acute Subdural Hematoma in Infant Piglets
- 1995 SEYED M. EMADIAN Correlation of Chromosome 17p Loss with Clinical Outcome in Patients with Primitive Neuroectodermal Tumors
- 1996 JOHN PARK Platelet Derived Growth Factor Induces Differentiation of Neuroepithelial Stem Cells into Neurons
- 1997 MICHAEL J. DREWEK Quantitative Analysis of the Toxicity of Human Amniotic Fluid to Rat Fetal Spinal Cord Cultures
- 1998 ADRIANA RANGER Implantation of Medulloblastoma Cells into Collagen Type I Gels: Invasiveness, Enzymatic Characterization, and the Impact of Surgical Excision and Radiation
- 1999 SUSAN DURHAM The Surprisingly Sturdy Infant Brain: Why Is It More Resistant to Focal Injury?
- 2000 KETAN R. BULSARA Novel Findings in the Development of the Normal and Tethered Filum Terminale
- 2001 DAVID I. SANDBERG Convection Enhanced Delivery into the Rat Brain Stem: A Potential Delivery for the Treatment of Diffuse Pontine Gliomas

Award Recipients

 Traveling Fellowship Awards

Domestic
JOHN DAVID MORENSKI
PRITHVI MORENSKI

International
CHARITY CORDERO

- 1989 ERIC ALTSCHULER Management of Persistent Ventriculomegaly Due To Altered Brain Compliance
- 1990 S.D. MICHOWIZ High Energy Phosphate Metabolism in Neonatal Hydrocephalus
- 1991 NESHER G. ASNER Venous Sinus Occlusion and Ventriculomegaly in Craniectomized Rabbits
- 1992 MARCIA DASILVA Reversal of High Energy Phosphate Metabolism Changes in Experimental Hydrocephalus after CSF Shunting
- 1993 CHARLES BONDURANT The Epidemiology of Cerebrospinal Fluid Shunting
- 1994 MONICA C. WEHBY-GRANT The Rabbit Model for Infantile Hydrocephalus: Regional Differences in the Cortical Metabolic Response to Hydrocephalus and Shunting
- 1995 RICHARD J. FOX Cerebrospinal Fluid Absorptive Site of the Parasagittal Dura: A Cadaveric Study
- 1996 MARTHA J. JOHNSON Reactive Astrocytosis in a New Model of Obstructive Hydrocephalus
- 1997 No Prize Awarded
- 1998 DANIEL LIEBERMAN In Vitro Detection of Fluid Flow in Ventriculoperitoneal Shunts (VPS) Using Contrast Enhanced Ultrasound
- 1999 KIMBERLY BINGAMAN Hydrocephalus Induces the Proliferation of Cells in the Subventricular Zone
- 2000 No Prize Awarded
- 2001 JAKE TIMOTHY Treatment of Hydrocephalus Using a Choroid Plexus Specific Immunotoxin: An In Vitro Study
- 2002 To Be Announced

Program Schedule

Wednesday, December 4

11:00 AM-7:00 PM

Registration

Noon-5:00 PM

Coding Course

Learning Objectives

Upon completion of this program, the participants should be able to:

1. Discuss the 2002 CPT, ICD-9 and Medicare updates.
2. Choose the correct code(s) for frequently performed pediatric and Neurosurgical cases—simple and complex.
3. Apply modifiers appropriately for streamlined reimbursement.

Noon-5:00 PM

Nurses' Seminar

Learning Objectives

Upon completion of this program, the participants should be able to:

1. Describe current treatment strategies in pediatric brain tumors, normal pressure hydrocephalus and in-utero myelomeningocele closure.
2. Describe cerebral spinal fluid physiology.
3. Identify current research in pediatric head injury.

2:00-6:00 PM

Poster Set-Up for Medical Registrants

6:00-7:30 PM

Opening Reception in the Palm Grove Courtyard at the Hyatt Regency Scottsdale

Thursday, December 5

7:00 AM-5:15 PM

Registration

7:00-8:00 AM

Continental Breakfast with Exhibit Viewing

7:00-8:00 AM

Poster Set-Up for Medical Registrants

7:00 AM-4:00 PM

Exhibit & Poster Viewing

8:00-8:10 AM

Welcome Remarks

Moderators: Harold L. Rekate, MD;

Thomas Luerssen, MD

8:10-10:05 AM

SCIENTIFIC SESSION I— BASIC SCIENCE AND VASCULAR LESIONS

Moderators: Rick Abbott, MD;

David Gruber, MD

Discussants: James Rutka, MD;

William Shapiro, MD

Learning Objectives

Upon completion of this program, the participants should be able to:

1. Identify the most current research as it relates to pediatric neurosurgery

8:10-8:25 AM

1. Immuno-Gene Therapy for Malignant Pediatric Brain Tumors

Roberta P. Glick, MD; Terry Lichtor, MD, PhD; Edward P. Cohen, MD; Henry Lin, BA; Kimberly Dean, BA (Chicago, IL)

8:25-8:35 AM

2. The Efficacy of Prophylactic Spinal Fusion in the Prevention of Post-Surgical Spinal Deformity in Pediatric Intramedullary Spinal Cord Tumors

Leslie N. Sutton, MD; Scott L. Simon, MD; Mark G. Burnett, MD; Sumeet Garg, BA; Albert Telfel, MD, PhD; John Dormans, MD (Philadelphia, PA)

8:35-8:45 AM

3. Restoration of Neurodevelopment after a Prenatal Ischemic Insult in Rats

Shenandoah Robinson, MD; Kathryn H. O'Keefe, BA; Qing Li, MD; Nanor Tabrizi, BA; Robert H. Miller, PhD (Cleveland, OH)

8:45-8:55 AM

4. Observation of CSF Bulk Flow in the Undisturbed Subarachnoid Space Over the Convexity of the Rat Brain

Shinya Yamada, MD; Takahashi Kouichi, MD; George McNamara, PhD; Gordon McComb, MD (Los Angeles, CA)

8:55-9:05 AM

5. Radiosurgery for Intracranial Arteriovenous Malformations in Children

Hans G. Eder, MD, PhD; Klaus A. Leber, MD; Senta Kurschel, MD (Graz, Austria)

9:05-9:15 AM

6. Young Age Moyamoya Disease: Role of Aggressive Surgical Treatment

Kyu-Chang Wang, MD, PhD; Ho-Jun Seol, MD; Seung-Ki Kim, MD, PhD; You-Nam Chung, MD; Hee-Soo Kim, MD, PhD; Byung-Kyu Cho, MD, PhD (Seoul, Korea)

9:15-9:25 AM

7. Serial MRS Metabolite Changes in Treated Pediatric Acute and Sub-Acute Hydrocephalus

Miriam Scadeng, MD; Stefan Bluml, PhD; Rex A. Moats, PhD; Marvin D. Nelson, MD; Gordon J. McComb, MD (Los Angeles, CA)

9:25-9:35 AM

8. Restrictive Periventricular Gliosis—Fact or Fiction

Sandeep Sood, MD; Jaliya Lokuketagoda, MD; Steven D. Ham, DO (Detroit, MI)

9:35-9:45 AM

9. Oxidative Metabolism Correlated to Symptomatology and Prognosis of Tethered Cord Syndrome

Shokei Yamada, MD, PhD; Daniel J. Won, MD (Loma Linda, CA); Margaret Wacker, MD (Riverside, CA); Santiago Figueroa, MD (Loma Linda, CA)

9:45-9:55 AM

10. The Role of Anti-Copper Antiangiogenic Agent Tetrathiomolybdate in Treatment of Human Medulloblastomas

Sami Khoshyomn, MD; Erica F. Bisson, MD; Samuel B. Braff, BA; Bruce I. Tranmer, MD (Burlington, VT)

9:55-10:05 AM

11. Pediatric Neurosurgery Research: A Survey Study

Todd A. Maugans, MD (Burlington, VT)

10:05-10:30 AM

Beverage Break in Exhibit Hall with Exhibit and Poster Viewing

10:30 AM-Noon

SCIENTIFIC SESSION II— CRANIOSYNTOSIS AND CRANIOFACIAL SURGERY

Moderators: Ann Marie Flannery, MD;

James Goodrich, MD

Learning Objectives

Upon completion of this program, the participants should be able to:

1. Differentiate various approaches to the management of craniosynostosis and craniofacial surgery and distinguish appropriate applications.

10:30-10:40 AM

12. Minimally Invasive Correction of Craniosynostosis: Definitive Osteotomies and Immediate Fixation with Resorbable Devices

Burak M. Ozgur, MD; Hal S. Meltzer, MD; Ralph E. Holmes, MD; Steven R. Cohen, MD (San Diego, CA)

10:40-10:50 AM

13. Fussiness and Irritability in Patients with Craniosynostosis

Cathy C. Cartwright, MSN; David F. Jimenez, MD (Columbia, MO)

10:50-11:00 AM

14. Scaphocephaly Without Synostosis: The Sticky Sagittal Suture

James E. Baumgartner, MD; John F. Teichgraeber, MD; Kelly Seymour-Dempsey, MD (Houston, TX)

11:00-11:10 AM

15. The Sagittal Suture May Close First in Late Premature Closure of Multiple Sutures: Oxicephaly

Donna C. Wallace, MSN, NP; Harold L. Rekate, MD, FACS; Edward F. Joganic, MD, FACS; Stephen P. Beals, MD, FACS (Phoenix, AZ)

11:10-11:20 AM

16. Multiple-Revolution Spiral Osteotomy for Cranial Reconstruction: A Report of Long Term Outcomes on 29 Patients

Farrokh R. Farrokhi, MD; Micam W. Tullous, MD; Peter T.H. Wang, MD; Patricia A. Mancuso, MD; Dennis G. Vollmer, MD (San Antonio, TX)

11:20-11:30 AM

17. Experience with Frontolateral Keyhole Craniotomy through a Superciliary Skin Incision in Children

Laszlo Bogner, MD, PhD; Gabor Madarassy, MD (Budapest, Hungary)

11:30 AM-Noon

Review of On-Going Studies

Brain Tumors: Jeffrey Wisoff, MD

Shunt Infection: John Kestle, MD

Noon-1:15 PM

Lunch in the Exhibit Hall and Poster Viewing

1:15-3:15 PM

SCIENTIFIC SESSION III— DYSRAPHISM AND CONGENITAL ANOMALIES

Moderators: John Grant, MD;

Cheryl Muszynski, MD

Discussants: Ann Marie Flannery, MD;

Mark Dias, MD

Learning Objectives

Upon completion of this program, the participants should be able to:

1. Examine the diagnosis and treatment of dysraphic syndrome and congenital anomalies.

1:15-1:30 PM

18. Prenatal Magnetic Resonance Imaging in the Diagnosis of Fetal CNS Abnormalities

Kimberly D. Bingaman, MD; Juliette A. Prust; Kim M. Walker, NP; Dorothy I. Bulas, MD; Robert F. Keating, MD; Philip H. Cogen, MD, PhD (Washington, DC)

1:30-1:45 PM

19. The Fetal Myelomeningocele Trial

Leslie N. Sutton, MD (Philadelphia, PA); Noel Tulipan, MD (Nashville, TN); Nalin Gupta, MD (San Francisco, CA)

1:45-1:55 PM

20. Inclination of the Odontoid Process in the Pediatric Chiari I Malformation

R. Shane Tubbs, PhD, MS, PA-C; Matthew D. Smyth, MD; John C. Wellons, III, MD; Jeffrey P. Blount, MD; Paul A. Grabb, MD; W. Jerry Oakes, MD (Birmingham, AL)

1:55-2:05 PM

21. Management of Cervical Spine in Klippel-Feil Patients

Fassett R. Daniel, MD, MBA; Brockmeyer L. Douglas, MD (Salt Lake City, UT)

2:05-2:15 PM

22. Treatment of Basilar Invagination in the Pediatric Population Using Cervical Reduction and Posterior Occipital-Cervical Fusion

Louis J. Kim, MD; Harold L. Rekate, MD, FACS; Jeffrey D. Klopfenstein, MD; Volker K. H. Sonntag, MD (Phoenix, AZ)

2:15-2:25 PM

23. Vestibular Function in Adolescent Idiopathic Scoliosis

Matthew T. Provencher, MD; Derin C. Wester, PhD (San Diego, CA); Bruce L. Gillingham, MD (Coronado, CA); Kim L. Gottschall, PhD; Michael Hoffer, MD (San Diego, CA)

2:25-2:35 PM

24. Improvement in Brainstem Auditory Evoked Potentials after Suboccipital Decompression in Patients with Chiari I Malformation

Richard C. Anderson, MD; Ronald G. Emerson, MD; Kathryn C. Dowling, EdD; Neil A. Feldstein, MD (New York, NY)

2:35-2:45 PM

25. Preliminary Analysis of Genetics of Lipomyelomeningocele

Timothy M. George, MD; Marcy Speer, PhD (Durham, NM); NTD Collaborative Group

2:45-2:55 PM

26. Atlantoaxial Distances in Children During Cervical Spine Flexion

Matthew D. Smyth, MD; R. Shane Tubbs, PhD, MS, PA-C; John C. Wellons, MD; Jeffrey P. Blount, MD; Paul A. Grabb, MD; W. Jerry Oakes, MD (Birmingham, AL)

Program Schedule

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2:55-3:05 PM
27. Children with Growth Hormone Deficiency and Chiari I Malformation: A Morphometric Analysis of the Posterior Cranial Fossa
R. Shane Tubbs, PhD, MS, PA-C; Matthew D. Smyth, MD; John C. Wellons, III, MD; Alfred A. Bartolucci, PhD; Jeffrey P. Blount, MD; Jerry W. Oakes, MD; Paul A. Grabb, MD (Birmingham, AL)

3:05-3:15 PM
28. Acquired Chiari I Malformation and Slit-Ventricle Syndrome
Concezio Di Rocco, Prof; Francesco Velardi, MD (Rome, Italy)

3:15-3:45 PM
Beverage Break with Exhibitors and Poster Viewing

3:45-5:15 PM SCIENTIFIC SESSION IV— FUNCTIONAL AND EPILEPSY

Moderators: Glenn Morrison, MD; Fredrick Boop, MD

Learning Objectives
Upon completion of this program, the participants should be able to:

1. Integrate concepts related to functional and epilepsy surgery in the pediatric patient.

3:45-3:55 PM
29. Complications of Vagal Nerve Stimulation for Epilepsy in Children
Matthew D. Smyth, MD; R. Shane Tubbs, PhD, MS, PA-C; Paul A. Grabb, MD; Jeffrey P. Blount, MD (Birmingham, AL)

3:55-4:05 PM
30. Changes in the Gross Motor Function Measure (GMFM) Following Selective Dorsal Rhizotomy (SDR)
Jack Ensberg, PhD; Sandy Ross, PT; T.S. Park, MD; Jeffrey Ojemann, MD (St. Louis, MO)

4:05-4:15 PM
31. Long Term Follow-up of Selective Dorsal Rhizotomy: Does Multi-level Laminotomy Influence the Risk of Scoliosis?
J. A. Hall, MD, MSc; J.P. Farmer, MD; A. O'Gorman, MD (Montreal, Canada)

4:15-4:25 PM
32. Surgical Management of Pediatric Epilepsy: Bringing Treatment Home to Indiana's Children
Jodi L. Smith, MD, PhD (Indianapolis, IN)

4:25-4:35 PM
33. Epilepsy Surgery in Tuberous Sclerosis
Howard L. Weiner, MD; Werner K. Doyle, MD (New York, NY); Pantaleo Romanelli, MD (Palo Alto, CA); Souhel Najjar, MD; Daniel Miles, MD; Orrin Devinsky, MD (New York, NY)

4:35-4:45 PM
34. Rate of Orthopedic Surgery After Selective Dorsal Rhizotomy in Relation to Ambulatory Status in Spastic Diplegia
Donncha F. O'Brien, MD; T.S. Park, MD; Eric C. Leuthardt, MD; Jeffrey R. Leonard, MD; Jeffrey G. Ojemann, MD (St. Louis, MO)

4:45-4:55 PM
35. Febrile Convulsions, Cortical Dysplasia, and Intractable Temporal Lobe Epilepsy in Children
Ann-Christine Duhaime, MD (Lebanon, NH); Brenda E. Porter, MD; Alex R. Judkins, MD; Robert R. Clancy, MD; Dennis J. Dlugos, MD; Jeffrey A. Golden, MD (Philadelphia, PA)

4:55-5:05 PM
36. Temporal Lobe Tumor Related Epilepsy in Childhood
Oguz Cataltepe, MD; Nejat Akalan, MD; Meral Topcu, MD; Serap Saygi, MD; Guzide Turanli, MD; Dilek Yalnizoglu, MD; Nese Dericioglu, MD (Ankara, Turkey)

5:05-5:15 PM
37. High Resolution MRI Enhances Identification of Lesions Amenable to Surgical Therapy in Children with Refractory Epilepsy
Natasha McKay, MD; Shenandoah Robinson, MD; Monisha Goyal, MD; Barbara Bangert, MD; Max Wiznitzer, MD; Mark Scher, MD; Barbara Swartz, MD (Cleveland, OH)

5:15-5:45 PM
Annual Business Meeting

Friday, December 6

7:00 AM-4:00 PM
Registration

7:00-8:00 AM
Continental Breakfast with Exhibit and Poster Viewing

7:00 AM-4:00 PM
Exhibits and Poster Viewing

8:00-10:00 AM SCIENTIFIC SESSION V— HYDROCEPHALUS

Moderators: Mark Luciano, MD; Joseph Piatt, MD

Discussants: Jogi Pattisapu, MD; Harold L. Rekate, MD

Learning Objectives
Upon completion of this program, the participants should be able to:

1. Distinguish the benefits of the evaluation and treatment of hydrocephalus and related disorders.

8:00-8:15 AM
38. In Vitro Testing of Current Spread During Ventricular Catheter Coagulation Using Diathermy
Frederick A. Boop, MD; John Honeycutt, MD; Jeffrey Sorenson, MD (Memphis, TN); Bruce Cherny, MD (Mesa, AZ)

8:15-8:30 AM
39. Use of a Novel Continuous Long-Term Intracranial Pressure Monitor in Ambulatory Hydrocephalus Patients
David M. Frim, MD, PhD (Chicago, IL); David Adelson, MD (Pittsburgh, PA); John Kestle, MD; Marion L. Walker, MD (Salt Lake City, UT); Keith Miesel, PhD; Lee Stylos, PhD (Minneapolis, MN); Richard Penn, MD (Chicago, IL)

8:30-8:40 AM
40. The Impact of Shunting in Congenital and Perinatal Hydrocephalus: Long-term Outcome in Adult Survivors
Venkat Sadanand, MD, MS, PhD (Saskatoon, Canada); Fred Gentili, MD, FRCS(C), MSc (Toronto, Ontario, Canada)

8:40-8:50 AM
41. Diffusion Weighted Imaging in Acute and Treated Raised Pressure Pediatric Hydrocephalus
Miriam Scadeng, MD; Marvin D. Nelson, MD; Gordon J. McComb, MD (Los Angeles, CA)

8:50-9:00 AM
42. What is the Ideal Initial Valve Pressure Setting in Infants with Programmable Hakim Valves?
Marcus C. Korinth, MD; Martin R. Weinzierl, MD; Joachim M. Gilsbach, MD (Aachen, Germany)

9:00-9:10 AM
43. "Quick Brain" MRI Versus CT Scan for Evaluating Shunted Hydrocephalus
Joshua E. Medow, MD; Bermans J. Iskandar, MD; Rowley Howard, MD (Madison, WI)

9:10-9:20 AM
44. Mortality After Ventricular Shunt Placement: A Retrospective Cohort Study
Edward R. Smith, MD; William E. Butler, MD; Fred G. Barker, MD (Boston, MA)

9:20-9:30 AM
45. The Predictive Value of a Normal Shuntogram
Jeffrey G. Ojemann, MD; Michael Taylor, PA-C; T.S. Park, MD (St. Louis, MO)

9:30-9:40 AM
46. Retrograde Venography in the Evaluation of Pseudotumor Cerebri in the Pediatric Population
Vivek R. Deshmukh, MD; Felipe C. Albuquerque, MD; Cameron C. McDougall, MD; Harold L. Rekate, MD, FACS (Phoenix, AZ)

9:40-9:50 AM
47. Shunt Pseudotumor
Julian J. Lin, MD; Fredrick A. Boop, MD; Robert A. Sanford, MD (Memphis, TN)

9:50-10:00 AM
48. Low Pressure Hydrocephalus: Diagnosis and Treatment Issues
Mark G. Hamilton, MD, FRCS(C), MD (Calgary, Canada); Angela V. Price, MD, FRCS(C) (Vancouver, Canada)

10:00-10:30 AM
Beverage Break in the Exhibit Hall with Exhibit and Poster Viewing

10:30-11:20 AM SCIENTIFIC SESSION VI— NEURO-ONCOLOGY

Moderators: Karin Muraszko, MD; Herbert Fuchs, MD

Learning Objectives
Upon completion of this program, the participants should be able to:

1. Recognize the current best practices on the evaluation and treatment of pediatric brain tumor.

10:30-10:40 AM
49. MRI Study of the Natural History of Ventriculomegaly and Tonsillar Herniation in Children with Posterior Fossa Tumours
Kanna Gnanalingham, PhD; Jesus Lafuente, FRCS; Dominic Thompson, FRCS; William Harkness, FRCS; Richard Hayward, FRCS (London, United Kingdom)

10:40-10:50 AM
50. Myxopapillary Ependymomas in Children: Results of Surgery
George I. Jallo, MD; Sean Wilson, BA; Karl Kothbauer, MD; Fred Epstein, MD (New York, NY)

10:50-11:00 AM
51. Current Diagnosis and Treatment Strategies in Germ Cell Tumors of the Pineal and Hypothalamic Regions
Greg Olavarria, MD; Ramon Navarro, MD; Tadanori Tomita, MD; (Chicago, IL)

11:00-11:10 AM
52. Presenter Declined

11:10-11:20 AM
53. Independent Learning Modules for Pediatric Neurosurgery
Jonathan Martin, MD; Leon E. Moores, MD (Washington, DC)

11:20-11:25 AM
Introduction of 2002 Raimondi Lecturer by Harold L. Rekate, MD

11:25 AM-Noon
Raimondi Lecture—"Assessment of Competence in Neurosurgery"
by Volker K. H. Sonntag, MD Co-Director, Department of Neurosurgery Barrow Neurological Institute Chairman, ABNS Committee on Assessment of Competence

Moderator: Harold L. Rekate, MD

Learning Objectives
Upon completion of this program, the participants should be able to:

1. Recognize his or her responsibilities regarding individual maintenance of knowledge base and skills required to continue to practice neurosurgery.
2. Assess competency in neurosurgery.
3. Recognize the methods by which the American Board of Neurological Surgery will judge competence in the neurosurgeons it certifies.

Noon-1:15 PM
Lunch in Exhibit Hall and Poster Viewing

1:15-2:55 PM SCIENTIFIC SESSION VII— TUMORS

Moderators: Jeffrey Wisoff, MD; Leland Albright, MD
Discussants: Concezio Di Rocco, MD; Arnold Menezes, MD

Learning Objectives
Upon completion of this program, the participants should be able to:

1. Recognize the current best practices in the evaluation and treatment of pediatric brain tumor.

2:15-1:30 PM
54. Posterior Fossa Tumour Surgery in Children: Does Craniotomy Lead to Fewer Complications than a Craniectomy?
Kanna Gnanalingham, PhD; Jesus Lafuente, FRCS; Dominic Thompson, FRCS; William Harkness, FRCS; Richard Hayward, FRCS (London, United Kingdom)

Program Schedule

1:30-1:45 PM (S)

55. Cooperation for the Chiari Malformations
David J. Sacco, MD; R. Michael Scott, MD (Boston, MA)

1:45-1:55 PM
56. Craniopharyngiomas: Complications Related to Complex Approaches in Children
Sean McNatt, MD (Sherman Oaks, CA); Michael P. Kim, BS (Chapel Hill, NC); Michael L. Levy, MD, PhD (San Diego, CA)

1:55-2:05 PM
57. Juvenile Pilocytic Astrocytoma of the Brainstem
John R.W. Kestle, MD; Douglas D. Brockmeyer, MD; Marion L. Walker, MD (Salt Lake City, UT)

2:05-2:15 PM
58. NF1 and the Pediatric Neurosurgeon—20 Year Institutional Review
Jean Pierre Farmer, MD; Saad Khan, MD; Asad Khan, PhD; June Ortenberg, MD; Carolyn Freeman, MD; Augustin M. O'Gorman, MD; Jose Montes, MD (Montreal, Canada)

2:15-2:25 PM
59. The Far Lateral Approach to Cerebello-Pontine Angle Ependymomas
Sunita Bhamidipaty, MD (Sherman Oaks, CA); Danita Tom, BS (Madison, WI); Michael L. Levy, MD, PhD (San Diego, CA)

2:25-2:35 PM
60. Lessons Learned Managing Malignant Gliomas of the Thalamus in Children
F. A. Boop, MD; R. A. Sanford, MD; A. Dyrer, MD; M.M. Muhlbauer, MD; S.J. Einhaus, MD; L.E. Kun, MD; T. Merchant, MD; A. Gajjar, MD; J. Jenkins, MD (Memphis, TN)

2:35-2:45 PM
61. High Incidence of Tumor Dissemination in Myxopapillary Ependymoma in Pediatric Patients
Daniel R. Fassett, MD, MBA; James Pingree, MD; John Kestle, MD (Salt Lake City, UT)

COCPU 43343 (17)

COCPU 43443 (18)

(5)

2:45-2:55 PM
62. Malignant Rhabdoid Tumors of the Brain
Bradley E. Weprin, MD; Kenneth N. Shapiro, MD; Frederick Sklar, MD; Dale M. Swift, MD; Korgan Koral, MD; Lynn Gargan, PhD; Jill Mraz, ARNP; Shane Valles, CRA (Dallas, TX)

2:55-3:15 PM
Beverage Break in the Exhibit Hall with Exhibit and Poster Viewing

3:15-4:15 PM
SCIENTIFIC SESSION VIII—
DYSRAPHISM II
Moderators: Timothy George, MD; Michael Parrington, MD
Learning Objectives
Upon completion of this program, the participants should be able to:

3:15-3:25 PM
63. Epidemiological Changes in Spinal Dysraphism
Jeffrey W. Campbell, MD (Charleston, SC)

3:25-3:35 PM
64. Pediatric Hydrocephalus Treatment Using a Flow Regulating Shunt: A Prospective Five Years Shunt Survival Study (European OSV II Study)
Patrick W. Hanlo, MD (Utrecht, Netherlands); Giuseppe Cinalli, MD (Paris, France); Peter Vandertop, MD (Amsterdam, Netherlands); Lars Bogeskov, MD (Copenhagen, Denmark); Svend E. Borgesen, MD; Jürgen Boschert, MD (Mannheim, Germany); Paul Chumas, MD (Leeds, England); Hans Eder, MD (Graz, Austria); Ian Pople, MD (Bristol, England); Willy Serlo, MD (Oulu, Sweden)

3:35-3:45 PM
65. Isolated Lumbosacral Strawberry Nevi Do Not Indicate The Presence of Occult Spinal Dysraphism
Nathan R. Selden, MD, PhD (Portland, OR); Rebecca M. Allen, BA (Stanford, CA); Michael A. Sandquist, MD (Portland, OR); Joseph H. Piatt, MD, FACP (Philadelphia, PA)

3:45-3:55 PM
66. The Tethered Cord in Arthrogyposis Multiplex Congenita
Kimberly D. Bingaman, MD; Laura L. Tosi, MD; Philip H. Cogen, MD, PhD; Robert F. Keating, MD (Washington, DC)

3:55-4:05 PM
67. Syringobulbia in a Pediatric Population
Jeremy D.W. Greenlee, MD; Kathleen A. Donovan, ARNP; Arnold H. Menezes, MD (Iowa City, IA)

4:05-4:15 PM
68. Mechanisms of Reclosure in a Surgical Model of Myelomeningocele: Implications for Fetal Surgery
David C. Adamson, MD, PhD; Timothy M. George, MD (Durham, NC)

4:15-5:30 PM
Wine & Cheese Reception with Exhibit and Poster Viewing

6:00-7:30 PM
Friday Night Social at the Lawn Court at the Hyatt Regency Scottsdale
A networking opportunity for Medical Registrants of the Pediatric Section Annual Meeting.

Saturday, December 7

7:00 AM-Noon
Registration

7:00-8:00 AM
Continental Breakfast with Exhibit and Poster Viewing

7:00-11:00 AM
Exhibit and Poster Viewing

8:00-10:00 AM
SCIENTIFIC SESSION IX—
ENDOSCOPY AND TRAUMA

Moderators: Andrew Parent, MD; Alan Cohen, MD
Discussants: Ann-Christine Duhaime, MD; Alan Cohen, MD

Learning Objectives
Upon completion of this program, the participants should be able to:
1. Identify the current trends in intracranial endoscopy and pediatric trauma.

8:00-8:15 AM
69. Reducing the Incidence of Abusive Head Injuries Through Hospital-Based Parent Education: The Upstate New York Shaken Baby Syndrome Education Program
Mark S. Dias, MD (Hershey, PA); Kim Smith, RNC; Kathy deGuehery, RNC; Diana Kachurek, RNC (Buffalo, NY); Veetai Li, MD (New York, NY)

8:15-8:30 am
70. Neuroendoscopic Findings in Patients with Intracranial Germinomas Correlate with Diabetes Insipidus
John C. Wellons, MD; Richard S. Tubbs, PhD, PA-C; Alyssa T. Reddy, MD; Hussein Abdullatif, MD; Walter J. Oakes, MD; Jeffrey P. Blount, MD; Paul A. Grabb, MD (Birmingham, AL)

8:30-8:40 AM
71. Failed Endoscopic Third Ventriculostomy in Children: Management Options
Aaron Mohanty, MD; Thimappa Hegde, MD; S. Sampath, MD; M. K. Vasudev, MD; Sastry V. R. Kolluri, MD (Bangalore, India)

8:40-8:50 AM
72. Results of Endoscopic Septal Fenestration in the Treatment of Isolated Ventricular Hydrocephalus
Philipp R. Aldana, MD; John R.W. Kestle, MD; Douglas L. Brockmeyer, MD; Marion L. Walker, MD (Salt Lake, UT)

8:50-9:00 AM
73. Management of Brain Tumor Related Obstructive Hydrocephalus with Endoscopic Third Ventriculostomy
Karl F. Kothbauer, MD (New York, NY); Bong Soo Kim, MD (Chicago, IL); George I. Jallo, MD; Rick Abbott, MD (New York, NY)

9:00-9:10 AM
74. Frameless Stereotactic Endoscopic Plate and Nail Approach to the Third Ventricle
John A. Lancon, MD (Jackson, MS)

9:10-9:20 AM
75. Sports and Recreational Head and Spine Injuries in Children
David J. Yeh, MD; Scott Y. Rahimi, MD; Ann Marie Flannery, MD; Mark R. Lee, MD, PhD (Augusta, GA)

9:20-9:30 AM
76. Long Term Outcome Following Gunshot Wounds to the Spine in Children and Adolescents
Anthony Kim, MD, PhD (Sherman Oaks, CA); Michael L. Apuzzo, MD (Los Angeles, CA); Michael L. Levy, MD, PhD (San Diego, CA); Rod Adkins, PhD (Los Angeles, CA)

9:30-9:40 AM
77. Operative Compound Skull Fractures in Children
Richard B. Rodgers, MD; Stephen Kralik, BS; Thomas G. Luerssen, MD (Indianapolis, IN)

9:40-9:50 AM
78. Third Ventriculostomy Safety Device: A Preliminary Report
Todd A. Maugans, MD (Burlington, VT)

9:50-10:00 AM
79. Endoscopic Intracranial Cyst Fenestration in Neonates and Infants
Christian W. Sikorski, MD; Yamini Bakhtiar, MD; David M. Frim, MD, PhD (Chicago, IL)

10:00-10:30 AM
Beverage Break in the Exhibit Hall with Exhibit and Poster Viewing

10:30 AM-Noon
SCIENTIFIC SESSION X—
PEDIATRIC NEUROSURGERY
POTPOURRI
Moderators: Aurthur Marlin, MD; Paul Steinbok, MD

Learning Objectives
Upon completion of this program, the participants should be able to:
1. Examine current treatment and outcomes in a variety of Neurosurgical conditions.

10:30-10:40 AM
80. Aberrant Neuronal Development in Hydrocephalus
Jonathan P. Miller, MD; David W. Leifer, BS; David Lust, PhD; Alan R. Cohen, MD (Cleveland, OH)

10:40-10:50 AM
81. Pitfalls in the Use of a Hydroxyapatite/Absorbable Plate Construct to Repair Full Thickness Skull Defects in Children
Gerald F. Tuite, MD (Saint Petersburg, FL); Ernesto Ruas, MD (Tampa, FL); Carolyn Carey, MD (Saint Petersburg, FL)

10:50-11:00 AM
82. The Repair of Large (>5cm²) Skull Defects Using "Reinforced" Hydroxyapatite Cement: Technique and Complications
Susan Durham, MD (Oregon City, OR); Michael L. Levy, MD; J.G. McComb, MD (Los Angeles, CA)

11:00-11:10 AM
83. Cisterna Magna to Pleural Shunting in Pseudotumor Cerebri Patients Who Fail Lumbar or Ventricular Shunting
Bakhtiar Yamini, MD; David Frim, MD, PhD (Chicago, IL)

11:10-11:20 AM
84. Management of Arachnoid Cysts in Children with Refractory Epilepsy
James E. Baumgartner, MD; James W. Wheless, MD (Houston, TX)

11:20-11:30 AM
85. Algorithm for the Evaluation of the Pediatric Patient with a Spontaneous Intracranial Hematoma
Carrie L. Antone, MD (Beltsville, MD); Robert F. Keating, MD (Washington, DC)

11:30 AM-Noon
Open Discussion

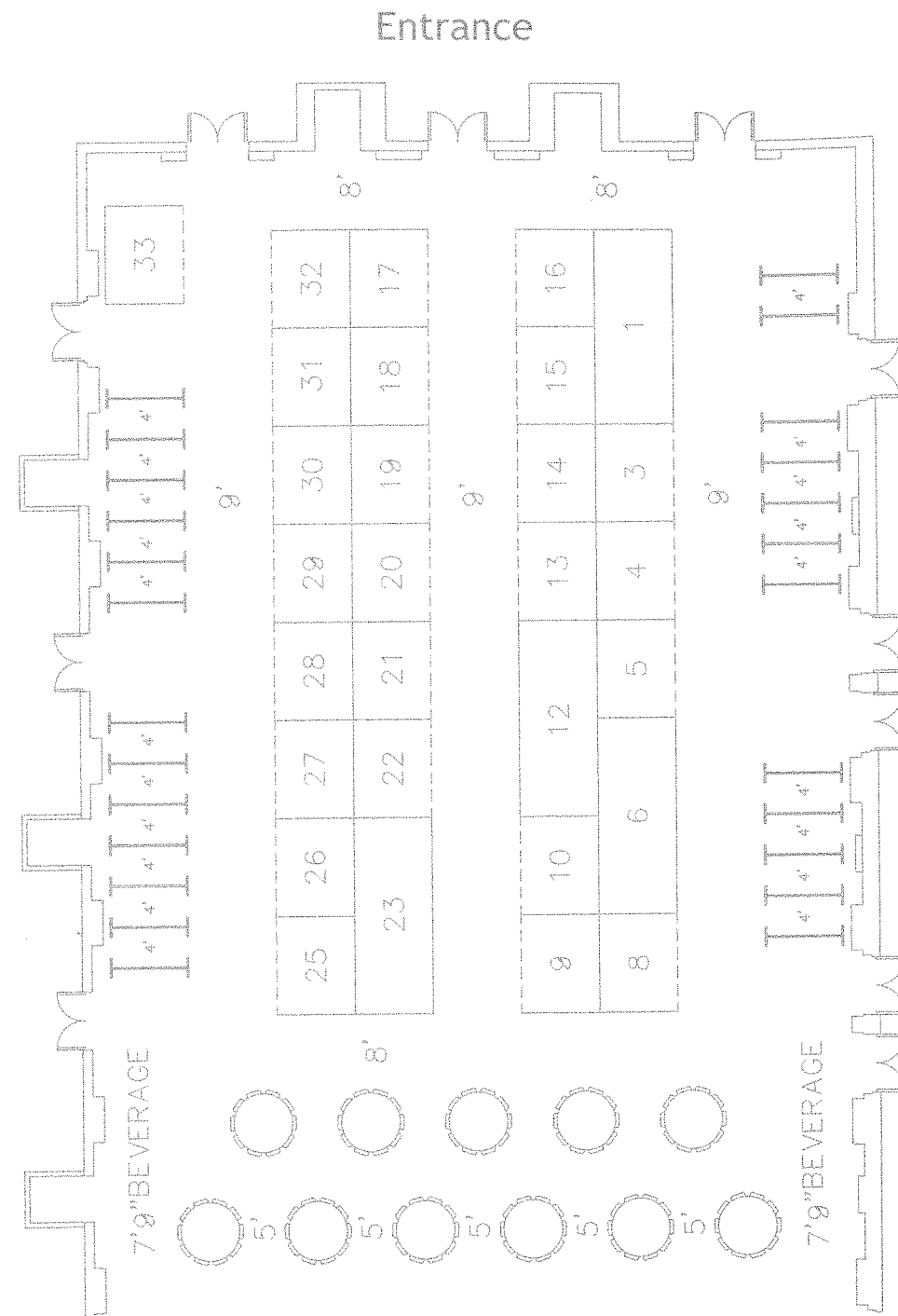
Noon-12:05 PM
Closing Remarks
Harold L. Rekate, MD

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 L.E. Kun, MD
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 Senta Kurschel, MD
 Yasuko Kusaka, MD, PhD
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 David W. Leifer, BS
 Jeffrey R. Leonard, MD
 Eric C. Leuthardt, MD

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 Michael L. Levy, MD, PhD
 Qing Li, MD
 Veetai Li, MD
 Terry Lichtor, MD, PhD
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 Jaliya Lokuketagoda, MD
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 Nanor Tabrizi, BA
 Michael Taylor, PA-C

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 Dominic Thompson, FRCS
 Jerome W. Thompson, MD, MBA
 John A. Thorne, FRCS
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 Tadanori Tomita, MD
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 Meral Topcu, MD
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 Christopher Troup, MD
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 Richard S. Tubbs, PhD, PA-C
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 David Wright, PhD
 David M. Wrubel, MD
 Dilek Yalmozoglu, MD
 Shinya Yamada, MD
 Shokei Yamada, MD, PhD
 Bakhitar Yamini, MD
 David J. Yeh, MD
 Takashi Yoshimoto, MD, PhD

1. Immuno-Gene Therapy for Malignant Pediatric Brain Tumors

Roberta P. Glick, MD; Terry Lichtor, MD, PhD; Edward P. Cohen, MD; Henry Lin, BA; Kimberly Dean, BA (Chicago, IL)

INTRODUCTION Brain tumors account for approximately 20% of all childhood neoplasms. Despite considerable progress in surgery, radiation and chemotherapy, local recurrence continues to be an important reason for treatment failure. Local recurrence rates can be as high as 80% for malignant gliomas. Thus, the current prognosis for pediatric patients with malignant brain tumors remains poor, and new and innovative forms of effective treatments are urgently needed. We have previously shown that intracerebral (i.c.) injection of allogeneic fibroblasts genetically engineered to secrete interleukin-2 (IL-2) results in prolongation of survival in an adult mouse with an i.c. glioma. Treated animals developed strong antitumor immunocytotoxic responses as measured by a Cr-release assay. No toxicity has been associated with i.c. immunization in adult mice.

METHODS The goal of this study was to determine if we could treat a malignant glioma (G1261) in a mouse (C57Bl/6) in the pediatric age group range (weanlings: 2-3 weeks, and adolescent: 3-4 weeks). In a series of experiments, mice were injected i.c. with a mixture of glioma cells and allogeneic fibroblasts genetically engineered to secrete IL-2. Control animals received either a mixture of glioma cells and non-secreting allogeneic fibroblasts, or glioma cells and media. Survival time and immune responses were investigated.

RESULTS The results demonstrate a significant prolongation of survival in animals with an i.c. glioma treated with i.c. injection of IL-2 secreting allogeneic fibroblasts ($P < 0.05$). There was no increased morbidity or mortality as a result of i.c. immunization. In addition, in the very young mouse experiments, there were several long term survivors who have been rechallenged with tumor to investigate the induction of immune memory by this treatment.

CONCLUSIONS These results demonstrate the efficacy and safety of using i.c. injection

with IL-2 secreting allogeneic fibroblasts as a tumor vaccine for the treatment of malignant glioma in mice in the pediatric age group.

2. The Efficacy of Prophylactic Spinal Fusion in the Prevention of Post-surgical Spinal Deformity in Pediatric Intramedullary Spinal Cord Tumors

Leslie N. Sutton, MD; Scott L. Simon, MD; Mark G. Burnett, MD; Sumeet Garg, BA; Albert Telfeian, MD, PhD; John Dormans, MD (Philadelphia, PA)

INTRODUCTION The increased risk of spinal deformity after resection of spinal cord tumors in children has been described. However, the efficacy of prophylactic spinal fusion at the time of, or soon after, surgical resection in order to prevent post-operative spinal deformity in this population is unclear.

METHODS A retrospective review was conducted on patients with intramedullary spinal cord tumors treated at the Children's Hospital of Philadelphia. The office charts, radiology reports, and tumor data bases were analyzed. The development and progression of post-operative deformity were studied and compared against the patient's age, presentation, diagnosis, pre-operative alignment, procedure (laminectomy, laminoplasty, fusion), levels, post-operative radiation, and the timing, type and extent of fusion and fixation.

RESULTS Of the 25 patients treated, 8 were females and 17 were males. The mean age was 8.7 years (0.1-14.0 years) and the mean duration of follow-up was 56.7 months (1-268 months). Seven patients underwent posterior fusion at the time of resection, 1 patient had a two level anterior cervical fusion 1 month after his resection, and 3 patients had laminoplasties preformed. Of these 25 patients, 12 developed significant post-operative spinal deformities, 2 of which had laminoplasties, and 1 of which had a posterior fusion without instrumentation at the time of their resections. Two of these patients experienced progression of their deformity even after subsequent posterior fixation

and fusions. None of the patients who underwent prophylactic instrumented fusions progressed to develop significant spinal deformities.

CONCLUSIONS Prophylactic instrumented spinal fixation and fusion decreases the risk of post-surgical spinal deformity in pediatric spinal cord tumors.

3. Restoration of Neurodevelopment after a Prenatal Ischemic Insult in Rats

Shenandoah Robinson, MD; Kathryn H. O'Keefe, BA; Qing Li, MD; Nanor Tabrizi, BA; Robert H. Miller, PhD (Cleveland, OH)

INTRODUCTION Children that suffer perinatal insults frequently develop cerebral palsy, epilepsy, and cognitive delay. Perturbation of oligodendrocyte development causes white matter lesions (WMLs) and cerebral palsy, and neuronal development causes epilepsy and cognitive delay. We propose that because oligodendrocytes and GABAergic interneurons arise from the same stem cells, that both of their lineages are disrupted by perinatal insults. Second, we hypothesize that perinatal insult-induced pro-inflammatory cytokines mediate this disruption. Previously we used a rat prenatal ischemic model to induce lesions that mimic human WMLs. Here we investigated whether GABAergic neurons are also affected, and present preliminary data describing an intervention to restore neurodevelopment, namely, a systemic pro-inflammatory cytokine suppressant, metallothionein (MT).

METHODS Uterine arteries of pregnant rats were occluded for 45 minutes on E18, and the pups were born at term. In sham controls the arteries were exposed, but not occluded. Cell counts were performed on at least three sections from each rat, and from at least three rats per group. The t-test was used to identify significant differences.

RESULTS Immunohistochemistry demonstrated minimal change in CA1 neuronal density in post-insult P13 rats. By contrast, a significant reduction of CA1 GAD (glutamic-acid-decarboxylase)-positive

GABAergic neurons was present in post-insult rats (0.148 cells/mm³), compared to shams (0.106 cells/mm³, $p < .004$), suggesting that GABAergic neurons were particularly susceptible to injury. These findings will be correlated with post-mortem human infant brains. To test the intervention, intraperitoneal metallothionein was administered on P0 and P1. MT-treated P9 post-insult rats had significantly more periventricular CC1+ oligodendrocytes ($p < 0.02$) and CXCR2+ cells ($p < 0.0002$) than saline-control post-insult rats, and similar numbers to sham surgery controls.

CONCLUSIONS These results support the hypotheses that 1) perinatal insults affect both oligodendrocytes and GABAergic neurons, and that 2) insult-induced pro-inflammatory cytokines disrupt neurodevelopment. They will guide development of novel interventions to restore the neonatal brain after injury.

4. Observation of CSF Bulk Flow in the Undisturbed Subarachnoid Space Over the Convexity of the Rat Brain

Shinya Yamada, MD; Takahashi Kouichi, MD; George McNamara, PhD; Gordon McComb, MD (Los Angeles, CA)

INTRODUCTION To directly observe cerebrospinal fluid (CSF) flow over the convexity of the undisturbed subarachnoid space (SAS) of the rat brain.

METHODS These experiments were done using anesthetized male Wistar rats weighting 350–400g. The skull was exposed via midline incision with the left side of the skull being thinned with a high speed drill to make it transparent but keeping enough bone present so as not to alter the intracranial contents. A total volume of 100ml of fluorescent microbeads (1mm dia.) was introduced into the lumbar subarachnoid space (SAS) via a catheter. A stereotactic fluorescent microscope with a digital CCD camera was used to capture serial images of the microbeads in the CSF at the rate of 0.03 sec/frame. The direction and rate of CSF flow were estimated by analyzing the movements of the microbeads. The brain was removed and inspected at the end of 4 hours.

RESULTS Most microbead movement in the SAS took place adjacent to blood vessels. Beads did not flow to the superior sagittal sinus (SSS). The CSF flow direction was consistent even if the intracranial pressure was raised by Valsalva maneuver. The average CSF flow rate in the SAS exposed by the cranial window at normal CSF pressure was 7.6 ± 6.6 mm/sec. Inspection of the brain removed at the end of the experiment showed that the microbeads accumulated in the olfactory lobes, basal cisterns, and along the larger blood vessels on the brain surface.

CONCLUSIONS Microbeads were observed moving in the undisturbed SAS. The microbeads did not move toward nor accumulate along the SSS but rather in the basal cisterns and olfactory regions indicating CSF drainage in those areas.

5. Radiosurgery for Intracranial Arteriovenous Malformations in Children

Hans G. Eder, MD, PhD; Klaus A. Leber, MD; Senta Kurschel, MD (Graz, Austria)

INTRODUCTION We describe our experience treating AVMs in the pediatric population with Gamma Knife radiosurgery (GKRS).

METHODS Between March 1992 and October 2001, 23 children (6 to 17 years, mean 13.4 years) were treated with GKRS for AVMs. GKRS was performed as primary treatment in 12 children and in 4 children after partial resection of the AVM. Partial endovascular embolization was performed in 5 cases before GKRS, in the remaining 2 children endovascular treatment, partial resection and GKRS was combined. Two patients underwent repeated GKRS and four a staged procedure, because of large volume AVM. The volume of the lesions ranged from 0.2–25 ccm (mean 5.0 ccm). A mean dose of 20 Gy (range, 12.5–28 Gy) was delivered to a mean peripheral isodose of 50% (range, 40–70%).

RESULTS Follow-up of 19 children ranged from 10 to 73 months (mean 36.2 months). Fourteen oblations (74%) were confirmed by angiography, 2 years after GKRS. Partial obliteration was seen in the remaining 5

children (26%) with a follow-up period less than 2 years. Two patients were lost for follow-up and in another two follow-up was too short. Two children developed undue effects without permanent deficits, hemorrhage was noted in one child, 10 months after GKRS. Karnofsky performance rate were stable in all patients.

CONCLUSIONS Review of the literature and our results suggest that GKRS represents a safe and effective technique in the treatment of AVMs. Larger AVMs should be treated by a staged and multimodality procedure to avoid radiation-induced adverse effects.

6. Young Age Moyamoya Disease: Role of Aggressive Surgical Treatment

Kyu-Chang Wang, MD, PhD; Ho-Jun Seol, MD; Seung-Ki Kim, MD, PhD; You-Nam Chung, MD; Hee-Soo Kim, MD, PhD; Byung-Kyu Cho, MD, PhD (Seoul, Korea)

INTRODUCTION The clinical course of young age moyamoya disease (MMD) were known to be worse than those of older patients. The aim of this study was to investigate the surgical outcome of young age (< 5 years) MMD.

METHODS The authors reviewed 204 MMD children (age < 15) who underwent unilateral or bilateral indirect revascularization surgery from 1988 to 2000. Mean duration of follow-up was 39.3 months (0.25–173). We classified the patients into two groups (A: age \leq 5, B: age > 5), and Group A was classified into Subgroups A1 (age \leq 2) and A2 (2 < age \leq 5). Surgery was performed in two stages for bilateral indirect revascularization. The number of patients was 23 in Group A1, 50 in A2, and 131 in B.

RESULTS Clinical types with infarction (type 3, 4, 5 of Matsushima classification) were more common in Group A than in B ($p = 0.001$). The incidence of postoperative infarction including minor asymptomatic cases was not significantly different in each group. In all the groups, such complications developed more frequently after the first surgery than the second surgery. The outcome was excellent or good in 58%

of A1, in 84% of A2, and in 86% of B (Matsushima classification). On the last follow up (\geq 24 months after two staged surgery) SPECT, cerebral hemodynamics were improved or stabilized in 67% of A1, 84% of A2, and 82% of B.

CONCLUSIONS Young age MMD, especially in Group A1, showed more aggressive clinical course than older age MMD (A2, B). Active surgical treatment could stabilize the rapid progression of natural course.

7. Serial MRS Metabolite Changes in Treated Pediatric Acute and Sub-acute Hydrocephalus

Miriam Scadeng, MD; Stefan Bluml, PhD; Rex A. Moats, PhD; Marvin D. Nelson, MD; Gordon J. McComb, MD (Los Angeles, CA)

INTRODUCTION Little work has been done to evaluate the effects of a severe or acute episode of hydrocephalus on the developing human brain. Our aim is to non-invasively determine the long-term effects of treated acute hydrocephalus in children using MRS.

METHODS Eight children (0–14 yrs) with raised pressure hydrocephalus had single voxel MRS prior to and at 2–4yrs after surgery. They were assigned to one of two groups depending on acuteness and duration of their initial presenting symptoms: 1. Acute group, 2. Chronic group.

RESULTS Acute group (n=5)—Initial N-acetyl aspartate/Creatine (NAA/Cr) was reduced in 4/5 patients and was normal in one. On follow-up in these 4 patients, NAA/Cr remained the same in 2 patients and improved in 2. The MRS on the patient with normal initial NAA/Cr remained normal. On the post treatment studies there was a slight reduction in Choline/Creatine (Cho/Cr) in the periventricular white matter in 2/5 patients. Chronic group (n=3)—2/3 patients had normal NAA/Cr both pre and post treatment though both had marked reduction in Cho/Cr in the periventricular white matter post treatment. One patient (premature neonate with neonatal intra ventricular haemorrhage) had a 20% drop in NAA/Cr.

CONCLUSIONS 1. The changes in white matter NAA/Cr at presentation indicate neuronal vulnerability. This may be reversed

or improved with normalising the intraventricular pressure. 2. Delayed Cho/Cr changes in the periventricular white matter is seen in patients with more longstanding hydrocephalus and is suggestive of gliosis. This appears to be irreversible.

8. Restrictive Periventricular Gliosis—Fact or Fiction

Sandeep Sood, MD; Jaliya Lokuketagoda, MD; Steven D. Ham, DO (Detroit, MI)

INTRODUCTION Pathophysiological mechanisms that prevent the ventricles from enlarging following shunt malfunction in chronically shunted patients have perplexed investigators for along time. Engel et al had proposed that periventricular gliosis as a result of chronic shunting prevents ventricular enlargement at the time of shunt malfunction. Although morphologically demonstrable, there is no direct or experimental evidence that periventricular gliosis can restrict ventricular enlargement in humans. In this study, by simultaneously measuring pressure changes in the ventricle and the brain parenchyma we demonstrate evidence against restriction from periventricular gliosis.

METHODS In 21 patients who had presented with slit ventricles at the time of shunt revision, pressure measurements were obtained from the ventricle through a fluid coupled transducer and from the brain parenchyma using Codman ICP monitor. Change in ventricular pressure was compared to the change in the parenchymal pressure after bolus infusion of fluid into the ventricle.

RESULTS There was no significant difference on t-test in the change in pressure in the ventricle compared to that in the parenchyma suggesting absence of restriction from periventricular gliosis. The ratio compliance of the extraventricular system to that of the ventricular system was 1.1.

CONCLUSIONS This study strongly suggests that periventricular gliosis does not restrict ventricular enlargement and other mechanism should be sought to explain failure of ventricles to enlarge after shunt malfunction in chronically shunted patients.

9. Oxidative Metabolism Correlated to Symptomatology and Prognosis of Tethered Cord Syndrome

Shokei Yamada, MD, PhD; Daniel J. Won, MD (Loma Linda, CA); Margaret Wacker, MD (Riverside, CA); Santiago Figueroa, MD (Loma Linda, CA)

INTRODUCTION The purpose of this paper is twofold: 1) to discuss whether the intra-operative oxidative metabolism is correlated with the severity and the progression of symptoms of TCS; 2) whether the post-untethering oxidative metabolic improvement is correlated with postoperative neurological improvement.

METHODS The authors analyzed signs and symptoms of 35 patients who presented with Tethered Cord syndrome (N=25: 0–9 years of age; N=10: 10–16 years). Oxidative metabolism was determined by redox changes of cytochrome a, a 3 which were measured by reflectance spectrophotometry. The results of redox studies were correlated with the signs and symptoms of these patients before and after all untethering procedures retrospectively. The patients were divided into three groups: 1) those with only mild redox changes; 2) those with moderate redox changes, and 3) those with severe redox changes before untethering.

RESULTS The group-1 patients showed rapid neurological improvement with redox normalization after untethering. Group-2 patients showed recovery to nearly normal neurological condition associated with redox improvement to normal control after untethering. Group-3 patients showed limited recovery from neurological deficits associated with partial recovery of redox status.

CONCLUSIONS The severity of neurological signs and symptoms before and after untethering procedures will be tabulated and correlated to the redox changes of cytochrome a, a 3. The authors extend their discussion to the controversial subjects such as occult tethered cord syndrome and prophylactic release of the tethered cord.

10. The Role of Anti-Copper Antiangiogenic Agent Tetrathiomolybdate in Treatment of Human Medulloblastomas

Sami Khoshyomn, MD; Erica F. Bisson, MD; Samuel B. Braff, BA; Bruce I. Tranmer, MD (Burlington, VT)

INTRODUCTION Recent experimental data using the novel anti-copper antiangiogenic agent, tetrathiomolybdate (TTM), has shown striking effectiveness in palliative treatment of a number of adult tumors. The efficacy of this non-toxic oral agent has not been investigated in angiogenesis-dependent pediatric solid tumors such as medulloblastomas. In this study the effect of oral TTM on growth and angiogenesis of the human medulloblastoma line HTB-186 was investigated in a nude mice model.

METHODS Human medulloblastoma cell line HTB-186 was implanted subcutaneously in the Nu/Nu immunosuppressed mouse model. Mice in the treatment group were fed with the anti-copper agent TTM at 0.75 mg/day for a period of one month. Serum copper and ceruloplasmin levels were measured in control and treatment groups to monitor the effect of TTM on the copper status of the animals. Tumor volume and vascular density per microscopic high-power field, as measure of angiogenesis, were measured in both groups at the conclusion of the treatment interval.

RESULTS Treated animals exhibited a 70% reduction in serum copper and ceruloplasmin levels, and a 94.8 +/- 4.8% inhibition of tumor growth as compared to the control animals. This growth inhibitory effect significantly correlated with a reduction in tumor vascular density: 15.67 +/- 1.53 in control animals vs. 0.67 +/- 0.58 in TTM-treated animals. The treated animals displayed nearly avascular microscopic colonies of tumor cells. Histopathological examination of brain, liver and kidney did not reveal any abnormalities.

CONCLUSIONS In the present study we show a striking inhibition of human medulloblastoma growth using an anti-copper antiangiogenic agent. The prospect of using a potentially non-toxic oral agent such as

TTM for palliative treatment of angiogenesis-dependent pediatric tumors is encouraging and deserves further investigation.

11. Pediatric Neurosurgery Research: A Survey Study

Todd A. Maugans, MD (Burlington, VT)

INTRODUCTION The survey was designed to ascertain the prevalence and nature of clinical and basic science investigation by pediatric neurosurgeons; elucidate facilitators and barriers to research; explore possibilities for increasing research activities and collaboration.

METHODS A 23 item questionnaire was administered to the entire North American membership of the AANS/CNS Section on Pediatric Neurological Surgery via two mailings during 2001. Results were tabulated and analyzed using basic qualitative and quantitative methods.

RESULTS 154 members responded for a response rate of 62%. 82% indicated past or present research activities. The three commonest barriers were: time limitations, access to funding, and human resources/personnel. 31% had participated in research sponsored by recognized neurosurgical organizations. 70% had received funding from or participated in research sponsored by a biomedical or pharmaceutical corporation. 41% indicated that for-profit corporations are appropriate funding sources, while 48% conveyed conflict of interest issues pertaining to such arrangements. Non-biomedical academic departments, for-profit medical/biotech corporations, and private donors were identified by the majority of responders as appropriate none-traditional collaborator/sponsors of pediatric neurosurgical research. 58% indicated support for a pediatric neurosurgical research clearinghouse. 71% support an internet-based form for discussion of research. Hydrocephalus, CNS neoplasms, and trauma were identified as the highest priority issues for future research and funding.

CONCLUSIONS The survey study has identified a high prevalence of and interest in pediatric neurosurgical. Time, finances,

and human resources represent significant barriers. National organizations with pediatric neurosurgical ties are potentially underrecognized and underutilized resources.

Respondents are ambivalent about the role of for-profit organizations in the conduct and sponsorship of research. A robust Internet-based research discussion forum and research clearinghouse should be developed. Collaboration with a variety of non-neurosurgical entities must be promoted. Networking amongst pediatric neurosurgeon offers hope for overcoming barriers.

12. Minimally Invasive Correction of Craniosynostosis: Definitive Osteotomies and Immediate Fixation with Resorbable Devices

Burak M. Ozgur, MD; Hal S. Meltzer, MD; Ralph E. Holmes, MD; Steven R. Cohen, MD (San Diego, CA)

INTRODUCTION We have developed minimally invasive techniques for the correction of sagittal, metopic, and coronal synostosis which address each condition's specific deformity with definitive osteotomies and immediate reconstruction utilizing resorbable devices.

METHODS Utilizing small scalp and, when necessary, superior tarsal incisions, we perform cranial vault and orbital osteotomies in concert with suturectomies. Epidural endoscopy is used to prevent inadvertent dural injury. Immediate correction with absorbable plate fixation is employed as needed to minimize or eliminate postoperative cranial banding.

RESULTS 24 infants over a two year period have undergone correction of their craniosynostosis with these techniques. This includes 14 patients with sagittal, 5 with metopic, and 5 with unilateral coronal synostosis. There has been no significant peri-operative morbidity noted. Postoperative cranial banding, when needed, has been less than 3 months in all but one patient. Results to date have been acceptable and appear comparable to that of children undergoing standard surgical correction.

CONCLUSIONS Our techniques for minimally invasive, endoscopically assisted craniosynostosis permit immediate reconstruction with acceptable short-term outcomes. Long-term follow-up data will be needed to further assess and refine these procedures.

13. Fussiness and Irritability in Patients with Craniosynostosis

Cathy C. Cartwright, MSN; David F. Jimenez, MD (Columbia, MO)

INTRODUCTION The goal is to assess the frequency of irritability and fussiness in infants with craniosynostosis and to evaluate the effects of surgical release on this condition.

METHODS A total of 89 patients were studied, 87 with single suture involvement and 2 with bicoronal synostosis. Ages ranged from 1.1 months to 9.2 months. There were 56 males and 33 females. There were 48 patients with sagittal synostosis, 21 with metopic and 20 with coronal suture involvement. The parents completed the Fussiness Rating Scale, assessing the amount and intensity of fussiness/irritability and hours of unexplained crying (scale 0-6). Rating was done for one month preoperatively and one month postoperatively following treatment with endoscopic strip craniectomies of the involved sutures.

RESULTS For the entire cohort group, the total number of hours of unexplained fussiness preoperatively was 344 hours, the amount of unexplained fussiness (scale >3) was 62%, and the intensity (scale>3) was 65%. Postoperatively, the total number of hours decreased to 105 hours (-31%), the amount of fussiness decreased to 20% and the intensity to 26%. There were no differences between suture type, or patient's age (Kruskal-Wallis p>0.89). However, the decreases in amount, intensity and number of hours were significant between preoperative and postoperative ratings (Wilcoxon Signed Rank test p<0.0001).

CONCLUSIONS Our results indicate that craniosynostosis is associated with unexplained fussiness/irritability in a significant number of untreated patients. Surgical

release of the stenosed suture is associated with a significant decrease in irritable behavior in these patients. Although the etiology is unclear, local or global intracranial pressures may play a role.

14. Scaphocephaly without Synostosis: The Sticky Sagittal Suture

James E. Baumgartner, MD; John F. Teichgraber, MD; Kelly Seymour-Dempsey, MD (Houston, TX)

INTRODUCTION Scaphocephaly is usually associated with sagittal craniosynostosis, and is treated surgically. Rarely, screening CT scans in scaphocephalic infants reveal open sagittal sutures. We chose to attempt cranial molding in this unusual subset of scaphocephalic patients.

METHODS From 1997 to 2001, 58 patients presented to the University of Texas-Houston Craniofacial Center with scaphocephaly. The diagnosis was documented with morphometric head measurements. All underwent screening CT scans, and six were found to have open sagittal sutures. Five patients, ages three to five months at presentation, were managed with cranial molding helmets. A sixth patient who presented at 13 months of age was managed surgically. Serial morphometric head measurements were obtained on each patient.

RESULTS Four of the five patients managed with molding helmets, achieved correction of their cranial deformities. The fifth patient did not correct with molding, and repeat CT scanning demonstrated synostosis of the sagittal suture. The patient underwent surgical correction of his scaphocephaly at age 6 months. The final patient demonstrates the natural course of an uncorrected sticky sagittal suture. The morphometric correction achieved at 13 months of age was less than that seen in the patients treated before 6 months of age.

CONCLUSIONS Scaphocephaly without synostosis (or sticky sagittal suture) can be corrected with cranial molding, when diagnosed early. If cranial molding is unsuccessful or the patient presents late, surgical correction is necessary.

15. The Sagittal Suture May Close First in Late Premature Closure of Multiple Sutures: Oxicephaly

Donna C. Wallace, MSN, NP; Harold L. Rekate, MD, FACS; Edward F. Joganic, MD, FACS; Stephen P. Beals, MD, FACS (Phoenix, AZ)

INTRODUCTION We define the clinical findings and natural history of late premature closure of multiple sutures.

METHODS A retrospective review of a single craniofacial center was performed to study the clinical presentation, radiographic findings, and the outcome of patients with late pansutural closure.

RESULTS Eleven patients were identified as fulfilling inclusion criteria. Four patients were normal at birth and progressed to complete closure of coronal, lambdoid, and sagittal sutures with a pointed area at the level of the anterior fontanel. Seven other patients presented early with the typical clinical picture of sagittal synostosis. Open coronal and lambdoid sutures were seen on early CT scans. Following cranial remodeling, from three months to three years of age, the children developed increased intracranial pressure, a pointed head, and prominent digital markings as seen on CT scan.

CONCLUSIONS Craniosynostosis of the sagittal suture may be the first sign of progressive closure of all sutures. A bulging pointed region at the level of the anterior fontanel is the hallmark of this condition.

16. Multiple-revolution Spiral Osteotomy for Cranial Reconstruction: A Report of Long Term Outcomes on 29 Patients

Farrok R. Farrokhi, MD; Micam W. Tullous, MD; Peter T.H. Wang, MD; Patricia A. Mancuso, MD; Dennis G. Vollmer, MD (San Antonio, TX)

INTRODUCTION Many cranial remodeling techniques have been employed in attempts to provide optimal expansion and cosmetic results. The authors present long term outcomes in the treatment of various cranial deformities by the technique of multiple-revolution spiral osteotomy.

METHODS A series of 29 patients, ages ranging from 6–95 months (avg=24) underwent treatment of cranial deformity by reconstruction with this technique. Follow-up cranial computed tomography was performed at 6, 12 and 24 months. Increases in head circumference, bone formation at osteotomy sites, cosmetic results and increase in cranial index (for sagittal synostosis patients, n=13) were used as measures of outcome.

RESULTS Mean follow-up was 12 months (range 1–28 months). An average increase of 2.8cm was noted in head circumference before and after surgery. Complete or near complete bone formation was noted in all patients by 12 month follow-up CT Cranial index was noted to increase from a pre-operative average of 64 to 76 after surgery, with a mean increase of 12.2. No immediate complications were noted. Delayed formation of cystic granulomas around the absorbable plates occurred in four patients, with two requiring surgical excision.

CONCLUSIONS Multiple-revolution spiral osteotomy is a safe and effective method for correction of cranial deformity. This method offers an effective means for achieving increases in head circumference, cranial index and bone formation as well as an excellent cosmetic result when compared to traditional techniques. It also shows promise for older children, and may serve as a potential treatment option for patients who come to neurosurgical attention long after infancy.

17. Experience with Frontolateral Keyhole Craniotomy through a Superciliary Skin Incision in Children

Laszlo Bogner, MD, PhD; Gabor Madarassy, MD (Budapest, Hungary)

INTRODUCTION During the last four years 24 superciliary frontolateral keyhole craniotomies were carried out on pediatric patients at the Department of Pediatric Neurosurgery of the National Institute of Neurosurgery, Budapest, Hungary. 20 children presented with suprasellar, parasellar or frontobasal tumor, 3 children with frontal or temporal arachnoid cyst, and 1 child with frontobasal liquor fistule.

METHODS The frontolateral keyhole craniotomy is a modification of the generally used pterional approach. The operation were carried out through an approximately 2.5 x 3 cm frontolateral miniaturized craniotomy after a skin incision just above the eyebrow.

RESULTS Despite the small size of the craniotomy, the exploration allows enough room for intracranial manipulation with maximal protection of the brain and other intracranial structures. One patient from the presented series had a wound infection that was cured with good result, but we did not have any other craniotomy-related complication.

CONCLUSIONS In our experience, the frontolateral keyhole craniotomy in children, together with the advent of modern neuroanaesthesia, cerebrospinal fluid drainage, and microsurgical techniques, is a safe approach for an experienced neurosurgeon to use in the treatment of the supratentorial tumors or arachnoid cysts of the anterior fossa and sellar regions.

18. Prenatal Magnetic Resonance Imaging in the Diagnosis of Fetal CNS Abnormalities

Kimberly D. Bingaman, MD; Juliette A. Prust; Kim M. Walker, NP; Dorothy I. Bulas, MD; Robert F. Keating, MD; Philip H. Cogen, MD, PhD (Washington, DC)

INTRODUCTION The use of magnetic resonance imaging (MRI) as an adjunct to prenatal ultrasound has recently become more prevalent. However, its utility in the diagnosis of fetal cerebral and spine abnormalities has not been well established.

METHODS This study retrospectively examined all cases in which MRI was used when sonographic findings suggested CNS anomalies from 1990 to 2002 at Children's National Medical Center. 46 pregnant women, ages 20 through 45, underwent fetal MRI (gestational ages ranged from 18 to 35 weeks, average of 26 weeks).

RESULTS 46 pregnancies underwent 49 fetal MRI's. Sonographic indications for fetal MRI included: aqueductal stenosis (2), ventriculomegaly (15), posterior fossa abnormality (11), mass (2), myelomeningo-

cele (5), vein of Galen malformation (1), arachnoid cyst (2), agenesis of the corpus calosum (4), microcephaly (1), hemorrhage (1), and family history of CNS anomaly (2). In 34 cases (73.9%), the findings of the MRI correlated with the ultrasound findings. In 6 cases (13.0%), the diagnosis was changed after an MRI was performed, and in 2 cases (4.3%), the MRI ruled out abnormalities originally thought to be present by ultrasound.

CONCLUSIONS Because of its superior imaging, prenatal MRI can confirm, refute or alter the diagnosis of fetal CNS abnormalities discovered by ultrasound. Other diagnoses suspected by ultrasound, such as Vein of Galen aneurysm, can be verified by MRI so that definitive treatment can be coordinated and optimized. Prenatal MRI can be a useful tool in prenatal counseling and management.

19. The Fetal Myelomeningocele Trial

Leslie N. Sutton, MD (Philadelphia, PA); Noel Tulipan, MD (Nashville, TN); Nalin Gupta, MD (San Francisco, CA)

INTRODUCTION The early experience with fetal myelomeningocele closure at 4 institutions suggest possible benefit for selected groups of patients. To date, comparison has only been performed with matched historical controls, which is subject to bias. The NIH has recently approved funding for a multi-center prospective trial clinical trial to be conducted at 3 centers. The aim of this presentation is to inform the pediatric neurosurgical community about the design of the trial so that they can inform prospective subjects and care for the patients who may be in their communities.

METHODS The study is an unblinded randomized controlled clinical trial of 200 patients. Patients diagnosed with myelomeningocele at 16 to 25 weeks gestation will be referred to a central Data and Study Coordinating Center (DSCC) for initial screening and information. Those eligible and interested will be assigned by the DSCC to a Fetal Surgery Unit where final evaluation and screening will be carried out.

Patients who satisfy the eligibility criteria and consent to randomization will be centrally randomized to one of the following two management protocols:

- Intrauterine repair of the myelomeningocele at 180 to 256 weeks, discharge to nearby accommodation on tocolytics when stable for preterm labor, weekly prenatal visits and biweekly ultrasounds conducted at to the FSU; cesarean delivery at 37 weeks following demonstration of lung maturity.
- Return to local perinatologist for prenatal care, with monthly ultrasounds reported to the FSU; return to the Fetal Surgery Unit at 37 weeks gestation for cesarean delivery following demonstration of lung maturity; neonatal repair of the myelomeningocele.

RESULTS The study is expected to recruit 200 patients, and to end in 2006.

CONCLUSIONS It is hoped that the neurosurgical community will support this effort.

20. Inclination of the Odontoid Process in the Pediatric Chiari I Malformation

R. Shane Tubbs, PhD, MS, PA-C; Matthew D. Smyth, MD; John C. Wellons, III, MD; Jeffrey P. Blount, MD; Paul A. Grabb, MD; W. Jerry Oakes, MD (Birmingham, AL)

INTRODUCTION The quantitative analysis of odontoid process angulation has had scant attention in the Chiari I malformation population. This study seeks to elucidate the correlation between a posteriorly tilted odontoid process and patients with tonsillar ectopia.

METHODS MR images of the craniocervical junction were analyzed in 100 children with a Chiari I malformation and in 50 children with normal intracranial anatomy used as controls. Measurements focused on the degree of angulation of the odontoid process and the assignment of various degrees to a grading scale (0–III). Postoperative outcome following posterior cranial fossa decompression was then correlated to grades of angulation. Other measurements included midsagittal lengths

of the foramen magnum and basiocciput, our institution's previously documented pB-C2 line, level of the obex from a midpoint of McRae's line, and the extent of tonsillar herniation.

RESULTS Higher grades of odontoid angulation (retroflexion) were found to be more frequently associated with syringomyelia and particularly holocord syringes. Higher grades of angulation were more common in females and were often seen to have obices that were caudally displaced greater than three standard deviations below normal. Grades of angulation were not found to correlate with postoperative outcome.

CONCLUSIONS This study not only confirms prior citations of an increased incidence of a retroflexed odontoid process in the Chiari I malformation but quantitatively defines grades of inclination. Grades of angulation were not found to correlate with postoperative outcome. Our hopes are that these data add to our current limited understanding of the mechanisms involved in hindbrain herniation.

21. Management of Cervical Spine in Klippel-Feil Patients

Fassett R. Daniel, MD, MBA; Brockmeyer L. Douglas, MD (Salt Lake City, UT)

INTRODUCTION Klippel-Feil syndrome and Klippel-Feil variant are complex disorders of vertebral segmentation. The natural history of this disorder and surgical indications for instability are incompletely understood.

METHODS A retrospective chart review identified twenty-one patients who had been evaluated and followed at our institution since 1990. Radiographic studies and clinical history were reviewed. Data was collected for each patient regarding symptoms, location of abnormality, number of abnormal segments, degenerative changes, instability, and treatment.

RESULTS Thirteen of the 21 patients were symptomatic with neck pain, sensory changes, motor changes, or torticollis. Seven patients had degenerative changes in their cervical spine as noted by osteophytic spurs on plain films or disc desiccation on MRI imaging.

CONCLUSIONS Klippel-Feil patients should be followed closely for degenerative spine disease and instability in the cervical spine. Patients with more than two fused vertebrae or separate block deformities at multiple levels in the cervical spine appear to be a greater risk for instability than patients with a single block level deformity. Therefore, these factors should be taken into account in regard to timing for clinical follow-up and imaging studies.

22. Treatment of Basilar Invagination in the Pediatric Population Using Cervical Reduction and Posterior Occipital-Cervical Fusion

Louis J. Kim, MD; Harold L. Rekate, MD, FACS; Jeffrey D. Klopfenstein, MD; Volker K.H. Sonntag, MD (Phoenix, AZ)

INTRODUCTION Standard treatment in the pediatric population for basilar invagination (BI) includes anterior, posterior or combined decompression with stabilization. We propose that occipital-cervical (OC) reduction using accentuated cervical extension and distraction followed by posterior OC fusion is a viable stand-alone treatment that may obviate the need for transoral odontoidectomy.

METHODS From January 1994 to May 2002, nine pediatric patients presented with clinically and radiographically significant BI. Suboccipital decompression was performed initially, then patients underwent manual reduction intraoperatively with evoked-potentials monitoring, fluoroscopic guidance, and direct visualization. The endpoint position required axial distraction and cervical extension to maximally reduce the BI. All patients then were fused posteriorly using autologous bone grafts, threaded Steinman pin, and suboccipital/sublaminar wires. Early and long-term clinical and radiographic follow-up were reviewed retrospectively.

RESULTS Age at presentation ranged from 18 months to 17 years (n=9). Five patients were male. All patients presented with concomitant Chiari malformations. Immediately postoperatively, 7 were significantly improved, 2 unchanged. Over long term follow-up (3 to 76 months,

mean 23.1), 7 were good or excellent, 1 unchanged, and 1 worse than preoperative. One patient required subsequent odontoidectomy due to continued progression of BI. The remaining eight patients all demonstrated stable fixation on MRI and solid fusion on plain films or CT.

CONCLUSIONS Posterior OC fusion after realignment using cervical distraction and extension technique provided good or excellent outcomes in 77% of patients. Careful clinical and radiographic follow-up permits detection of those patients that subsequently require odontoidectomy.

23. Vestibular Function in Adolescent Idiopathic Scoliosis

Matthew T. Provencher, MD; Derin C. Wester, PhD (San Diego, CA); Bruce L. Gillingham, MD (Coronado, CA); Kim L. Gottschall, PhD; Michael Hoffer, MD (San Diego, CA)

INTRODUCTION Theories for adolescent idiopathic scoliosis (AIS) have focused on the pathology of cerebral asymmetry and asymmetrical vestibular-ocular reflex function. The relationship between scoliosis and vestibular-spinal function can be determined from a state-of-the-art vestibular balance assessment.

METHODS Patients with AIS and matched controls underwent a comprehensive orthopaedic scoliosis evaluation followed by a standardized vestibular-balance assessment to include, vertical axis rotation (VAR at 0.08 Hz, 0.32 Hz, and 0.64 Hz), eccentric vertical axis rotation (EcVAR), computerized oculomotor testing (OM) and dynamic posturography (CPD), spinal reflex testing (Fukuda Test), rotation chair testing for vestibular-visual interaction (VFX and VVOR) and VORTEQ testing for high-frequency motor tracking.

RESULTS Thirty-four patients were enrolled, 21 with AIS and 13 as controls. There were no differences in clinical oculomotor function, positional testing, computerized dynamic posturography (CDP), VFX, and VVOR. A significant difference was found for vertical axis rotation chair (VAR) at 0.32 Hz ($p < 0.0001$) with left rotational asymmetry,

but was not significant at 0.08 or 0.64 Hz ($p > 0.05$). As angle of the spine curve increased (Cobb Angle), so did the overall central vestibular dysfunction ($p = 0.045$), with the right curves showing a left-sided deficit. As adolescents, both the AIS and control patients demonstrated a higher threshold for vestibular gain.

CONCLUSIONS The data suggests that a central vestibular function is worse with larger curves, and the dysfunction is opposite to the curve. First-time data is presented to demonstrate that adolescents (both AIS and controls) have an overall vestibular function either less well developed or less sensitive than normal adults. Further data collection is still needed to test for differences; however, these results support a vestibular dysfunction in patients with AIS.

24. Improvement in Brainstem Auditory Evoked Potentials after Suboccipital Decompression in Patients with Chiari I Malformation

Richard C. Anderson, MD; Ronald G. Emerson, MD; Kathryn C. Dowling, EdD; Neil A. Feldstein, MD (New York, NY)

INTRODUCTION The optimal treatment for symptomatic patients with Chiari I malformation remains controversial. Although a suboccipital decompression with duraplasty is most commonly performed, there may be a subset of patients who improve with bony decompression alone. In an initial attempt to identify such patients, we performed continuous intraoperative brainstem auditory evoked potentials (BAEPs) in patients undergoing a standard decompression with duraplasty and compared conduction at three different time points: (1) baseline in supine position (before positioning), (2) immediately after bony opening and release of the atlanto-occipital membrane (i.e., dural band), and (3) after dural opening.

METHODS 11 children (mean age 9.8 years) with symptomatic Chiari I malformation underwent a suboccipital decompression and duraplasty with intraoperative BAEP and somatosensory evoked potential (SSEP) monitoring. Six patients (55%) had associated syringohydromyelia.

RESULTS At baseline, the I-V interpeak latencies (IPL) for both sides (total of 21 BAEPs) were 4.19 +/- 0.22 ms (mean +/- standard deviation). After complete bony decompression and prior to any dural opening, the I-V IPL decreased to 4.03 +/- 0.25 ms ($P = 0.0005$). When the dura was opened, however, no further decrease in the I-V IPL was detected (4.03 +/- 0.25 ms; $P = 0.6$). SSEPs remained stable throughout the procedure.

CONCLUSIONS In pediatric patients undergoing suboccipital decompression with duraplasty for Chiari I malformation, the vast majority of improvement in conduction through the brainstem occurs after bony decompression and division of the atlanto-occipital membrane rather than after dural opening. Further studies are needed to establish whether the improvement seen with BAEP monitoring during bony decompression will predict long-term improvement in these patients.

25. Preliminary Analysis of Genetics of Lipomyelomeningocele

Timothy M. George, MD; Marcy Speer, PhD (Durham, NC); NTD Collaborative Group

INTRODUCTION Given the similarities in timing of development and vertebral anomalies between open and closed neural tube defects (NTD), the question arises as to whether these two types of disorders result from the similar genetic mechanisms. The genetic contribution to closed NTD development has not been investigated.

METHODS Probands from fifty-seven families were diagnosed with a lipomyelomeningocele (53 prior to age ten and 4 as adults). A standard three-generation family history, general medical records and imaging studies were collected to confirm the presence and level of the lipo(myelo)meningocele. Recurrence risks and evidence of familial clustering were analyzed.

RESULTS A total of 57 families with at least one proband with a lipomyelomeningocele were found. There were 29 females and 28 males. There were no sibling recurrences of NTDs for a recurrence risk of 0.0 (95% c.i.). However, there were five pedigrees that indicated familial aggregation. There

was either a lipoma or other NTD in the family. There were no instances of a multiplex family that exhibited complete concordance.

CONCLUSIONS We attempted to establish evidence in support of a genetic hypothesis for lipomyelomeningocele. There was no sibling recurrence risk for the series of families in which the proband was affected with lipomyelomeningocele. We have, however, demonstrated evidence of familial clustering of lipomyelomeningocele with other types of open neural tube defects. These data suggest that a closed form of NTD, lipomyelomeningocele, may be genetically related to the open form of NTD, which has been established to have a genetic component.

26. Atlantoaxial Distances in Children During Cervical Spine Flexion

Matthew D. Smyth, MD; R. Shane Tubbs, PhD, MS, PA-C; John C. Wellons, MD; Jeffrey P. Blount, MD; Paul A. Grabb, MD; W Jerry. Oakes, MD (Birmingham, AL)

INTRODUCTION Analysis of the atlantoaxial interlaminar distance in children has not been previously performed. This study seeks to determine the age-matched relationship between the posterior elements of C1/C2 in children during cervical flexion, to be used as an adjunct to the atlanto-dental interval in common clinical use.

METHODS Lateral radiographs of the cervical spine in full flexion were analyzed in 74 children during the initial evaluation of isolated Chiari I malformation. The atlantoaxial interlaminar distance (ILD) was defined as the distance between midpoints of the anterior cortices of the posterior arch of the atlas and axis during full cervical flexion. This line roughly correlated to the spinolaminar line.

RESULTS The ILD was measured in 74 children (32 M, 42 F, mean age 9.1 yrs). The range for the entire group was 8.0-30 mm. When stratified by age, the mean ILD was 12.3 mm (n=15, SD=3.0 mm) in children aged 3 or less, and 20.5 (n=59, SD=4.7mm) in children aged > 3. Further

stratification of the groups yielded a mean ILD of 10.4 mm (n=8, SD=1.4) in children aged 1 to 2 years and 14.4 mm (n=7, SD=4.7) in children aged 3 years. Children greater than 3 years old consistently had a mean ILD of approximately 20 mm (SD 5.0 mm) regardless of age.

CONCLUSIONS From our study, children less than three years of age routinely had an ILD of less than 20 mm (mean 12.3 mm) and children older than three consistently had a mean ILD of approximately 20 mm. Knowledge of the distance between the posterior elements of the atlas and axis during full flexion should enhance the clinician's ability to accurately diagnose atlantoaxial instability on lateral radiographs. Identification of children with atlantoaxial interlaminar distances falling outside of these parameters should raise the suspicion for atlantoaxial instability.

27. Children with Growth Hormone Deficiency and Chiari I Malformation: A Morphometric Analysis of the Posterior Cranial Fossa

R. Shane Tubbs, PhD, MS, PA-C; Matthew D. Smyth, MD; John C. Wellons, III, MD; Alfred A. Bartolucci, PhD; Jeffrey P. Blount, MD; Jerry W. Oakes, MD; Paul A. Grabb, MD (Birmingham, AL)

INTRODUCTION The posterior fossa (PF) has been found to be small in various forms of Chiari I malformation. Anecdotes regarding a connection between growth hormone deficiency (GHD) and Chiari I Malformation (CIM) have been proposed. However, to date, no quantitative analysis of the PF of patients with the CIM and GHD has been performed. Our study was performed to determine the geometry of the PF in children with GHD and CIM.

METHODS Morphometric analysis of the PF was performed in ten children with GHD and a CIM (Group I), 20 children with GHD and no CIM (Group II), and 50 controls.

RESULTS PF volumes for Group I ranged from 128-259 ml +/- 33 ml and for Group II ranged from 115-186.2 ml +/- 25.4. Lengths of the foramen magnum for Groups I and II

had means of 36 mm and 38 mm respectively. The basiocciput length and tentorial angle means for Groups I and II were 20 mm/19 mm and 89/87.5 degrees respectively.

CONCLUSIONS We have determined that children with GHD +/-CIM have no significant difference in their PF volume compared to controls. However, our data demonstrate significant underdevelopment of portions of the bony PF in both patients with GHD alone and in patients with GHD and CIM. Tentorial angles were elevated in noncontrol groups. We propose that this association is not due to an increased rate of "midline" defects seen in GHD but rather a structurally distorted PF that is not capacious enough to house the entire developing rhombencephalon. These data will hopefully aid in the further understanding of the pathophysiology of the CIM.

28. Acquired Chiari I Malformation and Slit-ventricle Syndrome

Concezio Di Rocco, Prof; Francesco Velardi, MD (Rome, Italy)

INTRODUCTION The pathogenesis of cerebellar tonsils acquired herniation in children with extrathecal CSF shunt is subject of debate. Two recent cases of adolescents presenting with clinical and neuroimaging findings of slit-ventricle syndrome and acquired Chiari I suggest the progressive reduction in intracranial volume due to the CSF shunt as cative factor of both phenomena.

METHODS Seriated MR examinations of two subjects harboring CSF shunt devices demonstrated progressive cerebellar tonsils herniation and increasing calvarial thickening. Abnormal CSF pressures were detected in both patients.

RESULTS The first case was a 16 year old boy, who received a cysto-ventriculo-peritoneal CSF shunt when 10 months old, because of a suprasellar arachnoid cyst. Postoperative course was uneventful by excepting cerebellar tonsils progressive herniation in the last 3 years associated with recurrent headaches, vomiting, and impaired consciousness. The second subject,

a 15 year old girl, developed her symptomatology 12 years after lumbo-peritoneal shunt because of pseudotumor cerebri. She also showed reduced intracranial volume accompanied by cerebellar tonsils herniation. Both patients underwent cranial vault expansion with immediate clinical improvement and ascent of cerebellar tonsils. CSF shunt devices were left untouched.

CONCLUSIONS Acquired herniation of cerebellar tonsils is a reversible complication of extrathecal CSF shunting due to secondarily reduced intracranial volume. It differs from congenital Chiari I malformation as the entire skull volume is involved. Consequently it should be treated by enlarging cranioplasty rather than limited posterior fossa decompression (to further support this conclusion the authors present a similar patient operated on in a different institution who developed acute tetraplegia following posterior fossa decompression secondary to hindbrain herniation through the craniolacunia). As the complication was observed after shunting both intracranial and intraspinal CSF spaces, acquired Chiari I does not appear related to negative pressure gradients in spinal compartment as propounded to occur in case of lumboperitoneal shunting.

29. Complications of Vagal Nerve Stimulation for Epilepsy in Children

Matthew D. Smyth, MD; R. Shane Tubbs, PhD, MS, PA-C; Paul A. Grabb, MD; Jeffrey P. Blount, MD (Birmingham, AL)

INTRODUCTION Vagal nerve stimulation (VNS) emerged during the past decade as an option for treating medically refractory epilepsy. Although many published series exist addressing the efficacy of VNS, relatively little data have been published regarding surgical complications of this procedure, especially in children.

METHODS We performed a retrospective review of 73 consecutive patients less than 18 years of age implanted with a VNS from 1998 to 2001 with a minimum follow-up of one year (mean=2.23 yrs).

RESULTS Of the 73 patients treated, (41 males and 32 females; mean age 8.75 years; range 11 mo to 18 yrs), 7 (9.6%) had a complication ultimately resulting in device removal. The overall infection rate was 7.1% (6 of 73 patients undergoing 84 implantation/revision procedures). Two stimulators were salvaged with IV antibiotics alone and one with operative debridement and IV antibiotics. Three devices were removed because of infection (3.6% of 84 procedures). All infections presented within the first few weeks of implantation or revision. An additional four stimulators (5.5%) were removed because of lack of clinical benefit and device intolerance (one patient with symptomatic tachycardia, two with hardware intolerance, and one with an inactivated device complicating the workup of fever). Two devices were revised because of lead fracture (2.7%). Amongst the cohort thus far, 10 battery changes have been performed, none less than 3 years after initial implantation. Two patients had stimulation-induced symptoms (periodic hoarseness or brief outbursts of laughter) which did not require device removal. Ipsilateral vocal cord paralysis was identified in one patient.

CONCLUSIONS VNS remains a viable option for improving seizure control in difficult to manage pediatric epilepsy patients. Non-preventable surgical complications such as hardware failure (2.7%) or deep infection (3.6%) resulting in device removal or revision were seen. Occasional stimulation-induced symptoms (tachycardia, hoarseness, laughter) may be expected.

30. Changes in the Gross Motor Function Measure (GMFM) following Selective Dorsal Rhizotomy (SDR)

Jack Ensberg, PhD; Sandy Ross, PT; T.S. Park, MD; Jeffrey Ojemann, MD (St. Louis, MO)

INTRODUCTION This investigation compared the effects of SDR with intensive physical therapy (PT), intensive PT alone, and standard therapy in subjects with cerebral palsy (CP) on the GMFM.

METHODS Ninety-seven subjects with spastic diplegia CP were assigned to 3

groups; SDR group (n=31), PT group (n=37) and CP group (n=29). The mean age for all groups was 9+5 years in this repeated measures semi-randomized design. The SDR group underwent SDR and intensive PT (4 times/week). The PT group had only intensive PT, and the CP group maintained their existing level of PT (0-2 times/week). All subjects were evaluated using the GMFM initially and 8 months later. Subjects were placed into one of the 5 Gross Motor Function Classification System (GMFCS) levels. Regression equations were used to calculate the natural change in function as a consequence of development over the 8 month period (Palisano et al., 2000). This predicted change was subtracted from the 8 month postop GMFM score. ANOVA with repeated measures was used to determine significant differences (p<0.05).

RESULTS The SDR group indicated a significant improvement in function from pre- to postoperative (86.4+10.0 to 87.5+9.4). No significant changes in function were observed for the PT (88.7+7.2 to 89.2+7.1) and CP (80.6+11.1 to 80.7+11.7) groups over the intervention period.

CONCLUSIONS GMFM results with predicted change in function due to development parceled out, demonstrated significant improvements in function for the SDR group with no improvements in function for the PT and CP groups. The data suggest beneficial effects of SDR on function in children with spastic diplegia.

31. Long Term Follow-up of Selective Dorsal Rhizotomy: Does Multi-level Laminotomy Influence the Risk of Scoliosis?

J. A. Hall, MD, MSc; J.P. Farmer, MD; A. O'Gorman, MD (Montreal, Canada)

INTRODUCTION Selective dorsal rhizotomy (SDR), a procedure used for the relief of spasticity, involves surgically exposing and sectioning the lumbosacral roots. This requires a significant bony opening which has traditionally been accomplished via multi-level laminectomy from L2 to S1. Both cerebral palsy and surgery on the pediatric spine are independent risk factors for scoliosis. To reduce the possibility of

future spinal deformity, the technique of laminotomy with total reconstruction of the posterior spinal arch has been proposed. We have used this procedure in all of our patients. It is hypothesized that laminotomy results in a lower incidence of scoliosis than that associated with multi-level laminectomy.

METHODS Pre- and post-operative charts and radiographs were examined for one hundred patients with more than one year follow-up from SDR. Scoliosis curves were measured using Cobb's Angles. There was independent verification by a second observer of all cases with a scoliotic curve of >10%. The rates of scoliosis were compared to those in the literature using laminectomy.

RESULTS There were no patients who required instrumentation for correction of severe scoliosis. Eight patients had scoliotic curves. Mild scoliosis, defined as between 11 and 20 degrees, was found in six patients; moderate scoliosis, defined as between 21-40 degrees, was noted in two patients. None of the patients who developed scoliosis was an independent ambulator prior to surgery.

CONCLUSIONS Laminotomy for SDR appears to be associated with a lesser incidence of scoliosis than laminectomy at comparable follow-up times. Independent ambulation prior to surgery seems to confer some protection against scoliosis following SDR. Longer follow-up is needed to confirm the stability of these findings but initial data suggests an advantage of multi-level laminotomy over laminectomy, at least in this population.

32. Surgical Management of Pediatric Epilepsy: Bringing Treatment Home to Indiana's Children

Jodi L. Smith, MD, PhD (Indianapolis, IN)

INTRODUCTION Approximately 20-25 percent of pediatric epilepsy patients have inadequate seizure control with pharmacotherapy. Persistent seizures refractory to medical management can lead to severe neurocognitive impairment. Seizure surgery

provides an alternative therapeutic option for pediatric patients who suffer from recurrent seizures. This study represents a retrospective analysis of 24 consecutive pediatric patients who underwent surgical management of their seizures at Riley Hospital for Children (Indiana University Medical School) during the first 18 months after establishing a seizure surgery program at this institution.

METHODS Clinical, radiographic, and electroencephalographic records of pediatric epilepsy patients operated on from August 1, 2000 to January 31, 2002 were reviewed. All patients underwent a comprehensive presurgical evaluation. Patients with a clearly defined focal area of seizure onset underwent resection of their seizure focus; all others underwent vagal nerve stimulator implantation.

RESULTS Twenty-four patients were operated on, 10 females and 14 males, with ages ranging from 0.2-15.3 years. Twelve of 24 patients underwent surgical resection. All twelve had a structural brain abnormality on MRI (e.g., tumor, cortical dysplasia, and/or hamartoma), which corresponded to the location of their epileptic focus on EEG. Eight patients had temporal lobe lesions (four right and four left), three had frontoparietal lesions, and one had a frontal lobe lesion. Gross total resections were achieved in eight; near total resections in four. No new postoperative neurological deficits were identified. Two patients were lost to follow-up, nine are seizure-free on the same or less anticonvulsants, and one had no seizures until four months after surgery but now is having 1-2 seizures/day compared with 5-30 seizures/day preoperatively.

CONCLUSIONS Proper patient selection is essential for achieving a successful outcome in seizure surgery.

33. Epilepsy Surgery in Tuberous Sclerosis

Howard L. Weiner, MD; Werner K. Doyle, MD (New York, NY); Pantaleo Romanelli, MD (Palo Alto, CA); Souhel Najjar, MD; Daniel Miles, MD; Orrin Devinsky, MD (New York, NY)

INTRODUCTION Tuberous sclerosis is often associated with medically-refractory epilepsy, usually resulting from cortical tubers, areas of cortical dysplasia surrounded by reactive gliosis. There is a strong association between mental retardation with early age of seizure onset and greater seizure burden in childhood. Effective seizure control is thought to reduce significantly the adverse developmental effects of chronic epilepsy. However, multifocal ictal and interictal activity on video-EEG recordings may falsely suggest that surgery may not control or significantly reduce seizure activity. Further, seizure foci in eloquent areas often preclude surgery for fear of unacceptable neurological deficits. In other cases, seizures persist after surgery because residual epileptogenic cortex was left due to its proximity to eloquent cortex, or because a distant independent focus was not addressed. Multistage invasive monitoring can detect residual adjacent or distal epileptogenesis, and is especially useful when pre-surgical data suggest eloquent cortex or bihemispheric involvement. We report the safety and efficacy of staged surgical approaches for epileptogenic foci involving either eloquent areas of the dominant hemisphere or both hemispheres.

METHODS We have recently treated 6 patients (2 males, 4 females), mean age 4.2 years, utilizing a novel, staged surgical approach. Three patients underwent two-stage procedures, and three patients underwent three-stage procedures.

RESULTS At a mean follow-up of 25.2 months, three patients are seizure-free, and three have rare, non-disabling seizures. Moreover, all have made significant developmental progress since surgery. Individual cases will be illustrated.

CONCLUSIONS These data suggest that a select group of TS patients may benefit from a more aggressive surgical approach.

The presence of multiple or bilateral ictal foci should not be considered a contraindication to surgery if the severity of seizures, the ineffectiveness of medical therapy, and progressive neurodevelopmental delay dictate a more aggressive course of action.

34. Rate of Orthopedic Surgery After Selective Dorsal Rhizotomy in Relation to Ambulatory Status in Spastic Diplegia

Donncha F. O'Brien, MD; T. S. Park, MD; Eric C. Leuthardt, MD; Jeffrey R. Leonard, MD; Jeffrey G. Ojemann, MD (St. Louis, MO)

INTRODUCTION Although selective dorsal rhizotomy (SDR) in children with spastic cerebral palsy may reduce the need for future orthopedic surgery, the relationship between ambulatory status prior to SDR and the incidence of post SDR orthopedic surgery is unknown. In this study, we examined 1) whether spastic diplegic children who walk independently prior to dorsal rhizotomy require less orthopedic surgeries than children who walk with a walker or crutches; 2) what type of orthopedic surgeries are needed.

METHODS A total of 159 children with spastic diplegia who underwent SDR between 1990–1994 were studied. Age at the time of SDR ranged from 2 to 19 years. The postoperative follow-up period was 5–9 years. All patients were surveyed by mail or telephone. All orthopedic procedures after SDR were performed by local orthopedic surgeons.

RESULTS Of the 159 patients, 52 were independent ambulators and 107 patients were assisted ambulators. Of the independent ambulators, 25% required orthopedic surgery, including releases of heel cord (7) and hamstring (3), iliopsoas (1), muscle transfer (1) and foot ankle surgery (1). Of the assisted ambulators, 43% required orthopedic surgery, including releases of heel cord (18), hamstrings (12), adductors (7), foot ankle surgery (1) and derotational osteotomy (4).

CONCLUSIONS After SDR for spastic diplegia, independent ambulators have a lower rate of orthopedic surgery and require only soft tissue surgery relative to the assisted ambulators.

35. Febrile Convulsions, Cortical Dysplasia, and Intractable Temporal Lobe Epilepsy in Children

Ann-Christine Duhaime, MD (Lebanon, NH); Brenda E. Porter, MD; Alex R. Judkins, MD; Robert R. Clancy, MD; Dennis J. Dlugos, MD; Jeffrey A. Golden, MD (Philadelphia, PA)

INTRODUCTION A high percentage of patients with temporal lobe epilepsy have a history of febrile convulsions, but it is not known whether febrile convulsions cause later epilepsy or whether an underlying condition predisposes children to both febrile convulsions and epilepsy. We report a consecutive series of 33 children with intractable temporal lobe epilepsy who were analyzed for a history of febrile convulsions and neuropathologic findings of cortical dysplasia.

METHODS Data were collected prospectively on 33 consecutive children with intractable temporal lobe epilepsy who underwent temporal lobectomy, including patient demographics, age of febrile convulsions if present, age of seizure onset, age at surgery, neuropsychological data and seizure outcome. Temporal cortical tissue was resected en bloc and was examined using light microscopy and neurofilament immunohistochemistry with RMDO20.

RESULTS Fifteen patients had febrile convulsions and eighteen patients did not. There was no difference in gender, age of epilepsy onset (mean 3.6 years), or age at surgery (mean 11.9 years), but patients with febrile convulsions had a lower rate of mental retardation than those without ($p=0.018$). Ten of 18 children without febrile convulsions and 11 of 15 children with febrile convulsions had cortical dysplasia (total 21/33 patients). Patients with cortical dysplasia had a lower rate of seizure freedom postoperatively (58%) compared to those without (80%).

CONCLUSIONS This rate of cortical dysplasia is higher than that usually reported in adult series, and may represent differences in neuropathologic techniques and classification. The high rate in patients both with and without febrile seizures suggests that cortical dysplasia may predispose both to febrile convulsions and to intractable epilepsy.

36. Temporal Lobe Tumor Related Epilepsy in Childhood

Oguz Cataltepe, MD; Nejat Akalan, MD; Meral Topcu, MD; Serap Saygi, MD; Guzide Turanlı, MD; Dilek Yainizoglu, MD; Nese Dericioglu, MD (Ankara, Turkey)

INTRODUCTION We retrospectively studied 15 pediatric age and 15 adult patients whom underwent resective surgery because of intractable seizures secondary temporal lobe tumors at Hacettepe University Hospitals, Ankara, between 1995 and 2002.

METHODS All patients had comprehensive pre- and post-surgical work-up including a detailed seizure history, neurological examination, neuroimaging studies, neuropsychological tests, WADA test and underwent non-invasive continuous EEG/Video monitoring. Pre- and postoperative tests, surgical and pathological findings and seizure outcome were documented and compared in adult and pediatric age groups.

RESULTS Mean ages of the patients were 11 and 26 years in pediatric and adult groups respectively. Eighty five percent of the patients had a history of seizure more than 5 years. NE was normal in 74% of the patients. Interictal epileptogenic abnormalities were evident in 90% of the patients. Ictal activity was recorded in 55% of the patients and localizing value was 84%. Forty percent of the tumors involved mesial temporal structures and 30% in lateral temporal neocortex. Surgical gross total tumor removal was achieved in 72% of the patients. Surgical approach was tumor removal alone in 24% of the patients and tumor removal anterior temporal lobectomy with amygdalohippocampectomy in 44% percent of the patients. DNET was the most common histopathological diagnosis as seen in 66% of the patients. Eighty four percent of the patients was Class I, 8%

Class II and 8% Class III according to Engel seizure outcome classification.

CONCLUSIONS This group of epilepsy patients constitute a specific clinicopathological group with an excellent chance of postoperative seizure control rate.

37. High Resolution MRI Enhances Identification of Lesions Amenable to Surgical Therapy in Children with Refractory Epilepsy

Natasha McKay, MD; Shenandoah Robinson, MD; Monisha Goyal, MD; Barbara Bangert, MD; Max Wiznitzer, MD; Mark Scher, MD; Barbara Swartz, MD (Cleveland, OH)

INTRODUCTION Many children with refractory epilepsy can achieve better seizure control with surgical therapy. An abnormality on MRI, along with corroborating localization from other modalities, markedly increases the chances of a successful surgical outcome. We present our initial results using a four-coil phased-surface-array MRI (Quad Coil)(MRI Devices, Waukesha, Wisconsin) to identify lesions in children previously thought to have non-lesional epilepsy on standard MRI. Compared to standard MRI, the Quad Coil has a signal-to-noise ratio that is one-third greater at the center of the head, and over 100% greater at the periphery, providing improved cortical resolution.

METHODS Quad Coil MRIs were obtained as part of the comprehensive pre-surgical evaluation in 13 children ages 5–19 years during the first half of 2002.

RESULTS Five of nine children (56%) that did not have abnormalities identified on standard MRIs within the last year had lesions found using the Quad Coil. Three children had temporal lobe hamartomas and two had hippocampal atrophy. The newly identified lesions visualized on the Quad Coil MRI shifted the weight of the evaluation towards surgery. Two patients with hamartomas underwent surgical resection. One was Engel I and one Engel III at follow-up. Two of the children with newly identified lesions are awaiting surgery. Two children without lesions underwent

vagal nerve stimulator implantation. For the remaining four children that had lesions previously identified on standard MRI, the lesions were confirmed on the Quad Coil MRI, and two of them have undergone surgery (one Engel I and one Engel II).

CONCLUSIONS High resolution MRI identified lesions in over half of the children with refractory epilepsy who previously had no lesions identified by recent traditional MRI technology. MRI advances increase the number of children with refractory seizures who are candidates for successful epilepsy surgery.

38. In Vitro Testing of Current Spread During Ventricular Catheter Coagulation Using Diathermy

Frederick A. Boop, MD; John Honeycutt, MD; Jeffrey Sorenson, MD (Memphis, TN); Bruce Cherny, MD (Mesa, AZ)

INTRODUCTION Since its introduction by Chambi and Hendrick in 1988, the use of monopolar coagulation of ventricular catheters embedded in choroids plexus has become widespread. The authors had previously reported an anterior cerebral artery pseudo-aneurysm which developed and hemorrhaged following monopolar coagulation of a ventricular catheter lodged in the interhemispheric fissure. This complication lead to the development of a simple bench test which can be performed by any neurosurgeon utilizing this technique to determine the safest coagulation parameters for his own diathermy unit.

METHODS Utilizing a beaker of egg whites, the authors placed a modified ground pad into the protein solution. Ventricular catheters were then placed into the solution and the stylette touched with the monopolar coagulator at varying wattages and for varying lengths of time.

RESULTS Coagulation at 40 Watts current (Valley Lab) produced flame around the tip of the catheter emanating from the pores in the catheter (Medtronic PS Medical). Flash flames also occurred at 35 Watts, forming a coagulum of egg white for 1 cm distant from the tip. All heat was dissipated

through the holes in the first 16 mm of the catheter. At 20 Watts, flame was minimal. At 15 Watts, only bubbles around the tip were seen with minimal coagulum formation.

CONCLUSIONS The authors will present a simple technique for defining the optimum wattage to be used during catheter coagulation for a given diathermy unit. For our own ValleyLab units, we now lower the current to 15–20 Watts for coagulation of ventricular catheters.

39. Use of a Novel Continuous Long-Term Intracranial Pressure Monitor in Ambulatory Hydrocephalus Patients

David M. Frim, MD, PhD (Chicago, IL); David Adelson, MD (Pittsburgh, PA); John Kestle, MD; Marion L. Walker, MD (Salt Lake City, UT); Keith Miesel, PhD; Lee Stylos, PhD (Minneapolis, MN); Richard Penn, MD (Chicago, IL)

INTRODUCTION Monitoring ICP in patients is usually limited to single measurements or short periods of time in a hospital setting. The dynamic changes in pressure and how they correlate with patient symptomatology are not known. The purpose of this study was to test a novel, implantable ICP monitor and recording system in patients who have undergone multiple shunt revisions.

METHODS Eleven patients underwent implantation of the Medtronic InSite® ICP device at the time of shunt revision. Tandem pressures were correlated during surgery from the ventricular catheter and the InSite® monitor in all patients and post-operatively from the InSite(r) and implanted Radionics TeleSensor devices in some patients. Ongoing data for activity, temperature, and ICP was downloaded at intervals. Data samples of ICP were taken and recorded over 2-second intervals every five minutes. Patients carried a barometric pressure monitor at all times for correlation with ICP and kept a written diary of clinical symptoms such as headache.

RESULTS ICPs were recorded beyond 8 months in several patients and at least 1 month in all patients. Tandem manometric or TeleSensor ICPs were consistent with the InSite derived data in all cases.

Maneuvers to vary ICP (postural changes or Valsalva) were appropriately reflected in the ICP records. Marked circadian variation in ICP was noted in all patients. Clinical observations were also made: in two patients, headache pain previously attributed to occlusive shunt malfunction (leading to revision several times in the past) was found to be associated with low rather than high ICP. One InSite device had to be removed after several weeks due to infection and 2 have malfunctioned.

CONCLUSIONS Initial tests indicate that ICP can be accurately and continuously monitored in out-patients by the InSite device and that such information can be correlated with patient activity and symptoms. This information may prove valuable in understanding baseline CSF dynamics and correlating ICP with symptomatology associated with shunt function and malfunction.

40. The Impact of Shunting in Congenital and Perinatal Hydrocephalus: Long-term Outcome in Adult Survivors

Venkat Sadanand, MD, MS, PhD (Saskatoon, Canada); Fred Gentili, MD, FRCS(C), MSc (Toronto, Ontario, Canada)

INTRODUCTION Most data on hydrocephalus is based on either the pediatric population when the majority of shunts are inserted or on shunting in adult-onset hydrocephalus. The purpose of this study was twofold: First, to determine the long term impact of patient and shunt specific factors in congenital and perinatal hydrocephalus and the probability of subsequent shunt revision in adulthood. Second, to determine the long term impact of CSF shunting on the neuropsychological function in adult survivors.

METHODS In this 35 year retrospective and prospective study, records of 127 patients were statistically analyzed. The patients were initially seen and treated at a pediatric hospital and followed until their 18th birthday. Subsequently, the patients were managed by one surgeon at an adult hospital until the present date. The present age of the

patients ranged from 18–45 years and the age of first shunting ranged from 2 days to 18 years.

RESULTS Our data showed that 50% of the patients have attended high school, 39% have attended college and 11% have a university degree. Of the different types of shunts used initially only lumboperitoneal (LP) and ventriculoperitoneal (VP) shunts had incidences of 'no revisions'. The lowest percentage of revisions was associated with Spina Bifida while the highest percentage was with congenital hydrocephalus. A younger age at first shunting was associated with a larger expected number of revisions. Patients with no revisions tended to have their first shunt placed at an older age. Neuropsychological development was inversely related to the number of revisions.

CONCLUSIONS The long term functional outcome of patients with hydrocephalus is generally good with many patients being capable of leading normal, independent and productive lives.

41. Diffusion Weighted Imaging in Acute and Treated Raised Pressure Pediatric Hydrocephalus

Miriam Scadeng, MD; Marvin D. Nelson, MD; Gordon J. McComb, MD (Los Angeles, CA)

INTRODUCTION Diffusion Weighted Imaging (DWI) allows the dynamic study of brain water movement in and around cells. Diffusion is described in terms of the apparent diffusion coefficient (ADC). In tissue where proton movement is restricted by boundaries such as cell membranes ADC is reduced. Interstitial and vasogenic edema results in an increase in water protons outside the cell and ADC is increased. In the brain, grey matter (GM) has a slightly higher ADC value than white matter (WM) due to higher spatial organization in WM. Our aim is to characterize the pattern of ADC values in 1. acute raised pressure hydrocephalus, and 2. after treatment when the ventricular pressure is normal, in functionally normal children.

METHODS All patients had presented with a CSF obstructing brain tumor. The same diffusion pulse sequence parameters

(b=1000) were used for all scans. ROI were taken from multiple regions of interest including Periventricular white matter (PVWM) (edema on imaging), WM, GM and thalamus.

RESULTS Acute hydrocephalus n=5 (mean age 3.7y), ADC-PVWM 0.0018 (thin rim of low ADC 0.00066), WM 0.0012, GM 0.001, Thalamus 0.00088. Post treatment group n=5 (mean age 10.2y). ADC-Post treatment WM 0.0008 GM 0.0008, thalamus 0.00075. Normal control group =13 (mean age 6.8y). ADC-WM 0.0008. GM 0.00089, thalamus 0.00077.

CONCLUSIONS In acute hydrocephalus extra cellular fluid is increased as CSF is forced between cells leading to a raised ADC. The thin rim of reduced ADC in the PVWM may be due to acutely compressed periventricular tissue. After treatment, ADC values for GM returns to normal, however WM ADC remains slightly elevated suggesting some axonal/cell loss, and an increase in extracellular space.

42. What is the Ideal Initial Valve Pressure Setting in Infants with Programmable Hakim Valves?

Marcus C. Korinth, MD; Martin R. Weinzierl, MD; Joachim M. Gilsbach, MD (Aachen, Germany)

INTRODUCTION In neonates and infants less than 1 year of age who are treated with a ventriculoperitoneal shunt non-infectious complications are almost equally frequent and dangerous to infectious complications. While the incidence of infections can be reduced using perioperative antibiotics, special surgical techniques and perioperative care, non-infectious complications like wound break down, cerebrospinal fluid (CSF) fistula and subcutaneous CSF collections are preventable but seem difficult to manage, especially in the susceptible age group of patients.

METHODS The authors present their experience with the use of the programmable Hakim valve in 47 infants less than 1 year of age, who were treated with de novo implantation of a ventriculoperitoneal shunt

due to various pathologies. An uneventful wound healing during the first weeks after shunt implantation, avoiding the above mentioned non-infectious complications was supported by initial, temporary overdrainage (initial valve pressure setting in neonates (< 5 weeks of age) of 30–40 mmH₂O and 50-60 mmH₂O in older infants (2–12 months)) and readjustment of the programmable valve after completed wound healing.

RESULTS All patients tolerated this procedure well and showed no pathological signs and symptoms of overdrainage like premature closure of cranial sutures, clinical low-pressure syndrome, slit ventricle syndrome (SVS), subdural fluid collection and brain collapse during the average follow-up period of 2.6 years (2-62 month). Infectious and other mechanical, non-infectious complications were analyzed as well during the follow-up period.

CONCLUSIONS These results suggest that an initial, temporary overdrainage in infants and newborn with shunted hydrocephalus may contribute to further lower the incidence of non-infectious complications like wound break-down, CSF-fistula or subcutaneous CSF accumulation, without negative side effects. This technique could be a valuable option in the regimen of shunt-treatment of this age group in order to optimize the overall success rate and lower the general complication rate.

43. "Quick Brain" MRI versus CT Scan for Evaluating Shunted Hydrocephalus

Joshua E. Medow, MD; Bermans J. Iskandar, MD; Rowley Howard, MD (Madison, WI)

INTRODUCTION Shunted hydrocephalic patients are routinely exposed to significant doses of radiation from repeated CT scans. We report a new MRI sequence that shows substantial promise in evaluating the shunted hydrocephalic patient while minimizing many of the disadvantages seen with CT.

METHODS We have prospectively followed 50 consecutive pediatric patients who

underwent a "Quick Brain" MRI scan, and compared them to 50 consecutive patients who had CT scans. The MRI technique consisted of a Single Shot Fast Spin Echo (SSFSE) sequence. Meta analysis of cost, image definition and clarity was performed.

RESULTS Compared to CT, the SSFSE MRI had the following advantages: 1) Absence of radiation exposure; 2) Multiplanar visualization of the ventricles, especially helpful in cases of multiloculated hydrocephalus; 3) A significant decrease in motion artifact; this technique offers exceptional accuracy and speed (<15 sec per entire brain). The acquisition of each slice is so quick that movement does not significantly alter captured images, a problem that has plagued both MRI and CT since their inception. As a result, the child does not require sedation. The main disadvantages of SSFSE MRI compared to CT have been a decreased ability to visualize hemorrhage and ventricular catheter position postoperatively. Cost was comparable between CT and SSFSE MRI.

CONCLUSIONS "Quick Brain" MRI offers significant advantages in shunted hydrocephalic patients with respect to CT in image quality and scan time, at a comparable cost, and without recurrent doses of radiation or need for sedation.

44. Mortality After

Ventriculoperitoneal Shunt Procedures: The Impact of Hospital and Surgeon Volume

Edward R. Smith, MD; William E. Butler, MD; Fred G. Barker, MD (Boston, MA)

INTRODUCTION Prior studies have shown lower mortality when complex medical or surgical procedures are performed at high-volume centers or by high-volume providers. We studied in-hospital mortality after pediatric ventriculoperitoneal shunt procedures using a national database.

METHODS We used the HCUP Nationwide Inpatient Sample to identify all admissions for VP shunt procedures (placement or revision, as principal procedure) on patients age 1–18 at a 20% sample of all US hospi-

tals during 1999–2000. In-hospital mortality was related to hospital and surgeon volume and other factors using multivariate logistic regression.

RESULTS 3202 admissions were identified (184 hospitals, 257 surgeons). Mortality was lower at high-volume centers and for high-volume surgeons. For hospital volume, mortality was 1.4% (10/693) at lowest-quartile-volume centers (<27 procedures/yr) and 0.2% (2/905) at highest-quartile-volume centers (>121/yr). For surgeon volume, mortality was 1.2% (5/410) for lowest-quartile-volume providers (<10 procedures/yr) and 0.2% (1/566) for highest-quartile-volume providers (>127/yr). After multivariate adjustment for emergency admission (P=0.03) and presence of infection (P=0.006), hospital volume remained a highly significant mortality predictor (P=0.003). Surgeon volume was statistically significant in a similar multivariate model (P=0.009). Length of stay was also significantly shorter at higher-volume centers (P<0.001), after adjustment for age, infection, emergency admission and geographic region. Hospital charges were lower at high-volume centers (P<0.001), after adjustment for age, presence of infection, and geographic region.

CONCLUSIONS High-volume hospital care for pediatric shunt procedures was associated with lower mortality, shorter length of stay and lower hospital charges in a nationwide sample.

45. The Predictive Value of a Normal Shuntogram

Jeffrey G. Ojemann, MD; Michael Taylor, PA-C; T.S. Park, MD (St. Louis, MO)

INTRODUCTION Radionuclide injection of shunts ("shuntogram") can assist the evaluation of possible shunt malfunction. In our institution, shuntograms are performed when patients present with symptoms suggestive, but inconclusive for, shunt malfunction, without CT or shunt series evidence of malfunction. Knowing the predictive value of apparently normal flow (a 'negative' study) influences the decision making process.

METHODS Shuntograms over the past 3 1/2 years were reviewed. In nearly all, the injected reservoir was integral to the (PS Medical) valve—a 'negative' study refers to a functioning shunt determined by flow to the peritoneum within 15 minutes. Patient records were reviewed for revision in proximity to a negative study.

RESULTS 115/149 tests were negative. 34 surgeries (in 32 patients) occurred within 1–7 days of a negative shuntogram. In 19/34 revisions the shunt was functional: 15 surgeries for overdrainage, three for unrelated reasons with shunt function confirmed incidentally, and one exploration for cognitive deterioration. In the remaining 15 cases (13 patients) the shunt was not functional. They were revised for persistent symptoms (headache, nausea, neck pain) or subsequent deterioration (diplopia). Ten of 15 had proximal obstruction. The remaining had valve malfunction, atrial terminus requiring revision despite good flow, undiagnosed disconnection, peritoneal catheter occlusion, and one distal malfunction that proved to be infection. All improved following revision.

CONCLUSIONS In this series, the false negative rate for shuntograms was 15/115 (13%) with proximal occlusion most common. This estimate of the predictive value of a normal flow study may influence the decision to revise a shunt in certain situations.

46. Retrograde Venography in the Evaluation of Pseudotumor Cerebri in the Pediatric Population

Vivek R. Deshmukh, MD; Felipe C. Albuquerque, MD; Cameron C. McDougall, MD; Harold L. Rekate, MD, FACS (Phoenix, AZ)

INTRODUCTION We retrospectively reviewed our experience with retrograde venography in managing pseudotumor cerebri (PTC) in children. In adults with PTC, elevated intracranial venous pressure is universal. In children, the etiology of PTC is variable. We postulated that venography would help to identify patients with PTC and to delineate subtype. Treatment would be tailored to subtype.

METHODS All patients underwent retrograde venography to identify venous stenosis/gradient or elevated venous pressures. The records for 18 patients were retrospectively reviewed to determine the etiology of the pseudotumor and to formulate a treatment protocol. All patients met clinical criteria for PTC.

RESULTS Eighteen patients (10 males, 8 females) were treated between 1996 and 2001. Mean age was 9.7 years. Sixteen patients had abnormal venograms demonstrating either a venous gradient or elevated right atrial pressures. Twelve of these 16 patients had primary PTC; four were patients with syndromic hydrocephalus and secondary PTC. All of these patients benefited from LPS. The two patients with normal venograms were found to have cephalocranial disproportion and were advised to undergo a cranial remodeling procedure.

CONCLUSIONS Three subtypes of PTC exist in children. Prototypical pseudotumor patients present with increased right atrial pressure or venous stenosis. Normal volume hydrocephalus may develop in patients with ventriculomegaly treated by shunting (secondary PTC). Patients in both subtypes respond to lumboperitoneal shunting. The third subtype, cephalocranial disproportion, is suggested by normal venography in the setting of documented intracranial hypertension. These patients should undergo cranial vault reconstruction. Retrograde venography is valuable in distinguishing between these forms of pseudotumor cerebri.

47. Shunt Pseudotumor

Julian J. Lin, MD; Fredrick A. Boop, MD; Robert A. Sanford, MD (Memphis, TN)

INTRODUCTION Increased intracranial pressure in hydrocephalic children with functioning shunts is an uncommon problem. Slit ventricle syndrome (episodic transient symptoms of shunt malfunction with a functioning shunt system and slit ventricles) is a well-recognized phenomenon. Less well described is the child with a functioning shunt, chronic increased intracranial pressure manifest by headache and papilledema.

METHODS Two recent problematic cases provoked a retrospective review of a previously reported series. This retrospective review of 1400 children treated at LeBonheur Children's Hospital from 1985 through 2001 located 4 additional cases. Five of the 6 cases presented with recent onset of chronic headaches and papilledema with normal head growth. CT scan confirmed long-standing bilateral slit ventricles. The 6th patient presented with acute onset of symptoms of shunt malfunction and papilledema. The shunt was explored, proved to be functioning, the child was treated emergently with drain proceeding to LP shunt and suffered significant neurologic impairment.

RESULTS All 6 patients had Camino intracranial pressure monitoring and demonstrated normal pressure when awake and alert, but elevation of pressure within the 40–80 mm/mercury range during deep sleep. Five of the 6 patients were treated with vault expansion with resolution of papilledema and headache. Of the 4 children prior to 2001, one had a proximal shunt malfunction one year after the vault expansion, and the other 3 children had no subsequent shunt malfunction for 5 years.

CONCLUSIONS Discussion of possible mechanisms of this clinical phenomenon will be presented.

48. Low Pressure Hydrocephalus: Diagnosis and Treatment Issues

Mark G. Hamilton, MD, FRCS(C), MD (Calgary, Canada); Angela V. Price, MD, FRCS(C) (Vancouver, Canada)

INTRODUCTION The majority of patients with acute hydrocephalus have ventriculomegaly and high intracranial pressure (ICP). However, there is a small but significant subset of patients who have acute hydrocephalus with ventriculomegaly and inappropriately low intracranial pressure.

METHODS A prospective database has been maintained since the first patient was identified in 1997. Each patient experienced clinical deterioration that included a significant decrease in level of consciousness with new and significant ventriculomegaly

with inappropriately low ICP. A review of the medical literature was performed to identify other similar patients.

RESULTS Ten such patients have been diagnosed and treated at the University of Calgary between 1997 and 2002. Insertion of an external ventricular catheter (external ventricular drain: EVD) revealed ICP < 5 cm H₂O in all patients. The treatment instituted consisted of either wrapping the patient's neck with a tensor bandage, in order to increase brain turgor, or lowering the EVD to well below head level to facilitate drainage of cerebrospinal fluid (CSF) and encourage re-expansion of the cortical mantle. In each patient, treatment resulted in clinical improvement, normalized ICP, and resolution of the ventriculomegaly. A review of the medical literature revealed 5 reports with 21 other patients.

CONCLUSIONS Low-pressure hydrocephalus is a little recognized entity in adults and children. The hypothesized pathophysiology is decreased brain turgor resulting in ventriculomegaly and cerebral/cortical dysfunction. A discussion about the hypothesized pathophysiology and a resultant treatment algorithm will be presented utilizing this combined experience of 31 reported patients.

49. MRI Study of the Natural History of Ventriculomegaly and Tonsillar Herniation in Children with Posterior Fossa Tumours

Kanna Gnanalingham, PhD; Jesus Lafuente, FRCS; Dominic Thompson, FRCS; William Harkness, FRCS; Richard Hayward, FRCS (London, United Kingdom)

INTRODUCTION Posterior fossa tumours in children predispose to hydrocephalus, although the natural history is unclear and the need for drainage of the ventricles is controversial. We report on the natural history of ventriculomegaly and tonsillar herniation, as seen on serial MRI scans in children with posterior fossa tumours.

METHODS Hospital Case notes and MRI imaging of 89 children with posterior fossa tumours were reviewed retrospectively.

RESULTS On pre-operative MRI, ventricular size was assessed by measurement of the ventricular index (VI) and 59 patients (66%) had VI greater than 0.4. There was a progressive decrease in the mean VI from pre-operative to post-operative MRI scans ($p=0.0001$). Eighteen (20%) patients required permanent CSF drainage in the form of VP shunt (15) or third ventriculostomy (3). Patients requiring permanent CSF drainage had a greater VI pre-operatively and at 3-9 months post-operatively ($p<0.05$). On pre-operative MRI scan, 75 patients (84%) had greater than 5 mm herniation of the cerebellar tonsils below the level of the foramen magnum. There was a progressive decrease in the mean tonsillar herniation from pre-operative to post-operative MRI scans, with time ($p=0.0001$), although this did not relate to the need for CSF drainage. On multivariate analysis, the only risk factors for the need for permanent CSF drainage was pre and intra-operative CSF drainage (Odds ratio = 23.3; $p=0.0001$) and incomplete surgical excision of tumor (Odds ratio = 7.7; $p=0.006$).

CONCLUSIONS Hydrocephalus and tonsillar herniation are common in children with posterior fossa tumours, although post-operatively there is a natural tendency for it to resolve. With a policy of commencing steroids on admission and early surgical debulking, only a fifth of patients needed permanent CSF drainage. Patients at risk are those with severe symptoms, needing peri-operative CSF drainage and those with subtotal resection of tumour. We recommend selective drainage of CSF in children with posterior fossa tumours.

50. Myxopapillary Ependymomas in Children: Results of Surgery

George I. Jallo, MD; Sean Wilson, BA; Karl Kothbauer, MD; Fred Epstein, MD (New York, NY)

INTRODUCTION Although myxopapillary ependymomas are generally benign with a tendency for slow growth and local recurrence, there is no report about the long-term outcome for these tumors in children. The authors report their experience with 14 children and discuss their management strategy.

METHODS A retrospective review was conducted of 14 cases of myxopapillary ependymoma with a mean age of 12.5 years (range, 7–18 years). The male:female ratio was 2.5:1. The follow-up period for this study group was 3.9 years (range, 0.5–8.8 years).

RESULTS Patients with myxopapillary ependymomas have a variety of clinical symptoms. The most common was pain seen in 93%, motor deficit 46%, sensory 31%, and bladder difficulty 23%. The duration of symptoms averaged 14.8 months prior to intervention. A total removal was accomplished in 86% of patients at our center. Adjuvant radiotherapy was administered to 5 children prior to referral. Three patients had recurrent disease which required reoperation, mean 63 months. These children had surgery alone without adjuvant treatment. At last follow-up all children are alive and free of active disease.

CONCLUSIONS Patients with myxopapillary ependymomas have an indolent course. Children tend to have a better progression free survival as compared to older patients. Adjuvant radiotherapy does not seem to prolong progression-free survival. Patients who are treated with radical surgery alone have a good outcome and long progression free survival.

51. Current Diagnosis and Treatment Strategies in Germ Cell Tumors of the Pineal and Hypothalamic Regions

Greg Olavarria, MD; Ramon Navarro, MD; Tadanori Tomita, MD; (Chicago, IL)

INTRODUCTION We present our series of children with germ cell neoplasms treated at our institution from 1981 to the present. We present a discussion addressing controversies in diagnosis, method of biopsy, and treatment regimens.

METHODS 36 children with pineal region and hypothalamic germ cell tumors were treated by one neurosurgeon at one institution. All except six had tumor markers, CSF cytology and biopsy. Tumor locations included 19 pineal region, 6 hypothalamic, and 11 involving both areas. Total resection was performed in 15 patients. Pathologic

diagnoses were consistent with 15 germinomas, 7 teratomas, and 14 mixed germ cell tumors. Adjunctive therapy included entire neuroaxis radiation or limited field radiotherapy in 12 patients, and 15 patients had radiation and chemotherapy.

RESULTS The 5 year progression free survival was 72% for all germ cell tumors. Specifically, germinomas had the best outcomes at 88%, followed by teratomas at 84%, while mixed germ cell tumors had the poorest prognosis (56% 5-year survival). A significant number of patients in our series had mixed germ cell tumor pathology, with tumors present in both pineal and hypothalamic locations.

CONCLUSIONS Although therapy has been advocated based solely on radiologic diagnosis and tumor markers, we recommend active histological diagnosis for more prompt tailored therapy. Craniotomy is the preferred and safest method of tumor biopsy. The incidence of concurrent lesions is 33% in our series. Germinomas responded equally well to cranio-spinal axis radiation and chemotherapy/radiation combinations. Non-germinomatous tumors require both radiation and chemotherapy. Second-look surgery following chemotherapy should be considered.

52. Declined to Present

53. Independent Learning Modules for Pediatric Neurosurgery

Jonathan Martin, MD; Leon E. Moores, MD (Washington, DC)

INTRODUCTION Decreased resident and medical student work hours will result in decreased trainee contact time with both faculty and patients. Additionally, outcome data for training techniques will soon become a requirement during programmatic evaluations. Novel teaching methods which increase efficiency and provide measurable trainee outcomes for both individual and program evaluation and feedback will be essential for advancing GME.

METHODS Independent learning modules comprised of a variable combination of written, multimedia, and hands-on skills acquisition may increase the efficiency of academic faculty while simultaneously enhancing the learning and satisfaction of the trainee. We have developed independent learning modules for medical student and junior resident levels in several pediatric neurosurgical areas of interest. Specifically, didactic presentations of shunt malfunction and posterior fossa tumor presentation and management are now in use. Pre- and post-study test scores are collated for evaluation of the trainee and the methodology.

RESULTS We are continuing to gather data on the test scores achieved by trainees using this technique and will report available preliminary results.

CONCLUSIONS These independent learning modules are interesting, challenging, and focused. They allow for efficient, independent study. They can be stored easily on electronic media or transmitted via the Internet and may have applicability to other teaching programs and community hospital settings for use by professional and para-professional personnel.

54. Posterior Fossa Tumour Surgery in Children: Does Craniotomy Lead to Fewer Complications Than a Craniectomy?

Kanna Gnanalingham, PhD; Jesus Lafuente, FRCS; Dominic Thompson, FRCS; William Harkness, FRCS; Richard Hayward, FRCS (London, United Kingdom)

INTRODUCTION Traditional approach to the posterior fossa involved a suboccipital craniectomy. More recently posterior fossa craniotomies have been described, although the long-term benefits of this technique are not clear. We compared the post-operative complications of craniectomies and craniotomies in children with posterior fossa tumours.

METHODS We retrospectively reviewed the case notes and MRI imaging of 110 Children who underwent surgery for posterior-fossa tumours between 1985 and 1999.

RESULTS From a total of 110 children, 56 underwent craniectomy and 54 craniotomy. The mean duration of hospital stay was longer in the craniectomy group (17.5 versus 14 days). At operation, similar numbers of patients in both groups had total macroscopic clearance of tumor, completeness of the dural closure and duraplasty.

Post-operatively, more patients in the craniectomy group were noted to have CSF leak (27% versus 4%; $p < 0.01$) and pseudomeningoceles (23% versus 9%; $p < 0.05$). There was no difference between the two groups in the numbers of patients with CSF infections, wound infections or hydrocephalus, requiring permanent CSF drainage. Patients with CSF leak had a longer duration of hospital stay (20.7 versus 14.9 days; $p < 0.01$), and were more likely to have CSF (35% versus 12%; $p < 0.01$) and wound infections (24% versus 1%; $p < 0.01$), than those patients without CSF leak. Post-operatively, wound exploration and re-closures for CSF leak were more likely in the craniectomy group (11% versus 0%; $p < 0.01$). Multivariate analysis revealed that the only predictor of CSF leak post-operatively was the type of surgical approach (i.e. craniotomy versus craniectomy; Odds ratio=10.8; $p = 0.03$).

CONCLUSIONS Craniectomy was associated with post-operative CSF leak, pseudomeningocele, increased wound re-closures and thus prolonged hospital stay. CSF leak in turn was associated with infections of the CSF and wound. We propose mechanisms that could explain why CSF leak is less likely if the bone flap is replaced.

55. Reoperation for the Chiari Malformations

David J. Sacco, MD; R. Michael Scott, MD (Boston, MA)

INTRODUCTION This study was undertaken to determine if it might be possible to identify those patients who might be more likely to require reoperation for Chiari malformation, and to determine if modifications in surgical technique at the initial procedure might have obviated the need for repeat surgery.

METHODS We reviewed the hospital records, imaging studies, operative reports, and follow-up data of a consecutive operative series of Chiari decompressions carried out by one neurosurgeon over a 14 year period.

RESULTS Of 100 operations for Chiari I malformation, 15 were reoperations; of 33 operations for Chiari II malformation, 6 were reoperations. Of 33 operations for Chiari I in patients 5 years of age or younger, 7 were reoperations (21%); of 67 patients over age 5, 8 were reoperations (12%). Of the 33 operations for Chiari II malformation, 10 were reoperations, 4 of 10 who had undergone their initial surgery at the age of 5 or younger (40%), and 2 of 23 who had undergone their initial surgery over the age of 5 (9%). Reoperation was required because of failure to treat adequately an associated syrinx (10), because of possible faulty IVth ventricular stent placement (2), and because of persisting neurologic symptoms or headache (3). 5 of 9 patients in the operative series with craniosynostosis represented reoperations (55%), and all three of the patients whose syrinx persisted after reoperation had craniosynostosis syndromes.

CONCLUSIONS Reoperations will be required in approximately 20% of patients with Chiari I and Chiari II malformations. Factors which predicted a higher than usual likelihood of reoperation included a young age at initial surgery, the presence of complex bony anatomy at the foramen magnum, an association with syndromic craniosynostosis, and failure of the surgeon at the initial operation either to assess patency of the foramen of Magendie or correctly place a IVth ventricular stent.

56. Craniopharyngiomas: Complications Related to Complex Approaches in Children

Sean McNatt, MD (Sherman Oaks, CA); Michael P. Kim, BS (Chapel Hill, NC); Michael L. Levy, MD, PhD (San Diego, CA)

INTRODUCTION Craniopharyngiomas are benign intracranial tumors, which can represent complicated management problems due to the invasive nature of the lesion

and the poor response to adjuvant therapy. Given that optimal cure rates are related to the completeness of resection, considerations of surgical approach are significant determinants of potential outcome. An additional consideration is the manner in which such approaches are practiced and perfected.

METHODS We evaluated our series of children presenting with craniopharyngioma and the anatomical patterns and decision making process utilized in determining the operative approach. Approaches reviewed included the transphenoidal (6/18), subfrontal (5/18), orbitozygomatic (3/18), and transbasal/interhemispheric (4/18).

RESULTS Mean age was 3.4 years (+ 4.8 yrs). Eight of the patients were female and 10 male. In 18 patients 11 had diminished vision at presentation. Following surgery at three months 37% improved and 18% deteriorated. At one year an additional 27% improved and none deteriorated. 11% had Diabetes Insipidus at presentation and 67% following surgery. None cranial nerve abnormalities at presentation and 28% following surgery. 33% had nausea/vomiting at presentation and none following surgery.

CONCLUSIONS The incidence of injury to the hypothalamic-pituitary axis is increased in complex approaches. No significant difference in vision was noted. Our results suggest that even with maximal exposure, complications related to aggressive surgical resection remain significant and the integration of less aggressive management protocols needs to be considered.

57. Juvenile Pilocytic Astrocytoma of the Brainstem

John R.W. Kestle, MD; Douglas D. Brockmeyer, MD; Marion L. Walker, MD (Salt Lake City, UT)

INTRODUCTION Surgery is now advocated for focal brainstem tumors. The literature often presents results by tumor location with different histologic types grouped together. In order to determine prognosis after resection, histology specific data may be helpful.

METHODS The medical records at Primary Children's Medical Center were searched for juvenile pilocytic astrocytomas of the brainstem. Tumors were excluded if they were reported as "low grade" (without reference to juvenile pilocytic), fibrillary, anaplastic or higher grade.

RESULTS Eleven patients (2 midbrain tegmentum, 3 pons, 6 medulla) were found. All were alive at last follow-up (0.3-11.7 yrs, mean 5 yrs). Recurrence occurred in 6/11 patients. All presented with focal deficit. Initial treatment was resection in 10/11, biopsy + shunt + radiation in one. There was postop residual linear enhancement in 4, solid residual tumor in 6. Two of the 6 with solid residual were observed: the residual disappeared in one and is stable in the other. The other 4 had postop radiation (2) or reoperation + radiation (2) and all 4 developed recurrent tumor. Of the 4 patients with residual linear enhancement only, 3 had no further treatment and have no recurrence at 1, 6 and 10 years. The other patient had postop radiation and developed a PNET 6.6 years later. Neurologic deficits after resection were improved in 4, unchanged in 4 and worse in 3 patients.

CONCLUSIONS Long term survival after resection of pilocytic astrocytomas of the brainstem has been observed. Recurrence is common and appears to be related to the extent of initial resection.

58. NF1 and the Pediatric Neurosurgeon—20 Year Institutional Review

Jean Pierre Farmer, MD; Saad Khan, MD; Asad Khan, PhD; June Ortenberg, MD; Carolyn Freeman, MD; Augustin M. O'Gorman, MD; Jose Montes, MD (Montreal, Canada)

INTRODUCTION Children with Neurofibromatosis type 1 (NF1) undergo costly surveillance scanning for a variety of asymptomatic CNS lesions whose natural history is poorly understood.

METHODS We performed a 20-year retrospective chart review of 25 patients with clinically proven NF1 who required surgery

(group A) and contrasted this cohort with 150 NF1 patients who did not require surgery (group B).

RESULTS In group A, 52% of patients underwent multiple procedures for more than one lesion ($p=0.043$). Group A patients were further distinguished from those in Group B by exhibiting a greater number of optic gliomas ($p=0.015$), non-optic intracranial tumors ($p=0.006$), cranial nerve ($p=0.000$), paraspinal ($p=0.0062$), craniofacial ($p=0.001$), and visceral ($p=0.03$) neurofibromas, and Moya Moya disease ($p=0.00$), as well as a higher frequency of seizure disorder, sphenoid-wing dysplasia and poor academic performance. Gadolinium enhancement occurred in 43% of optic gliomas, 50% of parenchymal gliomas, 100% of cranial nerve, 100% of plexus, 67% of paraspinal, 50% of craniofacial and 50% of visceral neurofibromas in Group A, while only 1 Group B tumor enhanced. In Group A, radiological progression occurred after a median of 4 years from initial diagnosis for optic gliomas as well as cranial nerve, plexus and visceral neurofibromas, 2 years for paraspinal neurofibromas and brainstem gliomas, and 2.7 years for craniofacial neurofibromas. Only one tumor progressed in Group B.

CONCLUSIONS Therefore, only 12.5% of NF1 patients who require treatment are at risk of subsequently needing further neurosurgical attention, whereas 87.5% carry an indolent form of NF1. We recommend imaging for asymptomatic, gadolinium-enhancing lesions every 2 years for optic pathway and parenchymal gliomas, and cranial nerve, and visceral neurofibromas, and every year for brainstem gliomas and paraspinal as well as craniofacial neurofibromas. Non-enhancing optic pathway lesions could be followed radiographically much less often since they do not progress.

59. The Far Lateral Approach To Cerebello-Pontine Angle Ependymomas

Sunita Bhamidipaty, MD (Sherman Oaks, CA); Danita Tom, BS (Madison, WI); Michael L. Levy, MD, PhD (San Diego, CA)

INTRODUCTION Management of ependymomas extending into the CP angle remains controversial. The majority of surgeons currently employ a midline approach despite significant extension thru Luschka into the angle. In the midline approach, the approach to this lateral aspect is difficult given the angle of the approach and restrictions of the operative field. We suggest the use of a far lateral approach (either isolated or with a midline exposure) to allow for aggressive resection while diminishing complications.

METHODS Sixty-one patients underwent 82 procedures. We used a far lateral approach in six. Mean age was 33.5 months (range = 11 to 87 months). There were 6 males and no females. Isolated lateral approaches represented 33% of cases while combined procedures represented 67%. The lateral decubitus position was used in all of cases. Pathology for this series included ependymoma in 50% and anaplastic ependymoma in 50%.

RESULTS Gross total resections were achieved in 83% with one failure in a patient with an anaplastic variant. Four patients have required multiple resections, with one free of recurrence to date. Two others remained free of recurrence following their initial surgery. No long-term eye movement abnormalities were noted following surgery. Two patients had transient diplopia and three had transient nystagmus. Ataxia improved in two patients and was exacerbated in three. Two patients required ventilator support following surgery, one transient and one permanent. One patient required long-term NG feeds following surgery.

CONCLUSIONS We suggest that far-lateral approaches allow for a more extensive approach to diffuse CPA tumors while minimizing the potential complications of surgical resection. Despite apparent total resections in 5 patients only two have remained recurrence free to date. No

patients had evidence of spinal instability as the result of the procedure. We advocate that the far lateral approach is a safe alternative and provides ample exposure for tumor resection.

60. Lessons Learned Managing Malignant Gliomas of the Thalamus in Children

F. A. Boop, MD; R. A. Sanford, MD; A. Dyrer, MD; M.M. Muhibbauer, MD; S.J. Einhaus, MD; L.E. Kun, MD; T. Merchant, MD; A. Gajjar, MD; J. Jenkins, MD (Memphis, TN)

INTRODUCTION Malignant gliomas of the thalamus (MGT) in children, although rare, present particular treatment conundrums. Despite improvements in surgical and adjunctive treatments, these children have a rapid decline in performance status and mean survival is just beyond a year.

METHODS The authors review a series of 20 children with MGT treated between 1995 and 2000. Patients presented a mean age of 11 years (Range 2–21 years), typically with symptoms of raised ICP secondary to ventricular obstruction. 10 were male and 10 female. Pathology as anaplastic astrocytoma in 5, glioblastoma in 12 and malignant glioma in 2. In most instances, patients were treated with stereotactic or endoscopic biopsy followed by biventricular shunting. Four patients underwent frameless stereotactic craniotomy and resection. All children were treated with conformal RT and chemotherapy.

RESULTS In 4 cases of biopsy and 1 of shunting, life-threatening intra-tumoral hemorrhage occurred. Two perioperative deaths occurred due to hemorrhage. In 4 cases open resection was performed at presentation, but this did not confer a survival advantage over biopsy and shunting. Karnofsky at presentation was 75 but dropped to 55 by 6 months and 25 by 1 year.

CONCLUSIONS This study suggests no survival advantage to craniotomy and tumor resection over biopsy and shunting except in children with raised intracranial pressure from mass effect. Biopsy of these tumors

carries a 22% risk of significant intratumoral hemorrhage. The authors now place these children in ICU following biopsy and routinely perform a post procedural CT following biopsy.

61. High Incidence of Tumor Dissemination in Myxopapillary Ependymoma in Pediatric Patients

Daniel R. Fassett, MD, MBA; James Pingree, MD; John Kestle, MD (Salt Lake City, UT)

INTRODUCTION Myxopapillary ependymomas usually occur in adults and have been regarded as benign tumors. Only 8–20% of these tumors occur in the first two decades making this tumor rare in pediatric neurosurgery.

METHODS Our records contained five cases of intraspinal myxopapillary ependymoma from 1992 to present. The literature yielded three pediatric case series of myxopapillary ependymoma and an additional two case series from which data regarding pediatric patients could be extracted. These five case series were combined with our series giving a total of 26 cases of pediatric myxopapillary ependymoma for review.

RESULTS Four of our 5 cases (80%) had disseminated disease. When combined with the literature, the average age at presentation was 11.1 years (range 8–17) with a male predominance of 1.89:1.0. Twenty-two of the 26 cases were lumbar (typical conus-cauda-filum lesions), three did not have tumor location reported, and one was cervical. Nine patients (35%) had disseminated disease. All patients with disseminated disease were treated with surgical resection of their primary tumor followed by radiation therapy. Two patients also received chemotherapy. Tumor control appeared to be excellent with all patients having stable disease or no apparent disease at follow-up.

CONCLUSIONS Myxopapillary ependymoma in pediatric patients has a relatively high propensity to spread throughout the CNS via cerebrospinal fluid pathways. Despite the aggressive nature of this tumor in regard to dissemination, metastatic disease

can be very well controlled with radiation therapy. Therefore, screening of the entire CNS axis with MRI is recommended at the time of presentation and follow-up.

62. Malignant Rhabdoid Tumors of the Brain

Bradley E. Weprin, MD; Kenneth N. Shapiro, MD; Frederick Sklar, MD; Dale M. Swift, MD; Korgan Koral, MD; Lynn Gargan, PhD; Jill Mraz, ARNP; Shana Valles (Dallas, TX)

INTRODUCTION The malignant rhabdoid tumor of the brain (MRT) is a rare tumor that has recently been established as a distinct clinicopathologic entity. It is an aggressive neoplasm that is associated with a poor prognosis in spite of multiple management regimens. In an attempt to guide future therapy, we have reviewed the mode of presentation, radiographic findings, clinical course and outcome of individuals with MRT treated at a single institution.

METHODS The medical records and radiographic imaging studies of children diagnosed with MRT of the CNS were reviewed.

RESULTS Between 1985 and 2002, 16 children were diagnosed with malignant rhabdoid tumors out of the 1139 individuals with brain tumors evaluated at our institution. The male: female ratio was 1:1. The mean age at diagnosis was 1.08 ± 0.21 years (range: 0.22-2.73 years). The mean period of follow-up was 0.65 ± 0.16 years. Thirteen children presented with unifocal disease while three exhibited evidence of multifocal disease at presentation. Location at presentation included 9 with infratentorial lesions, 5 with supratentorial lesions, and 2 with combined supra- and infratentorial lesions. Treatment consisted of a variable combination of surgery, chemotherapy and radiation. A subtotal resection was performed in 14 and a gross total resection accomplished in 2.

Disease progression occurred in 9/16 patients with the mean time to progression calculated as 0.32 ± 0.11 years. The overall survival was 18.75% with the mean time to expiration calculated to be 0.71 ± 0.19 years

(range: 0.02–2.19 years). The mean time to expiration was calculated for the following treatments: surgery alone (0.08 ± 0.03 years), surgery plus chemotherapy (0.62 ± 0.18 years), and surgery plus chemotherapy plus radiation therapy (1.13 ± 0.43 years).

CONCLUSIONS MRT seems to most often develop in children under three. The mode of presentation and radiographic findings are often nonspecific. The outcome is extremely poor in spite of multidisciplinary approaches. A search for more effective therapies is necessary.

63. Epidemiological Changes in Spinal Dysraphism

Jeffrey W. Campbell, MD (Charleston, SC)

INTRODUCTION The incidence of live births with myelomeningoceles fell in the 1990's, in part due to public health initiatives such as introduction of folic acid into breakfast cereals and education regarding prenatal vitamins. It is unclear whether these interventions have resulted in a lower incidence of occult spinal dysraphism such as spinal lipomas or split cord malformations.

METHODS The Nationwide Inpatient Sample of the Healthcare Cost and Utilization Project (HCUP-3) provides detailed information on inpatient services rendered in a representative sample of U.S. hospitals with weighting factors that allow reasonable estimates of services rendered in the entire country. Roughly 20% of inpatient admissions are captured in this database, providing valuable epidemiological information on all inpatient healthcare services. This database was used to compare the relative incidence of myelomeningocele closures to the surgical treatment of other forms of spinal dysraphism between the years 1988–1998.

RESULTS The relative incidence of surgical treatment of occult spinal dysraphism compared to myelomeningoceles rose by roughly fourfold over this 10 year period. In addition, there were moderate differences in the gender and racial makeup of these two forms of spinal dysraphism. The average length of stay and hospital costs

were roughly twofold higher in patients undergoing closure of a myelomeningocele compared to surgical treatment of occult spinal dysraphism.

CONCLUSIONS The incidence of treated occult spinal dysraphism appears to be increasing in the United States, suggesting that the public health initiatives thought helpful in prevention of myelomeningoceles are not effective with occult spinal dysraphism. Since the true incidence of occult spinal dysraphism is not known, some of this increase may be related to changes in diagnosis or surgical indications.

64. Pediatric Hydrocephalus Treatment Using a Flow Regulating Shunt: A Prospective Five Years Shunt Survival Study (European OSV II Study)

Patrick W. Hanlo, MD (Utrecht, Netherlands); Giuseppe Cinalli, MD (Paris, France); Peter Vandertop, MD (Amsterdam, Netherlands); Lars Bogeskov, MD (Copenhagen, Denmark); Svend E. Borgesen, MD; Jürgen Boschert, MD (Mannheim, Germany); Paul Chumas, MD (Leeds, England); Hans Eder, MD (Graz, Austria); Ian Pople, MD (Bristol, England); Willy Serlo, MD (Oulu, Sweden)

INTRODUCTION To evaluate long-term results of a flow-regulating shunt (OSV II, NMT Neuroscience Implants (SA), Sophia Antipolis, France) in the treatment of pediatric hydrocephalus, whether it was a first insertion or a revision of another type of shunt, in everyday clinical practice in a prospective multicenter study (9 European centers).

METHODS Patients with hydrocephalus, at any age, with different etiologies, were treated by implanting an OSV II system (ventriculo-peritoneal shunts: 95.2%). Primary endpoint was defined as any shunt related surgery. The secondary endpoint was the type of mechanical complication (shunt obstruction, overdrainage, catheter misplacement, migration or disconnection) or infection. The overall five-years shunt survival and the survival in the different subgroups were assessed.

RESULTS Two-hundred-and-ninety (290) patients were selected for OSV II implantation, 110 patients reached an endpoint. Shunt obstruction occurred in 42 (14.5%), overdrainage in 1 (0.3%), and infection in 25 (8.6%). The probability of having a shunt failure-free interval at 1 year was 71%, 67% at 2 years, and then remained stable in following years (to 61% at 5 years follow up). Shunt survival was significantly lower in children under six months of age at 5 years follow up (54%). In this group shunt obstruction occurred in 20.8% (with 14.6% valve obstruction), infection in 9.4%, underdrainage in 8.3% and no overdrainage. There was no significant difference regarding shunt survival whether it was a first insertion of an OSV II or a revision of another type of shunt. Using Cox Regression analysis it was demonstrated that only age was the most important factor of influence on the primary endpoint.

CONCLUSIONS This prospective study demonstrates the effectiveness of flow-regulation in the treatment of hydrocephalus in children. Flow-regulating shunts significantly limit the incidence of overdrainage and shunt related complications. The overall five-years survival rate (61%) compares favorably to other recent published series.

65. Isolated Lumbosacral Strawberry Nevi Do Not Indicate The Presence of Occult Spinal Dysraphism

Nathan R. Selden, MD, PhD (Portland, OR); Rebecca M. Allen, BA (Stanford, CA); Michael A. Sandquist, MD (Portland, OR); Joseph H. Piatt, MD, FACP (Philadelphia, PA)

INTRODUCTION The presence of cutaneous strawberry nevi commonly prompts a search for underlying occult spinal dysraphism. Nevertheless, the risk of occult spinal dysraphism in patients with strawberry nevi alone or in combination with other cutaneous markers is largely unknown.

METHODS 20 consecutive newborns with lumbosacral strawberry nevi at our institution were investigated with spinal ultrasound and/or MRI. Fifteen patients had strawberry nevi in isolation and five had strawberry nevi plus other associated cutaneous markers of occult spinal dysraphism.

RESULTS None of the patients with isolated nevi, but four of five patients with other cutaneous markers, harbored occult spinal dysraphism, as confirmed by ultrasound, MR imaging and surgical exploration.

CONCLUSIONS Our findings indicate that strawberry nevi in isolation do not appear to indicate underlying dysraphic states. The sparse clinical literature on this topic confirms an association between occult spinal dysraphism and strawberry nevi presenting in conjunction with other cutaneous signatures. Prospective study of larger numbers of patients with isolated strawberry nevi is necessary to conclude whether radiographic screening of these patients is indicated.

66. The Tethered Cord in Arthrogyrosis Multiplex Congenita

Kimberly D. Bingaman, MD; Laura L. Tosi, MD; Philip H. Cogen, MD, PhD; Robert F. Keating, MD (Washington, DC)

INTRODUCTION Spinal cord tethering in association with anorectal and urogenital abnormalities has been well described. Arthrogyrosis multiplex congenita is a symptom complex characterized by multiple joint contractures that are present at birth. Various etiologies for arthrogyrosis have been hypothesized, but it appears that the final common pathway involves decreased fetal movement in utero. The only spinal cord abnormalities previously reported in patients with arthrogyrosis are those involving the anterior horn cells.

METHODS A retrospective chart review was performed which examined all cases of patients referred to the neurosurgery service at Children's National Medical Center from October, 1997 to May, 2002 with the diagnosis of both arthrogyrosis and tethered cord.

RESULTS 6 patients, who ranged in age 5 weeks to 12 years (2 female, 4 male), were identified who carry the diagnoses of both tethered cord and arthrogyrosis. 5 patients underwent MRI of the spine because of spinal cord tethering symptoms such as progressive scoliosis, back pain, or increased lower extremity tone. One patient underwent an MRI because of a

sacral dimple, but was subsequently noted to have increased tone in the lower extremities. MRI findings demonstrated a low-lying cord (below L3) in each patient. Some patients also had a hypertrophied or lipomatous filum terminale. All 6 patients underwent untethering procedures and all improved or stabilized function and/or their scoliosis.

CONCLUSIONS Patients with arthrogyrosis who develop progressive neurologic decline or orthopedic deformities should be examined for the presence of a tethered cord. These patients can improve from untethering procedures.

67. Syringobulbia in a Pediatric Population

Jeremy D.W. Greenlee, MD; Kathleen A. Donovan, ARNP; Arnold H. Menezes, MD (Iowa City, IA)

INTRODUCTION Syringobulbia in the pediatric age group is a very rare condition, and little is published on the subject. In an effort to better understand the condition, we reviewed our series of patients.

METHODS The University of Iowa pediatric neurosurgery database was searched for patients under the age of 18 with a diagnosis of syringobulbia. The patients' records were then reviewed for demographic data, chief complaint and presenting symptoms, neurologic and radiographic findings, treatment, outcome, and complications.

RESULTS Six pediatric patients were identified. The average age at time of surgery was 14.8 years. The chief complaints were vision impairment in 3 patients, and numbness, gait instability, and headache worsened with valsalva in one patient each. Other prominent symptoms included sleep apnea and weakness. Four patients showed at least one cranial nerve palsy. Radiographs revealed hindbrain herniation and syringomyelia in all cases. Two patients had scoliosis. Treatment was posterior fossa decompression with tonsillar shrinkage, opening of foramen of Magendie, and duraplasty. The cavity of syringobulbia communicated with syringomyelia and the fourth-ventricle (V4) in all children, but was

distinct from V4. Two patients received fourth ventricle to subarachnoid shunts. Follow-up averaged 2.6 years, and all patients clinically improved. MRI documented resolution of syringobulbia in all cases, with syringomyelia improving in all cases. There was no permanent morbidity or mortality in the series.

CONCLUSIONS Syringobulbia is associated with Chiari malformation and syringomyelia, and patients usually present due to cranial nerve palsies. Posterior fossa decompression is a safe and effective treatment.

68. Mechanisms of Reclosure in a Surgical Model of Myelomeningocele: Implications for Fetal Surgery

David C. Adamson, MD, PhD; Timothy M. George, MD (Durham, NC)

INTRODUCTION Surgical models have been used to help study the pathophysiology of open neural tube defects. A major drawback of the surgical models has been that the neural tube will reclose if the lesion is performed early in gestation. This study attempts to determine if the reclosure is a recapitulation of primary neurulation or by another mechanism.

METHODS Chick embryo were grown until stages 10 to 14 then explanted into an ex ovo culture system. A sharpened tungsten wire was used to lesion the roof plate of the closed spinal neural tube in a rostral to caudal manner for a length of 2 to 9 somites. Embryos were either lesioned only or lesioned and treated with a sense, antisense or missense oligonucleotides to Pax-3, and/or citral and retinoic acid.

RESULTS Embryos tended to reclose regardless of the treatment. However, other embryonic defects were noted in response to the specific treatment.

CONCLUSIONS Spontaneous healing of the early surgical defect implies that the reclosure is likely due to a type of healing mechanism and is not a recapitulation of primary neurulation. Further studies on the mechanism of healing may be important in the future management of open neural tube defects.

69. Reducing the Incidence of Abusive Head Injuries Through Hospital-Based Parent Education: The Upstate New York Shaken Baby Syndrome Education Program

Mark S. Dias, MD (Hershey, PA.); Kim Smith, RNC; Kathy deGuehery, RNC; Diana Kachurek, RNC (Buffalo, NY); Veetai Li, MD (New York, NY)

INTRODUCTION A hospital-based parent education program was begun in December 1998 to educate both parents (mothers and, whenever possible, fathers or father figures) of all infants born in an 8 county region of Western New York (WNY), before discharge from the hospital, about the dangers of violent infant shaking, and asked that they, in return, voluntarily sign a commitment statement affirming their receipt and understanding of the information. The premise was that parents needed to be reminded at the correct time (upon the birth of a child) about SBS, and that educated parents could be advocates to disseminate this information to all who care for their child.

METHODS Parents of newborn infants received written and video information about violent infant shaking, and were asked to voluntarily sign a commitment statement affirming their receipt and understanding of the materials. Commitment statements were returned monthly to the study coordinators and tracked. The incidence of abusive head injuries was tracked during the program and compared with the incidence during a 6 year control period immediately preceding the program.

RESULTS All 16 regional hospitals participated in the program. A total of 30,345 commitment statements were returned, representing 58% of the live births during this 3 year period. The incidence of abusive head injuries declined 62% from an average of 7 cases per year (range 4-8) during the control period, to 2.7 cases per year (average 2-3) during the three years of the study ($p < 0.05$). Of the 8 cases identified during the three years of the study, the parents of at least 5, and perhaps as many as 7, had not participated in the program.

CONCLUSIONS Hospital-based parent education at the time of a child's birth can significantly reduce the incidence of violent infant shaking.

70. Neuroendoscopic Findings in Patients with Intracranial Germinomas Correlate with Diabetes Insipidus

John C. Wellons, MD; Richard S. Tubbs, PhD, PA-C; Alyssa T. Reddy, MD; Hussein Abdullatif, MD; Walter J. Oakes, MD; Jeffrey P. Blount, MD; Paul A. Grabb, MD (Birmingham, AL)

INTRODUCTION Central nervous system germinomas commonly occur in the pineal region and the floor of the third ventricle. Presenting symptoms include headache, vomiting, Parinaud's sign, or diabetes insipidus (DI). We have found that preoperative DI predicts the presence of tumor on the third ventricular floor visualized endoscopically despite nonvisualization of such tumor on magnetic resonance imaging (MRI).

METHODS The hospital records and imaging from patients who underwent third ventricular tumor biopsy with or without endoscopic third ventriculostomy (ETV) at Children's Hospital of Alabama from May 1998 to January 2002 were reviewed. Nine patients with the diagnosis of pure germinoma were identified. Preoperative MRI and presenting symptoms were correlated with intraoperative neuroendoscopic findings.

RESULTS Of these nine patients with biopsy-proven pure germinomas, six presented with symptomatic hydrocephalus and underwent concomitant ETV. Five patients presented with DI and MRI evidence of involvement of the third ventricular floor. Two patients presented with DI, and no involvement of the third ventricular floor by MRI. Both had clearly detectable disease, however, involving the floor of the third ventricle seen during endoscopic biopsy of the pineal lesion. The one child who presented without DI had no involvement of the third ventricular floor by both MRI and direct endoscopic observation.

CONCLUSIONS Neuroendoscopic tumor biopsy and ETV have simplified the diagnosis and surgical management of CNS germinomas. In our experience, preoperative DI is a predictor of involvement of the third ventricular floor regardless of MRI. Neuroendoscopy may be a more accurate means of determining disease extent.

71. Failed Endoscopic Third Ventriculostomy in Children:

Manoj K. Mishra, MD; Aaron Mohanty, MD; Thimappa Hegde, MD; S. Sampath, MD; M. K. Vasudev, MD; Sastry V. R. Kolluri, MD (Bangalore, India)

INTRODUCTION Endoscopic third ventriculostomy (ETV) for obstructive hydrocephalus has a failure rate of 20–50% in various series. Of the various causes for failure, closure of the third ventriculostomy site by scarring has been observed in 6–15% of cases. Proper identification of this group of patients who would benefit from a repeat third ventriculostomy procedure is of considerable importance.

METHODS 72 children underwent ETV between January 2000 to December 2001 of which 13 failed. These patients were studied with MRI with sequences to identify flow at the ventriculostomy site. Patients with no flow were considered for repeat ETV. Patients with a good flow void underwent shunt placement without an endoscopic exploration.

RESULTS Of the 13 failures, in 7 it manifested within 1 month and in 5 others it failed between 1–2 months. Another child had a delayed failure after 2 years of the initial surgery. Of these, in 12 patients no flow could be demonstrated. One patient had a good flow. Of the 12 undergoing endoscopic exploration, in 7 the stoma was patent, necessitating a VPS insertion. In the rest 5, the stoma had closed by gliosis and a repeat ETV was performed. Ventriculoperitoneal shunt insertion was carried out in the patient with suggestion of a good flow.

CONCLUSIONS In failed ETV, MRI with flow studies is essential to identify the possible cause of failure. Endoscopic exploration is

indicated for patients with no evidence of flow. A repeat ETV is indicated in patients with a closed stoma. Patients with patent stoma would require a shunt insertion.

72. Results of Endoscopic Septal Fenestration in the Treatment of Isolated Ventricular Hydrocephalus

Philipp R. Aldana, MD; John R.W. Kestle, MD; Douglas L. Brockmeyer, MD; Marion L. Walker, MD (Salt Lake City, UT)

INTRODUCTION Although a common indication for neuroendoscopy has been the treatment of isolated lateral ventricular hydrocephalus (ILVH), surgical series detailing the results and complications of this treatment are lacking. We examine our experience at the Primary Children's Medical Center with endoscopic fenestration of the septum pellucidum or septostomy for the treatment of ILVH.

METHODS We studied a series of 32 consecutive patients in whom an endoscopic septostomy was performed between 1993 and 2001. The patients were identified from a search of our computerized database.

RESULTS Forty-three septostomies were performed in 32 patients, with a mean follow-up was 30.9 months. Fifty-three per cent of initial septostomies remained patent. Ten patients had at least one more septostomy performed after the initial septostomy failure. All but one were successful. Including repeat septostomies, 81% of the patients had relief of their ILVH on last follow-up. No septostomy failures occurred later than 6 months postoperatively. A history of multiple previous shunt procedures was significantly predictive of initial septostomy failure, increasing this risk 4.5 times. Four procedures (9.3%) were complicated by significant intraventricular hemorrhage, wound breakdown, shunt infection and sterile meningitis. Two septostomies were aborted due to ventricular anatomy unfavorable for septal fenestration.

CONCLUSIONS Endoscopic septostomy is a reasonable treatment option for ILVH, avoiding additional cerebrospinal fluid shunts. Outcome is negatively affected

by multiple prior shunt procedures. Favorable results can be achieved with repeat septostomies in patients who have failed similar treatment previously. Lasting results are expected for septostomies that remain patent after 6 months.

73. Management of Brain Tumor Related Obstructive Hydrocephalus with Endoscopic Third Ventriculostomy

Karl F. Kothbauer, MD (New York, NY); Bong Soo Kim, MD (Chicago, IL); George I. Jallo, MD; Rick Abbott, MD (New York, NY)

INTRODUCTION Braintumor-related obstructive hydrocephalus may be treated with a ventriculoperitoneal shunt or an endoscopic third ventriculostomy. The purpose of this study is to review the authors' series of patients treated endoscopically to assess outcome and identify factors influencing it.

METHODS 61 patients with tumor-related obstructive hydrocephalus were treated with an endoscopic third ventriculostomy. Clinical and radiographic data were analyzed from the available documentation and from new patient follow-up when appropriate. The following factors were analyzed in the entire patient group and comparatively studied in subsets of patients: age at the time of ventriculostomy, sex, type and location of tumor, prior shunting, success rate defined as long-term shunt independence, as well as the frequency and nature of ventriculostomy-related complications.

RESULTS There were 31 male and 32 female patients with a mean age of 15.5 years. Seventeen patients had tectal midbrain tumors, 5 tegmental midbrain tumors, 3 pineal region tumors, 7 midbrain/basal ganglia tumors, 5 thalamic tumors, 1 3rd ventricle tumor, 6 diffuse pontine gliomas, 3 circumscribed brainstem tumors, 6 fourth ventricular tumors, 1 cerebellar and 1 frontal tumor; 2 patients had disseminated tumors. There were 38 low grade astrocytomas, 3 anaplastic astrocytomas, 5 glioblastomas, 5 PNETs, 5 Ependymomas, and 6 had other tumors. The overall success rate was 79% (48 patients with a mean follow-up 20.4 months). Patients with

midbrain tumors had an 86% success rate. Patients with deep basal ganglia neoplasms had 91% success rate. Mild transient complications occurred in 5 patients and a ventriculitis 1.

CONCLUSIONS Based on the premise that endoscopic third ventriculostomy provides an inherent advantage over shunting because of the relative normalcy of CSF flow and the absence of implanted foreign materials the success rate in this series is considered favorable for the endoscopic approach.

74. Frameless-Stereotactic Endoscopic Placement of Third to Fourth Ventricular Stent

John A. Lancon, MD (Jackson, MS)

INTRODUCTION Advances in neuroendoscopy and intraoperative image-guidance have facilitated the management of multiloculated hydrocephalus. However, the management of the isolated fourth ventricle remains challenging. A conventional approach is the creation of a ventriculo-cystoperitoneal shunt with two proximal catheters (placed into a lateral ventricle and the fourth ventricle) connected via a Y-connector to a single valve and distal catheter. Although this is effective in many instances, complications associated with placement of the fourth ventricular catheter are relatively common. These include infection, cerebrospinal fluid leak, pseudomeningocele formation, brainstem injury, and torquing of the fourth ventricular catheter with secondary scarring to the ventricular wall. As an alternative management strategy, a technique for frameless-stereotactic endoscopic placement of a third-to-fourth ventricular stent was developed.

METHODS A fiducial MRI is obtained and presurgical planning completed. An instrument-tracker is attached to a 1.2 millimeter endoscope which is registered into the workstation. Navigation based on surface landmarks is confirmed. A trajectory is selected to allow a direct path through the interventricular foramen to the aqueductal orifice. A stent introducer is fashioned from a ventricular catheter and loaded into the

endoscope followed by the stent. The endoscope-introducer-stent construct is advanced into the frontal horn under image-guidance. The endoscope tip is advanced to the aqueductal orifice under endoscopic guidance. The stent is delivered into the aqueduct using the introducer. Placement is confirmed with direct visualization.

RESULTS This technique has been used to place a third-to-fourth ventricular stent in four cases of symptomatic isolated fourth ventricle. There were no perioperative complications. Postoperative imaging studies have confirmed decompression of the fourth ventricular cyst. There have been no infections or stent occlusions in the first year of follow-up.

CONCLUSIONS Frameless-stereotactic endoscopic placement of a third to fourth ventricular stent appears to be a useful alternative to direct shunting of an isolated fourth ventricular cyst in some patients.

75. Sports and Recreational Head and Spine Injuries in Children

David J. Yeh, MD; Scott Y. Rahimi, MD; Ann Marie Flannery, MD; Mark R. Lee, MD, PhD (Augusta, GA)

INTRODUCTION Recreational head and spine injuries in children sustained during organized and extemporaneous community sports activities frequently call for neurosurgical evaluation.

METHODS We reviewed office charts spanning from 1992 to the present and identified recreational and sports injuries which either required outpatient follow-up or inpatient treatment under the primary care of the pediatric neurosurgical service. We excluded all injuries associated directly with the use of motorized vehicles with the exception of battery-powered golf carts.

RESULTS A total of 2546 charts were retrospectively reviewed. 82 patients who sustained recreational head, spine, or brachial plexus injuries were identified. The most common activities included bicycling (34.1%), golfing (19.5%), baseball (11.0%), and gymnastics (8.5%). Unusual sport injuries were also seen with trampolining (4.9%), skateboarding/rollerskating (2.4%),

and hockey (1.2 %). Surprisingly few injuries were recorded from football (4.9%). Most common injury mechanisms involved falls, missiles or blunt objects striking the calvarium, or collisions. Head injuries accounted for 77% of cases and most commonly consisted of skull fracture (45.0%), concussion (33.3%), and intracranial hemorrhage (18.1%). Spine injuries accounted for 19.2% of injuries and most commonly consisted of cervical fractures or sprain (60.0%). Brachial plexus injuries accounted for 2.6% of injuries. 21.3% of patients required surgical intervention (19.5% craniotomy). Good recovery to baseline function was seen in 77.5% of patients. There were 2 deaths secondary to severe closed head injury, neither of which underwent craniotomy. The most frequent untoward sequelae observed were psychiatric or cognitive dysfunction (7.0%). Over 70% of injuries occurred during the spring and summer months of March through August, and over 70% of all injuries occurred in male children.

CONCLUSIONS Pediatric sports and recreational head and spine injuries requiring neurosurgical evaluation and/or surgical intervention are frequent. We review our experience with this series of 82 patients and describe observed patterns of injury occurrence.

76. Long Term Outcome Following Gunshot Wounds to the Spine in Children and Adolescents

Anthony Kim, MD, PhD (Sherman Oaks, CA); Michael L. Apuzzo, MD (Los Angeles, CA); Michael L. Levy, MD, PhD (San Diego, CA); Rod Adkins, PhD (Los Angeles, CA)

INTRODUCTION The incidence of gunshot wounds (GSW) to the spine in children has increased from 6 to 38% over a 15-year period. A review was undertaken to assess the treatment and long term outcome of this group of patients. The patients were subsequently transferred to a rehabilitation facility for further treatment. Exams were completed as part of a prospective analysis utilizing the ASIA score and Frankel scales. The assessment parameters 1) degree of neurological damage and 2) degree of pre-

served neurologic function were employed to describe the severity of injury ($p < 0.05$) and were assessed at admission and the time of definitive discharge from the SCI care system.

METHODS Identified were 60 patients whose mean age was 17.6 years (± 1.5 yrs) of whom 55 were males. The distribution of injuries was as follows: Cervical—12, thoracic—31, lumbosacral—17, of which 34 presented with complete injuries. Acute hospitalization lasted for 21.1 days (± 22.8 days) and the rehabilitation stay was 86.3 days (± 48.9 days) for a total mean hospitalization of 107.4 days (± 65.9 days).

RESULTS At five year follow-up there were 19 ambulators and 53 patients were autonomous. Significant improvement in the exam was noted at one-year follow-up ($p < 0.0001$). Scores improved by 11 and 41 (complete vs. incomplete) in patients with cervical injuries, .7 and 13.5 in thoracic injuries, and 7 and 14 in lumbar injuries. There was also a significant improvement between complete and incomplete injuries ($p < 0.0001$).

CONCLUSIONS Patients with spinal cord injuries resultant from GSW's spent less time in rehab than those with other types of SCI's.

77. Operative Compound Skull Fractures in Children

Richard B. Rodgers, MD; Stephen Kralik, BS; Thomas G. Luerksen, MD (Indianapolis, IN)

INTRODUCTION Compound skull fractures in children, although relatively uncommon, pose interesting clinical dilemmas, especially in regards to operative treatment and prevention of infection. We discuss our experience with operative open fractures at a Pediatric Level One Trauma Center.

METHODS A Retrospective review of records (1989-present) of patients with open skull fractures requiring operative treatment.

RESULTS 1162 patients with head trauma requiring admission are included in our database. 544 (47%) had skull fractures, and 43 (3.7%, mean age 6.2 years) of

these suffered open fractures. 22 patients required operative treatment, based on depression, evidence of dural violation, foreign body or mass lesion. Mechanism of injury varied. Mean presenting GCS was 14 (range 9–15). Treatment consisted of operative debridement, irrigation, dural repair, and immediate reconstruction with all available skull fragments. Time from injury to operation varied, with 75% treated within 8 hours. All received general peri-operative antibiotics, with special consideration from the Infectious Disease service for deeply penetrating wounds. Mean hospital stay was 6 days (range 1–14). GOS was 1 in 21/22 patients. All patients with neurologic/cognitive deficits improved in long-term follow-up (mean 7.9 years). Complications included one patient with post-operative CSF leak requiring re-exploration and lumbar drainage, and one reoperation for enlarging hematoma versus infection.

CONCLUSIONS Compound skull fractures in children frequently require management in the operating room. Debridement, irrigation and immediate reconstruction without craniectomy within a reasonable time period, along with a short course of antibiotics is our usual practice. The complication rate is low. The presenting GCS and outcomes were indicative of the focality of injury.

78. Third Ventriculostomy Safety Device: A Preliminary Report

Todd A. Maugans, MD (Burlington, VT)

INTRODUCTION Third ventriculostomy is becoming an increasingly common technique employed in the management of hydrocephalus. Despite its high success rate in appropriately selected patients, both early and late failures have been recognized. These patients may develop symptoms attributed to elevated ICP. Worse, patients may experience death if the failure is unrecognized and/or not rectified in a timely fashion. Access to the ventricular system is paramount to assess intracranial pressure and divert CSF if necessary.

METHODS A novel device has been developed that consists of a ventricular catheter with integrated reservoir and a detachable,

externalizable tubing segment. The device is placed at the time of third ventriculostomy and the externalized tubing is transduced and/or used for temporary CSF diversion. After assuring success of the procedure, the tubing is manually separated from the reservoir which remains as a subcutaneously located access portal for the later assessment/management of ICP issues as clinically indicated. An information kit was developed to be distributed to patients, families and providers outlining the indications and technique for accessing the device emergently.

RESULTS A prototype of the device was developed and tested in a laboratory setting. The final product has been implanted into one patient to date. A male teenager was operated upon for PNET with obstructive hydrocephalus. The patient's ICP was normal immediately postoperatively, however, he developed symptoms suggestive of elevated ICP months later. Mild intracranial hypertension was confirmed by percutaneous access of the device. The device was subsequently converted to a ventriculoperitoneal shunt. An infection of the device necessitated its removal.

CONCLUSIONS A novel device has been developed to offer immediate and long term ventricular access following third ventriculostomy. Preliminary experience in a single patient confirmed its benefits and revealed potential liabilities that may be similar to shunts. A multi-institutional prospective study of the use of this device and an associated educational program is being planned.

79. Endoscopic Intracranial Cyst Fenestration in Neonates and Infants

Christian W. Sikorski, MD; Yamini Bakhtiar, MD; David M. Frim, MD, PhD (Chicago, IL)

INTRODUCTION Intracranial cyst as a cause of hydrocephalus in infants is common. Historically, these lesions have been treated with open fenestration or shunting. More recently, however, endoscopic fenestration has become a viable initial intervention. While there is a significant amount of data regarding endoscopic fenestration of cysts

in older children and adults, there is not similar information regarding outcomes in neonates and young infants. We present a retrospective analysis of 7 infants with intracranial cysts treated with fenestration at our institution.

METHODS Patients ranged in age from 3 days to 6 months at time of surgery. Mean follow-up was 36 months. The locations of the cysts were: suprasellar, lateral ventricle (2), foramen of Monro, third ventricle and pineal region. All were treated with rigid or flexible endoscopic fenestration using unipolar electrocautery or blunt technique from standard frontal or occipital approaches.

RESULTS 3 patients avoided extracranial shunting completely. In 1 case the need for a second intraventricular shunting catheter was avoided. The remaining infants required cyst or ventricular shunts within several months of the initial fenestration.

CONCLUSIONS Initial endoscopic fenestration of intracranial cysts that cause mass effect or hydrocephalus succeeded in avoiding a shunt in 3 out of 7 and prevented a second shunting catheter in another infant. 3 of 7 infants failed fenestration and required shunting. Despite this low success rate, we argue that due to the morbidity associated with lifelong shunting from infancy, there is a strong consideration for such a shunt-preventing procedure.

80. Aberrant Neuronal Development in Hydrocephalus

Jonathan P. Miller, MD; David W. Leifer, BS; David Lust, PhD; Alan R. Cohen, MD (Cleveland, OH)

INTRODUCTION Although hydrocephalus has been studied extensively, little is known about neuronal development in this condition. We hypothesize that congenital hydrocephalus is associated with unique patterns of neuronal migration and development. In the present study, we show that congenital hydrocephalus is associated with increased apoptotic activity as well as disruption of neuronal migration, which may explain the depletion of cortical neurons in hydrocephalus. Furthermore, both processes affect the same region of the

developing brain, suggesting that there may be an association between the two.

METHODS We examined neuronal migration and apoptosis in a transgenic model of hydrocephalus in animals overexpressing the cytokine, transforming growth factor beta-1. In the present report, we used bromodeoxyuridine (BrdU) labeling to evaluate abnormalities of neuronal migration, and caspase-3 immunostaining to evaluate apoptosis. By injecting pregnant mice with BrdU on day E15 we selectively identified dividing neurons and trace their subsequent migration. Pups were sacrificed at appropriate developmental stages, fixed and cryopreserved. Polymerase chain reaction was used to ascertain the status of the transgene. Heads were sectioned and stained for BrdU and caspase-3 and visualized on a Leica fluorescent microscope.

RESULTS Equal numbers of BrdU-positive neurons were seen throughout the cerebral cortex of hydrocephalics vs. controls at age E18. Significantly fewer BrdU-positive neurons were seen in the cerebral cortex and hippocampus of hydrocephalics vs. controls at age P2. An abnormal accumulation of neurons was seen around the ventricles in the hydrocephalics, with an aberrant pattern of migration to the cortex. Caspase-3 staining revealed increased apoptotic activity in the same distribution in hydrocephalics.

CONCLUSIONS In this animal model of hydrocephalus, abnormalities of neuronal migration and survival precede the onset of ventriculomegaly and may account for the depletion of cortical neurons. Further elucidation of the unique abnormalities of neuronal development may lead to novel methods for neuronal protection in hydrocephalus.

81. Pitfalls in the Use of a Hydroxyapatite/Absorbable Plate Construct to Repair Full Thickness Skull Defects in Children

Gerald F. Tuite, MD (Saint Petersburg, FL); Ernesto Ruas, MD (Tampa, FL); Carolyn Carey, MD (Saint Petersburg, FL)

INTRODUCTION Hydroxyapatite cement has been used for a variety of surgical

indications over the past several years. Experimental and early clinical results have been favorable, but long term follow-up data is limited.

METHODS Our craniofacial team has utilized a combination of an absorbable plate and hydroxyapatite cement to fill full thickness bone defects over the past three years. The surgical technique involves the placement of an absorbable plate in the epidural plane, under the bone defect. This creates a relatively deep trough which is then filled with hydroxyapatite cement to create a thick cranioplasty. Patients were followed clinically and radiographically to analyze the integrity of the construct.

RESULTS Between 1999 and 2001, twenty children had their cranial defects repaired using the aforementioned method. Their ages ranged from two to twelve years, with a median age of 7 years. Most patients who underwent the procedure had undergone a craniofacial repair as an infant and were left with a defect that did not fill in completely. The size and number of the defects varied, with most patients having a single defect that was less than five centimeters in diameter. However, two children required reconstruction of very large bifrontal areas. Our average follow-up period was 20 months.

There were no significant intraoperative difficulties or perioperative complications. Each patient had a satisfactory cosmetic result, and the immediate patient satisfaction was high. In all but two patients, the bone source has remained solid and the cosmetic result has endured. However, the two patients who had very large defects have required revision of their cranioplasties: After several months, the constructs fractured and failed.

CONCLUSIONS An absorbable plate/hydroxyapatite construct has advantages over some other methods for the repair of small cranial defects in children. However, we would not recommend its use for repair of very large defects.

82. The Repair of Large (>25cm²) Skull Defects Using "Reinforced" Hydroxyapatite Cement: Technique and Complications

Susan R. Durham, MD (Oregon City, OR); Michael L. Levy, MD; J G. McComb, MD (Los Angeles, CA)

INTRODUCTION Hydroxyapatite cement is both biocompatible and osteoconductive and lacks significant toxic or immunogenic properties making it an ideal substrate for the repair of cranial defects. However, due to its putty-like composition, the repair of large cranial defects can be difficult as significant settling occurs as the cement hardens. We describe a technique using hydroxyapatite cement, reinforced with tantalum mesh and titanium miniplates, for the repair of large (>25 cm²) cranial defects.

METHODS After the margins of the cranioplasty are delineated, tantalum mesh is placed under the edges of the defect. Titanium miniplate single-hole bars are used to criss-cross the defect and secured to the surrounding bone using screws. The mesh is secured to the bars using 28-gauge stainless steel wire. Hydroxyapatite cement is applied in the defect and contoured appropriately.

RESULTS We have performed nine cranioplasties in eight patients ranging in age from 1.5 years to 35 years (mean 12.2 + 10.1 years). Etiologies included cranial defect from prior trauma (n=4), fibrous dysplasia (n=2), infected bone flaps (n=2) and tumor (n=1). The cranioplasties ranged in size from 40 cm² to 196 cm² (mean 128.3 + 56.9 cm²). Follow-up ranged from 2 to 33 months (mean 11.4 + 12.8 months). Two cranioplasty constructs were removed at 1 and 3 months postoperatively secondary to infection.

CONCLUSIONS The use of hydroxyapatite cement with mesh and miniplates provides internal structural support and increased stability of the construct. While providing an excellent cosmetic result and no evidence to date of bony resorption, the rate of infection is alarmingly high in these large constructs.

83. Cisterna Magna to Pleural Shunting in Pseudotumor Cerebri Patients Who Fail Lumbar or Ventricular Shunting

Bakhtiar Yamini, MD; David Frim, MD, PhD (Chicago, IL)

INTRODUCTION Patients suffering from pseudotumor cerebri often require multiple shunting procedures and frequently fail ventricular shunting due to slit ventricles and lumbar shunting due to obesity. We have found that cisterna magna to pleural shunting, with a programmable valve, in these patients can be a solution.

METHODS 5 patients, 1 male and 4 female, presented after failing either lumbar shunting (n=4) or lumbar and ventricular shunting (n=1) and were treated by cisterna magna to pleural shunting. Patients ranged from 20-42 years in age and follow-up was from 3 months to 3 years. A Radionics TeleSensor and Codman-Medos programmable valve with anti-siphoning component were implanted in each patient.

RESULTS 4 patients continue to have their shunts in place; 1 shunt was removed due to infection and 1 cisternal catheter was revised due to occlusion. 1 pleural catheter was moved to the cardiac atrium due to pleural malabsorption. All patients have avoided optic nerve decompression and remain with stable disease. 2 cisternal catheters required repositioning: 1 for partial facial nerve palsy and 1 for trigeminal neuralgia. Telemetric ICP monitoring revealed postural CSF dynamics identical to those seen in ventriculopleural shunting.

CONCLUSIONS Cisterna magna to pleural shunting is a viable option in pseudotumor cerebri patients when other choices for cerebrospinal fluid diversion have been exhausted.

84. Management of Arachnoid Cysts in Children with Refractory Epilepsy

James E. Baumgartner, MD; James W. Wheless, MD (Houston, TX)

INTRODUCTION Although an association between arachnoid cysts and epilepsy is well known, the role of arachnoid cysts in children with refractory epilepsy is poorly described. This study reports phase 1 and phase 2 experiences with six children with refractory epilepsy and arachnoid cysts.

METHODS From 1993 to 2001, six children with arachnoid cysts and refractory epilepsy presented to the Texas Comprehensive Epilepsy Program. All patients underwent Phase 1 evaluation including long term video EEG monitoring, neuropsychology testing, and neuroimaging (CT, MRI, SPECT and/or MEG). One patient underwent monitoring with intracranial electrodes. Patients subsequently underwent treatment consisting of cyst fenestration or cyst-to-peritoneal shunt placement. All patients had post operative follow-up of at least one year.

RESULTS One patient had a left frontal convexity arachnoid cyst, the remaining patients had right choroidal fissure arachnoid cysts. All patients had frequent complex partial seizures in spite of adequate antiepileptic drug regimens. One patient experienced frequent secondary generalization of his seizures. Phase 1 EEG data suggested a seizure focus near the arachnoid cyst. Invasive monitoring, in the patient with the convexity arachnoid cyst, confirmed the cyst as the focus of epileptogenic activity. In one case cyst fenestration was transiently effective in controlling seizure activity, but the cyst recurred, and the cyst was shunted. The remaining patients all underwent cyst-to-peritoneal shunt placement. Four patients are seizure free off all medications, one patient is seizure free on one AED, and the final patient has had a > 90% reduction in seizures since surgery on a 2 AED regimen.

CONCLUSIONS Arachnoid cysts can cause refractory epilepsy. If phase 1 evaluation suggests a seizure focus in the region of an arachnoid cyst, invasive monitoring is unnecessary. Surgical treatment can be curative, and shunt placement appears to be more effective than cyst fenestration.

85. Algorithm for the Evaluation of the Pediatric Subdural Hematoma

Carrie Antoline, MD (Beltsville, MD); Robert F. Keating, MD (Washington, DC)

INTRODUCTION Current standard of care necessitates that a child with presentation of a subdural hematoma (SDH) be evaluated for history of non-accidental trauma (NAT). There are cases which have an immediately apparent cause, such as overshunting within the setting of ventriculo-peritoneal shunting, obviating a child protective investigation. Other cases need some evaluation but perhaps with development of an algorithm, full investigation can spare both medical resources and parental duress. We propose such an algorithm.

METHODS We compiled a representative case series from retrospective chart and imaging review of subdural hematomas presenting in children. These include most demonstrably subdural hematomas in the setting of non-accidental trauma, overshunting, a history of benign extra-axial collections of infancy and macrocephaly. Within our institution, the mere presence of the SDH prompted a thorough investigation for NAT, which was frequently negative but was continued even in the presence of 1. other causative factors and 2. early negative indices.

RESULTS We present a proposed algorithm based on clinical setting, neurosurgical factors and early investigative findings. The ophthalmologic finding of retinal hemorrhages is classified as either peripheral or central. This is a newly documented edification able to differentiate retinal hemorrhage secondary to either a central increased intracranial pressure issue or to a peripheral, "shaken baby" etiology. This may assist in the correlation of SDH due to NAT versus, for example, overshunting. The algorithm also classifies etiology including a history of excessive extra-axial space.

CONCLUSIONS Given recent advances in the delineation of retinal hemorrhages as well as the compilation of other criteria, we can classify subdural hematomas as more likely traumatic versus non. We propose an initial algorithm to facilitate the evaluation of SDHs, perhaps alleviating some of the burden on both resources and families that is created by redundant investigation.

86. Declined to Present

87. Declined to Present

Poster Index

101. Trilateral Retinoblastoma?

Francesco T. Mangano, DO; Peter M. Farmer, MD (Manhasset, NY); Steven J. Schneider, MD; Mark A. Mittler, MD (New Hyde Park, NY)

102. Real-Time Rendered 3D Perspective Volumetric Image Guidance, Pre-Planned With Holographic Co-Navigation, Guides Transventricular Excision of a Dominant Temporal Horn Meningioma Without Disruption of Overlying Primary Speech Cortical Function

John Collins, MD (Morgantown, WV); Steven Hart, BSc (Provo, UT)

103. Post-Operative Swallowing Function After Posterior-Fossa Craniotomy in Pediatric Patients

Frederick A. Boop, MD; Lisa A. Newman, DSc; Jerome W. Thompson, MD, MBA; Carrie K. Temple, MA (Memphis, TN)

104. Surfactant Poloxamer Protects Against Hemorrhage-Associated Neuronal Loss in the Rat Brain

Sandra B. Cadichon, MD; Hoang M. Le, MD; David Wright, PhD; Un Kang, MD; David Frim, MD, PhD (Chicago, IL)

105. Subependymal Hemangioblastomas of the Brain Stem: Lessons Learned in the Management of 5 Cases

John S. Winestone, BS (Memphis TN); Julian Lin, MD (Peoria, LA); Robert A. Sanford, MD (Memphis, TN)

106. Subfascial Implantation of a Vagal Nerve Stimulator through a Single Cervical Incision.

C. Corbett Wilkinson, MD; John J. Collins, MD (Morgantown, WV)

107. Posterior Fossa Tumors: Importance of Surgical Resection

Julian J. Lin, MD (Memphis, TN); Leanna M. Lugo (Aquadila, Puerto Rico); Robert A. Sanford, MD (Memphis, TN)

108. Internal Graft Closure Technique for Intradural Spinal Surgery to Prevent Re-Tethering

Karl F. Kothbauer, MD; George I. Jallo, MD; Fred J. Epstein, MD (New York, NY)

109. Folate Receptor Expression in CNS Injury and Regeneration

Aaron Nelson, BS; Bermans J. Iskandar, MD (Madison, WI)

110. Assessment of Successful Endoscopic Third Ventriculostomy with Third Ventricular Tumors

Yu-Hung Kuo, MD, PhD; Mark M. Souweidane, MD (New York, NY)

111. The Technique of Cyst to Ventricle Stenting for Treatment of Non-neoplastic Intracranial Cyst

Kevin M. Jackson, MD; Joel C. Boaz, MD (Indianapolis, IN)

112. Intrathecal Catheter Placement for Continuous ITB Therapy in Children with Prior Spinal Fusion

Jason E. Tullis, MD; John A. Lancon, MD (Jackson, MS)

113. Baclofen Pump Placement Through the Foramen Magnum

Kristine Dziurzynski, MD; Bermans J. Iskandar, MD (Madison, WI)

114. Aggressive Psammomatoid Ossifying Fibroma: Combined Craniofacial and Endoscopic Endonasal Resection

Adair Blackledge, MD; Amanda L. Ellis, BA, CNOR, RN; Mark A. Reed, MD; John A. Lancon, MD (Jackson, MS)

115. Kyphotic Thoracic Cord Injury as a Complication of Cervical Surgery in Mucopolysaccharidosis

Ian D. Kamaly-Asl, FRCS (Manchester, United Kingdom); George Malcolm, FRCS (Bristol); John A. Thorne, FRCS (Manchester, United Kingdom)

116. Rapid Correction of Coagulopathy with Recombinant Factor VIIa

John D. Morenski, MD, Joseph D. Tobias, MD (Columbia, MO); David F. Jimenez, MD

117. The Role of PCV Chemotherapy in the Treatment of Central Neurocytoma

Cornelia S. von Koch, MD, PhD (San Francisco, CA); Meic H. Schmidt, MD (Salt Lake City, UT); Jane H. Uyehara-Lock, MD; Mitchel S. Berger, MD; Susan. Chang, MD (San Francisco, CA)

118. Diagnostic Challenges of Primitive Neuroectodermal Tumors: Extraskelatal Peripheral Ewing's Sarcoma

Francesco T. Mangano, DO; Peter M. Farmer, MD; Albert E. Stanek, MD (Manhasset, NY); Steven J. Schneider, MD (New Hyde Park, NY); Mark A. Mittler, MD (New York, NY)

119. Use of an Intravenous Pump to Provide Controlled Drainage for External Ventricular Drains

Michael S. Turner, MD (Indianapolis, IN)

120. Current Management Strategy with Embolization, Resection, and Reconstruction for Aneurysmal Bone Cysts of the Pediatric Spine

L. Madison Michael II, MD; Frederick A. Boop, MD; Kevin T. Foley, MD; Robert A. Sanford, MD (Memphis, TN)

121. Management Strategy for Failed Chiari Type I Decompression

David C. Adamson, MD, PhD; Timothy M. George, MD (Durham, NC)

122. Loss of Vision in MPS VI as a Consequence of Increased Intracranial Pressure and Hydrocephalus

James T. Goodrich, MD (Bronx, NY); P. Harmatz, MD (Oakland, CA); Paul A. Levy, MD; Robert W. Marion, MD (Bronx, NY)

123. Mutism After Posterior Fossa Tumor Resection in Children: Incomplete Recovery on Longterm Follow-up

Paul Steinbok, FRCS(C), MBBS; David D. Cochrane, MD, FRCS(C); Richard Perrin, MD; Angela Price, MD, FRCS(C) (Vancouver, BC, Canada)

124. CSF Pulsatility: Patho-Hydraulics—Clinical Use

Eldon L. Foltz, MD, Prof (Orange, CA)

125. Closure Technique for Intradural Spinal Surgery to Prevent Re-Tethering

Karl F. Kothbauer, MD; George I. Jallo, MD; Fred J. Epstein, MD (New York, NY)

126. Endoscopic Corpus Callosotomy: a Feasibility Study

Matthew D. Smyth, MD; R. Shane Tubbs, PhD, MS, PA-C; George Salter, PhD; Kyle Doughty, MD; Jeff P. Blount, MD (Birmingham, AL)

127. Aplasia Cutis Congenita: Three Cases in Eighteen Months

David M. Wrubel, MD; George T. Burson, MD (Little Rock, AR)

128. A Pitfall in Radiographic Assessment of Progression of Incompletely Resected Low Grade Astrocytomas

Michael H. Handler, MD; Nicholas Foreman, MD; K. Wendorf, MD; Laura Fenton, MD (Denver, CO)

129. Immediate Post-Operative Correction in Head Shape Following Calvarial Vault Remodeling for Sagittal Synostosis

Susan Durham, MD (Portland, OR); J.G. McComb, MD; Michael L. Levy, MD, PhD (Los Angeles, CA)

130. Al-Anazi Ventriculo-Uterine Shunt

Abdul Rahman AL-Anazi, MD (Al Khobar, Saudi Arabia)

131. The Effect of Change in Technique on Complication Rate for Baclofen Pump Procedures

Michael S. Turner, MD (Indianapolis, IN)

132. A Single-Pass Tunneling Technique for CSF Shunting Procedures

Michael A. Sandquist, MD; Nathan R. Selden, MD, PhD (Portland, OR)

133. Diagnosis of Two Brain Tumors In-Utero: Case Reports

Marcus L. Ware, MD, PhD; Errol R. Kolen, MD; Andrew Horvaic, MD; Victor Perry, MD; Nalin Gupta, MD, PhD (San Francisco, CA)

134. Internal Carotid Artery Dissection in an Infant: Case Report and Literature Review

Rebekah C. Austin, MD; Steven S. Glazier, MD (Winston-Salem, NC)

135. Hydrocephalus and Chronic Epidural Hematoma

Mansour Parvaresh-Rizi, MD (Esfahan, Iran); Hossein Ghalea-Novei, MD; Farzaneh Chitsaz, MD (Tehran, Iran); Mohsen Dalvandi, MD (Arak, Iran)

136. Lumboperitoneal Shunting Preferentially Drains the Subarachnoid Space

Leila Khorasani, BS; Christian W. Sikorski, MD; David M. Frim, MD, PhD (Chicago, IL)

137. Meningococcal Meningitis in a Shunted Patient Presenting with Paraplegia

David H. Harter, MD; Michael Gewitz, MD; Bruce Roseman, MD; Chiedozie Nwagwu, MD (Valhalla, NY)

138. Pilocytic Astrocytomas in Two Siblings

David M. Wrubel, MD; Georget T. Burson, MD (Little Rock, AR)

139. Chiari II Malformation and Foramen of Magendie Choroid Plexus Papilloma: A Case Association?

Todd A. Maugans, MD (Burlington, VT)

140. Multicentric Primitive Neuroectodermal Tumor: Evaluation, Diagnosis and Management

Victor L. Perry, MD; Marcus L. Ware, MD, PhD; Anuradha Banerjee, MD; Nalin Gupta, MD, PhD (San Francisco, CA)

141. Cervical Meningocele and Associated Spinal Abnormalities, Case Report and Review of the Literature

Carlos H. Feltes, MD; Vassilios Dimopoulos, MD; Rostislav Davydov, MD; Angel Boev, BS; Debbie Melone, RN; Christopher Troup, MD (Macon, GA)

142. Management of Herniated Disc of Lumbar Spine in Teenagers

S.M. Rezaian, MD, PhD, FICS, FRCS, (Beverly Hills, CA)

143. Pediatric High Grade Astroblastoma: A Case Report and Review of the Literature

Francesco T. Mangano, DO; Mark A. Mittler, MD; Elsa Valderrama, MD; Steven J. Schneider, MD (New Hyde Park, NY)

144. Surgical Treatment of Tethered Cord Syndrome

Yasuko Kusaka, MD, PhD; Reizo Shirane, MD, PhD; Takashi Yoshimoto, MD, PhD (Sendai City, Japan)

145. Surgical Treatment of Late Infantile Moyamoya Disease

Yasuko Kusaka, MD, PhD; Reizo Shirane, MD, PhD; Takashi Yoshimoto, MD, PhD (Sendai City, Japan)

145. Endoscopic Ventriculostomy in Management of Hydrocephalus in Paediatric and Adolescent Patients

Francis H. Tomlinson, MD, PhD, FRACS (Brisbane, Queensland, Australia); Terrence J. Coyne, FRACS (Herston, Queensland, Australia); Michael J. Bryant, MBBS

101. Trilateral Retinoblastoma?

Francesco T. Mangano, DO; Peter M. Farmer, MD (Manhasset, NY); Steven J. Schneider, MD; Mark A. Mittler, MD (New Hyde Park, NY)

INTRODUCTION We report a 16-month-old male who presented to a pediatric ophthalmologist with right eye twitching. Physical examination revealed leukocoria and lesions in the optic globes that were consistent with bilateral retinoblastoma. Screening magnetic resonance imaging revealed a 2cm enhancing suprasellar mass. After cryosurgery for one of the optic tumors, the patient underwent a pterional craniotomy. The tumor was found to be invasive of the pituitary stalk and a subtotal resection was obtained. With the exception of transient diabetes insipidus, his neurologic and endocrinologic status remained normal. Histologically the tumor consisted of small cells with neuronal differentiation and focal calcifications. Immunological stains for keratins, neurofilament, vimentin, glial fibrillary protein, CD99, and PLAP were all negative, while synaptophysin and S-100 protein were positive. Cytogenetic analysis of the tissue revealed a normal karyotype. A deletion of the chromosome band 13q14 was not detected using fluorescence in-situ hybridization. By histologic and cytogenetic criteria the tumor was not a retinoblastoma. Since the original description of the trilateral retinoblastoma in 1977, differing theories have been postulated as to how the midline neuroblastic intracranial tumor arises. Those have included metastasis, ectopic nests of retinal cells, direct spread through optic nerve invasion, and the two "hit" genetic mutation theory. Our data would suggest that this intracranial tumor associated with bilateral retinoblastoma is a central primitive neuroectodermal tumor with evidence of neuronal differentiation, and no evidence of the classic mutation associated with retinoblastoma. This lends favor to the theory that it may arise from the germinal layer of primitive cells (the subependymal plate).

102. Real-time Rendered 3D Perspective Volumetric Image Guidance, Pre-planned with Holographic Co-navigation, Guides Transventricular Excision of a Dominant Temporal Horn Meningioma Without Disruption of Overlying Primary Speech Cortical Function.

John J. Collins, MD (Morgantown, WV); Steven Hart, BSc (Provo, UT)

INTRODUCTION A right-handed boy presented with left temporal horn tumor extending directly beneath primary speech cortex. To avoid speech deficit, volumetric image guidance was applied within a digital hologram to plan and successfully carry out entirely trans-ventricular excision through entry at the temporal tip.

METHODS With fiducials, the patient underwent gadolinium brain MRI, from which a digital hologram (Voxgram by Voxel, Inc) was generated. This is a translucent, life-size, "sculpture out of light" in real space. The same data was imported to a new volumetric image guidance system (SAVANT by CBYON, Inc.). This system presents the 3D volumetric perspective appearance of ventricle, tumor and feeding vessels continuously, in real-time during navigation, from the probes-eye vantage point. The navigation system and hologram were co-registered using fiducials. The hologram was then explored with the navigation probe to until a linear trajectory was found that entered the ventricle through the temporal tip and extended directly to the feeding vessels of the tumor. That trajectory was saved as the operative approach plan. Surgery recapitulated the pre-determined trajectory by means of a right temporal craniotomy with orbitozygomatic osteotomy. Volumetric images guided microsurgery through the ventricle directly to the tumor feeding vessels. These were coagulated and cut first. Tumor was then resected.

RESULTS The result was complete trans-ventricular tumor removal with minimal bleeding and no post-operative speech deficit. Pathology revealed meningioma.

CONCLUSIONS This case demonstrates how volumetric image guidance within holographic image space can "flight simulate" novel surgical approaches that may provide innovative surgical solutions to challenging clinical situations.

103. Post-operative Swallowing Function After Posterior-Fossa Craniotomy in Pediatric Patients

Frederick A. Boop, MD; Lisa A. Newman, DSc; Jerome W. Thompson, MD, MBA; Carrie K. Temple, MA (Memphis, TN)

INTRODUCTION Swallowing difficulties are prevalent post-operatively following posterior-fossa craniotomies impacting when children can safely begin eating. The purpose of this retrospective study was to quantify post-operative swallowing function in these children.

METHODS 24 children (10 females, 14 males, mean age 5.5 years, range= 4 months-17.8 years) were seen for video-fluoroscopic evaluations of swallowing (VFS) when the neurosurgeon determined the child was stable at a mean of 6 days post-op (range=2-52 days). The VFS was analyzed for the following variables: oral and pharyngeal transit anomalies, laryngeal penetration, aspiration, naso-pharyngeal backflow, and pharyngeal residue.

RESULTS 20 (87%) out of 23 children who took barium orally had oral transit anomalies, 2 so severe they were unable to move material into the pharynx. Of the remaining 21, 10 (47%) had pharyngeal transit anomalies. Six children (32%) experienced laryngeal penetration and 4 (18%) aspirated. Only 1 of 4 children coughed and none cleared their airway. Children with brainstem involvement had significantly greater aspiration (p=.05). 7 (29%) had material enter the nasopharynx. Children who aspirated were referred for VFS significantly later (mean days=10.7) than those who did not (mean days=4.9) (t=-2.84, p=.01).

CONCLUSIONS Most children demonstrated swallowing anomalies following posterior-fossa craniotomies. However, 46% did not experience pharyngeal transit deficits and 75% were able to swallow safely within

the first week of surgery. Children with brainstem involvement had worse aspiration. Children who aspirated had a more difficult post-operative course and were referred for swallowing evaluations 1-2 weeks after surgery with greater swallowing disorders.

104. Surfactant Poloxamer Protects Against Hemorrhage-Associated Neuronal Loss in the Rat Brain

Sandra B. Cadichon, MD; Hoang M. Le, MD; David Wright, PhD; Un Kang, MD; David Frim, MD, PhD (Chicago, IL)

INTRODUCTION Neuronal injury remains a leading cause of morbidity in both neonates and adults with injuries induced from intracranial hemorrhage, ischemia-reperfusion, and excitotoxic cell damage. A number of therapies have been attempted with little if any benefit. Poloxamer 188 (P-188), a triblock copolymer, has been studied extensively as a cell membrane sealant (Lee, et al. 2000). Recently, we have demonstrated its ability to protect neurons from excitotoxic injury induced via glutamate excitation. In this study, we use an animal model to study the neuroprotectant effects of P-188 after iatrogenic induction of intraparenchymal hemorrhage.

METHODS Sprague-Dawley rats were divided into 4 groups: Group 1, n=7, and Group 3, n=6: "treated" received IC injection of P-188 10 min after striatal infusion of 50 microL of autologous blood. Group 2, n=8, and group 4, n=7: "control" received IC injection of vehicle 10 min after striatal infusion of 50 microL of autologous blood. Group 1 and 2 animals were sacrificed after 2 days; group 3 and 4 animals were sacrificed after 7 days. Striatal tissues were histologically analyzed for neuronal loss by NeuN staining and lesions were volumetrically calculated. In a separate experiment, 18 rats were divided between daily injections of vehicle or poloxamer for 5 days after blood infusion; rats were sacrificed after 7 days.

RESULTS The volume of the lesions was significantly (p=0.04) larger in the control group (31.4+/-4.3, mean +/- SEM), than the P-188 group (18.3+/-4.3) for groups 1 and 2, sacrificed at 2 days. For groups 3 and 4, no statistical difference was observed. There

was a trend toward protection in the animals where the P-188 was injected daily for 5 days before sacrifice on day 7 after hemorrhage.

CONCLUSIONS A single P-188 dose protects against early neuronal loss after hemorrhage but has no effect on long-term hemorrhage induced neuronal injury or death. However, preliminary data suggests that repeated daily injections of P-188 intracisternally for several days after hemorrhage may be a more efficacious protective paradigm.

105. Subependymal Hemangioblastomas of the Brain Stem: Lessons Learned in the Management of 5 Cases

John Winestone, BS; Julian Lin, MD (Peoria, LA); Robert A. Sanford, MD (Memphis, TN)

INTRODUCTION Hemangioblastomas are an uncommon tumor of the CNS, with an incidence of von Hippel Lindau (VHL) being 1/43,000 births and a spontaneous mutation rate of 1/780,000. These lesions are known for their vascularity, making resection a surgical challenge. Hemangioblastomas arising within the brain stem presented with headaches, intractable vomiting, proprioceptive abnormalities and gait instability. In the past, surgical mortality for these lesions was 30-50%. Surgical resection may be complicated by vascular instability associated with manipulation of the medullary vasomotor centers. Postoperative complications may include central hypoventilation, sleep apnea (Ondine's curse), permanent neurogenic hypertension, dysphagia, and aspiration.

METHODS The authors present a series of 5 patients with hemangioblastomas of the medulla/cervicomedullary junction. Four presented with headaches, vomiting and gait ataxia. One was detected by routine screening for VHL.

RESULTS Gross total resection was accomplished without mortality. Two patients have maintained an ataxic gait. Complications include transient central hypoventilation (2), aspiration pneumonia (1) and permanent neurogenic hypertension (2).

CONCLUSIONS It is critical that the vascular nidus be recognized on neuroimaging so that resection proceeds without biopsy and catastrophic hemorrhage be avoided. The neuroimaging characteristics of these tumors will be presented. A postoperative management paradigm assessing respiratory drive, vocal cord function and pharyngeal motility will be suggested as a means of complication avoidance. Finally, the extent of evaluation necessary in the child who presents with an isolated hemangioblastoma and no family history of VHL will be discussed.

106. Subfascial Implantation of a Vagal Nerve Stimulator through a Single Cervical Incision.

C. Corbett Wilkinson, MD; John J. Collins, MD (Morgantown, WV)

INTRODUCTION Vagal nerve stimulators are generally implanted via two incisions, one cervical and one thoracic, with the pulse generator placed subcutaneously in the chest. This technique can lead to two problems: 1) wound breakdown in cognitively-delayed children who either pick at their thoracic incision or disrupt it with abnormal arm movements, and 2) undesirable cosmetic appearance due to two separate incisions and, especially noticeable in children, bulging skin over the subcutaneous pulse generator. We describe a technique to lessen rates of wound breakdown and improve cosmetic appearance. In this technique, the entire stimulator is implanted through a single cervical incision with the pulse generator placed beneath the pectoral fascia.

METHODS This is a retrospective review of 16 consecutive vagal nerve stimulators implanted by the senior author using our technique. Briefly, a transverse cervical incision is followed by subplatysmal dissection to the clavicle. The pectoral fascia is then separated from the clavicle and the pulse generator implanted between the fascia and pectoralis major. Vagal nerve electrodes are applied using standard techniques.

RESULTS Of 16 implantations, 14 were performed in cognitively-delayed children. Postsurgery, there was one superficial incisional cellulitis that resolved with antibiotics. There were no wound breakdowns. One incision was revised to remove a noticeable electrode-wire strain-relief bracket. Cosmetic appearance of the subfascially-placed pulse generators was excellent.

CONCLUSIONS Implanting vagal nerve stimulators through a single cervical incision with the pulse generator beneath the pectoral fascia may lower rates of wound breakdown and improve cosmetic appearance, especially in children.

107. Posterior Fossa Tumors: Importance of Surgical Resection

Julian J. Lin, MD (Memphis, TN); Leanna M. Lugo (Aquadria, Puerto Rico); Robert A. Sanford, MD (Memphis, TN)

INTRODUCTION The importance of resection in pediatric posterior fossa tumors, astrocytomas, ependymomas, and medulloblastomas is well documented. However, it is unclear in what percentage of cases in a nonselected series is it possible to obtain a gross total or near total resection of posterior fossa tumors.

METHODS A retrospective review of the senior author's personal series of posterior fossa tumors from 1985 to 2001 revealed 234 children (2 months–20 years). The pathology was medulloblastomas 125, low grade gliomas 57, ependymomas 35, anaplastic ependymomas 10, and high grade gliomas 7.

RESULTS Independent neuroimaging review by neuroradiologists at 2 institutions and pediatric neurosurgery documented gross total resection (no tumor) in 192 cases (82%), near total resection <1.5 cm in 19 cases (8%), and subtotal resection >1.5 cm in 23 cases (10%); 14 of whom had metastatic disease. The largest residual was 2 cases <2.5 cm. Tumor pathology did not impact on the ability of the surgeon to achieve a gross total resection except in the cases of preoperative metastatic disease 12/19 cases in which the goal of surgery was to open the CSF pathways. Of the 192 gross total resections, 36 patients (18.75%)

were dead at the time of review in comparison to 14 deaths (60.8%) of 23 patients with subtotal resection.

CONCLUSIONS Gross total resection/near total resection is possible in 90% of pediatric posterior fossa tumors and strongly correlates with surgical.

108. Internal Graft Closure Technique for Intradural Spinal Surgery to Prevent Re-Tethering

Karl F. Kothbauer, MD; George I. Jallo, MD; Fred J. Epstein, MD (New York, NY)

INTRODUCTION Retethering is a common occurrence after surgery for spinal dysraphism, or intradural spinal surgery. The pia of the cord becomes densely adherent to the inner surface of the dural sac, particularly at a suture line.

METHODS A simple closure technique using a synthetic dural substitute is presented. The intent is to prevent scarring, retethering and its associated morbidity. A DuraGuard synthetic dural graft is placed over the spinal cord, or the cauda equine to cover the entire dural opening. The graft is tailored so that it covers the intradural structures beyond the suture line. The dura is then closed over the graft. The graft is secured to the dura using a few tangential stitches, enough to keep it from migrating craniocaudally, or compressing the intradural structures. If the dura is not intact for complete closure over the DuraGuard, as is often the case in tethered cord surgery, a local fascial graft is used in an insert-fashion.

RESULTS Over a period of the past 2 years, the technique was used in 15 patients. Two had spinal cord lipomas, 4 intramedullary spinal cord tumors, and 9 had tethered cord surgery. There were no technical problems at surgery, and no postoperative morbidity that would be attributable to the graft or the closure technique. There was one CSF leak in a patient with recurrent tethered cord syndrome, where a second operation was necessary.

CONCLUSIONS The proposed closure technique is relatively simple and appears at least as safe as closure using a direct dural-insert graft. The follow-up interval is too short to evaluate whether this indeed prevents anatomic and/or symptomatic tethering or re-tethering.

109. Folate Receptor Expression in CNS Injury and Regeneration

Aaron Nelson, BS; Bermans J. Iskandar, MD (Madison, WI)

INTRODUCTION We have previously shown that folic acid supplementation significantly improves CNS regeneration after injury, which suggests that the folate pathway is altered at the time of injury to support neuronal growth and repair. Folic acid is taken up by neurons through 2 types of receptors: the reduced folate carrier 1 (RFC-1) and folate binding protein á (FBP- á). Since folic acid is intimately dependent on receptor availability to function, we studied the activity of the RFC-1 and FBP- á with injury.

METHODS Sharp transection injuries were made in the dorsal columns and sciatic nerve of adult rats. Using *in situ* hybridization, we studied the expression of the RFC-1 and FBP- á genes in the dorsal root ganglia (DRG), in which the cell bodies of both the dorsal column and sciatic nerve axons are located. This was accomplished using RFC-1 and FBP- á specific oligonucleotide probes, which we labeled using an *in vitro* transcription method.

RESULTS While RFC-1 mRNA levels were no different from uninjured controls, there was a significant increase in FBP- á mRNA in the injured neurons. A variety of negative and positive controls confirmed the specificity of the results.

CONCLUSIONS The FBP- á receptor is highly unregulated with neuronal injury. We propose that this high affinity folate receptor plays an important role in the regenerative response of neurons to folic acid supplementation.

110. Assessment of Successful Endoscopic Third Ventriculostomy with Third Ventricular Tumors

Yu-Hung Kuo, MD, PhD; Mark M. Souweidane, MD (New York, NY)

INTRODUCTION A turbulent flow pattern within the third ventricle on standard MRI sequences is typically used as an indicator of a patent fenestration following endoscopic third ventriculostomy (ETV). In patients with third ventricular tumors, this turbulent flow pattern may be difficult to distinguish from tumor mass and result in misinterpretation of the study.

METHODS From January 2001–July 2002, 35 patients underwent ETV at our institution, 8 of whom had a third ventricular tumor. MRI including long TR and FLAIR sequences were performed. Preoperative and postoperative MRI scans were reviewed in an effort to define those features which best demonstrate a functional fenestration.

RESULTS All postoperative MRI's revealed changes consistent with a patent third ventriculostomy. Comparison of the preoperative and postoperative MRI's revealed a new turbulent flow pattern in both the third ventricle and the prepontine cistern. However, the intraventricular flow pattern was sometimes difficult to distinguish from tumor mass whereas the prepontine flow void was unambiguous in patients with successful ETV. Cine MRI studies further supplemented the radiographic assessment of the fenestration in two patients.

CONCLUSIONS MRI assessment of ETV in patients with third ventricular tumor can be difficult given the heterogeneous MRI characteristics of the mass. Comparison of the postoperative MRI with preoperative studies can assist in distinguishing flow artifact from tumor mass. A turbulent flow pattern in the prepontine cistern on long T2 or FLAIR sequences is unambiguous for determination of patency. This finding is not dependent upon comparison with the preoperative MRI. Finally, cine MRI can complement standard sequences.

111. The Technique of Cyst to Ventricle Stenting for Treatment of Non-neoplastic Intracranial Cyst

Kevin M. Jackson, MD; Joel C. Boaz, MD (Indianapolis, IN)

INTRODUCTION Arachnoid and ependymal cysts are benign, extra axial, mass-occupying lesions that can present with sign or symptoms of increased intracranial pressure, seizures, or focal neurological deficits. Treatment options are controversial. Long-term success rates for cyst wall fenestration is only 73%. Cyst shunting carries the risk of over drainage. This paper will describe our technique of cyst to ventricle stenting.

METHODS Placement of the cyst puncture catheter was performed using the CRW stereotactic frame. Coordinates were obtained from within the cyst and also on the skull that would give a trajectory to traverse the ventricle. The catheter was fenestrated in a manner that left holes in the cyst and the ventricle. An intracatheter was used to selectively aspirate and decompress the cyst. A Salmon-Rickham reservoir was connected to the end of the catheter.

RESULTS Two children were evaluated for headaches, ventriculomegaly, and intracranial cyst: a two-year-old with a pineal region cyst, and an eight-year-old with a cyst within the right lateral ventricle. Both patients underwent cyst to ventricle stenting, as described above. Ventricular size decreased postoperatively. They remain symptom free two years post-op.

CONCLUSIONS Cyst to ventricle stenting is effective for the treatment of non-neoplastic intracranial cyst. The technique involves stereotatic placement of a catheter into the cyst with holes also in the ventricle. A Salmon-Rickham reservoir facilitates future percutaneous interrogation of the system, or conversion into a conventional cyst-peritoneal shunt, if necessary. The complications associated with open fenestration and permanent shunts are avoided. Good results obtained with this procedure; follow up > two years.

112. Intrathecal Catheter Placement for Continuous ITB Therapy in Children with Prior Spinal Fusion

Jason E. Tullis, MD; John A. Lancon, MD (Jackson, MS)

INTRODUCTION The placement of an intrathecal catheter for continuous ITB therapy in a child with spasticity who has previously undergone spinal fusion presents several technical challenges that predispose the child to postoperative morbidity including infection and cerebrospinal fluid leak. In the presence of a prior spinal fusion, the steps involved in placement of the intrathecal catheter must be modified to eliminate the resultant dead space that develops around the intrathecal catheter.

METHODS A midline incision is carried down to the lumbodorsal fascia and one side is undermined in the suprafascial plane. The dissection proceeds to the dorsal surface of the spinal fusion mass. An oval osteotomy is drilled down to the dural surface. Fluoroscopic guidance may be useful in the absence of identifiable osseous landmarks. Reapproximation sutures are placed in interrupted fashion and tagged but not secured. The spinal access needle is directed via a paramedian approach from the lumbodorsal fascia to the entrance of the osteotomy. The access needle is advanced into the lumbar cistern and the intrathecal catheter is advanced into position. A 4-0 braided nylon encircling stitch is placed around the access needle in the lumbodorsal fascia. The access needle and guide wire are withdrawn and fibrin sealant is instilled into the osteotomy while awaiting the backflow of cerebrospinal fluid through the intrathecal catheter. The reapproximation and encircling sutures are then secured.

RESULTS The results with respect to placement of an intrathecal catheter for continuous ITB therapy in children with spasticity and dystonia who have undergone prior spinal fusion are presented. The surgical steps are clarified using intraoperative photographs and line drawings.

CONCLUSIONS Single technical modifications in the placement of the intrathecal catheter for continuous ITB therapy in

children with prior spinal fusion are effective in preventing complications associated with the greater amount of tissue dissection necessary in these cases.

113. Baclofen Pump Placement through the Foramen Magnum

Kristine Dziurzynski, MD; Bermans J. Iskandar, MD (Madison, WI)

INTRODUCTION The baclofen pump has been utilized in children with refractory spasticity. However, in children with prior lumbar fusion, such device implantation is difficult and fraught with complications. As an alternative to placing the pump catheter through the lumbar spine, we report our experience with placement of the catheter in the spinal canal via the foramen magnum.

METHODS We have followed three patients with prior lumbar fusion and refractory spasticity, who also had VP shunts. Each of these patients has undergone intrathecal placement of a baclofen pump through the foramen magnum. With the patient in the lateral position, the procedure was carried out through a vertical incision extending from the occiput to C2, followed by a C1 laminectomy to access the epidural space. Once the foramen magnum was identified, a small incision was created in the dura and arachnoid to insert the catheter. Appropriate positioning of the catheter tip at the T6 spinal level was determined using intra-operative fluoroscopy. The catheter was then tunneled to the pump in the abdominal wall.

RESULTS Each patient experienced significant decrease in the severity of spasticity with pump placement. One patient required pump removal because of a concomitant VP shunt infection. One patient required revision of the spinal catheter that had pulled out and caused a subcutaneous CSF collection in the setting of a VP shunt malfunction. He and the third patient continue to have appropriate pump functioning at an average followup period of 1 and 2.5 years, respectively.

CONCLUSIONS Placement of the baclofen pump intrathecally via the foramen magnum is a viable alternative for patients with spastic quadriplegia with prior lumbar fusion. The tip of the spinal catheter can be positioned at T6 or elsewhere in the spinal canal similarly to the traditional lumbar pumps. There were minimal adverse sequelae related to pump positioning.

114. Aggressive Psammomatoid Ossifying Fibroma: Combined Craniofacial and Endoscopic Endonasal Resection

Adair Blackledge, MD; Amanda L. Ellis, BA, CNOR, RN; Mark A. Reed, MD; John A. Lancon, MD (Jackson, MS)

INTRODUCTION Aggressive psammomatoid ossifying fibroma (APOF) is a biologically benign neoplasm of the sinonasal region that may be locally aggressive and result in significant craniofacial deformity. Definitive treatment of an APOF requires complete surgical resection.

METHODS We describe an 11-year-old boy who presented with nasal congestion. He was initially thought to have a large nasal polyp and underwent endonasal biopsy at an outside facility. Following histopathological identification of an APOF, the boy was referred to the University of Mississippi Medical Center for further evaluation. At the time of referral, he complained of persistent nasal congestion but denied headache or facial pain. Computed tomography of the craniofacial skeleton showed an expansile mass extending from the posterior right cribriform plate and filling portions of the sphenoid, ethmoid and right maxillary sinuses. The right medial orbital wall was thinned and displaced laterally. Magnetic resonance imaging showed a homogeneous, well-demarcated mass with minimal enhancement. The mass appeared hyperintense to brain parenchyma on T1 and T2 weighted images.

RESULTS A combined craniofacial (bandeau) and endonasal endoscopic approach allowed complete resection of the mass. There was no perioperative morbidity.

CONCLUSIONS The technical steps necessary for complete resection of the tumor using this combined approach are reviewed. Options for reconstruction of the anterior skull base and medial orbital wall are discussed.

115. Kyphotic Thoracic Cord Injury as a Complication of Cervical Surgery in Mucopolysaccharidosis

Ian D. Kamaly-Asl, FRCS (Manchester, United Kingdom); George Malcolm, FRCS (Bristol); John A. Thorne, FRCS (Manchester, United Kingdom)

INTRODUCTION Patients with mucopolysaccharidosis are prone to spinal disease and deformity. Progressive myelopathy secondary to either atlantoaxial instability or cervical canal stenosis is common. There is often varying degrees of associated thoracic or thoracolumbar kyphosis/scoliosis.

METHODS Two case reports presented.

RESULTS The first child was diagnosed with atlantoaxial instability on screening scans. He was neurologically normal on examination. He underwent an atlantoaxial fusion at the age of four years eleven months. The cervical surgery itself was uneventful but the child awoke with only a flicker of movement in his lower limbs. On follow up MR scans there is evidence of change within the cord at the thoracic kyphotic level. The second child presented for surgery age eleven with progressive cervical myelopathy. Examination confirmed a spastic tetraparesis. MR scan pre operatively demonstrated cervical cord compression with an upper thoracic kyphoscoliosis. Following a cervical decompression he awoke with improved upper limb power but a complete paraplegia with mid thoracic sensory level.

CONCLUSIONS These children suffered cord injuries at the level of their thoracic deformities during cervical surgery. The aetiology is presumably a vascular event. There are implications for positioning of patients and per operative management of blood pressure.

116. Rapid Correction of Coagulopathy with Recombinant Factor VIIa

John D. Morenski, MD; Joseph D. Tobias, MD (Columbia, MO); David F. Jimenez, MD

INTRODUCTION Brain injury remains one of the leading causes of death and disability in the pediatric patient. Therapy involves aggressive management of intracerebral pressure (ICP) cerebral perfusion pressure (CPP), which often requires invasive procedures from placement of an intraparenchymal ICP monitor or intraventricular catheters. These potentially life-saving interventions require normal coagulation function; however, several factors may lead to coagulopathy in the head injured patient. The standard therapy which includes often multiple doses of fresh frozen plasma (FFP) has a number of drawbacks in the pediatric patient. FFP requires time to type and cross-match, thaw, and administer. This imposes a significant volume load on a child in whom cerebral edema remains a problem. Success in the hemophilic population with recombinant factor VIIa (rVIIa) suggests an alternative therapy.

METHODS We present three patients who suffered severe coagulopathy after cerebral injury. One received rVIIa after repeat doses of FFP failed to correct their coagulopathies, while the other two received rVIIa as initial therapy. Treatment with rVIIa consisted of a bolus of 90mg/kg.

RESULTS rVIIa rapidly corrected the patients' coagulopathies which allowed placement of intraventricular catheters to monitor and relieve ICP.

CONCLUSIONS Brain injury-induced coagulopathy threatens significant secondary injury through hemorrhage and preventing invasive therapies, and it can be refractory to FFP. FFP takes time to administer, may require repeat doses of significant volume for the pediatric patient, and may ultimately fail. Our preliminary data suggests that rVIIa proves provided a rapid and successful therapy.

117. The Role of PCV Chemotherapy in the Treatment of Central Neurocytoma

Cornelia S. von Koch, MD, PhD (San Francisco, CA); Meic H. Schmidt, MD (Salt Lake City, UT); Jane H. Uyehara-Lock, MD; Mitchel S. Berger, MD; Susan M. Chang, MD (San Francisco, CA)

INTRODUCTION Most central neurocytomas follow a benign clinical course, however, more aggressive variants have been described requiring additional surgical resection, radiation- or chemotherapy. Chemotherapy has rarely been used as an adjuvant therapy for central neurocytomas.

METHODS We report a case of a 17 year old girl, who underwent four subtotal resections, over the course of 3 years, for a large central neurocytoma that continued to progress. To avoid radiation injury in a young patient, she was treated with six cycles of chemotherapy including procarbazine, CCNU and vincristine. Procarbazine was stopped after 2 cycles because of the development of a rash.

RESULTS After 2-3 cycles her tumor started to reduce in size and remained stable for at least 12 months after completion of chemotherapy. Serial magnetic resonance imaging was used to follow treatment response.

CONCLUSIONS To our knowledge, this is only the 4th report describing the use of chemotherapy for progression of central neurocytomas as a treatment alternative to radiation therapy. The use of procarbazine, CCNU and vincristine has not been previously described for the treatment of a central neurocytoma and presents an alternative treatment option.

118. Diagnostic Challenges of Primitive Neuroectodermal Tumors: Extraskelatal Peripheral Ewing's Sarcoma

Francesco T. Mangano, DO; Peter M. Farmer, MD; Albert E. Stanek, MD (Manhasset, NY); Steven J. Schneider, MD (New Hyde Park, NY); Mark A. Mittler, MD (New York, NY)

INTRODUCTION A previously healthy 6 year-old female was referred to the pediatric neurosurgical service with a 5-week history of thoracic back pain. The physical examination revealed thoracic tenderness, decreased strength of the right lower extremity and a spastic asymmetric gait. Emergent magnetic resonance imaging revealed an extra-dural extra-medullary mass extending from T5 to T9 causing significant spinal cord compression. A bilateral osteoplastic laminectomy was performed and a soft pink-white mass was resected. Microscopically the tumor consisted of solid sheets of small round cells with little cytoplasm. Although thought to be a neuroblastoma initially, the diagnosis came into question when immunohistochemical studies for synaptophysin, chromogranin, neuron specific enolase, vimentin and neurofilament were all negative. Conversely, the cells contained cytoplasmic glycogen and showed positivity for CD99, and CD57 immunostaining. Cytogenetic analysis revealed a translocation T (11;22) (q24;12) characteristic of peripheral neuroectodermal tumor/Ewing's sarcoma (PPNET/ES). PPNET/ES were originally described in bone and later in soft tissue, and found to be identical to primitive neuroectodermal tumors. The differential diagnosis for such small round cell tumors includes lymphoma, neuroblastoma, and osteoblastoma. The diagnosis of extraskelatal Ewing's sarcoma has been facilitated by the identification of its characteristic translocation and identification of the MIC2 gene as evidenced by CD99 expression.

119. Use of an Intravenous Pump to Provide Controlled Drainage for External Ventricular Drains

Michael S. Turner, MD (Indianapolis, IN)

INTRODUCTION Management of patients on external ventricular drainage requires extreme care and often ICU admission due to the risk from undetected obstruction, overdrainage, and backflow of CSF into the ventricle. The patient must be kept at bedrest, and the relationship of the patient and the drip chamber of the drainage system must remain constant. The use of a routine intravenous pump to provide controlled drainage of the ventricular system has eliminated many of the risks of external drainage, and allows the care of the patient to be done safely on the routine ward.

METHODS The system currently used will be described and its effectiveness over 2 years will be described.

RESULTS Use of a routine IV pump system has been safe, with no significant adverse effects, and has allowed patients with EVD and lumbar drains to be routinely and safely cared for on a general ward. This allows the child to have his parents with him at all times and significantly reduces the cost of shunt infections.

CONCLUSIONS The drain pump is a safe and effective way to control drainage from external ventricular drains.

120. Current Management Strategy with Embolization, Resection, and Reconstruction for Aneurysmal Bone Cysts of the Pediatric Spine

L. Madison Michael II, MD; Frederick A. Boop, MD; Kevin T. Foley, MD; Robert A. Sanford, MD (Memphis, TN)

INTRODUCTION Aneurysmal bone cysts in the pediatric population are extremely rare, representing only 1% of all primary bone tumors. Of these, only 10%–30% arise in the mobile spine. Although benign, the usual clinical course is that of ultimate expansion and destruction of the spine with compression of neural elements. To date, a majority of the literature consists of only single case reports. The authors report a

series of six patients treated with a comprehensive, multi-disciplinary approach consisting of extensive neuro-imaging, pre-operative embolization, aggressive surgical resection, and spinal stabilization as indicated.

METHODS A retrospective chart review was performed for all six patients. Pre- and post-operative imaging was obtained in all patients in addition to long-term follow-up.

RESULTS The diagnosis of aneurysmal bone cyst was made pre-operatively in all patients secondary to the pathognomonic imaging characteristics. Three lesions involved the cervical spine, one was located in the thoracic spine, and 2 were found in the sacrum. Three patients underwent selective arterial embolization, and all patients were treated with aggressive surgical resection. A gross total resection was ultimately achieved in all patients, with one patient requiring a second procedure for residual tumor. Spinal reconstruction was necessary in two patients.

CONCLUSIONS We feel aggressive surgical resection preceded by pre-operative embolization offers the pediatric patient the best chance for a cure while minimizing morbidity. If spinal instability results from the resection, modern reconstructive techniques enable the surgeon to effectively restore biomechanical stability.

121. Management Strategy for Failed Chiari Type I Decompression

David C. Adamson, MD, PhD; Timothy M. George, MD (Durham, NC)

INTRODUCTION Chiari decompression has proven to be a very successful procedure for Chiari Type I Malformation; however, there is a subset of patients who do not respond for unclear reasons. The goal of our study was to define this "Failed Chiari Type I Syndrome" and offer management strategies.

METHODS We retrospectively examined all pediatric patients at a large academic institution over a five year period who underwent decompression for Chiari Type I Malformation alone, and/or Chiari Type I Malformation with hydrosyringomyelia,

and/or Chiari Type I Malformation with skull base anomalies. We identified and characterized all patients who did not respond and underwent secondary procedures.

RESULTS We found a high success rate for resolution of symptoms after an initial Chiari Type I decompression. However, we also identified a group of patients who had recurrence of preoperative headaches, return of neurologic symptoms (brainstem and/or myelopathy syndromes), or demonstrated a lack of syrinx resolution. Postoperative radiographic studies were performed and contributing factors such as hydrocephalus were not identified. These patients underwent secondary procedures consisting of secondary decompression, spinal fusion, or syrinx shunt.

CONCLUSIONS The goal of this study was to describe the subset of Chiari Type I pediatric patients who do not respond to standard decompression. Possible predictive factors and management strategies for patients with "Failed Chiari Type I Syndrome" will be discussed in detail.

122. Loss of Vision in MPS VI as a Consequence of Increased Intracranial Pressure and Hydrocephalus

James T. Goodrich, MD (Bronx, NY); P. Harmatz, MD (Oakland, CA); Paul A. Levy, MD; Robert W. Marion, MD (Bronx, NY)

INTRODUCTION Caused by a deficiency of N-acetylgalactosamine 4-sulfatase, Maroteaux-Lamy syndrome (MPS VI) is a rare, progressive autosomal recessively inherited disorder. Of the many clinical features associated with MPS VI, the sudden onset of permanent vision loss is one of the most devastating. The cause of this vision loss is not fully understood: theories include the effect of chronically increased intracranial pressure (IICP) and infiltration of the optic nerve with GAGs. We are currently following three patients with MPS VI who are blind. The youngest of these patients, KG, was diagnosed with MPS VI at birth, because of a positive family history. At four years of age, he presented with nystagmus; over the course of the next three weeks,

he lost all vision in both eyes. Examinations by an ophthalmologist and neuro-ophthalmologist failed to identify papilledema or any other significant ocular pathology. A CT scan and MRI showed findings consistent with MPS VI, but no apparent signs of IICP were seen. A lumbar puncture performed under general anesthesia revealed a markedly elevated opening pressure (>5000 mm H₂O). Based on this finding, a ventriculoperitoneal shunt was placed. Following the procedure, KG has regained some vision.

METHODS None.

RESULTS See conclusions.

CONCLUSIONS Our experience strongly suggests that blindness in MPS VI is caused by IICP that may not be detectable using standard exams and imaging studies. Patients with this condition require lumbar puncture for correct diagnosis. We urge that this course of action be taken in any patient with MPS VI showing signs or symptoms of IICP, even when exams and imaging studies reveal "normal" findings.

123. Mutism after Posterior Fossa Tumor Resection in Children: Incomplete Recovery on Longterm Follow-up

Paul Steinbok, FRCS(C), MBBS; David D. Cochrane, MD, FRCS(C); Richard Perrin, MD; Angela Price, MD, FRCS(C) (Vancouver, BC, Canada)

INTRODUCTION Mutism after posterior fossa tumor resection is generally said to be transient. Our experience suggested that speech did not usually normalize, and that mutism was associated with neurologic deficits that did not recover fully.

METHODS Children with complete mutism after posterior fossa tumor resection, and alive more than 2 years post-operatively, were reviewed retrospectively. Charts were reviewed and parents contacted to ascertain details about mutism, associated neurologic deficits, and the most recent speech and neurologic status.

RESULTS There were six children, with follow-up ranging from 2.5–13.1 years (mean 7.4 years). Tumors were midline, with four astrocytomas and two medulloblastomas. Mutism was noted immediately after postoperative extubation in all patients. Speech reappeared 1–14 weeks postoperatively, except for 1 patient, who remained mute at 2.5 years. Speech returned to normal in only one patient. Mutism was always accompanied by new or worsened cerebellar ataxia, which resolved incompletely in the long term. Sixth nerve palsies occurred in 3, and recovered incompletely. Seventh N paresis occurred in 2 and recovered completely.

CONCLUSIONS Mutism after posterior fossa tumor resection is associated with other neurologic deficits, particularly ataxia. Whereas speech usually returns, contrary to general opinion, speech rarely normalizes. Other associated deficits rarely resolve completely.

124. CSF Pulsatility: Patho-Hydraulics—Clinical Use

Eldon L. Foltz, MD, Prof (Orange, CA)

INTRODUCTION Eleven prior publications (Foltz) included 126 hydrocephalus or "suspect" patients with 38 normal controls; 183 analog CSF pulse/ICP records. CSF pulsatility (CSFp) is systole induced and markedly damped by inducing cerebral venous pulsed venting. CSFp average amplitude is ± 25 mm water with peak amplitude at $\pm 80\%$ of intersystole time. Normal inspiration causes increased slope (undamping <ETH> increased Venous venting to thorax); expiratory recovery. Mean ICP decreases with inspiration, -recovers on expiration. Intracranial single compartment volume increase follows Munro-Kellie Doctrine, i.e., pathologic volume increase causes progressive reduction of CSFp damping as the available venous volume decreases. The "water hammer" effect of undamped CSFp results with rising mean ICP leading to brain herniations, etc.

METHODS CSFp was evaluated by superimposing correlative triangles, -facilitating wave slope via peak amplitude and latency measurements, etc. Communicating and aqueduct stenosis hydrocephalus had similar undamped CSFp, the former was slower in developing. Rate of developing such is complicated—not studied.

RESULTS 1. CSFp systole generated, induces "compensatory" cerebral venous pulsatile venting into dural sinuses. Normal inspirations cause modest undamping (wave slope increase, less latency) recovered during expirations. 2. Intracranial lesions of increasing volume cause progressive undamping of CSFp, leading to extreme "Water Hammer" effect on brain due to progressive exhaustion of available cerebral venous volume for venting. Rising mean ICP occurs only after inspiration/expiration CSFp are equally undamped. A warning signal of mean ICP rise and possible crisis 3. All hydrocephalus patients showed CSFp undamping in variable degree, usually with mean ICP elevated, but some prior to mean ICP elevation.

CONCLUSIONS 1. "NPH Suspect" patients need a diagnostic CSFp analysis by L.P. Early undamping CSFp, unaffected by respirations seems diagnostic of "NPH", 2. CSF pulsatility analysis merits wide diagnostic use. It is more sensitive to expanding intracranial lesions than mean ICP.

125. Closure Technique for Intradural Spinal Surgery to Prevent Re-Tethering

Karl F. Kothbauer, MD; George I. Jallo, MD; Fred J. Epstein, MD (New York, NY)

INTRODUCTION Retethering is a common occurrence after surgery for spinal dysraphism, or intradural spinal surgery. The pia of the cord becomes densely adherent to the inner surface of the dural sac, particularly at a suture line.

METHODS A simple closure technique using a synthetic dural substitute is presented. The intent is to prevent scarring, retethering and its associated morbidity. A DuraGuard synthetic dural graft is placed over the spinal cord, or the cauda equine to

cover the entire dural opening. The graft is tailored so that it covers the intradural structures beyond the suture line. The dura is then closed over the graft. The graft is secured to the dura using a few tangential stitches, enough to keep it from migrating craniocaudally, or compressing the intradural structures. If the dura is not intact for complete closure over the DuarGuard, as is often the case in tethered cord surgery, a local fascial graft is used in an insert-fashion.

RESULTS Over a period of the past 2 years, the technique was used in 15 patients. Two had spinal cord lipomas, 4 intramedullary spinal cord tumors, and 9 had tethered cord surgery. There were no technical problems at surgery, and no postoperative morbidity that would be attributable to the graft or the closure technique. There was one CSF leak in a patient with recurrent tethered cord syndrome, where a second operation was necessary.

CONCLUSIONS The proposed closure technique is relatively simple and appears at least as safe as closure using a direct dural-insert graft. The follow-up interval is too short to evaluate whether this indeed prevents anatomic and/or symptomatic tethering or re-tethering.

126. Endoscopic Corpus Callosotomy: a Feasibility Study

Matthew D. Smyth, MD; R. Shane Tubbs, PhD, MS, PA-C; George Salter, PhD; Kyle Doughty, MD; Jeff P. Blount, MD (Birmingham, AL)

INTRODUCTION Here we describe a novel endoscopic technique for minimally invasive bisection of the corpus callosum.

METHODS Brow incisions with midline twist drill trephinations were performed in fifteen adult cadavers. An endoscope was introduced along the anterior falx cerebri in the subarachnoid space and used to transect the anterior two-thirds of the corpus callosum, taking care to avoid the anterior cerebral arteries and their branches. Specimens were then removed and inspected for adequacy of the bisection and for any evidence of neurologic or vascular injury.

RESULTS No complications such as injury to the superior sagittal sinus, anterior cerebral vessels, or cingulum were encountered in any of our cadaveric specimens. The corpus callosum was easily transected in each specimen. The frontal sinus was entered in one case.

CONCLUSIONS We believe this technique will provide a less invasive method for patients who require corpus callosotomy and minimize much of the morbidity associated with the traditional methods of open corpus callosotomy. The addition of frameless stereotactic navigation may aid in defining the intraoperative extent of the transection and the location of the frontal sinus for optimal entry site localization.

127. Aplasia Cutis Congenita: Three Cases in Eighteen Months

David M. Wrubel, MD; George T. Burson, MD (Little Rock, AR)

INTRODUCTION Cutis Aplasia is an extremely rare condition presenting at birth. Death from exsanguination is known to occur in these patients due to the location of the defect over the sagittal sinus and the small total blood volume in infants.

METHODS A multi-disciplinary approach was taken, involving neurosurgeons, plastic surgeons and neonatologists in the medical and surgical care of these complicated patients.

RESULTS All three patients did well. However, one patient required three operations along with multiple transfusions to close the defect.

CONCLUSIONS In the care of these complex patients, it is necessary to have multiple teams involved during their hospitalization in order to maximize their potential for a successful outcome. Six to eighteen month follow-up will be available.

128. A Pitfall in Radiographic Assessment of Progression of Incompletely Resected Low Grade Astrocytomas

Michael H. Handler, MD; Nicholas Foreman, MD; K. Wendorf, MD; Laura Fenton, MD (Denver, CO)

INTRODUCTION Incompletely resected low grade astrocytomas may progress, and are typically followed with serial MRI scans. Some may be treated with chemotherapy immediately at diagnosis, or at sign of progression.

METHODS Between 1995 and 2001, 18 children were consecutively treated with a regimen of Carboplatin and Vincristine. Each child was followed with MRIs at three month intervals, each compared to the previous scan. None was noted to have clinically significant changes in scans. We then retrospectively compared scans at initiation of therapy and at completion for each child.

RESULTS We noted 1 complete response, 3 partial responses, 7 minor responses, 5 with stable disease, and 2 who had substantial progression. The two children with progression had an alarming rapid growth of the tumor with discontinuation of therapy.

CONCLUSIONS When following children with low grade astrocytomas, serial MRIs must be compared to the original base-line study to assess reliably for progression. Subtle small interval changes may otherwise be missed, which may have prognostic significance at the ending of therapy.

129. Immediate Post-Operative Correction in Head Shape Following Calvarial Vault Remodeling for Sagittal Synostosis

Susan R. Durham, MD (Portland, OR); J.G. McComb, MD; Michael L. Levy, MD, PhD (Los Angeles, CA)

INTRODUCTION We present a prospective analysis of the effect of operative position (supine or prone) on the immediate correction in head shape following calvarial vault remodeling for sagittal synostosis.

METHODS Depending on the surgeon, the procedure was performed either supine or prone. In each operation, the sagittal suture was excised and barrel-staves performed from the coronal to the lambdoidal sutures. Posterior to the lambdoidal sutures, parietal/occipital remodeling was performed. If prone, the parietal/occipital remodeling was carried out to just below theinion. If supine, the remodeling was carried down to just above theinion and barrel-staving of the frontal bone was performed. In each child, the cranial index (cranial width/cranial length x 100) was measured immediately before and after surgery.

RESULTS Eight children were prospectively studied. Three operations were supine, and 5 were prone. The mean age was 4.5 months (supine) and 6.4 months (prone) (p=0.32). The estimated blood loss was 300 cc (supine) and 210 cc (prone)(p=0.088). Operative time was 110 minutes (supine) and 114 minutes (prone) (p=0.61). The pre-operative cranial index was 66 (supine) and 67 (prone) (p=0.64). The post-operative cranial index was 78 in the supine group and 74 in the prone group (p=0.04).

CONCLUSIONS The position the surgery is performed in does have a significant effect on immediate post-operative head shape. When the surgery is performed in the supine position, it allows for more extensive bony remodeling of the forehead and a significant improvement in immediate post-operative head shape as indicated by the cranial index. The addition of remodeling of the frontal bone did not significantly increase operative time or blood loss.

130. AL-Anazi Ventriculo-Uterine Shunt

Abdul Rahman AL-Anazi, MD (Al Khobar, Saudi Arabia)

INTRODUCTION Hydrocephalus is one of the commonest congenital anomalies especially in the developing countries for several reasons; one of them is the prohibition of abortion in most of them. The better understanding of the development of the brain and CNS showed that hydrocephalus and subsequently high intracranial pressure usually lead to devastating outcome such

as abnormal development of the brain. On the other hand, theoretically at least, they have the potential of recovery from hydrocephalus without the devastating effects, if they are treated early enough (before 32 gestational weeks). This potential is attainable due to the elasticity of the normal cells, and the fact, that a reduction in the ICP at this early stage may prevent cortical ischemia, as well as restoring synaptic, neurotransmitter formation and function.

The advances of methods of intrauterine diagnosis, US and MRI made it possible to detect early enlargement of the ventricles and diagnose hydrocephalus as early as 20 gestational weeks. There were several trails of intra-uterine shunting, but most of them ended up with complications; for example: shunt migration intra or extracranially, obstruction, infection and malposition, etc.

METHODS To overcome these complications, the author have developed a special ventriculo- uterine shunt device, which will be the first step in treating congenital hydrocephalus.

RESULTS AL-Anazi V-U Shunt is easy to implant, have a one way valve to prevent the amniotic fluid back flow during the uterine contraction stages, contained a special wings to prevent intra or extra-cranial migration, being relatively short and wide reduce the possibility of malfunction, and since there is no communication with the external environment, the risk of infection is reduced. Details of the device and implantation technique will be presented.

CONCLUSIONS AL-Anazi V-U Shunt is a promising shunt to be considered as the first step in treating congenital hydrocephalus.

131. The Effect of Change in Technique on Complication Rate for Baclofen Pump Procedures

Michael S. Turner, MD (Indianapolis, IN)

INTRODUCTION The implantation of a programmable pump for baclofen infusion has a complication rate due to catheter malfunction that varies between 5 and 50%. The author will use his database of

over 200 pumps over 8 years to identify changes in the type and rate of catheter complications as the technique for catheter implantation changed. The consensus of an expert panel will also be reviewed. Video on implantation of a new catheter system from Medtronic will also be presented.

METHODS Retrospective chart review using a database of over 200 pump implantations.

RESULTS Changes in technique were effective in eliminating certain common catheter complications.

CONCLUSIONS The complication rate related to catheter systems can be favorably altered by changes in techniques.

132. A Single-Pass Tunneling Technique for CSF Shunting Procedures

Michael A. Sandquist, MD; Nathan R. Seiden, MD, PhD (Portland, OR)

INTRODUCTION Implantation of ventriculo-peritoneal shunts in the pre-coronal position is generally accomplished using a retro-auricular incision for subcutaneous tunneling. Retro-auricular incisions may be associated with complications, including CSF leak and shunt infection. The occurrence of two retro-auricular incision complications in our institutional practice prompted us to develop a technique for 'single-pass' shunt tunneling from frontal to abdominal incisions.

METHODS Eleven consecutive children (age 2 days to 16 months) who underwent primary insertion of a ventriculo-peritoneal shunt in the pre-coronal position were studied prospectively. In each case, a malleable tunneling device with a disposable plastic sheath was contoured based on the individual patient anatomy and passed in the subcutaneous plane from the abdominal to cranial incision. The tunneler was then removed and the distal shunt system passed through the sheath. Eight patients were treated for congenital hydrocephalus (including four with myelomeningocele and one with post-hemorrhagic hydrocephalus) and three for hydrocephalus secondary to CNS tumors. Average length of clinical follow-up is currently 7 months (range: 2 to 11 months).

RESULTS There were no peri-operative or long-term complications of the single-pass technique. Six of eight patients with congenital hydrocephalus are currently well without any further medical or surgical intervention. Two underwent shunt revision for proximal obstruction, with an intact distal system. Three of three patients with hydrocephalus secondary to CNS tumor suffered secondary infectious shunt complications during periods of severe neutropenia from chemotherapy (6 weeks to 6 months after shunt insertion).

CONCLUSIONS For primary ventriculoperitoneal shunt insertion in infants and young children, the single-pass tunneling technique is safe and avoids one source of complications.

133. Diagnosis of Two Brain Tumors in Utero: Case Reports

Marcus L. Ware, MD, PhD; Errol R. Kolen, MD; Andrew Horvatic, MD; Victor Perry, MD; Nalin Gupta, MD, PhD (San Francisco, CA)

INTRODUCTION Although the prognosis for neonatal brain tumors is poor, early detection of intracranial lesions may lead to interventions that improve outcomes. We present two cases of pediatric brain tumors diagnosed in utero by fetal ultrasonography and MRI.

METHODS The first patient was a 2 day old male delivered at 36 5/7 weeks of gestation from a 20 year old G2P2 mother. Ultrasonography showed a 7 cm hyperechoic intracranial mass at 32 weeks of gestation. At 36 weeks, fetal MRI showed a 7 x 7 x 6 cm heterogeneous suprasellar mass with severe ventriculomegaly. Neurosurgical consultation was obtained. No intervention was offered. Pathology at autopsy showed an adamantinomatous craniopharyngioma. The second patient was a 1 day old female delivered to a 25 year old G4P3 mother. Ultrasonography at 31 5/7 weeks of gestation showed a large solid and cystic heterogeneous lobulated intracranial mass, extending above and below the tentorium. Fetal MRI showed a 9.5 x 9.1 x 7 cm mass and severe associated hydrocephalus. Neurosurgical consultation was obtained

and no intervention was offered. Pathology at autopsy showed an immature teratoma.

RESULTS At the time of diagnosis, these tumors were too large for safe intervention. In the near future, early diagnosis may allow brain tumors to be detected while small enough for surgical resection.

CONCLUSIONS The surgical repair of fetal myelomeningocele, has been shown to be a safe procedure that improves neurologic outcomes after delivery. Future studies on the role of fetal surgery for treatment of brain lesions are highly anticipated.

134. Internal Carotid Artery Dissection in an Infant: Case Report and Literature Review

Rebekah C. Austin, MD; Steven S. Glazier, MD (Winston-Salem, NC)

INTRODUCTION Spontaneous dissections of cerebral vessels are rare occurrences in any patient population. We present what we believe to be the first case of ruptured pseudoaneurysm from a dissected right internal carotid artery in an infant with radiographic imaging suggestive of non-accidental trauma.

METHODS A five month old male presented with lethargy, irritability, anorexia, and ptosis. Initial cranial CT revealed right sylvian fissure and tentorial subarachnoid hemorrhage, right lateral interventricular hemorrhage and early hydrocephalus. The pattern of subarachnoid hemorrhage was highly suggestive of non-accidental trauma. Evaluation included a negative skeletal survey and fundoscopic examination without evidence of retinal hemorrhages. MRI/MRA revealed a hypoplastic right A1 and slight attenuation of the right MCA consistent with mild vasospasm. No significant flow void or aneurysm was evident. The patient was discharged with a presumptive diagnosis of non-accidental trauma. The patient subsequently presented with similar symptoms and precipitous development of right ptosis and anisocoria. Recurrent subarachnoid hemorrhage was found on CT. Arteriography revealed a dissected right supraclinoid internal carotid artery aneurysm.

RESULTS Right pterional craniotomy was performed for trapping the aneurysm. Though the parent vessel was sacrificed, distal pulsatile flow was confirmed with intraoperative ultrasound. No post-operative neurologic deficits were found.

CONCLUSIONS We present what we believe to be the first case report of spontaneous dissection of the internal carotid artery with ruptured pseudoaneurysm in an otherwise healthy infant. Although investigation yielded no evidence of other injury, non-accidental trauma cannot be completely excluded due to the rarity of this clinical entity.

135. Hydrocephalus and Chronic Epidural Hematoma

Mansour Parvareh-Rizi, MD (Esfahan, Iran); Hossein Ghalea-Novaei, MD; Farzaneh Chitsaz, MD (Tehran, Iran); Mohsen Dalvandi, MD (Arak, Iran)

INTRODUCTION Complications of Shunting for hydrocephalus are numerous and would be expected in any patient. One of the extremely rare complications is chronic epidural hematoma. We report one case with that complication, its management and outcome.

METHODS A 5 year old boy who had known of Hydrocephalus and shunt inserted referred to us because of recurrent generalized tonic-clonic convulsion. General examination showed anemia, pectus excavatum and diffuse wheezing at chest. Neurological examination showed GCS=9 and generalized spasticity. No other deficit was found. Shunt reservoir filling and emptying was normal. CT Scan showed a large fronto parietal heterogen (mostly hypodense) extraaxial lentiform mass lesion with significant mass effect and midline shift.

RESULTS The patient operated through two burr holes and a mixed liquid and clot formed lesion removed totally. Specimen sent for pathology and bacteriology. Definite diagnosis was chronic epidural hematoma.

CONCLUSIONS One of the extremely rare complications after shunt insertion is chronic epidural hematoma that must be differentiated from other extraaxial collections. It causes brain compression syndrome and must remove totally. If the general condition of patient would not be bad, postoperative course will be excellent.

136. Lumboperitoneal Shunting Preferentially Drains the Subarachnoid Space

Leila Khorasani, BS; Christian W. Sikorski, MD; David M. Frim, MD, PhD (Chicago, IL)

INTRODUCTION Based on a proposed pathophysiology of SVS, Le et al (Pediatr Neurosurg 36:178-182, 2002) hypothesize that LP shunting exerts effects in SVS patients by increasing the buffering capacity for raised intracranial pressure (ICP) via an increase in cerebrospinal fluid (CSF) drainage potential within the subarachnoid space (SAS). We describe two SVS patients with functioning LP shunts but non-functioning VP shunts who presented with ventriculomegaly (not SVS), and persistence of shunt malfunction-like symptoms. Revision of the VPS resulted in complete resolution of symptoms. This supports the hypothesis that LP shunting preferentially drains the SAS over the intraventricular space and in these cases allowed the "SVS" ventricles to enlarge.

METHODS Two patients with SVS presented with symptoms consistent with malfunction of their VP shunts (headache, nausea and vomiting, lethargy and decreased cognitive skills) and were found to have elevated CSF pressure by lumbar puncture. They both underwent placement of LP shunts with resolution of symptoms. However, each patient subsequently presented with recurrence or persistence of their symptoms and ventriculomegaly, not slit ventricles. Lumbar punctures performed at that time revealed low ICP.

RESULTS Both patients required revision of the VPS, which was found not to be functioning. In the presence of functioning LPS and VPS both patients had complete resolution of signs and symptoms of shunt malfunction.

CONCLUSIONS Low-ICP related ventriculomegaly with a functioning LPS and malfunctioning VPS in a SVS patient likely indicates that LP shunting preferentially drains the SAS and allows the ventricles to enlarge. That VPS revision in this setting results in resolution of ventriculomegaly supports this hypothesis.

137. Meningococcal Meningitis in a Shunted Patient Presenting with Paraplegia

David H. Harter, MD; Michael Gewitz, MD; Bruce Roseman, MD; Chiedozie Nwagwu, MD (Vaihalla, NY)

INTRODUCTION We describe a patient with hydrocephalus and factor VIII deficiency presenting with paraplegia secondary to meningococcal meningitis and endocarditis. The exceptional presentation of meningococcal meningitis with paraplegia as well as factor VIII deficiency and endocarditis makes this a unique case.

METHODS This 18-year-old boy underwent ventriculoperitoneal shunting at the age of nine months. Upon initial evaluation he was toxic, the temperature was 100.5, he was awake and minimally responsive. There was complete paraplegia and a T6 sensory level. Initial treatment included factor VIII. CSF from a shunt tap had 1,861 WBC and Gram-negative diplococci. He received intravenous antibiotics. The shunt was replaced with an external ventricular drain. Methylprednisolone was begun.

RESULTS Blood and CSF grew N. meningitidis. Given his coagulopathy, a CT scan was obtained and ruled a spinal epidural hematoma. An MR scan was normal. The patient remained febrile, prompting multiple cultures. An echocardiogram demonstrated vegetations on the mitral valve. A left occipital ventriculoperitoneal shunt was inserted. The patient was eventually able to ambulate independently.

CONCLUSIONS Myelopathy is a rare complication of meningococcal meningitis. This is the sole reported case reversible paraplegia in a shunted patient due to meningococcal infection. Other unique aspects of this case are factor VIII deficiency and endocarditis.

138. Pilocytic Astrocytomas in Two Siblings

David M. Wrubel, MD; Georget T. Burson, MD (Little Rock, AR)

INTRODUCTION Pilocytic Astrocytomas are a common brain tumor in the pediatric population. Usually presenting as posterior fossa masses. Two half-sisters presented to our institution within a year with symptoms consistent with posterior fossa masses.

METHODS Both patients were worked up with diagnostic imaging and seen by our APNOC team which includes a pediatric neurosurgeon, pediatric neurologist, pediatric oncologist, and pediatric neuropsychologist. Both patients subsequently underwent craniotomy for resection of tumor.

RESULTS One patient had a complete resection based on follow-up imaging. The other patient has a small residual satellite nodule which has been stable for one year.

CONCLUSIONS We present an interesting case of two half-sisters who share a father. Both had pilocytic astrocytomas of the posterior fossa without any signs, symptoms or history of any inherited syndromes which have increased incidence of astrocytomas including neurofibromatosis. We feel that these are to sporadic cases which happened to occur in two half-sisters.

139. Chiari II Malformation and Foramen of Magendie Choroid Plexus Papilloma: A Ca I Association?

Todd A. Maugans, MD (Burlington, VT)

INTRODUCTION The Chiari II malformation is frequently associated with compression of the cervicomedullary junction. In certain patients, this may produce dysfunction of the lower brainstem or spinal cord. Frequently encountered at the time of decompression are dense arachnoidal adhesions; these possibly arise from chronic irritation of the leptomeninges. Choroid plexus papillomas are benign neoplasms that typically occur in the supratentorial compartment in children. The finding of this neoplasm at the site of cervicomedullary compression in a patient with Chiari II is unique and worthy of contemplation.

METHODS Case report and review of the literature.

RESULTS A 27 month female with Chiari II malformation was lost to care shortly after birth. She presented with signs of chronic malnutrition secondary to swallowing dysfunction associated with severe lower cranial neuropathies. An unenhanced MRI demonstrated a severe Chiari II malformation with compression of the upper cervical region and absence of surrounding CSF; no masses were appreciated. At the time of craniocervical decompression, a fleshy mass covered by dense arachnoid locations was encountered at the Foramen of Magendie. This was resected; the pathology was consistent with choroid plexus papilloma. The patient demonstrated clinical improvement in her swallowing dysfunction and has remained without evidence of tumor recurrence for 18 months.

CONCLUSIONS Neoplasms of the integument, respiratory and gastrointestinal tracts have been associated with chronic irritation. Neoplasms of the CNS have not been reported as caused by irritation or inflammation. Although the finding of a choroid plexus papilloma in our patient with severe cervicomedullary compression may be coincidental, it is plausible that the association is causal. The pathophysiology of chronic irritation and inflammation inducing chemical and genetic alterations in cell types normally found in the CNS warrants consideration and investigation.

140. Multicentric Primitive Neuroectodermal Tumor: Evaluation, Diagnosis and Management

Victor L. Perry, MD; Marcus L. Ware, MD, PhD; Anuradha Banerjee, MD; Nalin Gupta, MD, PhD (San Francisco, CA)

INTRODUCTION Primitive neuroectodermal neoplasms (medulloblastomas) remain one of the most common intracranial tumors of childhood. With advances in surgical technique as well as adjuvant therapeutic protocols, the prognosis for children diagnosed with these malignant lesions has improved markedly. We describe the evaluation, diagnosis and management of an infant presenting with a rare multicentric, desmoplastic variant of this neoplasm.

METHODS The patient is an 18 month old male who presented with progressive lethargy. Head CT on admission revealed marked hydrocephalus with the presence of diffuse isointense lesions throughout the posterior fossa extending to the midbrain. MR imaging with gadolinium contrast revealed minimally enhancing posterior fossa lesions. Staging spine imaging was negative. Following placement of an external ventricular drain, the patient underwent a midline posterior fossa craniotomy for diagnosis and debulking of the lesion in relief of the obstructive hydrocephalus. Given the size and extent of the lesion, as well as apparent brainstem involvement, a complete resection was not attempted. Pathologic examination revealed PNET/medulloblastoma WHO grade IV, desmoplastic variant. Follow up CT revealed improved hydrocephalus; the EVD was weaned. After further MR imaging, the child underwent a right suboccipital craniotomy utilizing intraoperative navigation for resection of the remaining segments of the neoplasm.

RESULTS The patient recovered well from both procedures with return to full neurologic function. Immediate and 3 month postoperative imaging studies revealed no residual neoplasm. Given the negative staging and no residual tumor, the child was eligible for and enrolled in NCI Protocol No. PBTC-001 involving systemic and intrathecal chemotherapy (mafosfamide) followed by conformal radiation.

CONCLUSIONS Primitive neuroectodermal neoplasms (medulloblastomas) represent the most common malignant pediatric brain tumor. Optimal surgical resection for even unusual, multifocal lesions as illustrated in this case, utilizing advanced intraoperative navigation techniques, and combined with novel adjuvant therapeutic protocols may result in improved long-term outcomes for these patients.

141. Cervical Meningocele and Associated Spinal Abnormalities, Case Report and Review of the Literature

Carlos H. Feltes, MD; Vassilios Dimopoulos, MD; Rostislav Davydov, MD; Angel Boev, BS; Debbie Melone, RN; Christopher Troup, MD (Macon, GA)

INTRODUCTION This case is a new born baby girl which at birth was diagnosed with a cervical meningocele, initially by prenatal ultrasound it was thought to be occipital but at birth it was confirmed to be confined to the mid-cervical spine, further workup by MRI revealed a coexisting thoracic diplomyelia and bilateral tethered cords, patient at birth was found to be neurologically intact.

METHODS At 4 months of age patient underwent surgery for simultaneous repair of cervical meningocele, exploration of diplomyelia, and release of tethered cords bilaterally.

RESULTS Patient has now over a 30 month follow-up, is ambulating with no bowel or bladder incontinence and has developed well according to her chronological age.

CONCLUSIONS These three entities separately are infrequent occurrences but the presence of all three in this particular patient makes this a unique clinical scenario, pertinent review of the literature will be discussed.

142. Management of Herniated Disc of Lumbar Spine in Teenagers

S.M. Rezaian, MD, PhD, FICS, FRCS, (Beverly Hills, CA)

INTRODUCTION Children low back pain is real and always one must take a serious look for the etiology and find the cause and implement the real treatment. In this paper, we present two cases.

METHODS First case: A 13 year old who after a failure to correct diagnosis in a large prestigious medical center was under psychiatrist and lost 18 months of her schooling. Second case: a 17 year old boy, who were treated in spine clinic, one of the famous university. During subsequent 12

months, his condition aggravated and progressed and led to three herniated disc of L3-L4, L4-L5 and L5-S1, with 60 scoliosis and 30 lumbar kyphosis. He was offered to have anterior and posterior spinal instrumentation and fusion.

RESULTS Both of these patient were diagnosed by correct history and physical examination. The diagnosis were reconfirmed by dynamic discography and both of them were successfully treated by percutaneous discectomy, as an out patient.

CONCLUSIONS Low back pain in children is real and deserve a full clinical examination and investigation by a specialist. Percutaneous discography and nowadays endoscopic discectomy is recommended as a safe treatment of choice, where conservative treatment fails.

143. Pediatric High Grade Astroblastoma: A Case Report and Review of the Literature

Francesco T. Mangano, DO; Mark A. Mittler, MD; Elsa Valderrama, MD; Steven J. Schneider, MD (New Hyde Park, NY)

INTRODUCTION We report a case of cerebral astroblastoma in an 8 year old boy who presented with signs and symptoms of raised intracranial pressure. Computerized tomography and magnetic resonance imaging demonstrated a large heterogenously enhancing solid mass in the left frontal lobe with calcifications and cystic regions. The tumor was removed yielding a histologic diagnosis of high grade astroblastoma. The patient's recovery was complicated by hydrocephalus requiring placement of a ventriculoperitoneal shunt. The review of the literature is limited by the rarity of the tumor. However, the extent of resection and histologic grade are both shown to be prognostic variables. Adjuvant radiation therapy for high grade lesions has become a standard, but doses have varied from 3800 cGy to 7200 cGy without significant difference of survival outcome. Adjuvant chemotherapy has been administered after resection and radiation and has not been shown to alter recurrence, prognosis, or long-term survival. Conversely, low grade lesions, if totally resected offer a better overall prognosis. Given the existing

literature, adjuvant radiation therapy was administered, and we chose to reserve chemotherapy in the event of recurrence. Presently the patient remains tumor free. Astroblastomas have been infrequently reported in the neurosurgical literature since the original description in 1926. A meta-analysis of the existing cases has provided insight on the prognosis associated with these tumors.

144. Surgical Treatment of Tethered Cord Syndrome

Yasuko Kusaka, MD, PhD; Reizo Shirane, MD, PhD; Takashi Yoshimoto, MD, PhD (Sendai City, Japan)

INTRODUCTION Early prophylactic untethering surgery using modern neuroradiological evaluation and surgical techniques has achieved beneficial results in infants and patients in early childhood with asymptomatic spinal lipoma. However, the physiological differences in tethered cord syndrome with various lesions make untethering surgery more difficult. There still remains controversy concerning the surgical treatment of tethered cord syndrome.

METHODS We retrospectively reviewed the surgical treatment and results of 148 patients of spina bifida. This study involves three groups: group A (forty-five asymptomatic spinal lipomas), group B (eighty-two spinal lipomas with tethered cord syndrome) and group C (twenty-two myelomeningocele with tethered cord syndrome over four-year-old). Patients were all examined neurology and urodynamic study pre and postoperatively. In group B, tethered cord syndrome was recognized in 64% patients with sacral agenesis as in 24% without sacral agenesis. All surgeries contained untethering, spinal conus reconstruction and dural sac reconstruction with Gore-tex sheet. The median follow-up period was 63.4 months after untethering surgery.

RESULTS Post operatively, group A remains asymptomatic and the improvement rate in group B was 28.6% in neurogenic bladder (NB), 78.9% in motor symptoms (M) and 90% in pain (P). In group C, 41.7% in NB, 63.6% in M and 87.5% in P.

CONCLUSIONS We concluded that the most effective therapy against tethered cord syndrome was prophylactic untethering in infant, especially patients with sacral agenesis. However, for patients with tethered cord syndrome, we recommend adequate surgical indication and strategy for symptom reduction.

145. Surgical Treatment of Late Infantile Moyamoya Disease

Yasuko Kusaka, MD, PhD; Reizo Shirane, MD, PhD; Takashi Yoshimoto, MD, PhD (Sendai City, Japan)

INTRODUCTION The efficacy of revascularization surgery for childhood Moyamoya disease is believed to have therapeutic effects against cerebral ischemic attacks. The authors present the results of surgical treatment of late infantile Moyamoya Disease whose prognosis said to be worse.

METHODS Between 1976 and July 2000, we followed 258 moyamoya disease [133 children including 43 infants]. Since 1989, we utilized our newly developed original bypass procedures (dural pedicle insertion over the brain surface combined with encephalo-galeo-myo-synangiosis) which can cover the hemodynamic compromised area for 68 patients. There were 23 late infantile (<5 years) [from 10 months to 5 years (average 3.6 years), 1-134 months of follow-up period (average 6.0 years)].

RESULTS Their clinical presentation were TIAs in 18, cerebral infarction in 4, cerebral hemorrhage with the opposite side infarction in 1. All infant cases with onset of convulsion revealed cerebral infarctions on MRI. At 30 times, our original bypass procedures were done. Combined procedures with STA-MCA anastomosis were done for 9 times of 5 patients under 5 years. The surgical field was determined by investigation of hemodynamic compromised area using 123I-iodoamphetamine-SPECT with acetazolamide. Postoperatively, there were no significant complications. TIAs sometimes occurred within 7 days of surgery, however, they did not progress to complete stroke. In all patients, cerebral hemodynamic compromise improvements were recognized.

CONCLUSIONS Consideration of the hemodynamic compromise of developmental brain and selection of adequate surgical field and methods are required for good prognosis of late infantile Moyamoya disease.

Notes

146. Endoscopic Ventriculostomy in Management of Hydrocephalus in Paediatric and Adolescent Patients

Francis H. Tomlinson, MD, PhD, FRACS (Brisbane, Queensland, Australia); Terrence J. Coyne, FRACS (Herston, Queensland, Australia); Michael J. Bryant, MBBS

INTRODUCTION The safety and clinical use of endoscopic ventriculostomy in hydrocephalus(HC) was evaluated.

METHODS Patient records (1994–2001) were reviewed. EV was performed with a rigid endoscope and No 4 Fogarty catheter.

RESULTS There were 49 patients (26M, 23F), 1 month to 19 years, meningomyelocele (MM) 8, tumour (T) 18, intraventricular hemorrhage (IVH) 4, arachnoid cyst (AC) 3, Aqueduct stenosis(AS) 5, Chiari malformation (CM) 4, other (O) 6. Four patients (T 3), (AC 1), underwent successful endoscopic septum pellucidotomy. 47 endoscopic third ventriculostomies (III EV) were performed in 44 patients. In 24 patients, III EV was successful—MM 2, T 12, IVH 2, AS 5, AC 11, O 1. 7 patients were previously shunt dependent. 2 patients were under 2 years of age. Twenty patients—MM 6, T 3, IVH 2, AC 1, CM 1, AS 2, O 5, required shunting after III EV. 7 patients were under 2 years of age. Shunting was performed within 48 hrs (3), 1 week (5), 2 months (6), 1 year (4) and 6 years (1). There were no deaths. Complications were infection (2), transient visual deficit (1) and subdural collection (1).

CONCLUSIONS EV using blunt dissection is safe but the technique may affect success. Failure of III EV occurred in MM and patients under 2 years of age. III EV in HC related to tumours were successful. III EV can also provide a window to allow CSF analysis and elective shunt revision. EV is an integral part of the management of hydrocephalus.

2002 Membership Roster

Rick Abbott, MD
Beth Israel Medical Center
170 E. End Ave./Neurosurgery
New York, NY 10128-7603

Jafri Malin Bin Abdullah, MD, PhD
Hosp. Univ. Sci/Neurosurgery/Neurosci.
16150 Kubang Kerian
Kelantan
Malaysia

P. David Adelson, MD, FACS
Children's Hosp. of Pittsburgh
3705 5th Ave./Neurosurgery
Pittsburgh, PA 15213-2583

Nejat Akalan, MD
Hacettepe University School of Med.
Dept. of Neurosurgery
Ankara 06100
Turkey

A. Leland Albright, MD
Children's Hosp. of Pittsburgh
3705 5th Ave./Neurosurgery
Pittsburgh, PA 15213-2524

Lance Altenau, MD
501 Washington, Ste. 700
San Diego, CA 92103-0000

A. Loren Amacher, MD
Geisinger Clinic
Dept. of Neurosurgery
Danville, PA 17822-0000

Luis V. Amador, MD
1440 Veteran Ave., Ste. 336
Los Angeles, CA 90024-4832

Ahmed S. Ammar, MD, PhD
El Salam Hospital
PO Box 3014
Cairo 11757
Egypt

Jim D. Anderson, MD
PO Box 658
San Carlos, CA 94070-0658

Brian T. Andrews, MD
2100 Webster St., Ste. 521
San Francisco, CA 94115-0000

Thomas J. Arkins, MD
Connecticut Neurosurgery PC
330 Orchard St., Ste. 316
New Haven, CT 06511-4417

Patricia A. Aronin, MD
Central Texas Neurosurgery for Children
1215 Red River St., Ste. 201
Austin, TX 78701-0000

Elaine J. Arpin, MD
S.W. Florida Neurosurgical Assoc.
413 Del Prado Blvd., Ste. 102
Cape Coral, FL 33990-5703

Wilson T. Asfora, MD, FRCSC F
North Center
1210 W. 18th St., Ste. 104
Sioux Falls, SD 57104-0000

Saleh S. A. Baeesa, MD
King Abdulaziz Univ. Med. Ctr.
PO Box 80215
Jeddah 21589
Saudi Arabia

Walter L. Bailey, MD
500 River St.
Minneapolis, MN 55401-0000

Gene A. Balis, MD
Neurological Surgeons Associates
3000 E. Fletcher Ave., Ste. 340
Tampa, FL 33613-4645

Steven J. Barrer, MD
Abington Neurosurgical Associates
2510 Maryland Rd., Ste. 185
Willow Grove, PA 19090-1134

Henry M. Bartkowski, MD, PhD
Akron Childrens Hospital
1 Perkins Square, Rm. 6301
Akron, OH 44308-1062

James E. Baumgartner, MD
3418 Georgetown St.
Houston, TX 77005-2910

Robert M. Beatty, MD
Neurosurgery of Kansas City
8919 Parallel Pkwy., Ste. 455
Kansas City, KS 66112-0000

William O. Bell, MD
Carolina Neurosurgical Associates
2810 N. Maplewood Ave.
Winston-Salem, NC 27103-4138

Mitchel S. Berger, MD
University of California—San Francisco
505 Parnassus Ave., Box 0112
San Francisco, CA 94122-2722

Jose A. Bermudez, MD
102 Thomas Rd., Ste. 107
West Monroe, LA 71291-0000

Karin S. Bierbrauer, MD
Temple University Hospital
3401 N. Broad St., Ste. 658
Philadelphia, PA 19140-5103

Peter McL. Black, MD, PhD
Children's/Brigham & Women's Hospital
300 Longwood Ave.
Boston, MA 02115-5724

Jeffrey P. Blount, MD
Children's Hospital of Alabama
1600 7th Ave., S. ACC 400
Birmingham, AL 35233-0000

John Scott Boggs, MD
1820 Barrs St., Ste. 104
Jacksonville, FL 32204-0000

Frederick A. Boop, MD
Semmes Murphey Clinic
1211 Union Ave., Ste. 200
Memphis, TN 38104-3562

William R. Boydston, MD
Pediatric Neurosurgery Associates
5455 Meridan Mark Rd., Ste. 540
Atlanta, GA 30342-1640

Bruce C. Bressler, MD
PO Box 8900
Green Bay, WI 54308-8900

Douglas L. Brockmeyer, MD
Primary Children's Medical Center
100 N. Medical Dr., Ste. 2400
Salt Lake City, UT 84113-1103

Jeffrey A. Brown, MD
Wayne State University
4160 John R Rd., Ste. 930
Detroit, MI 48201-0000

Derek A. Bruce, MD
78 Millbrook Rd.
Nantucket, MA 02554-0000

Michael James Burke, MD, FACS
Neurosurgery Institute of South Texas
3643 S. Staples
Corpus Christi, TX 78411-2456

George T. Burson, MD
Arkansas Childrens Hospital
800 Marshall St., Slot 838
Little Rock, AR 72202-3510

Leslie D. Cahan, MD
Kaiser Foundation Hospital
1505 N. Edgemont St., Rm. 4141
Los Angeles, CA 90027-5209

Jeffrey W. Campbell, MD
Medical University of South Carolina
96 Jonathan Lucas, Ste. 428CSB
Charleston, SC 29425-0000

Alexa Irene Canady, MD
6064 Forest Green Rd.
Pensacola, FL 32505-0000

Carolyn Marie Carey, MD
880 6th St. S.
St. Petersburg, FL 33701-0000

Peter W. Carmel, MD
UMDNJ-New Jersey Med. Sch.
90 Bergen St., Ste. 7300
Newark, NJ 07103-2425

Benjamin Carson, MD
Johns Hopkins University Hospital
600 N. Wolfe St., Harvey 811
Baltimore, MD 21287-8811

Michael J. Chaparro, MD
Palm West Pediatric Neurosurgery
3898 Via Poinciana, Ste. 18
Lake Worth, FL 33467-0000

Paul H. Chapman, MD
Massachusetts General Hospital
55 Fruit St., GRB502
Boston, MA 02114-2621

William R. Cheek, MD
3009 Robinhood
Houston, TX 77005-0000

Bruce Cherny, MD
Barrow Neurological Institute
2910 N. 3rd Ave.
Phoenix, AZ 85013-4466

Maurice Choux, MD
Hopital Des Enfants
De La Timone Cedex 5
Marseilles 13005
France

Samuel F. Ciricillo, Jr., MD
5238 Fair Oaks Blvd.
Carmichael, CA 95608-5766

David Douglas Cochrane, MD
Childrens & Womens Hlth. Ctr. of BC
B2 W. 4500 Oak St.
Vancouver, BC V6H-3N1
Canada

Philip Harry Cogen, MD, PhD
Children's National Medical Center
111 Michigan Ave. N.W.
Washington DC 20010-2916

Alan R. Cohen, MD
Rainbow Babies & Children's Hospital
11100 Euclid Ave., Rm. B501
Cleveland, OH 44106-1736

John J. Collins, MD
4759 Ridgetop Dr.
Morgantown, WV 26508-0000

Shlomo Constantini, MD
Dana Children's Hospital
6 Weizman St./Ped. Neurosurg.
Tel Aviv 64239
Israel

Richard A. Coulon, Jr., MD
Childrens Hospital/ Dept of Neurosurgery
200 Henry Clay Ave.
New Orleans, LA 70118-0000

Jeffrey W. Cozzens, MD
Evanston Northwestern Healthcare
2650 Ridge Ave., Ste. 4215
Evanston, IL 60201-1718

Kerry R. Crone, MD
Children's Hospital Medical Center
3333 Burnet Ave., Neurosurgery
Cincinnati, OH 45229-3026

Richard A. Day, MD
Montana Neurosurgery Center
2835 Fort Missoula Rd., Ste. 202
Missoula, MT 59804-7423

Concezio Di Rocco, MD
Univ. Cattolica/Neurochirurgia
Largo Gemelli 8
Rome, RO, 00168
Italy

Mark S. Dias, MD
Pennsylvania State Medical School
850 University Dr., Neurosurgery
Hershey, PA 17033-0000

Michael Dorsen, MD
Oregon West Neurosurgery PC
3615 N.W. Samaritan Dr., #210
Corvallis, OR 97330-3783

James R. Doty, MD
42 States St.
San Francisco, CA 94114-1402

James M. Drake, MD
Hospital for Sick Children
555 University Ave., #1504-D
Toronto, ON M5G-1X8
Canada

Ann-Christine Duhaime, MD
Dartmouth Hitchcock Medical Center
1 Medical Center Dr.
Lebanon, NH 03756-0000

Charles Cecil Duncan, MD
Yale-New Haven Medical Center
333 Cedar St., TMP 416
New Haven, CT 06520-8082

John A. Duncan, III MD
Rhode Island Hospital
2 Dudley St., Ste. 530
Providence, RI 02905-3236

Mary E. Dunn, MD
280 N. Smith Ave., Ste. 234
St. Paul, MN 55102-0000

Michael S. B. Edwards, MD
2800 L St., Ste. 340
Sacramento, CA 95816-5616

2002 Membership Roster

Michael R. Egnor, MD
NY Spine & Brain Surgery PC
Neurosurgery/HSC T12-080 SUNY
Stony Brook, NY 11794-8122

Stephanie L. Einhaus, MD
4425 Barfield Rd.
Memphis, TN 38117-2411

Howard M. Eisenberg, MD
University of Maryland Medical Center
22 S. Greene St., Ste. S12D
Baltimore, MD 21201-1544

Ibrahim Muftah El Nihum, MD
Scott & White Clinic
2401 S. 31st St.
Temple, TX 76508-0001

Richard G. Ellenbogen, MD
5616 N.E. 55th
Seattle, WA 98105-2835

Seyed M. Emadian, MD, PhD
Premier Neurosurgery & Spine Center
730 Kings Ln.
Tullahoma, TN 37388-0000

Fred J. Epstein, MD
Beth Israel Medical Center
170 E. End Ave.
New York, NY 10128-7603

Mark D. Erasmus, MD
New Mexico Neurosurgery P.C.
522 Lomas Blvd. N.E.
Albuquerque, NM 87102-2454

Walter J. Faillace, MD
University of Florida—Jacksonville
653 W. 8th St.
Jacksonville, FL 32209-6511

Neil Arthur Feldstein, MD
New York Neurological Institute
710 W. 168th St., Rm. 414
New York, NY 10032-0000

David A. Fell, MD
Neurosurgery Specialists
6767-A S. Yale
Tulsa, OK 74136-3302

Edwin G. Fischer, MD
Beth Israel Deaconess Medical Center
110 Francis St., Ste. 3B
Boston, MA 02215-0000

Ann Marie Flannery, MD
Medical College of Georgia
1120 15th St.
Augusta, GA 30912-4010

Eldon L. Foltz, MD
2480 Monaco Dr.
Laguna Beach, CA 92651-0000

Paul C. France, MD, PhD
Univ. of Oklahoma HSC/Neurosurgery
711 Stanton L. Young Blvd., #206
Oklahoma City, OK 73104-5021

Kathleen B. French, MD
3020 Hamaker Ct., B104
Fairfax, VA 22031-2220

Arno H. Fried, MD
Hackensack University Medical Center
30 Prospect Ave./WFAN Peds Ctr.
Hackensack, NJ 07601-0000

David M. Frim, MD
University of Chicago
5841 S. Maryland Ave., MC 4066
Chicago, IL 60637-1463

Herbert E. Fuchs, MD, PhD
Duke University Medical Center
Box 3272
Durham, NC 27715-3272

Joseph H. Galicich, MD
PO Box 276
Alpine, NJ 07620-0276

Sarah J. Gaskill, MD
Pediatric Neurosurgery of South Texas
4499 Medical Dr., Ste. 397
San Antonio, TX 78230-0000

Rosemaria Gennuso, MD
Scibetta & Gennuso PC
1661 Soquel Dr.
Santa Cruz, CA 95065-1709

Richard E. George, Jr., MD
3506 21st St., Ste. 400
Lubbock, TX 79410-0000

Timothy M. George, MD
Duke University Medical Center
Box 3272/Div. of Neurosurgery
Durham, NC 27710-0001

P. Langham Gleason, MD
Neurosurgery New Mexico PC
531 Harkle Rd., Ste. D
Santa Fe, NM 87505-0000

John C. Godersky, MD
Anchorage Neurosurgical Associates
3220 Providence Dr., Ste. E3-020
Anchorage, AK 99508-0000

James T. Goodrich, MD, PhD
Albert Einstein/Montefiore Medical Center
111 E. 210th St., Neurosurgery
Bronx, NY 10467-2401

Liliana C. Goumnerova, MD, FRCS
Childrens Hosp./Brigham & Womens Hosp.
300 Longwood Ave.
Boston, MA 02115-5724

Paul A. Grabb, MD
Children's Hospital of Alabama
1600 7th Ave. S., ACC 400
Birmingham, AL 35233-1711

Clarence S. Greene, Jr., MD
Memorial Medical Bldg.
2865 Atlantic Ave., Ste. 202
Long Beach, CA 90806-1740

David P. Gruber, MD
Neurosurgery Associates of Spokane
105 W. 8th Ave., Ste. 200
Spokane, WA 99204-2318

Laurance J. Guido, MD
30 Sutton Pl., Ste. 15A
New York, NY 10022-2365

Nalin Gupta, MD, PhD
University of California—San Francisco
505 Parnassus Ave., Rm. M-779
San Francisco, CA 94143-0112

Francisco A. Gutierrez, MD
201 E. Huron, Ste. 9-160
Chicago, IL 60611-0000

Yoon Sun Hahn, MD
University of Illinois at Chicago
912 S. Wood St., Ped. Neurosurg.
Chicago, IL 60612-7325

Stephen J. Haines, MD
Medical University of South Carolina
96 Jonathan Lucas, Ste. 428CSB
Charleston, SC 29425-0001

Sten E. Hakanson, MD
Lilla Ojelsbo S-590 98
Edsbruk
Sweden

Mark G. Hamilton, MD
Alberta Children's Hospital
1820 Richmond Rd. S.W.
Calgary, AB T2T-5C7
Canada

Lonnie L. Hammargren, MD
3196 S. Maryland Pkwy., Ste. 106
Las Vegas, NV 89109-2312

Mary Kathryn Hammock, MD
8620 Rolling Rd.
Manassas, VA 20110-3828

Michael H. Handier, MD
1010 E. 19th Ave., Ste. 605, Tammen Hall
Denver, CO 80218-1034

William C. Hanigan, MD, PhD
University of Illinois at Peoria
719 N. William Kumpf Blvd., #100
Peoria, IL 61605-0000

Michael D. Heafner, MD
Carolina Neurosurgery & Spine Assoc.
1010 Edgehill Rd. N.
Charlotte, NC 28207-1885

Michael A. Healy, MD
Neurosurgical Network Inc.
2222 Cherry St., Ste. M200
Toledo, OH 43608-0000

Dan S. Heffez, MD
CINN Specialty Care Pavilion
4550 N. Winchester Ave., 3rd Fl.
Chicago, IL 60640-5205

Leslie C. Hellbusch, MD
Midwest Neurosurgery
8005 Farnam Dr., Ste. 305
Omaha, NE 68114-4441

Robert W. Hendee, Jr., MD
10709 Rigsbee Ct.
Austin, TX 78739-0000

Thomas J. Holbrook, Jr., MD
Columbia Neurosurgical Associates PA
114 Gateway Corporate Blvd., #420
Columbia, SC 29203-0000

Robert D. Hollenberg, MD
McMaster University
1200 Main St. W., Rm. 4E8
Hamilton, ON L8N-3Z5
Canada

Gregory W. Hornig, MD
Children's Mercy Hospital
Sect. of Neurosurgery
2401 Gillham Rd.
Kansas City, MO 64108-0000

Roger Hudgins, MD
Pediatric Neurosurgery Associates
5455 Meridan Mark Rd., Ste. 540
Atlanta, GA 30342-1640

Stephen L. Huhn, MD
Stanford University Medical Center
300 Pasteur Dr., R203/Neurosurgery
Stanford, CA 94305-5327

Robin P. Humphreys, MD
Hospital for Sick Children
555 University Ave., #1504
Toronto, ON M5R-2Z8
Canada

Mark R. Iantosca, MD
Neurosurgeons of Central Connecticut
100 Retreat Ave., Ste. 705
Hartford, CT 06106-0000

Bermans J. Iskandar, MD
University of Wisconsin—Madison
600 Highland Ave., H4/334
Madison, WI 53792-0000

George I. Jallo, MD
Institute for Neurology & Neurosurgery
205 Commercial Ave., Ste. 37
Palisades Park, NJ 07650-0000

Hector E. James, MD
7930 Frost St., Ste. 103
San Diego, CA 92123-0000

John A. Jane, MD, PhD
University of Virginia Health System
Box 800212/Neurosurgery
Charlottesville, VA 22908-0000

David F. Jimenez, MD
University of Missouri—Columbia
1 Hospital Dr., N-521
Columbia, MO 65201-5276

Dennis L. Johnson, MD
408 Elm Ave.
Hershey, PA 17033-1751

John K. Johnson, MD
Southeastern Neurosurgical & Spine Inst.
20 Medical Ridge Dr.
Greenville, SC 29605-5605

Martin Johnson, MD
31870 S.W. Country View Ln.
Wilsonville, OR 97070-7476

Mary M. Johnson, MD
3223 Chatham Rd.
Atlanta, GA 30305-1101

Robert F. C. Jones, MD
Sydney Children's Hospital
21 Norfolk St.
Paddington, NSW, 2021
Australia

Allen S. Joseph, MD
Neuro Medical Center
7777 Hennessy Blvd., Ste. 10000
Baton Rouge, LA 70808-4300

John E. Kalsbeck, MD
Riley Hospital for Children
702 Barnhill Dr.
Indianapolis, IN 46202-5200

Paul M. Kanev, MD
Pennsylvania State Medical School
850 University Dr., Neurosurgery
Hershey, PA 17033-0850

Samuel S. Kasoff, MD
11 Sycamore Ln.
White Plains, NY 10605

2002 Membership Roster

Bruce A. Kaufman, MD
Children's Hospital of Wisconsin
9000 W. Wisconsin Ave., PO Box 1997
Milwaukee, WI 53201-1997

Robert F. Keating, MD
Children's National Medical Center
111 Michigan Ave. N.W.
Washington DC 20010-0000

David L. Kelly, Jr., MD
Wake Forest University
Medical Center Dr., Neurosurgery
Winston-Salem, NC 27157-1029

John R. W. Kestle, MD
Primary Children's Medical Center
100 N. Medical Dr., Ste. 2400
Salt Lake City, UT 84113-1103

David M. Klein, MD
690 Fearington Post
Pittsboro, NC 27312-8507

Laurence I. Kleiner, MD
Children's Medical Center
1 Childrens Plaza, Neurosurgery
Dayton, OH 45404-1815

David S. Knierim, MD
GED 4
9300 Valley Children's Pl.
Madera, CA 93638-8761

Edward J. Kosnik, MD
Columbus Children's Hospital
931 Chatham Ln.
Columbus, OH 43221-2417

Karl F. Kothbauer, MD
Beth Israel Medical Center
170 E. End Ave., Neurosurgery
New York, NY 10128-7603

Mark D. Krieger, MD
Mount Sinai School of Medicine
1 Gustave L. Levy Pl., Box 1136
New York, NY 10029-6500

Cornelius H. Lam, MD
University of Minnesota
420 Delaware St., S.E. MC 96
Minneapolis, MN 55455-0374

John P. Laurent, MD
Texas Children's Hospital
6621 Fannin MC-CC1710.05
Houston, TX 77030-2303

Mark Robert Lee, MD
Medical College of Georgia/Neurosurgery
1120 15th St., BI 3088
Augusta, GA 30912-0004

Michael Lee Levy, MD
8010 Frost St., Ste. 300
San Diego, CA 92123-0000

Veetai Li, MD
Children's Hospital of Buffalo
219 Bryant St., Neurosurgery
Buffalo, NY 14222-2006

Kenneth I. Lipow, MD
Connecticut Neurosurgical Specialists
267 Grant St.
Bridgeport, CT 06610-2805

John D. Loeser, MD
University of Washington
Box 356470
Seattle, WA 98195-0000

Morris D. Loffman, MD
17173 Strawberry Dr.
Encino, CA 91436-0000

Rafael Longo-Cordero, MD
Univ. Gardens
Calle Rochester 911
San Juan, PR 00927-4812

Ralph C. Loomis, MD
Mountain Neurological Center
7 McDowell St.
Asheville, NC 28801-4103

Kenneth M. Louis, MD
Neurological Surgeons Associates
3000 E. Fletcher Ave., Ste. 340
Tampa, FL 33613-4656

Mark G. Luciano, MD, PhD
Cleveland Clinic Foundation
9500 Euclid Ave., S80
Cleveland, OH 44195-0001

Thomas G. Luerssen, MD
Riley Hospital for Children
One Children's Sq., Ste. 1730
Indianapolis, IN 46202-5200

Joseph R. Madsen, MD
Childrens Hosp./Brigham & Womens Hosp.
300 Longwood Ave., Rm. 312
Boston, MA 02115-5724

Gail A. Magid, MD
241 Fourth Ave.
Santa Cruz, CA 95062-3815

Gary Magram, MD
Inova Children's Hospital
8505 Arlington Blvd., Ste. 100
Fairfax, VA 22031-0000

Kim Herbert Manwaring, MD
Phoenix Children's Hospital
909 E. Brill St.
Phoenix, AZ 85006-0000

Timothy B. Mapstone, MD
Emory Clinic
1365-B Clifton Rd., N.E. #6400
Atlanta, GA 30322-1013

Arthur E. Marlin, MD
Pediatric Neurosurgery of South Texas
4499 Medical Dr., Ste. 397
San Antonio, TX 78229-3713

Todd A. Maugans, MD
University of Vermont
111 Colchester Ave., Fletcher 5
Burlington, VT 05401-0000

John R. Mawk, MD
Iowa Clinic Neurosurgical Services
411 Laurel St., Ste. 2350
Des Moines, IA 50314-0000

James P. McAllister, II, PhD
Wayne State University
4201 St. Antoine UHC-6E
Detroit, MI 48201-0000

Jack E. McCallum, MD
SW Neurological Surgery Associates PA
800 8th Ave., Ste. 200
Fort Worth, TX 76104-0000

J. Gordon McComb, MD
University Children's Medical Group
1300 N. Vermont Ave., #1006
Los Angeles, CA 90027-6005

C. Scott McLanahan, MD
Carolina Neurosurgery & Spine Assoc.
1010 Edgehill Rd. N.
Charlotte, NC 28207-1885

Robert L. McLaurin, MD
2412 Ingleside Ave., Apt. 5C
Cincinnati, OH 45206-2185

David Gordon McLone, MD, PhD
Children's Memorial Hospital
2300 Children's Plaza, Ste. 28
Chicago, IL 60614-3318

John Mealey, Jr., MD
9315 Spring Forest Dr.
Indianapolis, IN 46260-1269

Michael Dean Medlock, MD
Massachusetts General Hospital
55 Fruit St., WANG-021
Boston, MA 02115-0000

Arnold H. Menezes, MD
University of Iowa Hospitals
200 Hawkins Dr., Neurosurgery
Iowa City, IA 52242-1009

Glenn A. Meyer, MD
Medical College of Wisconsin
9200 W. Wisconsin Ave.
Milwaukee, WI 53226-3522

W. Jost Michelsen, MD
Box 6978
Portsmouth, NH 03801-0000

Thomas H. Milhorat, MD
North Shore University Hospital
300 Community Dr., Neurosurgery
Manhasset, NY 11030-0000

Clinton F. Miller, MD
Coastal New Hampshire Neurosurgery
330 Bortwick Ave., Ste. 108
Portsmouth, NH 03801-7110

John I. Miller, MD, FACS
544 E. 86th St., Ste. 6W
New York, NY 10028-7536

Mark A. Mittler, MD
Long Island Neurosurgical Associates
410 Lakeville Rd., Ste. 204
New Hyde Park, NY 11042-1103

Richard H. Moiel, MD
3656 Ella Lee Ln.
Houston, TX 77027-4105

Jose L. Montes, MD
Montreal Children's Hospital
2300 Tupper St., Rm. C811
Montreal, PQ H3H-1P3
Canada

German Montoya, MD
Orlando Neurosurgical Associates
1801 Cook Ave.
Orlando, FL 32806-2913

Thomas M. Moriarty, MD, PhD
Kosiar Children's Hospital
210 E. Gray St., Ste. 1102
Louisville, KY 40202-3907

Michon Morita, MD
1319 Punahou St., Ste. 999
Honolulu, HI 96826-1032

William J. Morris, MD
1112 6th Ave., Ste. 302
Tacoma, WA 98405-0000

Glenn Morrison, MD
Miami Children's Hospital
3200 S.W. 60 Ct., Ste. 301
Miami, FL 33155-4071

S. David Moss, MD
Phoenix Children's Hospital
909 E. Brill St.
Phoenix, AZ 85006-2513

Kamel F. Muakkassa, MD
Center for Neurosurgery & Spine
157 W. Cedar St., Ste. 203
Akron, OH 44307-2564

Michael S. Muhlbauer, MD
Semmes Murphey Clinic
6325 Humphreys Blvd.
Memphis, TN 38120-2300

Michael G. Muhonen, MD
455 S. Main St.
Orange, CA 92868-3835

Karin M. Muraszko, MD
University of Michigan/MC TC2128
1500 E. Medical Center Dr.
Ann Arbor, MI 48109-0338

Awani F. Musharbash, MD
PO Box 910262
Amman 11191
Jordan

Cheryl A. Muszynski, MD, FACS
Children's Hospital of Wisconsin
9000 W. Wisconsin Ave., PO Box 1997
Milwaukee, WI 53201-1997

S. Terence Myles, MD
Alberta Children's Hospital
1820 Richmond Rd. S.W.
Calgary, AB T2T-5C7
Canada

John S. Myseros, MD
Children's Hospital Medical Center
3333 Burnet Ave., Neurosurgery
Cincinnati, OH 45229-3026

Joseph M. Nadell, MD
Children's Hospital
200 Henry Clay Ave.
New Orleans, LA 70118-5720

Mahmoud G. Nagib, MD
800 E. 28th St., 305 Piper Bldg.
Minneapolis, MN 55407-3799

Michael F. Nido, PA-C
Carolina Neurosurgery & Spine Associates
1010 Edgehill Rd. N.
Charlotte, NC 28207-1885

Mark Stephen O'Brien, MD
1900 Century Blvd., Ste. 4
Atlanta, GA 30345-3307

W. Jerry Oakes, MD
Children's Hospital of Alabama
1600 7th Ave. S., ACC 400
Birmingham, AL 35233-1711

Miguel A. Pagan, PA-C
19136 Cypress View Dr.
Fort Myers, FL 33912-4825

2002 Membership Roster

Larry Keith Page, MD
13845 S.W. 73rd Ct.
Miami, FL 33158-1213

Dachling Pang, MD
Kaiser Permanete Hospital
2025 Morse Ave.
Sacramento, CA 95825-2115

Andrew D. Parent, MD
University of Mississippi Medical Center
2500 N. State St.
Jackson, MS 39216-4500

Tae Sung Park, MD
St. Louis Children's Hospital
1 Children's Pl., Ste. 4S20
St. Louis, MO, 63110-1002

Michael David Partington, MD, FACS, FA
Gillette Children's Specialty Healthcare
200 E. University Ave.
St. Paul, MN 55101-0000

Jogi V. Pattisapu, MD
Pediatric Neurosurgery PA
22 W. Lake Beauty Dr., Ste. 204
Orlando, FL 32806-2037

Jerry O. Penix, MD
928 Holladay Point
Virginia Beach, VA 23451

Joseph A. Petronio, MD
Gillette Children's Specialty Healthcare
200 E. University Ave.
St. Paul, MN 55101-0000

Joseph H. Piatt, Jr., MD
St. Christopher's Hospital for Children
Erie Ave. at Front St.
Philadelphia, PA 19134-1095

Prem Kumar Pillay, MD
Asian Brain-Spine-Nerve Center
3 Mt. Elizabeth #15-03
Singapore 228510

Hal Watson Pittman, MD
44 E. Palm Ln.
Phoenix, AZ 85004-1529

Thomas Pittman, MD
University of Kentucky Medical Center
800 Rose St., Rm. MS105-A
Lexington, KY 40536-0000

Ian F. Pollack, MD
Children's Hospital of Pittsburgh
3705 5th Ave., Neurosurgery
Pittsburgh, PA 15213-2524

Harold D. Portnoy, MD
Oakland Neurological Clinic PC
44555 Woodward Ave., Ste. 506
Pontiac, MI 48341-2982

Antonio R. Prats, MD
3661 S. Miami Ave., Ste. 401
Miami, FL 33133-0000

Mark R. Proctor, MD
Childrens Hosp./Brigham & Womens Hosp.
300 Longwood Ave., Bader 3
Boston, MA 02115-5724

Corey Raffel, MD, PhD
Mayo Clinic
200 1st St., Neurosurgery S.W.
Rochester, MN 55905-0001

John Ragheb, MD
Univ. of Miami/Neurosurgery LPLC
1095 N.W. 14th Ter. D4-6
Miami, FL 33136-2104

Donald H. Reigel, MD
5178 Polo Field Dr.
Gibsonia, PA 15044-0000

Harold Louis Rekate, MD
Barrow Neurological Institute
2910 N. 3rd Ave.
Phoenix, AZ 85013-4434

Theodore S. Roberts, MD
Children's Hospital Medical Center
4820 Sand Point Way N.E. CH-50
Seattle, WA 98105-0000

Shenandoah Robinson, MD
Rainbow Babies & Children's Hospital
11100 Euclid Ave., RBC B501
Cleveland, OH 44106-1736

Walker L. Robinson, MD
602 University Ave.
Urbana, IL 61801-2594

Luis A. Rodriguez, MD
Memorial Healthcare
1150 N. 35th Ave., Ste. 300
Hollywood, FL 33021-5424

Bruce R. Rosenblum, MD
Riverview Medical Center
160 Ave. at The Commons
Shrewsbury, NJ 07702-4802

Alan Rosenthal, MD
Long Island Neurosurgical Associates
410 Lakeville Rd., Ste. 204
New Hyde Park, NY 11042-1101

Allen S. Rothman, MD, FACS
175 Memorial Hwy.
New Rochelle, NY 10801-5640

John R. Ruge, MD
630 S. Oak St.
Hinsdale, IL 60521-4634

James T. Rutka, MD, PhD, FRC
Hospital for Sick Children
555 University Ave., #1504
Toronto, ON M5G-1X8
Canada

Petr O. Ruzicka, MD
Children's Hospital of New Jersey
741 Northfield Ave., Ste. 208
West Orange, NJ 07052-1104

Robert A. Sanford, MD
Semmes Murphey Clinic
6325 Humphreys Blvd.
Memphis, TN 38120-0000

Osamu Sato, MD
1-5-40 Tamagawa Gakuen
Machida
Tokyo 1940041
Japan

Guy M. Sava, MD
Mayo Health System
501 Holly Ln., Ste. 50
Mankato, MN 56001-6800

Timothy B. Scarff, MD
33 S.W. Upper Riverdale Rd., Ste. 25
Riverdale, GA 30274-2626

Steven J. Schiff, MD
George Mason University
Rockfish Creek Ln., MS 2A1
Fairfax, VA 22030-4444

Steven J. Schneider, MD
Long Island Neurosurgical Associates
410 Lakeville Rd., Ste. 204
New Hyde Park, NY 11042-1101

Luis Schut, MD
Children's Hospital of Philadelphia
34th & Civic Center Blvd.
Philadelphia, PA 19104-0000

R. Michael Scott, MD
Childrens/Brigham & Womens Hosp.
300 Longwood Ave., Bader 319
Boston, MA 02115-5724

Nathan R. W. Selden, PhD, MD
Oregon Health Sciences University
3181 S.W. Sam Jackson Park Rd., L472
Portland, OR 97201-0000

Wan Tew Seow, MD
National Neuroscience Institute
11 Jalan Tan Tock Seng
Singapore

Ronald F. Shallat, MD
33 Evergreen Dr.
Orinda, CA 94563-0000

Kenneth N. Shapiro, MD
Neurosurgeons for Children
1935 Motor St., 3rd Fl.
Dallas, TX 75235-7701

John Shillito, MD
1109 Fearington Post
6 Caswell Sq.
Pittsboro, NC 27312-5014

Howard J. Silberstein, MD
1445 Portland Ave., Ste. 304
Rochester, NY 14621-3008

James C. Simmons, MD
190 Grove Park Rd.
Memphis, TN 38117-0000

Gary Robert Simonds, MD
PO Box 14
237 Sunbury Rd.
Riverside, PA 17868-0014

Frederick H. Sklar, MD
Neurosurgeons for Children
1935 Motor St.
Dallas, TX 75235-7701

Harold P. Smith, MD
Neurosurgical Group of Nashville
300 20th Ave. N., Ste. 106
Nashville, TN 37203-2131

Lenwood P. Smith, Jr., MD
Palmetto Neurosurgery & Spine
3 Medical Park Rd., Ste. 310
Columbia, SC 29203-6873

Sandeep Sood, MD
Childrens Hospital of Michigan
3901 Beaubien, 2nd Fl.
Detroit, MI 48201-2119

Mark M. Souweidane, MD
New York Hospital/Star Pav.
520 E. 70th St., Rm. 651
New York, NY 10021-9800

Phillip G. St. Louis, MD
Florida Medical Plaza
2501 N. Orange Ave., Ste. 540N
Orlando, FL 32804-4603

Sherman Charles Stein, MD
310 Spruce St.
Philadelphia, PA 19106-4201

Paul Steinbok, MD
British Columbia Children's Hospital
4480 Oak St., Rm. A325
Vancouver, BC V6H-3V4
Canada

Bruce B. Storrs, MD
University of New Mexico
2211 Lomas N.E., ACC-2
Albuquerque, NM 87131-5341

Douglas L. Stringer, MD
2011 N. Harrison Ave.
Panama City, FL 32405-4545

Merle Preston Stringer, MD
2011 N. Harrison Ave.
Panama City, FL 32405-4545

Michael H. Sukoff, MD
17602 E. 17th St., Ste. 102-118
Tustin, CA 92780-0000

Peter P. Sun, MD
Children's Hospital of Oakland
744 52nd St., Neurosurgery
Oakland, CA 94609-1810

Anthony F. Susen, MD
193 Old Glebe Point Rd.
Burgess, VA 22432-2006

Leslie N. Sutton, MD
Children's Hospital of Philadelphia
34th & Civic Center Blvd.
Philadelphia, PA 19104-0000

Dale M. Swift, MD
Neurosurgeons for Children
1935 Motor St., 3rd Fl.
Dallas, TX 75235-7701

Michael S. Taekman, MD
15 Oakmont Ct.
San Rafael, CA 94901-1235

Tadanori Tomita, MD
Children's Memorial Hospital
2300 Children's Plaza, Ste. 28
Chicago, IL 60614-3363

Eric R. Trumble, MD
Pediatric Neurosurgery PA
22 W. Lake Beauty Dr., Ste. 204
Orlando, FL 32806-2033

Gerald F. Tuite, Jr., MD
880 6th St. S., Ste. 450
St. Petersburg, FL 33701-0000

Noel Tulipan, MD
8533 McCrory Ln.
Nashville, TN 37221-5905

Michael S. Turner, MD
Indianapolis Neurosurgical Group
1801 N. Senate Blvd., Ste. 535
Indianapolis, IN 46202-1228

David D. Udehn, MD
4350 7th St., Unit E
Moline, IL 61265-6870

Membership Roster

Ronald H. Uscinski, MD
Mid-Atlantic Brain & Spine Institutes
3301 Woodburn Rd., Ste. 209
Annandale, VA 22003-1229

Michael Vassilyadi, MD
Children's Hospital East Ontario
401 Smyth Rd.
Ottawa, ON K1H-8L1
Canada

Joan L. Venes, MD
1831 North Bend Dr.
Sacramento, CA 95835-1218

Enrique C. Ventureyra, MD
Children's Hospital East Ontario
401 Smyth Rd.
Ottawa, ON K1H-8L1
Canada

John Kenric Vries, MD
University of Pittsburgh
217 Victoria Bldg.
Pittsburgh, PA 15261-0001

Steven L. Wald, MD
97 Grove Ln.
Shelburne, VT 05482-0000

John B. Waldman, MD
Albany Medical College
47 New Scotland Ave., MC-61
Albany, NY 12208-0000

Marion L. Walker, MD
Primary Children's Medical Center
100 N. Medical Dr., Ste. 2400
Salt Lake City, UT 84113-1103

John Wilson Walsh, MD
Tulane University School of Medicine
1430 Tulane Ave., SL47
New Orleans, LA 70112-0000

John D. Ward, MD
Medical College of Virginia
Box 980631
Richmond, VA 23298-0631

Daryl E. Warder, MD, PhD
Wainwright Bldg.
229 W. Bute St., Ste. 230
Norfolk, VA 23510-0000

Benjamin C. Warf, MD
PO Box 903
Mbale
Uganda

Howard L. Weiner, MD
New York University Medical Center
317 E. 34th St., #1002
New York, NY 10016-4974

Martin H. Weiss, MD
LAC-USC Medical Center
1200 N. State St., Ste. 5046
Los Angeles, CA 90033-1029

Jean K. Wickersham, MD
3030 Children's Way, Ste. 402
San Diego, CA 92123-4228

Philip J. A. Willman, MD
1415 3rd St., Ste. 102
Corpus Christi, TX 78404-2107

Ronald J. Wilson, MD
Neurosurgical Specialists of Austin
901 S. Mopac Expy., Bldg. 5 #110
Austin, TX 78746-5776

Joel W. Winer, MD
York Neurosurgical Associates PC
2319 S. George St.
York, PA 17403-5009

Jeffrey A. Winfield, MD, PhD
1000 E. Genesee St., Ste. 602
Syracuse, NY 13210-0000

Ken R. Winston, MD
1056 E. 19th Ave., Box B330
Denver, CO 80218-1007

Jeffrey H. Wisoff, MD
New York University Medical Center
317 E. 34th St., #1002
New York, NY 10016-6402

Daniel Won, MD
Pediatric Neurosurgical Associates
301 Vanderbilt Way, Ste. 305
San Bernardino, CA 92408-3520

Meredith V. Woodward, MD
Valley Children's Hospital
9300 Valley Children's Pl.
Madera, CA 93638-8761

Shokei Yamada, MD
5410 Via San Jacinto
Riverside, CA 92506-0000

Karol Zakalik, MD
William Beaumont Hospital
3535 W. 13 Mile Rd., Ste. 504
Royal Oak, MI 48073-6710

Ahmad Zakeri, MD
4235 Secor Rd.
Toledo, OH 43623-4231

Edward J. Zampella, MD
PO Box 808
10 Parrott Mill Rd.
Chatham, NJ 07928-2744

Luis Manuel Zavaia, MD
555 Mowry Ave., Ste. A-B
Fremont, CA 94536-4101

John G. Zovickian, MD
3000 Colby St., Ste. 101
Berkeley, CA 94705-2058