PROGRAM BOOK

of the
Joint Section on
Pediatric Neurological Surgery
of
The American Association of
Neurological Surgeons
and
Congress of Neurological Surgeons

25th Annual Meeting

The Mills House Hotel
Charleston, South Carolina

December 10–13, 1996

Jointly Sponsored by
The American Association of Neurological Surgeons
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PAOLO RAIMONDI LECTURERS
1978 E. Bruce Hendrick
1979 Paul C. Bucy
1980 Floyd Gilles
1981 Panel Discussion
1982 Panel Discussion
1983 Derek Harwood-Nash
1984 Anthony E. Gallo, Jr.
1985 Frank Nulsen
1986 William F. Meacham
1987 Dale Johnson

1988 Joseph J. Volpe
1989 Martin Eichelberger
1990 George R. Leopold
1991 Judah Folkman
1992 Olof Fodmark
1993 Maurice Albin
1994 Blaise F.D. Bourgeois
1995 Robert H. Pudenz
1996 Samuel S. Flint

KENNETH SHULMAN AWARD RECIPIENTS
1984 Arno Fried: A Laboratory Model of Shunt-Dependent Hydrocephalus
1985 Anne Christine Duhaine: 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 Falcì: 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. Helfner: Basilar Pons Attracts Its Cortical Innervation by Chemotrophic 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. Welby: 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

HYDROCEPHALUS ASSOCIATION AWARD RECIPIENTS
1989 Eric Altschuler: Management of Persistent Ventriculomegaly Due To Altered Brain Compliance
1991 Nesher G. Asner: Venous Sinus Occlusion and Ventriculomegaly in Cranietomized 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. Welby-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

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

ANNUAL WINTER 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 & Pebble Beach
1991 Boston
1992 Vancouver, BC
1993 San Antonio
1994 St. Louis
1995 Pasadena
1996 Charleston
EXHIBITOR LISTING

The AANS/CNS Joint Section on Pediatric Neurological Surgery gratefully recognizes the support of the following exhibitors:

Aera-Cut, Inc. Booth 3 Medtronic PS Medical Booths 4 & 5
989 Main Street
Acion, MA 01720
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870 Market Street, #955
San Francisco, CA 94102
(415) 732-7040

KLS-Martin, L.B. Booth 21 Sofamor Danek
PO. Box 50269
Jacksonville, FL 32250
(904) 641-7746

Leica, Inc. Booths 18 & 19 Walter Lorenz Surgical, Inc. Booth 16
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Allendale, NJ 07401
(800) 526-0355

Litvinou, division of Zimmer Booth 22
1311 Concept Boulevard
Largo, FL 34643
(800) 235-5713

OFFICERS OF THE JOINT SECTION ON PEDIATRIC NEUROLOGICAL SURGERY

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Joseph H. Piatt
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Committee for Child Advocacy:
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Committee for Guidelines:
Thomas Luerssen
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PROGRAM OF THE JOINT SECTION ON PEDIATRIC NEUROLOGICAL SURGERY

25TH ANNUAL MEETING

Jointly Sponsored by The American Association of Neurological Surgeons

PROGRAM SCHEDULE

MONDAY, DECEMBER 9

1:00 PM–5:00 PM  Adjudication Committee of the Shunt Design Trial - Rice Room
6:00 PM–8:00 PM  Pediatric Fellowship Directors Dinner - Middleton Room

TUESDAY, DECEMBER 10

8:00 AM–4:00 PM  Pediatric Fellowship Directors Meeting - Middleton Room
9:00 AM–5:00 PM  Adjudication Committee of the Shunt Design Trial - Rice Room
12:00 NOON–5:00 PM  Nurses Seminar - Indigo Room
3:30 PM–5:30 PM  Pre-Executive Committee Meeting of the Pediatric Section - Indigo Room
4:30 PM–8:00 PM  Registration - Outside Signer's Ballroom
5:00 PM–6:00 PM  Endoscopic Shunt Insertion Trial - Middleton Room
6:00 PM–8:00 PM  Welcoming Reception - Signer's Ballroom

WEDNESDAY, DECEMBER 11

7:00 AM–4:00 PM  Registration - Outside Hibernian Hall (Upstairs)
7:00 AM–8:15 AM  Breakfast Seminar: Management of Traumatic Brain Injury in Children - Hibernian Hall (Upstairs)
7:00 AM - 8:30 AM  Continental Breakfast and Exhibits - Hibernian Hall (Downstairs)
8:30 AM - 10:00 AM  Scientific Session I - Hibernian Hall (Upstairs)

8:30 AM–8:40 AM  1. Decrease in Number of Cholinergic and GABAergic Neurons in the Neostriatum Kaolin-Induced Hydrocephalic Rat
Yuzuru Tashiro, MD, Toshiaki Hattori, PhD, Shushovan Chakrabortty, MD, James M. Drake, FRCS (Toronto, ON)

8:40 AM–8:50 AM  2. Changes in Cholinergic and Noradrenergic Neurons Before and After Shunting in the Rat with Kaolin-Induced Hydrocephalus
Yuzuru Tashiro, MD, Shushovan Chakrabortty, MD, James M. Drake, FRCS, Toshiaki Hattori, PhD (Toronto, ON)

8:50 AM–9:00 AM  3. Third Ventriculostomy as the Initial Treatment of Hydrocephalus: A Survey of Opinions
John R.W. Kestle, MD, Rob Alisharan, BSc, D. Douglas Cochrane, MD (Vancouver, BC)

9:00 AM–9:10 AM  4. 101 Ways To Do a Shunt: An Analysis of a Practice Survey on the Management of Shunted Hydrocephalus
Mark S. Dias, MD, Veetai Li, MD (Buffalo, NY)

9:10 AM–9:20 AM  5. Shunt Malfunction in 1996*
W. Jerry Oakes, MD, Bermans J. Iskandar, MD, Colleen A. McLaughlin, RN, Paul A. Grabb, MD, Timothy B. Mapstone, MD (Birmingham, AL)

9:20 AM–9:30 AM  6. CSF Parameters Following a Reservoir Tap in Low Birth Weight Infants with IVH (Intraventricular Hemorrhage)/PHH (Posthemorrhagic Hydrocephalus): When to Convert a Permanent Shunt
Yoon S. Hahn, MD, FACS, FAAP (Oak Lawn, IL)

9:30 AM–9:40 AM  7. CSF Shunt Valve Performance Monitored Telemetrically in Patients with Shunted Hydrocephalus
David M. Prim, MD, PhD, Denise Fleig, RN, Patricia Reidy, RN, Liliana C. Goumnerova, MD (Chicago, IL)

9:40 AM–10:00 AM  Questions
10:00 AM–10:30 AM  Coffee Break - View Exhibits - Hibernian Hall (Downstairs)
10:30 AM–11:15 AM  Paolo Raimondi Lecturer
Needs of a Child in a Changing Healthcare Market
Samuel S. Flint, PhD
11:15 AM–12:30 PM Scientific Session II – Hibernian Hall (Upstairs) 
Moderator: James Rutka

11:15 AM–11:25 AM
8. The Effect of Shunt Treatment on Cortical Water and Electrolyte Changes in Hydrocephalic Infant Rats
   Hazel C. Jones, PhD, Robert W. Andersohn (Gainesville, FL)

11:25 AM–11:35 AM
9. Reactive Astrocytosis in a New Model of Obstructive Hydrocephalus*
   Martha J. Johnson, Mark G. Luciano, Igor Ayzman, Arcangelia S. Wood, J. James McAllister II (Cleveland, OH)

11:35 AM–11:45 AM
10. Cranial Expansion for slit Ventricles Syndrome with High ICP
    Ronald Young II, MD, Mohamed El-nabity, MD, Charles Teo, MD, Frederick A. Boop, MD (Little Rock, AR)

11:45 AM–11:55 AM
11. Differentiation Between Cortical Atrophy and Hydrocephalus Using 1H MRS
    Stefan Blum, MD, J. Gordon McComb, MD, Brian D. Ross, MD (Los Angeles, CA)

11:55 AM–12:05 PM
12. Endoscopic Retrieval of Retained Peritoneal Catheters
    S. David Moss, MD, Kim H. Manwaring, MD, Harold Rekate, MD (Phoenix, AZ)

12:05 PM–12:15 PM
13. Shunt Malfunction Associated with CSF Eosinophilia*
    Robin M. Bowman, MD, Joel C. Boaz, MD, Thomas G. Lahteen, MD, John E. Kalsbeck, MD (Indianapolis, IN)

12:15 PM–12:25 PM
14. MRI Assessment of Third Ventriculostomy
    Mark M. Soueidane, Christopher G. Gaposchkin, Linda Heier (New York, NY)

12:25 PM–12:35 PM
15. Propionibacterium Acnes Infections of Cerebrospinal Fluid Shunts*
    Ken S. Sato, MD, Joel C. Boaz, MD, Thomas G. Lahteen, MD, Martin Kleiman, MD, John E. Kalsbeck, MD (Indianapolis, IN)

*Considered for Shulman Award.
24. Syringomyelia of the Distal Spinal Cord in Children*
   Hal Meltzer, MD, Hector E. James, MD (San Diego, CA)

2:50 PM–3:00 PM
Questions

3:00 PM–3:30 PM
Coffee Break – View Exhibits – Hibernian Hall (Downstairs)

3:30 PM–5:30 PM
Scientific Session IV – Hibernian Hall (Upstairs)
Moderator: Mark Iatrosca

3:30 PM–3:40 PM

25. Urgent Radiation Therapy for Pineal Region Malignant Teratomas
   E. Christopher Troup, MD, Robert A. Sanford, MD, Larry Kyn, MD,
   Michael S. Muhlbaeur, MD, Jennifer Eldred, RN (Memphis, TN)

3:40 PM–3:50 PM

26. Superficial Neuroepithelial Tumors of Infancy and Childhood
   Richard Ellenbogen, Hernando Mena, Leslie Sutton, Lucy Rourke,
   Rocco Armonda, Ann-Christine Duhaime, Luis Schut (Washington, DC)

3:50 PM–4:00 PM

27. Anomalous Expression of Cell Cycle Regulatory Proteins in Pediatric
    Brain Tumors
   Joseph Petronio, MD, C. David James, PhD, Jing Ye, MD, Ju He, MD,
   James R. Allen, PhD (Atlanta, GA)

4:00 PM–4:10 PM

28. Pediatric Low Grade Astrocytoma – Results of Surgery and Multifactorial
    Analysis in Tumors Distributed Throughout the Entire Neuraxis
   Robert A. Sanford, Amar Gajjar, Michael S. Muhlbaeur, Larry E. Kun,
   E. Christopher Troup (Memphis, TN)

4:10 PM–4:20 PM

29. Platelet-Derived Growth Factor Induces Differentiation of Neuroepithelial
    Stem Cells Into Neurons*
   John K. Park, MD, PhD, Brenda R. Williams, PhD, Charles D. Stiles, PhD
   (Boston, MA)

4:20 PM–4:30 PM

30. The Relationship Between TP53 Mutations and Overexpression of
    p53 and Prognosis in Malignant Gliomas of Childhood
   Ian Pollack, Ronald Hamilton, Sydney Finkelstein, Jeffrey Campbell, A.
   Julio Martine, Richard Sherwin, Michael Bozik, Susanne Gallin
   (Pittsburgh, PA)

*Considered for Shulman Award.
35. A Randomized Clinical Trial to Compare Selective Posterior Rhizotomy Plus Physiotherapy with Physiotherapy Alone in Children with Spastic Diplegic Cerebral Palsy

36. Changes in Spasticity and Strength as a Result of Selective Dorsal Rhizotomy
Jack Engsberg, Kenneth Oltec, Sandy Ross, T. S. Park (St. Louis, MO)

37. Chronic Intrathecal Baclofen for Spasticity of Cerebral Origin
Jeffrey W. Campbell, A. Leland Albright, Margaret J. Barry (Pittsburgh, PA)

38. Effect on Ambulation of Intrathecal Baclofen Infusion for Lower Extremity Spasticity*
Peter C. Gerszten, MD, A. Leland Albright, MD (Pittsburgh, PA)

39. Continuous Intrathecal Baclofen for Generalized Dystonia
Jeffrey W. Campbell, A. Leland Albright, Margaret J. Barry, Barbara Shultz (Pittsburgh, PA)

40. Selective Dorsal Rhizotomy: Morbidity and Mortality
Jeffrey W. Campbell, A. Leland Albright (Pittsburgh, PA)

41. Image-Guided Applications in the Surgical Treatment of Epilepsy in Children
Roger H. Frankel, MD, Joseph Petronio, MD, Christine Narad, MSN, Roy A.E. Bakay, MD (Atlanta, GA)

42. Ultrasound Guided Modified Hemispherectomy for Seizure Control in Children
Paul Kanev, MD, Catherine Foley, MD, Dan Mikes, MD (Detroit, MI)

43. Multiple Subpial Transection: Application to Pediatric Epilepsies with Multifocal Onset*
Richard Kim, MD, MS, Rick Abbott, MD (New York, NY)

44. Medial Temporal Lobe Resections in Childhood Epilepsy

45. A Technique Allowing Awake Craniotomy in Children*
Michael H. Handler, MD, Geoffrey Lane, MB, FRCA, Rita Agarwal, MD, FAAP

46. Dysembryoplastic Neuroepithelial Tumor (DNET) as a Cause of Medically Intractable Epilepsy in Children
Antonio R. Prats, MD, Nolan Altman, MD, Carole D. Brainhaite, MD, Prasanna Jayakar, MD Raquel Prast, MSN, ARNP (Miami, FL)

47. Polymorphonuclear Leukocytes Exacerbate and Oxypurinol or Glutathione Protect Against Traumatic Injury in Cultured Cerebral Endothelial Cells
T. S. Park, Joel W. Beetsch, Jeffrey M. Giddand (St. Louis, MO)

48. Admissions for Uncomplicated Skull Fractures: Are They Necessary?
Michael A. Vogelbaum, Bruce A. Kaufman, T. S. Park (St. Louis, MO)

*Considered for Shulman Award.
11:50 AM–12:00 NOON
50. Piglet Brain Contusion: Does Maturaton Matter?
   A. C. Duhaime, R. Ragupathi, D. Meaney, D. Smith, T. McIntosh
   (Philadelphia, PA)

12:00 NOON–12:10 PM
51. Estimation of IQ Score Reduction Following Head Injury in Children*
    Mark R. Iantosca, MD, J. Gregory Javornisky, PhD (Hartford, CT)

12:10 PM–12:20 PM
52. Intracranial Pressure Monitoring After Severe Head Injury in Children: The Richmond Bolt Versus the Camino Catheter*
    James T. Rutka, MD, John S. Myserson, MD, Desmond Bohn, MD,
    Peter Cox, MD, Paul Chumas, MD (Toronto, ON)

12:20 PM–12:30 PM
53. Presentation, Treatment, and Outcome in a Large Series of Pediatric Patients with Aneurysmal Bone Cysts*
    Eric R. Trumble, MD, Frederick Sklar, MD (Dallas, TX)

12:30 PM–12:40 PM
54. Upper Cervical Spine Fusion in the Pediatric Population*
    David W. Lowry, Ian F. Pollack, Brent Clyde, P. David Adelson,
    A. Leland Albright (Pittsburgh, PA)

12:40 PM–12:50 PM
55. Pediatric Cl-2 Transarticular Screw Fixation for Atlanto-Occipital and Atlanto-Axial Instability
    Douglas Brockmeyer, MD, Ronald Apfelbaum, MD

12:50 PM–1:00 PM
Questions
1:00 PM–1:30 PM
Coffee Break – View Exhibits – Hibernian Hall
   (Downstairs)

1:00 PM–2:00 PM
North American Hydrocephalus Research Society – Indigo Room

1:00 PM–2:00 PM
Multi-Center Rhizotomy Study Group – Rice Room
1:00 PM–2:30 PM
Neurosurgery On-Call – Robert E. Lee Room
5:00 PM–6:00 PM
Carriage Tour to the Exchange
6:00 PM–7:00 PM
Reception – The Exchange
7:00 PM–7:30 PM
Bus Ride to the Mills House
7:30 PM–11:30 PM
Dinner – Hibernian Hall (Upstairs)

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**FRIDAY, DECEMBER 13**

7:00 AM–11:00 AM
7:00 AM–8:15 AM
Registration – Outside Hibernian Hall (Upstairs)
Breakfast Seminar: Communication in Pediatric Neurosurgery
   Faculty: Robin Humphreys, MD
            John Kestle, MD
            Harold Rekate, MD

7:00 AM–8:30 AM
Continental Breakfast and Exhibits – Hibernian Hall
   (Downstairs)
Scientific Session VII – Hibernian Hall (Upstairs)
   Moderator: Joseph Madsen

8:30 AM–8:40 AM
56. Virchow Among the Craniolestics: His Contribution to the Problem of Craniosynostosis
    Cary D. Alberstone, MD (Albuquerque, NM)

8:40 AM–8:50 AM
57. Occipital Plagiocephaly: Standards for Treatment
    Benjamin S. Carson, Carol S. James, Craig A. VanderKolk (Baltimore, MD)

8:50 AM–9:00 AM
58. Occipital Flattening: Indications for Intervention and Techniques for Repair
    Richard Polin, John Jane, Kant Lin (Charlottesville, VA)

9:00 AM–9:10 AM
59. Surgical Management of Sagittal Synostosis: A Comparative Outcome Analysis*
    Todd A. Maugans, MD, J. Gordon McComb, MD, Michael Levy, MD
    (Los Angeles, CA)

9:10 AM–9:20 AM
60. Endoscopic Cranietomy for Release of Stenosed Cranial Sutures
    David P. Jimenez, MD, Constance Barone, MD, Anthony Nobles
    (Columbia, MO)

9:20 AM–9:30 AM
61. The Transpalatal Approach for the Treatment of Infantile Transphenoidal Encephaloceles in Four Patients
    Kerry R. Crone, David P. Gruber, Erin M. Kennedy, David A. Billmire
    (Cincinnati, OH)

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*Considered for Shulman Award.
10:40 AM–10:50 AM
63. Treatment of Non-Communicating Hydrocephalus with
Ventriculoscopie Third Ventriculocisternostomy: Outcome and CSF
Flow Patterns
Liliana Goumnerova, MD, David Frim, MD, PhD, Sandra Rufo
(Boston, MA)

10:50 AM–11:00 AM
64. Complications of Neuroendoscopic Third Ventriculostomy
Steven J. Schneider, MD (New Hyde Park, NY)

11:00 AM–11:10 AM
65. Scopeless Endoscopy: The Use of Virtual Operating Environments in
the Ventricular Chamber*
Terri L. Harphold, MD, Michael L.J. Apuzzo, MD, J. Gordon McComb, MD,
Arun Amar, MD, SooHo Choi, MD, Michael L. Levy, MD (Los Angeles, CA)

11:10 AM–11:20 AM
66. Endoscopic Third Ventriculostomy and the Slit Ventricle Syndrome
Jonathan J. Baskin, MD, Kim H. Manwaring, MD, Harold L. Rekate, MD
(Phoenix, AZ)

11:20 AM–11:30 AM
67. Stereolithography and the Evaluation of Computer Image Based
Modeling in the Peri-Operative Management of Patients with Cerebral
Vascular Anomalies and Neoplasms*
Michael L. Levy, MD, Terry L. Harphold, MD, Joseph Chen, MD,
J. Gordon McComb, MD (Los Angeles, CA)

11:30 AM–11:40 AM
Questions

11:40 AM–11:50 AM
68. Surgery of Arachnoid Cysts Involving the Third Ventricle*
SooHo Choi, MD (Los Angeles, CA)

*Considered for Shulman Award.
1. A New Model of Human Medulloblastoma in the Nude Mouse
   Daniel R. LeMay, MD, PhD, Toby MacDonald, PhD, Ronnie I. Mimran, BS, Keith A. Hurvitz, BS, J. Gordon McComb, MD, Martin H. Weiss, MD, Berislav V. Zlokovic, MD, PhD (Los Angeles, CA)

2. Cerebellar Astrocytoma in Pediatric vs. Adult Patients
   Vittorio Morreale, Michael J. Ebersold, Lynn M. Quast, Joseph E. Parisi (Rochester, MN)

3. Should Age Be the Deciding Factor in Determining the Need for Arteriography to Exclude Aneurysm in Children With Oculomotor Palsy?
   Mark S. Dias, MD, Imtiaz A. Mehta, MD, Steven Awner, MD, Scott E. Olitsky, MD (Buffalo, NY)

4. Subdural Fluid as an Indication of Hydrocephalus
   Michael H. Handler, MD (Denver, CO)

5. Cerebral Gnathostomiasis
   John R. Mawak, MD (Portland, OR)

6. Intraventricular Urokinase For the Treatment of Posthemorrhagic Hydrocephalus: Does a Fibrinolytic State Prevent Need for Shunting?
   Joseph Madsen (Boston, MA)

7. A Novel Method of Cranioplasty Using Coralline Hydroxyapatite*
   SooHo Choi, MD, Michael L. Levy, MD, J. Gordon McComb, MD (Los Angeles)

8. Prevention of Spinal Cord Retethering Using Free Dermal Fat Grafts*
   Michael R. Egnor, MD, Mark D. Epstein, MD (Stony Brook, NY)

9. The Relationship Between Intraoperative Hypothermia and Ventricular Shunt Infections
   Peter C. Gerszten, MD, A. Leland Albright, MD, Ian E. Pollack, MD, P. David Adelson, MD (Pittsburgh, PA)

10. Use of the ISG Wand in the Posterior Fossa
    Rick Abbott, MD (New York, NY)

*Considered for Shulman Award.
Decrease in Number of Cholinergic and GABAergic Neurons in the Neostriatum of Kaolin-Induced Hydrocephalic Rat

Yuzuru Tashiro, MD, Toshiaki Hattori, PhD, Shushovan Chakraborty, MD, James M. Drake, FRCSC (Toronto, Ontario, Canada)

The basal ganglia, particularly the neostriatum, has an important role in the initiation and control of voluntary movement. Structural and/or functional injury in the neostriatum can lead to spasticity, abnormal gait and posture, and impaired cognition, abnormalities also frequently seen in hydrocephalus. Having previously demonstrated a reduction of dopaminergic immunoreactivity in the neostriatum in the progress of hydrocephalus, the present study was designed to investigate neostriatal cholinergic interneurons and GABAergic projection neurons.

Hydrocephalus was induced in 12 Wistar rats by intracisternal injection of 0.05ml volume of 25% kaolin solution under microscopic guidance. Four controls received an equal volume of normal saline. The animals were killed 2, 4 and 8 weeks after injection. The numbers of immunoreactive (IR) neostriatal neurons to Choline Acetyltransferase (ChAT) and glutamic acid decarboxylase (GAD), were counted in 60um-thick representative sections. The total number of all neostriatal neurons was also counted in 15um-thick sections stained by cresyl violet (Nissl stain).

The numbers of neostriatal ChAT-IR neurons was significantly and progressively reduced at 2, 4, and 8 weeks after the onset of hydrocephalus compared to controls (p<.05); GAD-IR neurons were dramatically decreased at 4 and 8 weeks (p<.05). There was a linear correlation between reduction in neuronal number and ventricular enlargement for ChAT-IR (p<.001), GAD-IR (p<.001) and total neostriatal neurons (p<.001). These findings indicate that progressive hydrocephalus results in functional impairments of neostriatal specific neurons. This may explain some of the motor and intellectual impairment occurring in this condition.

This project was supported by the Hydrocephalus Research Foundation.
Changes in Cholinergic and Noradrenergic Neurons Before and After Shunting in the Rat with Kaolin-Induced Hydrocephalus

Yuzuru Tashiro, MD, Shushovan Chakraborty, MD, James M. Drake, FRCSC, Toshiaki Hattori, PhD (Toronto, Ontario, Canada)

Loss of intellectual capacity with hydrocephalus has often been attributed to non-specific causes including white matter injury from ventricular dilatation, the etiological cause of hydrocephalus, etc. More recent investigations have demonstrated neuronal impairment in hydrocephalus implicating neuronal systems in the pathogenesis of this disorder. Two brain regions well established in intellectual function include the cholinergic basal forebrain nuclei (learning and memory), and the noradrenergic Locus Ceruleus (LC) (selective attention and vigilance). In this study we investigated the changes with hydrocephalus in these regions, as well as the response to CSF shunting, using immunohistochemistry.

Hydrocephalus was induced in 20 Wistar rats by intracisternal injection of 0.05ml volume of 25% kaolin solution under microscopic guidance. Four controls received an equal volume of normal saline. Ventriculoperitoneal shunts were inserted at 2 and 4 weeks after kaolin injection (shunted group). Non-shunted hydrocephalic animals were killed after 2, 4 and 8 weeks survival: shunted animals were killed at 8 weeks following injection. The morphology of immunoreactive (IR) neurons to Choline Acetyltransferase (ChAT) in cholinergic nuclei Ch1-6, and Tyrosine Hydroxylase (TH) in the LC, were compared between shunted and non-shunted groups, and the number of ChAT-IR neurons before and after shunting was counted in representative sections with 60um thickness.

The number and size of ChAT-IR neurons was progressively reduced in the process of ventricular dilatation in Ch1, 2, 3 and 4 (p < .005). Early (2 weeks), but not late (4 weeks), shunting restored ChAT immunoreactivity to control levels (p < .05). The TH-IR neurons in LC were remarkably compressed by dilated fourth ventricle. CSF shunting also restored TH immunoreactivity in LC.

These findings suggest that a progressive functional impairment occurs in the cholinergic and noradrenergic neuronal systems in the process of hydrocephalus and may contribute to intellectual impairment. Early shunting may prevent this functional impairment.

This project was supported by the Hydrocephalus Research Foundation.

Third Ventriculostomy as the Initial Treatment of Hydrocephalus: A Survey of Opinions

John R.W. Kestle, MD, Rob Alisharan, Bsc, D. Douglas Cochrane, MD (Vancouver, BC)

Third ventriculostomy is now performed in a number of conditions which have traditionally been treated with a VP shunt. A survey has been conducted to determine 1) whether the rate of Illostomy varies among surgeons, and 2) the factors that influence surgeons' decision to do a Illostomy. The survey was distributed by email and was available at the 1996 ISPAG meeting. To date, 41 responses have been received (32 North American, 6 European, 3 SE Asian). The proportion of new hydrocephalus patients treated with Illostomy varied widely (0-100%, mean 22%). Factors that increase the chance of a Illostomy: triventricular hydro on CT/MR (36/37), cerebellar hemisphere tumour (21/36), isolated aqueduct stenosis (35/37), thin ballonned floor (33/37), tectal tumour (34/37).

Factors that decrease the chance of a Illostomy: age <1 year (18/37), dilated subarachnoid spaces (22/37), MMC (24/37), presence of another brain malformation (19/37), meningitis (35/37), head injury (27/37). The areas of most disagreement were: pontine glioma (15/37 said this would increase the chance of them doing a Illostomy, 16/37 said decrease), 3rd/4th ventricle tumours. Pre-op imaging: most require MR (32/37). Technique: balloon dilation is most common (22/37)., hole size 2 - 8.5 mm (mean = 4.31), 23/37 penetrate arachnoid in interpeduncular cistern. Estimated success rate 68.9% (range 30-96%). If the Illostomy falls, 28/37 do shunt, 5 attempt a repeat Illostomy.

Conclusions: Variation in the rate of Illostomy as the first treatment for hydrocephalus is large. It is unlikely that this degree of variation can be explained by differences in practice patterns. Further work to refine the indications for Illostomy is warranted.
4 101 Ways to Do a Shunt: An Analysis of a Practice Survey on the Management of Shunted Hydrocephalus

Mark S. Dias, MD, Veeai Li, MD (Buffalo, NY)

Surgery for shunted hydrocephalus is the most common, and perhaps the most varied, pediatric neurosurgical procedure. In an effort to better understand how varied these practices are, we surveyed 261 neurosurgeons with an interested interest in pediatric neurosurgery of which 149 (57%) responded. The data presented reflects the distribution of answers expressed as a percentage of the number of responses to that particular question.

Most (90%) shave hair before surgery, and the majority shave along the entire course of the shunt. Prophylactic intravenous antibiotics are used by 97%, but only 28% utilized intraventricular antibiotics. The preferred approach for the proximal catheter was occipital (40%) and frontal (30%); very few routinely use endoscopic guidance. The peritoneal was the overwhelming site for the distal catheter. A medium pressure valve without an anti-siphon device was the preferred valve (41%).

The management of infected shunts varied, based upon the causal organism; the survey included inquiries about treatment plans for 8 common organisms. Intravenous antibiotics were the mainstay of treatment, and were used as the sole treatment by 20% and 24% for pneumococcal and H. influenzae, respectively. Intraventricular antibiotics were added by 12-38%, most commonly for gram-negative or fungal infections. Complete shunt removal, insertion of a ventriculostomy and subsequent reinsertion of a shunt was the favored surgical option (62-90%) whereas removal and replacement of the shunt at the same time was proposed by 5-18%.

Routine post-operative studies included computed tomographic (CT) scans (80%) and shunt continuity studies (35%). Subsequent CT and shunt continuity studies were obtained only in the presence of symptoms or signs by 47% and 70%, respectively. In rank order, the most important variables for diagnosing a shunt malfunction were clinical symptoms, increased ventricular size, and elevated pressure on a shunt tap.

These results indicate there is a wide variation in the management of shunted hydrocephalus.

5 Shunt Malfunction in 1996*

W. Jerry Oakes, MD, Bermans J. Islandar, MD, Colleen A. McLaughlin, RN, Paul A. Grabb, MD, Timothy B. Mapstone, MD (Birmingham, AL)

Shunt malfunction is one of the most common clinical problems in pediatric neurosurgery. The diagnosis can be both difficult and perplexing even for the experienced clinician. We evaluated all shunt revisions seen at our institution from January-July 1996. Sixty-eight patients underwent 100 operations for shunt malfunctions in this seven month time interval. The criteria for recommending surgery was evidence of raised intracranial pressure and/or headache. All operated patients had evidence of shunt blockage, disconnection, catheter malposition or value-pressure incompatibility. The prospective radiographic interpretation of preoperative scans (CT or MRI) was reviewed in each case. In the majority shunt failure was diagnosed or suggested. Evidence of subgaleal fluid was considered positive evidence for shunt failure even if malfunction was not diagnosed in the radiographic report. Twenty-seven percent of the reports made no mention of shunt malfunction or failure. In this group, the ventricular systems were described as “smaller, small”, “unchanged”, “negative” or “stable” with no other comment to support a diagnosis of shunt malfunction or raised intracranial pressure. In all patients within this group clinical symptoms improved following surgery. We conclude that as many as a quarter of patients presenting with shunt malfunction will not have the diagnosis supported by a prospective radiographic interpretation of brain imaging. Although the neurosurgical community can easily assess the clinical situation to determine the need for operation, other clinicians could easily be reassured by a radiographic report which does not mention or diagnosis shunt malfunction. Today more than ever non-neurosurgeons are called upon by health maintenance organizations to assess complex clinical situations and may rely too heavily on radiographic findings.
6 CSF Parameters Following a Reservoir Tap in Low Birth Weight Infants with IVH (Intraventricular Hemorrhage)/PHH (Posthemorrhagic Hydrocephalus): When to Convert to a Permanent Shunt

Yoon S. Hahn, MD, FACS, PAAP (Oak Lawn, IL)

The treatment of preemies with IVH remains a major problem particularly when birth weight is less than 1500gm. Current management includes treatment with acetazolamide or furosemide for temporary measures or lumbar punctures. Early shunting surgery is not recommended because of elevated RBCs and protein and the low birth weight. Subcutaneous reservoir is useful to tap and drain bloody CSF from the ventricle until the shunt is deemed necessary.

Questions are raised as to the optimum time to of shunt internalization. Factors for consideration include the severity of IVH/PHH, CSF chemistry and response to tapping. To answer the question, a prospective study was conducted in 68 consecutive preemies from 1988 to 1994 under strict protocol (to be presented). Patient data was collected including gestational age (23-28 weeks), body weight (550gm to 2740gm), severity of IVH/PHH, cumulative data on numbers of RBCs and protein from the "daily" taps until the shunt is internalized.

The following interim observations were obtained:
1. Initial RBCs of CSF ranged from 244 to 2 million and ranged from 0-34,200 at the time the shunt was internalized. Initial CSF protein ranged from 137-3600 and 54-844 at the time of the shunt surgery.
2. 8.8% (6/68 neonates) never required a permanent shunt and reservoir removed.
3. 22% (15/68) required shunt revisions in 2-8 years.
4. 13.2% (9/68) shunt infection occurred due to either staph epidermidis, Group B streptococcus, corynebacterium or pseudomonas.
5. The average number of shunt revisions was 2-3 per patient.
6. Based on these data, a treatment protocol will be presented.

7 CSF Shunt Valve Performance Monitored Telemetrically in Patients with Shunted Hydrocephalus

David M. Frim, MD, PhD, Denise Fleig, RN (Chicago, IL), Patricia Reidy, RN, Liliana C. Gourmerova, MD (Boston, MA)

Commercial availability of different types of shunt has raised controversy over the most appropriate shunt valve for hydrocephalic patients. Though their performance characteristics are well documented in the laboratory setting, there is little data that describes the in vivo dynamics of intraventricular pressure (IVP) after implantation of commonly used shunt systems in humans. We have had opportunity to couple telemonitoring devices (TeleSensor, Radiotronics) to different shunt systems (PS Medical flow-control valve, Delta valve, and siphon-control device, Cordis Hakim and Orbis-Sigma valves, Codman-Medos programmable Hakim valve, Heyer-Schulte low-profile valve- LPV) and to a shunt system implanted into the internal jugular vein against the direction of flow (El-Shaffey shunt). In this fashion we have measured the performance characteristics of these valve systems with respect to IVP at increments of head elevation and compared these pressures to monitored but unshunted patients. Eleven patients were implanted with 14 different shunt systems and telemetrically monitored for IVP at 0°, 15°, 30°, 45°, 60°, and 90° of head elevation. Two patients implanted with TeleSensors to measure IVP in situations other than hydrocephalus served as unshunted controls. We found that shunt function as a reflection of IVP fell into two distinct groups: shunts which caused a precipitous drop in IVP with minimal head elevation (PS medical flow control, Cordis Hakim, Codman-Medos, Heyer-Schulte LPV) and shunts which caused a gradual decline in IVP with head elevation that resembled the pressures seen in unshunted patients (PS Medical Delta valve and siphon control device, Cordis Orbis-Sigma, El-Shaffey). Antisiphon devices were frequently associated with IVP remaining above 0 even at 90° of head elevation. These data provide a basis for evaluating shunt valve performance and for predicting valve appropriateness where characteristics such as pressure dynamics, flow control, and cost are all weighed in the choice of a specific valve for implantation.
The Effect of Shunt Treatment on Cortical Water and Electrolyte Changes in Hydrocephalic Infant Rats

Hazel C. Jones, PhD, Robert W. Anderson (Gainesville, FL)

The H-Tx rat has inherited hydrocephalus caused by aqueduct stenosis in late gestation. There is rapid postnatal dilatation of the lateral ventricles and severe cortical thinning. Previously we have shown that rats with advanced hydrocephalus at 21 days after birth, have reduced cortical levels of many organic osmotics, indicative of substrate loss, decreased intracellular contents and cell damage. These changes are reversible with early (4-day) but not later (11-day) shunt treatment. To investigate this further, we have examined cortical water, Na, and K content at 21 days in hydrocephalic rats and in rats with ventriculosubcutaneous shunts placed at 4 or 11 days. The results were compared with cerebellar tissue and with tissue from age-matched control littersmates (n = 6-8 per group). In untreated rats, cortical H2O (ml/kg wet wt.) increased by 1.4%, and H2O (ml/kg dry wt.) increased by 8.4%. Wet weight concentration (mEq/kg wet wt.) of Na increased by 26.7% indicating a substantial increase in extracellular fluid. There was a small but significant decrease in dry weight concentration (mEq/kg dry wt.) of K (-6.0%), indicating a decrease in intracellular mass. Cerebellar tissue had small increases in H2O and Na but no change in K. The significant increases in H2O and Na were prevented by shunt-treatment but the decrease in K was unchanged in the 11-day shunt (-6.3%) and only partially avoided in the 4-day shunt (-3.6%). It is concluded that shunt treatment corrected the extracellular edema that occurs in hydrocephalus, but that the intracellular changes are not easily reversed.

Reactive Astrocytosis in a New Model of Obstructive Hydrocephalus

Martha J. Johnson, Mark G. Luciano, Igor Ayzman, Arcangela S. Wood, James P. McAllister II (Cleveland, OH)

In order to characterize histopathologic changes during the progression of ventriculomegaly, and to allow subsequent correlation with functional imaging and analysis of the efficacy of surgical treatments, we have developed a canine model of obstructive hydrocephalus.

Acute procedures on 13 animals allowed determination of location and size of the obstruction immediately after surgery. In 17 chronic animals, hydrocephalus was induced and the progression of ventriculomegaly was monitored using MRI. Hydrocephalus was induced by injecting cyanoacrylic glue into the fourth ventricle. Animals were monitored post-operatively for neurological changes and were scanned using MRI to measure progression of ventricular enlargement. At 5-10 weeks post-induction, animals were perfused with fixative and tissue from sensorimotor cortex was processed for Nissl staining or immunohistochemical analysis of glial fibrillary acidic protein (GFAP), an accepted marker of neurologic injury.

Hydrocephalus was successfully induced in 9 of 17 (53%) of the chronic animals. The severity of ventricular enlargement was determined by comparison of Evans' ratios. Nissl staining failed to reveal any apparent changes in the white matter of hydrocephalic animals relative to acute surgical controls. Gray matter regions stained for Nissl substance revealed mild disorientation of neurons. However, GFAP immunoreactivity in both gray and white matter was altered in hydrocephalic tissue relative to acute surgical controls and as a function of the degree of ventricular enlargement. Animals with moderate to severe hydrocephalus exhibited qualitative increases in the number of GFAP positive astrocytes. Morphological changes were manifested by increased astrocyte somal size, process thickness, number, and length. Apparent increases in the number of vascular endfeet were also observed in reactive astrocytes present in hydrocephalic brains.

Increases in GFAP content in hydrocephalic tissues relative to controls strongly suggest a process of traumatic brain injury as a function of ventricular size. The detected change may either reflect a secondary effect of impaired neuronal function, or it may be a cellular response to direct injury of astrocytes. In contrast to routine Nissl staining, immunohistochemical labeling of GFAP appears to be a more sensitive indicator of histopathology following chronic hydrocephalus. In order to further understand the consequences of histopathological change, the occurrence of reactive astrocytosis must be correlated with neurophysiological deficits, intracranial pressure, and brain compliance. The large animal model of obstructive hydrocephalus developed during this study will enable subsequent examinations of the temporal progression of pathophysiology during ventricular enlargement, and the efficacy of clinical intervention in reversing this process.

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10 Cranial Expansion for Slit Ventricle Syndrome with High ICP

Ronald Young II, MD, Frederick A. Boop, MD, Mohamed Elnabity, MD, Charles Teo, MD (Little Rock, AR)

Chronic headaches in shunt dependent children with slit-like ventricles is a common and difficult treatment problem in pediatric neurosurgery. Rekate has outlined five different classes of patients with slit-ventricle syndrome (SVS) according to findings during ICP monitoring. We have treated eight children with cranioectodermal disproportion as evidenced by intermittent rises in ICP with overnight monitoring, with cranial expansion procedures according to the algorithm presented in the aforementioned study. The male:female ratio was 1:1, mean age at time of shunting 4 months (one day to 1.2 years), mean age of cranial expansion 5.4 years, and mean time of follow-up of 2.1 years. On presentation the majority of patients presented with the typical signs and symptoms of intermittently increased ICP except that none had papilledema on fundoscopic exam. There were no complications of ICP monitoring nor of the cranial expansion other than blood transfusions. The authors will discuss the clinical presentation, radiographic findings, surgical treatment, results and long-term follow-up of these patients.

11 Differentiation Between Cortical Atrophy and Hydrocephalus Using ¹H MRS

Stefan Blum, MD, J. Gordon McComb, MD, Brian D. Ross, MD (Los Angeles, CA)

Quantitative ¹H MRS to determine cerebral metabolite patterns, and MRI to determine CSF flow, were applied to 12 patients with ventricular dilatation - Group A, cortical atrophy (N=5), or Group B, hydrocephalus (N=7) - and in 9 normal controls.

While mean brain water (Group A = 80% ± 6; Group B = 86% ± 5; normal = 95% ± 4) did not differ between the 2 groups of patients and controls, ¹H MRS distinguished those patients with cortical atrophy (Group A) (*NAA/Cr = 0.69 ± 0.17, vs. Normal = 1.06 ± 0.16; P < 0.002; [NAA] 5.9 ± 1.3, mmol/kg, vs normal 8.0 ± 1.4; P < 0.03) from those with hydrocephalus (Group B) (NAA/Cr=1.16 ± 0.11; [NAA] = 9.2 ± 1.2; P > 0.13 and P > 0.07). Lactate levels were elevated in 3/5 patients with cortical atrophy, but 0/7 of those with hydrocephalus. Mean absolute concentrations (mmol/kg) of the five major cerebral osmoles were 41 ± 4 (Group A), 43 ± 6 (Group B), and 42 ± 4 (normal), so that despite massive brain deformation, constant osmolality was maintained.

Thinning of the cortical mantle in hydrocephalus may result from osmotically driven reduction in individual cell-volumes, (shrinkage), rather than brain-compression.

*N-acetylaspartate/creatine
12 Endoscopic Retrieval of Retained Peritoneal Catheters
S. David Moss, MD, Kim H. Manwaring, MD, Harold Rekate, MD
(Phoenix, AZ)

Since the advent of cerebrospinal fluid shunts, every imaginable complication of shunt surgery has occurred. Shunts have obstructed, disconnected, fractured, migrated locations and frequently catheters are found to end up in the peritoneal cavity. Usually patients have been advised of the relative safety of leaving these catheters behind. Perforation of an abdominal viscous resulting in either shunt infection or passing the catheter through either the anus, vagina, or even the inguinal or femoral spaces have been described. Catheters left in the subcutaneous spaces have shown severe degradation and have virtually disintegrated into a calcified and granulomatous mass that prevents complete removal of the catheter. Similarly, catheters left in the peritoneal cavity may for tissue reactions and become encysted. They may be a source for reoccurrence of shunt infection. They may result in abdominal pain. Neurosurgeons, uncomfortable with retrieval of lost peritoneal catheters may enlist general surgical assistance. Larger laparotomy increases incidence of ileus and shunt infection. Shunt manufacturers have produced peritoneal catheters with smaller inside diameters maintaining the same inside diameter with smaller catheter profile. Fracture of these shunts have been described. General surgeons utilizing two laparoscopies through separate openings have inflated the cavity with CO2 to retrieve lost catheters.

We have removed 26 retained peritoneal catheters using endoscopic techniques through the same small opening used for trocar placement of the peritoneal shunt catheter and have encountered success in 26 of 27 attempts with no complications. The procedure is accomplished in minutes. The procedure and patient data are presented.

13 Shunt Malfunction Associated with CSF Eosinophilia
Robin M. Bowman, MD, Joel C. Boaz, MD, Thomas G. Luerssen, MD, John E. Kalsbeck, MD (Indianapolis, IN)

The significance and management of cerebrospinal fluid eosinophilia in patients with shunted hydrocephalus has been controversial. Between 1990-1996, we cared for 15 children who developed marked CSF eosinophilia (greater than 8% of total CSF WBC) after initial ventriculoperitoneal shunt placement. Ten of these 15 children were born prematurely (25-27 week estimated gestational age) and developed post-hemorrhagic hydrocephalus. The other 5 children had ventriculomegaly secondary to aqueductal stenosis or Dandy-Walker malformation. All of these children developed an early shunt malfunction rarely associated with an infection. All of their malfunctions were associated with CSF eosinophilia and elevated CSF protein. These children had a tremendously elevated mean revision rate (3.3 revisions per new shunt placement) when compared to the mean revision rate in our overall shunted population (0.93). Initially, this condition was managed with hardware removal and serial CSF aspirations via a ventricular reservoir until normalization of the CSF occurred. Recently, we have shown a trend toward a decreased revision rate by use of oral steroids. We plan to review this unique entity, our means of diagnosis, management strategies, and suggest reasons for its occurrence.
14 MRI Assessment of Third Ventriculostomy

Mark M. Souweidane, Christopher G. Gaposchkin, Linda Heier
(New York, NY)

Internal third ventriculostomy has become the treatment of choice for many patients with noncommunicating hydrocephalus. Our experience with postoperative MRI scanning to assess treatment adequacy in this situation has led to the recognition of a composite of findings that are present with a functioning fenestration. Seven patients were recently treated for non-communicating hydrocephalus with endoscopic third ventriculostomy. All had pre- and postoperative MRI scans. Of these seven patients, five were found to have been successfully treated based on clinical response. 100% of these five patients had similar, consistent findings on postoperative MRI scanning: (1) minimal or no reduction in lateral ventricle size, (2) turbulent flow patterns within the third ventricle with signal void at the fenestration site indicative of patent third ventriculostomy, (3) enlargement of the convexity subarachnoid spaces, and (4) a reduction of the subependymal periventricular high signal seen on the preoperative proton density image. Neither of the two patients who failed to respond clinically had these postoperative radiographic findings.

The postoperative radiographic finding after conventional VP shunting for treating hydrocephalus, namely a dramatic reduction in ventricular size, is an unreliable indicator after third ventriculostomy. The lack of this finding post third ventriculostomy must not be misinterpreted as inadequate treatment. Instead, we have found that a negligible reduction in ventricle size, turbulent third ventricular flow pattern with signal void, enlargement of convexity subarachnoid spaces, and diminished periventricular high signal all correlated with good clinical outcome, and are indicative of successful internal third ventriculostomy.

15 Propionibacterium Acnes Infections of Cerebrospinal Fluid Shunts*

Ken S. Sato, MD, Joel C. Boaz, MD, Thomas G. Luerssen, MD, Martin Kleiman, MD, John E. Kalsbeck, MD (Indianapolis, IN)

Propionibacterium acnes shunt infections are often difficult to detect or may be incorrectly dismissed as a contaminant. However, these infections were the leading cause of non-staphylococcal CSF shunt infections seen during a six year period of study. P. acnes infections generally presented over 6 months after the shunt was last operated. The most common presentation of P. acnes infection was low grade shunt malfunction with or without fever. Less frequently, P. acnes infections presented at the peritoneal end of a shunt with such findings as abdominal pain, intraperitoneal CSF collection, bowel obstruction or perforation. In cases of symptomatic P. acnes shunt infection the CSF usually showed less than 200 white cells with near normal glucose and protein. The organisms grow slowly in anaerobic medium requiring several days to be isolated. The treatment of P. acnes shunt infections at our institution has typically involved externalization of the peritoneal end of the shunt and treatment with vancomycin and penicillin. Subsequently, an entirely new shunt is placed. The clinical presentation, laboratory findings and treatment regimes of 11 cases of P. acnes infections of CSF shunts over the period of time from 9/89 to 10/95 will be described.
16 Hemodynamic Response to Fontanellae Compression in Neonatal Hydrocephalus: Resistive Index Changes Predict ICP and Need for Shunt

Joseph R. Madsen, MD, George A. Taylor, MD (Boston, MA)

Purpose: Compression of the anterior fontanellae during Doppler examination of the anterior cerebral artery results in an increase in resistive index (RI) in newborns with altered cranial compliance. RI is calculated from Doppler studies by dividing the difference between systolic and diastolic flow velocity by the systolic flow velocity. Our purpose was to test this technique in predicting intracranial pressure and need for surgical shunt placement.

Materials and methods: Sixty-eight Doppler studies were performed before and during fontanellae compression in 17 infants with hydrocephalus. Twenty-seven intracranial pressure (ICP) measurements were obtained during CSF removal by ventricular drain (10) or lumbar puncture (8) and correlated with percent change in RI (ΔRI) during compression in 13 patients.

Results: Baseline RI without fontanellae compression was not a strong predictor of ICP (r = .1, p = .63). A strong significant correlation was found between ΔRI during compression and elevated ICP (r = .8, P < .0001). Twelve infants required ventricular drainage catheters. Maximum ΔRI was significantly higher in infants subsequently requiring surgical intervention (mean 69 ± 8%, range 31%-132%), compared to maximum ΔRI in infants who did not require ventricular drainage (18 ± 5%, range 3%-29%, p < .002). All infants requiring ventricular drainage had a maximum pre-operative ΔRI of > 30%.

Conclusions: Hemodynamic response to fontanellae compression can be used as a non-invasive predictor of progressive or persistently elevated ICP in newborns with hydrocephalus and may be a helpful adjunct in predicting need for shunt placement.

17 Headache and Arnold-Chiari I Malformation in the Pediatric Population

Fred J. Epstein, MD, Jeffrey S. Weinberg, MD, Diana Freed, BS, James Saddock, BS (New York, NY), Michael Handler, MD (Denver, CO), Jeffrey Wisoff, MD (New York, NY)

Headache is one of a constellation of presenting symptoms in the pediatric population with Arnold-Chiari I malformation. There has been disagreement justifying surgical intervention to treat those patients with headache as a sole complaint. We believe these patients benefit from surgery. Therefore, we retrospectively reviewed our experience, over a 6 year period, with patients less than 5 years of age (mean = 34.8 months) with radiographically confirmed Arnold-Chiari I malformation operated on at NYU Medical Center. Our series consisted of 2 groups: Group A (n=7) presented with headaches as their only complaint; Group B (n=5) presented with cerebellar and long tract symptoms. No patient received prior intervention. All patients were treated with a suboccipital craniectomy, cervical laminectomy, duraplasty, and syringo-subarachnoid shunt when indicated. Follow-up was obtained in 12 patients and ranged from 6 months to 52 months with a mean of 33.9 months. All patients in group A were noted to have rapid clinical improvement (mean=11.6 weeks) and remain asymptomatic. Group B patients improved gradually: 1/5 had moderate improvement at 39 months, 1/5 had significant improvement at 52 months, and 3/5 were asymptomatic at a mean of 18 months. Therefore, we recommend decompression for all symptomatic patients with radiographically confirmed Arnold Chiari I malformations. We conclude that patients less than 5 years old, who present with headache, respond faster and more completely to decompression than patients with cerebellar or long tract complaints.
18 Surgery of Arachnoid Cysts Involving the Third Ventricle*

SooHo Choi, MD, Arun Amar, MD, J. Gordon McComb, MD,
Michael L.J. Apuzzo, MD, Michael L. Levy, MD (Los Angeles, CA)

Introduction: Despite the prevalence of studies regarding arachnoid cysts in children and adolescents, little is known about the presentation, treatment, outcome, and complications of patients harboring third ventricular region arachnoid cysts.

Methods: We reviewed our series of thirty-one patients with third ventricular arachnoid cysts presenting over a fifteen year period. There were twenty males and eleven females in the study with a mean follow up of seventy-eight months. A total of thirty-nine arachnoid cysts were present in thirty-one patients, with six individuals having multiple cysts. The majority of patients (17/31) had intraventricular lesions. Thirty patients were diagnosed by radiographic imaging following the progression of a neurological deficit. All cysts were found to be increasing in size on serial scans.

Results: Eighty-seven percent of patients (27/31) had hydrocephalus on their initial imaging studies; while, four patients had normal ventricular size. Five patients were treated with shunting and aspiration of the cyst, with two of the cyst catheters being placed stereotactically. Thirteen patients underwent fenestration alone; whereas, another thirteen patients had craniotomy along with temporary external drainage. Post-operatively, an additional sixteen patients eventually required shunting. Hyponatremia or hypernatremia occurred in nineteen percent of patients; while, less common complications included transient hemiparesis, subdural hygroma, seizure and progressive visual loss. Two patients had recurrence of their arachnoid cysts.

Conclusion: Third ventricular arachnoid cysts can be approached safely with a minimal amount of morbidity or mortality. Even after shunt placement for hydrocephalus, the cyst can progressively enlarge and cause neurologic deterioration. Therefore, fenestration is important in the long-term treatment of these cysts. With fenestration, thirty-nine percent (10/26) of the patients were able to avoid shunting.

19 Drop Attacks in Children with Chiari I Malformation—A Prelude to Sudden Death?

Sohaib Kureshi, MD, Timothy M. George, MD (Durham, NC)

Recently, it has been reported that Chiari I malformations may be a cause of sudden death in children. We report two children that presented with “drop attacks” who were found to have Chiari I malformations. No other cause for the brief episodic loss of consciousness could be found. Both underwent urgent posterior fossa decompression and duroplasty with resolution of their symptoms immediately postop. At follow-up, they continue to do well and have not had any more “drop” spells. We propose that Chiari I malformations in children can indeed be a cause of sudden death and that “drop attacks” are the early manifestation of impending doom. Urgent surgery is required to prevent death.
20 Benefits of Aggressive Fenestration Versus Limited Fenestration or Shunting of Enlarging Temporal Region Arachnoid Cysts*

Arun Amer, MD, Soo Ho Choi, MD, Michael Levy, MD, J. Gordon McComb, MD (Los Angeles, CA)

Introduction: The natural history of temporal region arachnoid cysts in children is variable. While some remain static, incidental findings, others may enlarge and exert local mass effects or contribute to hydrocephalus. The extent of communication between the cyst and the ventricles, which is similarly variable, is often difficult to assess preoperatively. This explains, in part, the poor results in some patients who merely undergo CSF diversion or limited fenestration into the Sylvian fissure. Recently, we have been aggressively treating these cysts by exposing the entire cyst wall and fenestrating them into the suprasellar and basilar cisterns. We compared the results with this approach to those with smaller fenestrations or initial attempts at shunting the cyst and/or ventricles.

Methods: Over the past 15 years, 42 children (31 males and 11 females) presented with enlarging temporal region arachnoid cysts. Six had associated hydrocephalus at presentation. The mean age was 66 months, and mean duration of follow-up was 56 months. 24 patients (group 1) were initially managed with shunting (5 patients) or limited fenestration (19 patients). The remaining 18 patients (group 2) underwent aggressive fenestration.

Results: Of the 5 patients undergoing initial shunting, 3 (60%) required subsequent fenestration. Of the 19 patients initially treated with limited fenestration, 10 (53%) required subsequent shunting, compared with only 2 of the 18 patients (11%) in group 2. Operative complications group 1 included 2 patients with transient hemiparesis and 1 patient requiring a subdural-peritoneal shunt. Those for group 2 included 2 patients with transient hemiparesis, 2 patients with transient CN III compromise, 1 post-operative seizure and 1 patient requiring a subdural-peritoneal shunt.

Discussion and conclusions: Aggressive fenestration of temporal arachnoid cysts minimizes the chance of needing a shunt and its attendant morbidity. Less aggressive attempts with initial shunting in the absence of hydrocephalus do not result in significant reduction of cyst size and may require subsequent craniotomy for fenestration. Patients with hydrocephalus at presentation, however, will likely need both a shunt and a fenestration.

21 Efficacy of Re-Operation for Symptomatic Recurrent Chiari Malformation

Charles Teo, Frederick Boop, Bruce Cherny (Little Rock, AR)

Suboccipital decompression with or without duraplasty and release of arachnoid adhesions is a well recognized treatment for symptomatic Chiari malformation in the Spina Bifida population. Studies showing long term follow-up have demonstrated excellent results from this procedure. However, the concept of reoperation for recurrent symptomatic Chiari malformation remains controversial. We present a retrospective study of 12 patients seen in the last 10 years at Arkansas Children's Hospital with Spina Bifida and previous surgery for Chiari malformation. The mean time between the first and second procedure ranged from 18 months to six years, and averaged 40 months. Results of surgery will be presented with particular reference to operative findings and technique employed. Complications after re-operation were no worse or more frequent than after the first procedure. The worthiness of re-operation will be discussed and recommendations will be made regarding the optimal technique required at the initial procedure to avoid early re-operation.
22 One-Stage Aggressive Surgical Management of Pediatric Arachnoid Cysts

Paul Jensen, MD, Edward Kosnik, MD (Columbus, OH)

The goal of surgical management of pediatric arachnoid cysts is to accomplish decompression by means of a single operative procedure that will not require additional revision and can be performed without morbidity. While it has been proposed that ideal management would leave a child treated for this disorder shunt independent, we feel that placement of cystoperitoneal shunt during initial cyst decompression is an important adjunct to the achievement of this goal.

We present a retrospective analysis of a large series of pediatric arachnoid cysts operated on by one neurosurgeon and describe the surgical strategy that has resulted in a near 100% long-term cyst decompression without need for subsequent reoperation. Between 1980 and 1996 over one hundred children with an arachnoid cyst were operated on by Dr. Edward Kosnik at Columbus Children's Hospital. All patients underwent craniotomy to gain access to the cyst. Excision of as much cyst wall as possible under microscopic vision was performed in most patients. Finally, cystoperitoneal shunt was performed under direct vision and magnification to assure ideal location that would not be subject to occlusion by residual cyst membrane or collapsing cyst. A smaller number of patients primarily underwent cyst excision alone, such as suprasellar and CP angle cysts, where shunt placement was not deemed optimal. All patients were followed up by CT scan analysis.

In our total series of pure arachnoid cyst patients, none required secondary surgery for shunt revision. In a few patients with other additional problems, a few required subsequent revision. There was no significant morbidity and no mortality.

We conclude that concomitant cystoperitoneal shunting with open arachnoid cyst decompression provides the optimal approach for treatment of pediatric arachnoid cysts.

23 Brainstem Width and Tentorial Angle Correlate with Outcome Following Decompression for Type I Chiari Malformation

Neil A. Feldstein, Tanvir F. Choudhri, Charles J. Prestigiacomo, Samuel C. Kim, E. Sander Connolly, Jr. (New York, NY)

A subset of Chiari I malformation (CM) patients tends to have worse outcome after surgical decompression. Previous studies have suggested that the ratio of posterior fossa volume to supratentorial volume on MRI correlates with benefit from surgery. To identify other radiological markers which may predict outcome we retrospectively reviewed the records of patients undergoing CM decompression at our institution between 1991 and 1996 and identified four patients with suboptimal post-operative courses. In addition to poor neurological recovery, these patients had impaired wound healing and all ultimately required ventriculoperitoneal (VP) shunting. As a control, we chose five patients with benign postoperative courses without need for VP shunting and with reversal of neurological deterioration. We reviewed the records and radiological studies for these 9 patients and for each computed the following variables from mid-sagittal MRI scans: tentorial angle (formed by the tentorium and a line from the torcula to the opisthion), and the sizes of the preopticine cistern, brainstem, and cerebelum at the pontomedullary line (PML), a line constructed parallel to the foramen magnum at the pontomedullary junction. The mean brainstem width in patients with poor outcome was 19.5±1.6% of the total posterior fossa width at the PML versus 25.8±2.0% in patients with good outcome (p=.05). The mean tentorial angle for patients with poor outcome (90.1±6.7°) trended higher than for patients with good outcome (80.4±2.9°). These findings suggest two radiological markers which may help identify a subset of CM patients prone to poor postoperative courses and assist intra- and post-operative management.
Syringomyelia of the Distal Spinal Cord in Children*

Hal Meltzer, MD, Hector E. James, MD (San Diego, CA)

We describe five children with syringomyelia of the distal cord. One child presented with abnormalities of bowel and bladder function, one with intermittent lower extremity pain and flexion spasms, two with an association with lipomeningocele, and one child had spina bifida occulta without symptomatology. All were imaged by spinal ultrasonography or MRI. None of the patients had hydrocephalus, Arnold Chiari malformation, or previous history of trauma. Four patients were treated with syrinx-peritoneal shunts, because of progressive enlargement of the syrinx. In the two symptomatic patients, there was a resolution of the clinical findings. All patients underwent follow-up spinal MRI in a period of three to six months after the shunt procedure and all had a collapse of the syringomyelia. The conservatively managed patient has not demonstrated any clinical progression or enlargement of the syrinx on yearly imaging in the four years of follow-up. We conclude that in otherwise healthy children who present with bowel or bladder dysfunction and/or lower extremity disturbances, the physician should consider in the differential diagnosis that of syringomyelia of the distal cord and neuro imaging of the spinal canal should be performed. Patients with lipomeningocele need to be monitored for the possibility of a syrinx formation after lipoma surgery, as has been previously described by others.

Urgent Radiation Therapy for Pineal Region Malignant Teratomas

E. Christopher Troup, MD, Robert A. Sanford, MD, Larry Kyn, MD, Michael S. Muhlbauer, MD, Jennifer Eldred, RN (Memphis, TN)

Reported is a series of 3 children from 1985 to 1995 with pineal region malignant teratomas who were urgently radiated following precipitous neurologic decline. All were male, age at diagnosis ranged from 8 to 15 years. All had ventriculoperitoneal shunts placed prior to evaluation at St Jude Children's Research Hospital (SJCRH), and their pineal tumor histology verified as malignant teratomas.

At the time of initial evaluation at SJCRH, each was neurologically intact with the exception of a Parinaud's. Prior to the onset of treatment, all had acute neurologic decline (obnuation with decerebrate posturing). Emergent neuroimaging revealed interval tumor growth, and functioning shunts. The patients were given intravenous steroids, and started urgently on a course of radiation therapy, followed by multi-drug chemotherapy. Neurologic deficits improved slowly. Follow-up neuroimaging demonstrated a decrease in tumor size.

Each subsequently underwent delayed (4-12 months) craniotomy for gross total resection, the tumor having converted to benign teratoma. The patients have remained neurologically intact and tumor free since resection ranging from 2 months to 10 years.

The rationale for avoiding surgical decompression at the time of neurologic deterioration in favor of emergent radiotherapy and later gross total resection after conversion from malignant to benign histology will be presented.
26 Superficial Neuroepithelial Tumors of Infancy and Childhood

Richard Ellenbogen, Hernando Mena (Washington, DC), Leslie Sutton (Philadelphia, PA), Lucy Rourke, Rocco Armonda (Washington, DC), Ann Christine Duhaime, Luis Schut (Philadelphia, PA)

We report a series of 12 superficial cystic neuroepithelial tumors of infancy and childhood collected from 1987-1995 from the Children’s Hospital of Philadelphia and Walter Reed Army Medical Center/Armed Forces Institute of Pathology. Patients ranged in ages from 4 months to 14 years with a mean age of 2 years. Presentation included hydrocephalus, focal motor deficits and seizures. All lesions were supratentorial with the parietal (n=7), temporal (n=2) and frontal (n=1) regions being affected. All lesions were cystic with a region of superficial cortical enhancement. Average lesion size was 6 by 5 cm, with a range of 3-10 cm. The cysts were separated from the ventricular system and typically had xanthochromic fluid. Pathologic review was divided into superficial cerebral astrocytomas (n=2) and desmoplastic gangliogliomas (n=10). No difference in survival or outcome was appreciated from these two subgroups. All but one patient had complete surgical resection with one patient who had subsequent malignant degeneration with disease progression and death from residual tumor. One pediatric patient who presented with focal motor seizures underwent functional magnetic resonance imaging which revealed bilateral motor representation for the contralateral hand supporting the concept that this was a long standing tumor. The pathologic review of these lesions revealed a spectrum of desmoplastic astrocytic components, neuronal components and neuropil with cortical disorganization.

27 Anomalous Expression of Cell Cycle Regulatory Proteins in Pediatric Brain Tumors

Joseph Petronio, MD, C. David James, PhD, Jing Ye, MD, Ju He, MD, James R. Allen, PhD (Atlanta, GA)

We have examined an unselected panel of 25 primary pediatric brain tumors of varying grades of malignancy for alterations in the pattern of expression of the retinoblastoma protein (pRb) and the proteins which modulate its activity, cyclin D1, cyclin-dependent kinase (cdk4), and p16INK4a. Immunoblotting of tumor lysates revealed high incidences of aberrant expression of these proteins in all classes of tumors. Lack of p16INK4a or pRb expression was evident in 17/25 (68%) and 9/25 (36%) tumors, respectively. High-level overexpression of cyclin D1 and cdk4 occurred in 7/25 (28%) tumors each. Anomalous expression of at least one of these proteins was found in 23/25 (92%) samples examined, and 16/25 (68%) tumors exhibited significant multiple anomalies of protein expression in this regulatory pathway. In contrast to other tumor, including malignant astrocytomas of adults, homozygous deletions of the P16INK4a gene were detected in only 2/25 (8%) tumors, a glioblastoma multiforme and a primitive neuroectodermal tumor. No amplifications of the genes encoding cyclin D1 or cdk4 were found among the 20 astroglial tumors or 5 primitive neuroectodermal tumors studied in this panel. Consequently, there were no detectable gene alterations in the majority of cases for which anomalous expression of p16INK4a, cyclin D1, and/or cdk4 was observed. Subsequent analysis using Southern hybridization with methylation-sensitive restriction endonucleases revealed that hypermethylation of the P16INK4a gene is not the mechanism of gene silencing in these tumors. Nonetheless, these data indicate that the abrogation of cell cycle regulatory mechanisms mediated through the pRb pathway is common in brain tumors in children.
28 Pediatric Low Grade Astrocytoma—Results of Surgery and Multifactorial Analysis in Tumors Distributed Throughout the Entire Neuraxis

Robert A. Sanford, Aimin Gajjar, Michael S. Muhlauer, Larry E. Kun, E. Christopher Troup (Memphis, TN)

Introduction: From January 1984 to July 1994, 142 patients (age 2 months to 19 years) with low grade astrocytoma were treated. Gross total resection (GTR) was accomplished in 79% of cerebellar and cerebral hemispheric tumors. For tumors in other sites, biopsy or less aggressive resection or stereotaxic biopsy was the standard.

Surgery was followed by observation in 107 cases, radiation therapy in 31, and chemotherapy in 4. Results: The overall survival is 90% ± 3% (SE) at 4 years. Progression free survival (PFS) is significantly better for patients with cerebellar and cerebral hemisphere tumors (n = 75) than those with tumors in other sites (P = 0.00006). At 4 years follow-up estimate, gross total resection resulted in progression free survival in 89% and incomplete resection resulted in 77%. Histology (juvenile pilocytic vs. astrocytoma NOS) has not been a significant correlate with progression free survival in an analysis controlled for tumor site and patient age. Patients less than 5 years at diagnosis had a poorer progression free survival than older children regardless of histology (P < 0.003) or tumor site (P < 0.0002). Treatment of progressive/recurrent disease is effective in a majority of patients, but appeared more successful in patients with hemispheric than thalamic or hypothalamic tumors.

Gross total resection of cerebral and cerebellar hemispheric low grade astrocytoma is clearly the treatment of choice; however, the optimal surgical approach to central midline tumors is less clear in our series.

29 Platelet-Derived Growth Factor Induces Differentiation of Neuroepithelial Stem Cells Into Neurons*

John K. Park, MD, PhD, Brenda P. Williams, PhD, Charles D. Steiles, PhD (Boston, MA)

Neuroepithelial cells are located in the ventricular zone of the embryonic brain. They are the precursor cells for the neurons, oligodendrocytes and astrocytes of the adult cerebral cortex. In examining the potential role of various cellular growth factors in brain development, we noticed that platelet-derived growth factor (PDGF) receptors were activated in embryonic rat brain tissue. We therefore investigated the effects of PDGF on neuroepithelial cell division and differentiation.

Neuroepithelial cells were isolated from embryonic rats and cultured overnight in PDGF-free medium. The following day, cells were pulsed with PDGF for time periods of 30 minutes to 12 hours, washed to remove any unbound PDGF, and then cultured in PDGF-free medium for an additional 7 days. At the end of the culture period, the resulting cells were labeled with cell-type specific antibodies to determine the distribution of neurons, astrocytes, and oligodendrocytes present.

Analyses of the cultures reveal that in the absence of PDGF, 32.5 ± 5.7% of the cells are neuronal while 67.5 ± 5.7% remain neuroepithelial. Following a 30 minute pulse of PDGF 62.8 ± 7.0% of the cells are neuronal while 37.3 ± 6.9% remain neuroepithelial (P<0.0001). Glial cells comprise <1% of both experimental and control cultures. The effects of longer PDGF pulses are similar. Further experiments indicate that RNA but not protein synthesis is required for the actions of PDGF.

The studies presented here indicate that PDGF may have a role in the early development of the cerebral cortex by regulating the formation of neurons. These findings have implications not only on developmental disorders of the cortex but also on the pathogenesis of primitive neuroectodermal and neural-derived tumors.
The Relationship Between TP53 Mutations and Overexpression of p53 and Prognosis in Malignant Gliomas of Childhood

Ian Pollack, Ronald Hamilton, Sydney Finkelstein, Jeffrey Campbell, A. Julio Martinez, Richard Sherwin, Michael Bozik, Susanne Gollin (Pittsburgh, PA)

High-grade astrocytomas have been reported to carry a better prognosis in children than in adults, although the prognosis for individual children with these tumors remains somewhat unpredictable. Whereas prolonged disease control is achieved in some pediatric patients after surgery and adjuvant therapy, others exhibit rapid disease progression that resembles the generally dismal outlook of these lesions in adults. Because recent studies have indicated that p53-dependent apoptosis mechanisms are involved in the cytotoxic effects of radiation and conventional chemotherapy, we questioned whether p53 status might be associated with outcome in childhood malignant gliomas. We therefore examined p53 status, both immunohistochemically and by direct sequencing of exons 5 to 8, in a series of 29 archival pediatric malignant non-brainstem gliomas treated consecutively at our institution between 1975 and 1992, in which clinical, histologic, diagnostic, and therapeutic parameters were previously defined. Eighteen tumors had dense p53 staining in the majority of cells, although only 11 had mutations of the p53 gene (TP53). On univariate analysis, there was a significant association between p53 overexpression as assessed immunohistochemically and a shorter progression-free survival (PFS) and overall survival (OS) (p = 0.019 and 0.013, respectively, rank sum test). In addition, there was a significant association between TP53 mutations and a poorer PFS (p = 0.04), and a strong trend towards a shorter OS among patients with TP53 mutations (p = 0.06). Median PFS and OS for patients with TP53-mutated tumors were 6 months and 16 months, respectively, and for those with p53 overexpression 5.5 months and 14 months, respectively, vs. 16 months and 25 months, respectively, for those without TP53 mutations and 25 months and > 4 years, respectively, for those without p53 overexpression. The percentage of patients in this series with TP53 mutations (37.9%) was substantially higher than in previous studies of childhood gliomas, comparable to the frequency of mutations noted in adult gliomas. TP53 mutation status did not correlate with either extent of resection or tumor histology (grade III vs. IV). However, there was a strong association between age and both TP53 mutation and p53 overexpression, which were less frequent in tumors from children younger than 4 than from older children (p = 0.02 and 0.01, respectively). These results indicate that, in contrast to the findings of prior reports, childhood malignant gliomas do not as a group differ significantly from adult malignant gliomas in terms of their incidence of TP53 mutations, although young children do appear to have a lower frequency of mutations. Moreover, p53 mutation and expression status appears to be associated with prognosis in childhood malignant gliomas and may provide a basis for stratifying patients biologically in future malignant glioma studies.

Survival and Prognostic Factors of Children with Ependymomas Treated on CCG-921

A. Leland Albright, MD (Pittsburgh, PA), Jeff Wisoff, MD (New York, NY), Patricia L. Robertson, MD (Ann Arbor, MI), Paul M. Zeltzer, MD (Orange, CA), James M. Boyett (Memphis, TN), Lucy B. Rorke, MD (Philadelphia, PA), Philip Stanley, MD (Los Angeles, CA)

We analyzed the correlation between prognostic factors and survival in 32 children with ependymomas, 21 benign and 12 malignant, who were treated in CCG-921 with resection, irradiation and chemotherapy, from 1986-1992. Children's ages ranged from 2-17.3 years, with a mean of 7.0 years. Overall survival and progression free survival were compared with the following prognostic factors: extent of resection, tumor site, pathology, age, and chemotherapy used (CCNU/Vincristine or the 8-drugs-in-1-day protocol).

Five year overall survival and progression-free survival were 64% and 50%, respectively. Neurosurgeons thought they performed total tumor resections in 47% and >90% resections in 34%. Postoperative imaging revealed <1.5cm3 residual in 93% of those with a "total" resection and 73% of those with a ">90%" resection.

Two factors were significantly associated with outcome: the surgical estimate of extent of resection and the amount of residual tumor visible on postoperative scans. Five year progression-free survival was 79% for those with total resections and 24% for those with less-than-total resections, p=0.0001. More than 50% of patients with >1.5cm3 residual tumor relapsed within one year. There was no correlation between survival and histopathology (anaplastic or benign), tumor location (supratentorial or infratentorial), M-stage (only 3 tumors were >M-0), race, age or gender. There was no difference in survival between the two chemotherapy regimens, and survival was comparable to historical treatment without chemotherapy.

Extent of resection is highly associated with survival in children with ependymomas and some risk of neurologic deficit is probably reasonable in order to achieve a complete resection.
32 Long-Term Follow-up of Resected Optic Nerve Gliomas: Influence of Residual Tumor on Outcome

Edgar M. Housepian, Kevin Yao, David W. Pincus, E. Sander Connolly, Jr., Davinder Singh (New York, NY)

Introduction: Considerable evidence supports a conservative approach to the treatment of optic pathway gliomas. For unilateral optic nerve tumors presenting with disfiguring proptosis, surgical excision results in excellent cosmetic results. However, the precise role of adjuvant therapy in incompletely resected tumors remains unclear due to the indolent, yet anecdotally unpredictable nature of these lesions. To address this issue, we obtained near 20 year follow-up of a consecutive series of patients with resections of solitary optic nerve gliomas (ONG).

Methods: A retrospective review of the management of all 35 patients who underwent resection of unilateral ONG by a single surgeon between 1963 and 1995 was performed. A transcranial approach with gross total resection was done in all cases. Hospital and office records were examined and long-term follow-up was obtained by telephone questionnaire.

Results: Records were available for 32/35 patients with long-term (>1 yr) follow-up for 29 of the 32 included in this series (91% of included subjects; 83% of entire series). Twenty-three patients were treated with resection alone and 6 received surgery plus radiation therapy. No patients were given chemotherapy. Review of pathology reports revealed no residual tumor in the optic nerve stump in 15 patients (52%), possible residual in 5 patients (17%) and definite residual in 9 (31%). There were no recurrences in any patients and one death of unrelated causes at most recent contact with a mean follow-up of 18 years.

Conclusions: Optic nerve gliomas causing proptosis may be resected with minimal morbidity. The presence of residual tumor in the optic nerve stump does not appear to be of prognostic significance and our data does not support the use of radiation therapy in these cases.

33 Immunotherapy for 9L Gliomas with IL-12*

Jeffrey W. Campbell, Ian F. Pollack, Hideko Okada, Hideaki Tahara, Michael E. Bozik, Michael T. Lotze (Pittsburgh, PA)

Conventional therapies for malignant gliomas have been unsuccessful in eradicating tumor in most patients. This has lead to considerable interest in novel strategies that target characteristics of the tumor which differ from the surrounding normal brain, including modulation of the immune system to enable it to recognize tumor cells as foreign. To this end, we are investigating IL-12 as a potent immunomodulator for malignant gliomas.

Method: 9L glioma cells were transfected with either the genes for IL-12 and neomycin resistance (neo) or neo alone. Our 9L-IL12 cell line produces 60-90 ng IL-12/10^6 cells/48 hours. Groups of six Fisher rats, syngeneic with the 9L tumor line, were injected subcutaneously with tumor in the right flank as follows: Group A- 2x10^6 9L-IL12; Group B- 2x10^6 9L-IL12; Group C- 2x10^6 9L-neo; Group D- 2x10^6 9L-neo. Tumor volume was calculated by the formula for the approximate volume of a spheroid (LxW^2/2). Animals were sacrificed when the tumor size significantly interfered with ambulation.

Results: The 9L-IL12 tumors were significantly smaller then the 9L-neo as early as 10 days after implantation. At 10 days, the tumor volume from groups A and C (2x10^6 cells implanted) were 76 cu mm and 181 cu mm respectively (p=0.004, t-test). Likewise, the tumor volumes from groups B and D (2x10^6 cells implanted) were 135 cu mm and 541 cu mm respectively (p=0.016, t-test). In addition, 6 of the 12 animals injected with 9L-neo tumors were sacrificed for large tumor volume while none of the animals injected with 9L-IL12 tumors were sacrificed (p=0.005, Chi-square). The tumorigenic dose for 9L-neo, where the majority of animals were sacrificed for tumor growth, was 2x10^5 cells. By contrast, the tumorigenic dose for naive 9L cells is less than 2x10^3 cells.

Discussion: Tumor cells that produce IL-12 appear to grow slower and regress to smaller volumes than cells with the neo gene alone. Animals injected with 9L-neo tumor cells who were not sacrificed within the first two weeks for large tumor volumes exhibited some regression of their tumors as well, likely because the neo gene itself is immunologically active, which emphasizes the importance of including appropriate controls in gene transfection studies. Studies utilizing IL-12 for intracranial gliomas are in progress.
Cerebello-Medullary Fissure Approach to the Fourth Ventricle: Resection of Fourth Ventricle Tumors Without Splitting the Vermis*

Jordi X. Kellogg, MD, Joseph H. Piatt, Jr., MD (Portland, OR)

Standard surgical practice for excision of fourth ventricle tumors entails splitting the inferior vermis, but incision of the vermis and lateral retraction on the dentate nuclei and their outflow tracts have been implicated in the development of the so-called cerebellar mutism syndrome. We describe a surgical approach in which the cerebellar vermis is preserved.

Clinical experiences with eight patients harboring fourth ventricle tumors were supplemented by fixed and fresh cadaver dissections. Anatomic illustrations, projections, and intraoperative photographs are presented. The senior author’s case material is tabulated, and clinical examples are discussed.

Dissection of the arachnoid membranes and division of filamentous arachnoidal attachments allow separation and elevation of the cerebellar tonsils and exposure of the tela choroidea along its attachment to the dorsal surface of the medulla at the taenia ventriculitis. The tela can be opened by sharp microdissection from the foramen of Magendie to the foramen of Luschka to expose the lateral recess of the fourth ventricle. Division of the tela allows additional elevation of the cerebellar tonsils, which can be mobilized further by opening of the tonsillo-vermian fissures. Performance of this dissection bilaterally opens the entire inferior end of the fourth ventricle and, particularly after excision of a large fourth ventricle tumor, gives a panoramic view from one lateral recess to the other and from the obex to the aqueduct without incision of the vermis.

The cerebello-medullary fissure approach yields exposure comparable to what can be achieved by splitting the vermis and may minimize the risk of neurological complications.

A Randomized Clinical Trial to Compare Selective Posterior Rhizotomy Plus Physiotherapy with Physiotherapy Alone in Children with Spastic Diplegic Cerebral Palsy


Selective posterior rhizotomy (SPR) is frequently done to treat spasticity in children with cerebral palsy, but the value of the operation has been challenged. A randomized controlled trial was carried out to determine whether SPR followed by intensive physiotherapy was more effective than intensive physiotherapy alone in improving motor function in children with spastic diplegic cerebral palsy.

The study comprised 30 children between 3 and 7 years, who had spastic diplegia with spasticity severe enough to impede gross motor function, and who had access to appropriate physiotherapy in their home community. Patients randomized to the surgical arm (n=15) had surgery within 1 month, and had intensive physiotherapy for 9 months postoperatively. Patients randomized to physiotherapy only (n=15) had identical intensive physiotherapy for 9 months. Patients were assessed by a blinded physiotherapist prior to initiation of therapy, and at 3, 6, and 9 months. The primary outcome measure was the change from baseline to 9 months in the Gross Motor Function Measure (GMFM), a quantitative and validated measure for use in children with spastic cerebral palsy.

There was a statistically significant and clinically important improvement in outcome for the children with rhizotomy, with a mean increase in GMFM score at 9 months of 11.3% for the surgical group compared to 5.2% for physiotherapy only (t=3.03, p=0.007).

This study shows that selective posterior rhizotomy improves function of children with spastic diplegic cerebral palsy, over and above the improvement that might be obtained by intensive physiotherapy alone, thus validating this surgical procedure.
36 Changes in Spasticity and Strength as a Result of Selective Dorsal Rhizotomy

Jack Engsberg, Kenneth Olvec, Sandy Rossa, T. S. Park (St. Louis, MO)

Selective dorsal rhizotomy (SDR) is performed on children with cerebral palsy to minimize or eliminate the influence of spasticity. A limitation associated with the surgery is muscle weakness. This investigation quantified changes in hamstring spasticity (i.e., velocity dependent resistance to passive stretch) and maximum active-joint torques as a function of SDR surgery. Sixteen children with spastic diplegic cerebral palsy (CP group) undergoing SDR were tested the day prior, and approximately 8 months post-surgery. Six children with able bodies (AB controls) made up a control group. For the spasticity measure, a dynamometer extended the passive leg through a range of motion at speeds of 10, 30, 60, and 90°/s while monitoring the resistive torque from the hamstrings. For the active torque measure, the machine flexed the leg at 10°/s while the child performed a maximum concentric contraction of the hamstrings. Work Values for the torque-angle data were calculated for each speed and child. For the spasticity measure, linear regression was used to derive the slope of the line of best fit for the work-velocity data. Work by the hamstrings was used for the strength measure. T-tests were employed for detecting significant differences (p<0.05).

Pre-surgery, the hamstring spasticity in the CP group (0.0300 J/(°/s), σ=0.0254) was significantly greater than that of the AB controls (0.0031 J/(°/s), σ=0.009). Post-surgery values (0.0089 J/(°/s), σ=0.0119) were significantly less than pre-surgery values and not significantly different than AB controls. Unexpectedly, post-surgery values for maximum flexion work of the hamstrings (0.33 J/kg, σ=0.25) were significantly greater than presurgery values (0.18 J/kg, σ=0.22), although both pre- and post-surgery work values were significantly less than similar values for the AB controls (0.62 J/kg, σ=0.17). It was concluded that SDR with post-surgery physical therapy reduced spasticity and increased ability to do work in the hamstrings. The first conclusion is supported by the literature, while the second is not. Results from a few individuals support literature stating that care must be taken in selecting patients for SDR. These methods could aid in the process.

37 Chronic Intrathecal Baclofen for Spasticity of Cerebral Origin

Jeffrey W. Campbell, A. Leland Albright, Margaret J. Barry (Pittsburgh, PA)

While intrathecal baclofen (ITB) infusion through an implantable pump is an effective treatment strategy for spinal spasticity, the morbidity exceeds 50% in some studies, with catheter dysfunction the most common culprit. Less is known about the efficacy and safety of ITB for spasticity of cerebral origin. The following is a review of efficacy and morbidity from ITB infused through programmable pumps for cerebral spasticity between 1989 and 1995 at the Children’s Hospital of Pittsburgh. Patients with cerebral spasticity are selected for a trial of ITB by a multi-disciplinary team. The trial is a double blind cross-over study with serial injections of baclofen and placebo through lumbar punctures and assessment of spasticity before and after each injection using the Ashworth spasticity scale. If a significant improvement in spasticity occurs with ITB, the patient is offered an implanted programmable pump for chronic ITB therapy. Complications from implanted pumps are prospectively maintained in a database.

Of 80 patients who completed a trial of ITB between 1986 and 1995, 52 had a positive response and received a programmable pump. The cause of spasticity was cerebral palsy in 46 patients (88%), head injury in 4 (8%), and single cases of a degenerative brain disorder and encephalitis. Median age at implantation was 13.7 years (range: 4.6-35.8) with a slight male predominance (32M:20F). During a median follow-up of 35.1 months (range: 0.6-75), 15 pumps (29%) were explanted for either lack of efficacy (n=9, 17%) or complications unacceptable to the patient (n=6, 11%). Kaplan-Meier analysis estimates 88%, 75%, 72%, 48%, and 38% of pumps remained implanted at 1, 2, 3, 4, and 5 years respectively. At the most recent evaluation, all of the remaining 37 patients (71%) had decreased spasticity in the lower extremities compared to prior to ITB, with the average Ashworth score falling from 3.0 to 1.7 (p<0.0001). 29 patients (56%) developed 51 complications: 18 catheters (26%) malfunctioned with 6 occlusions, 5 disconnections/breakages, 4 punctures, and 3 dislodgements; 9 pump related complications developed with 3 inverted pumps, one pump explanted for an infection in the abdominal pocket, and 5 episodes of misprogramming, two of which resulted in a baclofen overdose; 7 patients developed seromas in the abdominal pocket; 6 patients experienced a wound breakdown; 7 patients developed a postoperative cerebrospinal fluid leak from the incision; 4 patients developed meningitis. 36 operations were performed on 26 patients (49%) to treat these complications. Kaplan-Meier analysis estimates that 21% of pumps were removed by 5 years for complications unacceptable to patients who otherwise experienced a good therapeutic response to baclofen. A similar analysis reveals that 87%, 74%, 60%, 60%, and 19% of catheters functioned properly over 1, 2, 3, 4, and 5 years respectively.

Implantable pumps for ITB carry similar rates of morbidity for patients with either cerebral or spinal spasticity. The rate of catheter malfunction in patients with cerebral spasticity, while still unacceptably high, is somewhat less than that reported in patients with spinal spasticity. Thicker walled catheters have been used since November 1994 and appear to have a lower incidence of malfunction. While ITB is a successful management strategy for many patients with spasticity of any origin, patients should be informed of the possibility of complications.
Effect on Ambulation of Intrathecal Baclofen Infusion for Lower Extremity Spasticity*

Peter C. Gerszten, MD, A. Leland Albright, MD (Pittsburgh, PA)

Intrathecal baclofen (ITB) infusion has been shown to be an effective treatment for spasticity secondary to both cerebral palsy and spinal cord injury. Its effect on the ambulatory status of children with cerebral spasticity, however, has not previously been addressed. We retrospectively reviewed the effect of ITB on ambulation in 24 patients who were ambulatory, either with or without assistive devices. Twenty-one pumps were placed for spastic cerebral palsy and 3 for spasticity secondary to traumatic brain injury in 13 boys and 11 girls (mean age 18). Mean dose was 200 mg/day (range 22-550 mg/day) and the mean length of follow-up was 46 months. Ambulation was assessed individually by a physiatrist, physical therapist, orthopedic surgeon, and neurosurgeon. Ambulation was improved in 9 patients, unchanged in 12 patients, and worse in 3 patients. The 3 independent walkers remained so, 7 patients continued to use similar assistive devices, 3 patients progressed from walkers to crutches, 10 patients continued to mostly use their wheelchairs, and 1 patient went from a wheelchair to mostly using crutches. Overall functional improvement not directly related to ambulation, including increased agility, endurance, and independence, was found to be improved in 20 patients, unchanged in 2 patients, and worse in 2 patients. Subjectively, as determined by the patient and their family, ambulation was improved in 20 patients, unchanged in 3 patients, and felt to be worse in one patient secondary to increased fatigue. Nineteen patients used ankle orthoses prior to surgery. Three patients no longer required ankle orthoses following pump placement. We conclude that ITB allows for improved ambulation in a certain subset of patients with lower extremity spasticity. In addition, it should not be contraindicated in patients who are felt to rely upon their spasticity for support during ambulation. The baclofen dose may be titrated to balance between extensor tone for support and suppression of hyperactive reflexes which may impede normal locomotion.

Continuous Intrathecal Baclofen for Generalized Dystonia

Jeffrey W. Campbell, A. Leland Albright, Margaret J. Barry, Barbara Shultz (Pittsburgh, PA)

Conventional medical treatment of dystonia is unsatisfactory for most patients. Since 1993, suitable patients with generalized dystonia have been treated with intrathecal baclofen at the Children's Hospital of Pittsburgh. This is a report on the efficacy and complications associated with implantable programmable pumps for chronic ITB in appropriate patients with generalized dystonia. These patients undergo a trial of ITB through a percutaneous intrathecal lumbar catheter with continuous infusion of baclofen at increasing dosages to a maximum of 900mcg/day via an external pump. Patients with significant improvement in dystonia are offered an implanted programmable pump for chronic intrathecal baclofen infusion.

Fifteen patients completed a trial of ITB with twelve demonstrating significant improvement in dystonia at a median dose of 450mcg baclofen/day (range: 250-750mcg/day). Ten patients, whose modified Fahn-Marsden Dystonia Movement Scale (DMS) fell from an average of 24 to 12.2 (p=0.0004) during the trial, had programmable pumps implanted for ITB before September 1995. Eight (80%) remain with a good therapeutic result with an average DMS of 13 (p=0.0004) at a median follow-up of 16 months (range: 8-34mo.). Of the two patients with a poor therapeutic response, one developed unacceptable side effects from baclofen at subtherapeutic dosages, while the other had no significant improvement in dystonia with dosages as high as 985 mcg baclofen/day. Six patients (60%) developed 14 significant complications including five infections, three catheter dysfunctions, and single instances of wound breakdown, postoperative hypotension, generalized weakness, misprogramming of the pump, poorly positioned pump, and subcutaneous fluid above the pump. Eight operations were performed in treatment of the above complications with three pumps explanted for infection. To titrate baclofen dosages that provided good therapeutic relief without unacceptable side effects required an average of 7 changes in rate of infusion per year for each patient.

ITB is a successful management strategy for generalized dystonia in many patients, although associated with moderate morbidity. Frequent follow-up is required for the optimal dosage to decrease dystonia for ease of care, comfort, and improved function, without unacceptable side effects. Patients with appropriate expectations and a willingness to participate in frequent follow-up visits often benefit from ITB for generalized dystonia.
Selective Dorsal Rhizotomy: Morbidity and Mortality

Jeffrey W. Campbell, A. Leland Albright (Pittsburgh, PA)

Selective dorsal rhizotomy (SDR) is an effective treatment of spasticity, although associated with morbidity rates as high as 50% in published series. The following is a review of the complications from SDRs performed at the Children's Hospital of Pittsburgh (CHP) between 1986-1996. Most patients undergo an osteoplastic laminectomy from L2-L5 with SDR of bilateral L2-S1 roots using published electromyographic criteria. Patients receive prophylactic perioperative antibiotics and the anesthesia technique is modified only by the lack of paralytics during electrical stimulation. No perioperative bronchodilators are administered. Patients are maintained at bedrest for three days after surgery with epidural narcotics for pain relief. Patients who underwent SDR and the associated complications were identified from a prospectively maintained database and selective review of medical records.

189 SDRs have been performed at CHP since 1986 by a single surgeon (ALA). 16 complications (8.5%) were identified in 15 patients. Six patients (3.2%) suffered postoperative CSF leakage through the incision, associated with a gram negative meningitis in one patient and requiring an operative revision in another. This last patient returned three weeks later with progressive lower extremity (LE) weakness and underwent a second surgical exploration for a sterile epidural hygroma. She recovered her strength over a 3 month period. Three patients (1.6%) developed postoperative wound infections treated with antibiotics and requiring surgical debridement in two. Two patients (1.1%) developed a mild wound dehiscence, treated with local wound care. One patient suffered LE dysesthesias that lasted for six weeks after surgery. There was a single case each of aspiration secondary to postsurgical oversedation, intraoperative reintubation for decreasing oxygen saturation, and amputation of the left L2 root during the laminectomy none of which resulted in significant sequelae. No patient in this series suffered neurologic worsening from SDR.

Overall, six patients (3.2%) suffered significant morbidity, with two patients experiencing sequelae that extended past hospitalization. This compares favorably to the 17.5% rate of significant morbidity reported in the literature. In our series, four patients (2.1%) developed infections, and five patients (2.7%) required readmission for complications that developed after discharge from the hospital. Four operations were performed on three patients for treatment of complications. SDR is a highly successful procedure and has been associated with low morbidity at CHP.

Reduced Spasticity in Cerebral Palsy 12 Months After Selective Dorsal Rhizotomy

Theodore Roberts, Ross Hays, John McLaughlin, Kristie Bjornson (Seattle, WA)

This study evaluated efficacy of selective dorsal rhizotomy (SDR) in reducing spasticity in children with cerebral palsy (CP). We used an investigator-masked, randomized, controlled trial to compare SDR plus intensive physical therapy (SDR+PT) to intensive physical therapy alone (PT). Both groups received intensive physical therapy: 20 hrs/wk for one month, 4-5 hrs/wk the next five months and 1-2 hrs/wk for the subsequent six months. The spasticity management system (SMS), an electromechanical torque device designed to measure spasticity, and the modified Ashworth scale were administered at baseline and at 12 months. Thirty-eight children completed a 12 month follow-up. SMS change in SDR+PT (n=19, mean=13.6, SD=17.2) varied significantly (p=.04) from SMS change in PT (n=18, mean=-1.1, SD=9.4). Ashworth change in SDR+PT (n=19, mean=1.0, SD=0.5) varied significantly (p=.000) from Ashworth change in PT (n=19, mean=0.1, SD=0.5). There were no sensory losses or severe adverse events. Our data suggest that SDR reduces spasticity in children with spastic diplegia more than a regimen of physical therapy equivalent to that recommended after SDR.
42 Image-guided Applications in the Surgical Treatment of Epilepsy in Children

Roger H. Frankel, MD, Joseph Petronio, MD, Christine Narad, MSN, Roy A. E. Bakay, MD (Atlanta, GA)

Because of the preponderance of lesional epilepsy in children, image-guided surgical technology is quite well-suited to its management. Since January 1995, we have performed fifteen image-guided (frameless stereotactic) neurosurgical procedures in thirteen children for treatment of medically refractory epilepsy. The conditions treated included lesional epilepsy in eleven of thirteen children with six of these being temporal lobe lesions. One case each of non-lesional temporal lobe epilepsy (mesial temporal sclerosis), Rasmussen's encephalitis and nonlesional neocortical epilepsy were treated in this cohort. Eight of eleven cases of intractable lesional epilepsy were secondary to neoplasia, with seven low-grade or well-differentiated glial tumors and one glioblastoma. Other epileptogenic lesions included hemimegalencephaly in two children. Patients ranged in age from 5.0 months to 18.0 (median, 9.0) years at the time of surgery. The specific (and overlapping) frameless stereotactic applications included placement and localization of subdural electrode arrays in fourteen cases, placement and localization of depth electrodes in one, volumetric lesional resection in ten, neocortical resections in two, and cortical mapping in six cases. Of the eight children who underwent volumetric resections of tumor, there were six gross total resections and one near-gross total resection based on immediate post-operative imaging studies. Follow-up has ranged from 7.0 through 20.0 (median, 14.0) months from surgery. Seizure control has been excellent; seizure frequency was initially reduced or eliminated in all resected patients; there has been one early recurrence of infantile spasms in an infant. The surgical morbidity was minimal, and included three cases of transient oculomotor paresis, a transient hemiparesis following a hemispherectomy, and a single homonymous quadrantanopsia. There were no surgical mortalities. The present study demonstrates the feasibility and efficacy, even in infants, of image-guided surgical applications and frameless stereotaxy for the surgical treatment of childhood epilepsy.

43 Ultrasound Guided Modified Hemispherectomy for Seizure Control in Children

Paul Kanev, MD (Detroit, MI), Catherine Foley, MD, Dan Miles, MD (Philadelphia, PA)

Functional hemispherectomy techniques have been designed to minimize the long-term complications of anatomic resection without reducing the effectiveness of seizure control. Beginning with the Rasmussen modified hemispherectomy, there has been a steady trend towards maximum preservation of cortical tissue with each successive modification, exemplified by the recently reported peninsular hemispherotomy.

We have used an ultrasound guided tailored approach combining temporal lobectomy with frontal and occipital disconnections with a central toplectomy of the lateral, insular and interhemispheric cortex. This technique achieves a comprehensive functional disconnection and minimizes entrance and manipulation within the body of the lateral ventricle. Ten patients from 10 months to 23 years of age with congenital paresis and medically intractable seizures underwent functional hemispherectomy utilizing this technique. The average surgical time was 4½ hours and blood loss ranged from 90-400cc. All but one child was discharged after five days. Postoperative fever syndromes, septic meningitis and infection were avoided. On long-term follow-up (10-60 months, mean 42 months), 9 of 10 patients remain seizure-free off anticonvulsants. With this technique, the cortical resection remains largely extraventricular and patients have experienced a less stormy postoperative period and reduced postoperative complications than with other modified hemispherectomy procedures. We recommend ultrasound tailored modified hemispherectomy as an alternative to the anatomic or intraventricular resection of other techniques.
Multiple Subpial Transection: Application to Pediatric Epilepsies with Multifocal Onset

Richard Kim, MD, MS, Rick Abbott, MD (New York, NY)

Introduction: Multiple subpial transaction (MST) is a relatively new technique in which closely-spaced vertical incisions are made in epileptic cortex, interrupting the horizontal intracortical fibers which are thought to be necessary for the generation of epileptic activity. At the same time, it is thought that preserving the vertically oriented fibers spares primary cortical function. The technique was therefore developed as a means of surgically treating focal epilepsies originating in eloquent cortical areas. We report on the application of MST to children with seizures of multifocal onset and involvement of eloquent cortex.

Methods and patient population: MST was performed in 14 children with seizures of multifocal onset and involvement of eloquent cortical areas. Their ages ranged from 2 to 12 years. All patients had multiple seizure patterns, with some combination of complex partial and partial motor seizures. All patients demonstrated multiple or broad areas of epileptogenic cortex on invasive video EEG monitoring. All patients had focal neurologic deficits or evidence of developmental impairment. There were 2 patients with Landau-Kleffner syndrome and 1 with Sturge-Weber disease. 10 patients underwent resections combined with transactions. 3 patients had MST in speech areas, 9 in motor, 6 in sensory, and 1 in visual; 6 patients had MST in 2 eloquent areas. In 3 patients, persistent seizure activity following surgery prompted a second operation with additional MST.

Results: Follow-up duration is 9 to 34 months. Postoperative deficits attributable to the MST procedure occurred in 4 patients: 3 had mild hemiparesis, 2 had speech deficits, and 1 had a quadratic field defect. All have improved but demonstrate some residual deficit. In 2 patients, preoperative hemiparesis improved following MST. Seizure outcomes according to Engel’s classification were the following: Class 1, 1 patient; Class III, 6 patients; Class IV, 7 patients. All 3 patients who were taken back to surgery for more extensive MST demonstrated fewer electrographic and clinical seizures following the second procedure.

Discussion and conclusions: The incidence of postoperative deficits in our patients is higher than in other reports; one reason may be the pre-existence of developmental dysfunction in our patients. However, these deficits are mild and do not cause significant additional impairment. Moreover, in 2 patients preoperative hemiparesis improved following MST. Multiple subpial transaction is a useful adjunct to resective surgery in childhood epilepsies with multifocal onset. The technique may be applied to patients who normally would not be candidates for surgery. Further experience and critical analysis are needed to refine its application and to define its indications.

Medial Temporal Lobe Resections in Childhood Epilepsy

Steven J. Schiff, William D. Gaillard, Joan Conry, Steven Weinstein, L. Gilbert Vezina, Patricia Papero, William Theodore, Susumu Sato
(Washington, DC)

In adults with temporal lobe epilepsy, surgical outcomes are best when resections can be directed at epileptic foci associated with structural lesions of the medial temporal lobe. We report a series of 22 consecutive children presenting with temporal lobe epilepsy, and examined the outcomes from selective medial resections. The patients ranged in age from 9 months to 16 years, and follow-up was from 5-56 months. All patients underwent preoperative neuropsychological testing, MRI scanning, long term-video monitoring, and, when appropriate, WADA testing and PET scanning. Outcomes were based on the Engel Classification, where class I and II correspond to none or rare seizures postoperatively. All patients had follow-up neuropsychological testing for cognitive, language and memory deficits. Of 12 patients with lesions confined to the medial temporal structures (tumors or vascular malformations of amygdala or parahippocampal gyrus, or mesial temporal sclerosis - MTS) 11 (11/12) were class I/II postoperatively, without neurological or neuropsychological deficits. Five of these patients had MTS and underwent highly selective resections of the hippocampus and amygdala, and all 5 are class I (3/5 on the dominant side). Of 10 patients with less well defined lateral temporal lobe involvement, 3 were not offered resection on the basis of invasive electrode localization and functional mapping, and only 2/7 were class I/II following resection. We conclude that in childhood temporal lobe epilepsy, resection results mimic the adult experience, and that highly selective removal of mesial temporal lobe pathological substrates is appropriate for selected patients.
A Technique Allowing Awake Craniotomy in Children

Michael H. Handler, MD, Geoffrey Lane, MB, FRCA, 
Rita Agarwal, MD, FAAP (Denver, CO)

In children, the technical difficulties of an awake craniotomy makes it seldom used, even in resection of mass lesions in or near eloquent cortex. While in certain circumstances another preliminary operation to implant electrodes for mapping purposes is possible, the size of a mass may make it unwise. We have used a technique to allow five children, ages 9-14, to undergo an awake craniotomy. A combination of propofol anesthesia with small doses of fentanyl and midazolam, liberal use of local agents and airway support by the laryngeal mask airway (LMA), eliminating the potent stimulus of endotracheal intubation, allows the children to breathe spontaneously throughout the entire case. When dura has been opened, children are allowed to awaken and the LMA is removed. The child is awake and cooperative during the resection, then sedation with fentanyl and propofol resumed for closure. All children had complete resection of the lesion, with no neurological sequelae. One child had complete resection of a meningioma to spare eloquent cortex, but then required deeper anesthesia and endotracheal intubation to complete resection of the tentorial attachment when local anesthetics failed. One child had adequate mapping completed before being completely awakened. The children had full recall of intra-operative events without distress, and no recall of the opening or closure. Central to the success of the procedure is to minimize the time that the child has to be alert and cooperative, which is facilitated by the propofol and the LMA.

47 Dysembryoplastic Neuroepithelial Tumor (DNET) as a Cause of Medically Intractable Epilepsy in Children

Antonio R. Prats, MD, Nolan Altman, MD, Carole D. Braithwaite, MD, 
Prasanna Jayakar, MD, Raquel Pasaron, MSN, ARNP (Miami, FL)

Dysembryoplastic Neuroepithelial Tumor (DNET), is a rare, benign supratentorial brain tumor occurring most frequently in children. Patients with DNET present with partial complex seizures that are frequently intractable to antiepileptic medications. Their pathological features include multinodular tumors with nodules of astrocytic and/or oligodendrocytic cells associated with a glioneuronal element. They may also be associated with foci of cortical dysplasia.

Our series consists of 15 patients with DNET. All patients had medically intractable complex partial seizures and a normal neurological exam. The age of the patients were 0.5-20.9 years; mean of 9.9 years. There were 6 males and 9 females. The temporal lobe was most frequently involved (11), followed by the parietal lobe (2). The right hemisphere was involved in 9 cases, the left in 6.

All our patients underwent a two-staged surgical procedure consisting of implantation of subdural electrodes (Stage 1), and resection of tumor and surrounding epileptic tissue (Stage 2). There were no deaths and no neurological deficits. Of the 15 patients, 1 was lost to follow-up. Eleven of the remaining 14, were rendered seizure free or greater than 90% reduction in their seizures.

DNET has characteristic clinicopathological features. Resection of the tumor itself may not be sufficient for seizure control. Cortical resection surrounding the lesion, guided by subdural electrodes is occasionally required for seizure control.
48 Polymorphonuclear Leukocytes Exacerbate and Oxyturpinol or Glutathione Protect Against Traumatic Injury in Cultured Cerebral Endothelial Cells

T. S. Park, Joel W. Beetsch, Jeffrey M. Gidday (St. Louis, MO)

The cerebral vasculature is a prime target for cellular injury following trauma. Polymorphonuclear leukocytes (PMNs) and oxygen free radicals have been implicated as mediators of vascular injury following ischemia, but there is little direct evidence regarding their involvement in traumatic injury. We have developed an in vitro trauma model to examine the pathophysiology of PMN interaction with cerebral endothelial cells (CECs) following stretch-induced trauma. We have also examined the effect of oxyturpinol, a potent xanthine oxidase inhibitor, and glutathione, a hydrogen peroxide scavenger and general antioxidant, on CEC injury following trauma. Piglet CECs were grown to confluence in 25 mm tissue culture wells with silastic membrane bottoms (Flexcell International) and injury was induced by stretching the membrane with 50 msec pressure pulses of various intensities (Cell Injury Controller, Commonwealth Biotechnology). In the first study, PMN-CEC interaction was assessed by exposing injured CECs to isolated piglet PMNs (1 x 10^6 / well) for various times. In the second study, CECs were treated with 50 or 100 μM oxyturpinol or 2 mM glutathione for 2 hr prior to stretch-induced injury. CEC injury in both studies was determined by measuring lactate dehydrogenase (LDH) efflux into the culture medium. We observed significant stretch-dependent increases in CEC injury, all of which were higher than unstretched CEC controls. The addition of PMNs following trauma exacerbated trauma-induced injury by as much as 6-fold. In addition, increasing the time of PMN/CEC incubation from 1 to 4 hr elevated CEC injury 23-30%. Pretreatment with 50 or 100 μM oxyturpinol or 2 mM glutathione resulted in a 10-34% or 32% decrease, respectively, in CEC injury compared to untreated CECs. These results indicate trauma-induced CEC injury is due, in part, to oxygen free radical production from CECs. This radical production may induce PMN-CEC adherence, exacerbate traumatic CEC injury, and thus, lead to blood-brain barrier breakdown and subsequent edema characteristic of traumatic injury.

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49 Admissions for Uncomplicated Skull Fractures: Are They Necessary?

Michael A. Vogelbaum, Bruce A. Kaufman, T. S. Park (St. Louis, MO)

Introduction: Patients admitted with uncomplicated skull fractures were retrospectively reviewed to correlate their hospital course with the presumed indications for admission (observation for delayed injury, prevention of dehydration from emesis).

Methods: All children admitted in a 2 year period with a skull fracture without parenchymal brain injury by CT, and without other injuries on admission that required surgery or hospitalization were classified as "uncomplicated". Patient age, admission symptoms and signs, time from injury to ER evaluation to admission, length of hospitalization, intravenous hydration, and child abuse evaluation were determined.

Results: Forty-four patients were studied (mean age 23 months). Loss of consciousness had occurred in 7%, emesis in 20%. A scalp hematoma was present in 80% but only 7% had either a hemotympanum (1) or otorrhea/thororhea (2). No patient had a Battle's sign, seizure, or a focal neurological deficit. On average, patients were evaluated 3.25 hours after injury, spent 3.33 hours in the ER, and were hospitalized for 35 hours; 50% were hospitalized less than 24 hours. Child abuse was diagnosed in 16% and accounted for 5 of the 7 patients hospitalized for longer than 2 days. Intravenous lines were started on half the patients, but in only 18% was the rate at least 90% of calculated maintenance rate. Patients with emesis (9) were older (mean 41 months) and had longer hospital stays (mean 2.7 days), but only 67% were given IV fluids and only 55% received maintenance rates. No patient suffered a complication related to their skull fracture.

Conclusions: Children with uncomplicated skull fractures do not require routine admission for observation. Most do not require admission for IV hydration, although persistent emesis may require admission. No child with an uncomplicated skull fracture developed an injury requiring treatment during the evaluation and hospitalization. If child abuse screening could be accomplished in the ER, admissions could be further limited.
Piglet Brain Contusion: Does Maturation Matter?

A. C. Duhaime, R. Ragupathi, D. Meany, D. Smith, T. McIntosh
(Philadelphia, PA)

Little is known about the effect of immaturity on the brain’s response to mechanical trauma. Studies of hypoxic/ischemic insults in immature animals suggest that vulnerability to permanent damage varies as a function of insult type and age, and that certain regions or systems of the brain may be vulnerable at different times during development. These differences are likely explained by anatomic, cellular, molecular, and genetic changes early in life. To study whether the same changes might influence the brain’s response to mechanical trauma, we have developed a contusion model in the piglet in which mechanical force can be scaled to the total volume of the brain. Developmental levels in the piglets studied correspond to human infants, toddlers, and adolescents. Survival times of 24 hours and 7 days at each developmental level allowed for histologic analysis as well as changes in gene expression over time. Preliminary data from these experiments will be presented, showing that the response of the brain to injury changes with maturation. An understanding of these changes may lead to more specific neuroprotective strategies for the immature brain.

Estimation of IQ Score Reduction Following Head Injury in Children*

Mark R. Ianosco, MD, J. Gregory Javornisky, PhD (Hartford, CT)

Quantification of cognitive disability following head injury is frequently hampered by a lack of precise data concerning pre-injury cognitive functioning. The study reports on forty-six children and adolescents with Wechsler IQ scores obtained prior to and following head injury. Pre-injury evaluations were obtained to assess learning or emotional difficulties at an average age of 10.8 years (range 4-17 years), an average of 28.7 months (range 1 to 99 months) prior to head injury. The sample exhibited normal intelligence compared to standard values. Mean Pre-Injury scores for Verbal, Performance and Full Scale IQ were 103.3 (±16.6), 104.1 (±14.2), and 104.1 (±15.1) respectively. Post-injury evaluation occurred an average of 25.1 months (range 9 to 56 months) following head trauma with mean Post-Injury score of 92.8 (±12.0), 94.9 (±15.7), and 93.1 (±12.6). Injury characteristics associated with outcome in previous studies were compiled and multiple stepwise regression analyses performed to generate the following equations for estimating the difference between Pre- and Post-Injury IQ scores.

Difference in Full Scale IQ =
-2.9 + .3(Days to command following) + -5.4(Motor Score)

Difference in Verbal IQ =
-7.1 + .7(GCS) + -3.2(CT Scan Grade) + 10.1(Steroid Use) + -.2(Days to follow commands)

Difference in Performance IQ =
1.4 + -3(Days to follow commands) + -9.1 (Motor Score)

These equations account for 46% (p<.0001), 53% (p<.0001) and 41% (p<.0001) of the variance in the Full Scale, Verbal and Performance IQ declines post-injury. Estimates of IQ reduction and pre-injury IQ scores using these equations may provide a quantitative value for cognitive outcome following head injury.
52 Intracranial Pressure Monitoring After Severe Head Injury in Children: The Richmond Bolt Versus the Camino Catheter

James T. Rutka, MD, John S. Myseros, MD, Desmond Bohn, MD, Peter Cox, MD, Paul Chumas, MD (Toronto, Ontario, Canada)

Introduction: It is thought that prolonged intracranial hypertension after severe traumatic brain injury in children is associated with poor neurologic outcome. As such, attempts to control increased intracranial pressure (ICP) have been facilitated by the advent of invasive monitoring devices. The purpose of this study is to compare the efficacy and performance of two such devices, the conventional fluid-coupled Richmond bolt, and the more modern fiberoptic Camino catheter.

Methods: Eighteen patients with an average age of 8.8 years were monitored for a mean time of 4.9 days. The median Glasgow Coma Scale was 5. Each patient was monitored using both the Camino catheter and Richmond bolt placed side by side through the same scalp incision. Comparisons were sought between the two devices in the following categories: Fidelity of ICP waveforms, mean time to device malfunction, number of technical failures, number of re-calibrations or flushing, and dampening of waveforms.

Results: In the group of 18 patients monitored, one Camino catheter lost its ICP waveform compared to 4 Richmond bolts. One Camino suffered from a technical failure compared to 2 Richmond bolts. While no Camino catheters required re-calibration, 5 Richmond bolts required flushing to bring back adequate waveforms. Finally, there were no dampened waveforms associated with Camino catheter monitoring, the Richmond bolts in 4 patients became significantly dampened at ICPs readings > 40 mm Hg.

Discussion: Although both monitoring devices were consistent in maintaining adequate amplitude ICP waveforms with ICP values of 20-40 mm Hg, the Richmond bolt was associated with dampened waveforms in a significant number of patients whose ICPs were over 40 mm Hg. In addition, prolonged monitoring with the Richmond bolt was associated with device malfunction which frequently required recalibration. These data support the notion that the Richmond bolt (a low cost and reusable device) and the Camino catheter can provide comparable information in children expected to have a less protracted course of raised intracranial hypertension after head injury. However, our data suggest that children with more severe head trauma and acute brain swelling are better served by the single-use Camino catheter despite its higher cost than by the Richmond bolt.

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53 Presentation, Treatment, and Outcome in a Large Series of Pediatric Patients with Aneurysmal Bone Cysts

Eric R. Trumble, MD, Frederick Sklar, MD (Dallas, TX)

Introduction: Aneurysmal bone cysts (ABCs) comprise 5% of bony lesions in children. Although most ABCs occur in the long bones, up to 33% occur in the head and spine causing neurological symptoms. The mean age at presentation is 8, with few presenting after the third decade of life. Due to the relative paucity of data concerning the presentation and treatment of ABCs in the neurosurgical realm, we have undertaken this study and review of the literature to better delineate appropriate treatment and expected outcome.

Methods: Data was gathered concerning the presentation, treatment, and outcome of all the ABCs treated at Children's Medical Center of Dallas and Texas Scottish Rite Hospital since 1990. Radiographic and chart review were performed to capture all data. Imaging studies included plain radiographs, CTs, MRIs, and angiograms.

Results: Twenty-five patients were treated over the study period, 7 of whom had ABCs of the spine and 1 of whom had an ABC of the temporal bone, presenting with an ipsilateral CN III palsy. Spinal ABCs were most common in the thoracic spine, followed by lumbar, and cervical. Treatment modalities ranged from curettage only to cryotherapy to complete resection. There were no recurrences in the patients with complete resection with increasing recurrence rates from cryotherapy and curettage only (p<0.01). Pre-op particulate embolization was also selectively utilized.

Discussion: ABCs have been described throughout the spine, in the frontal, occipital, petrous, and ethmoidal regions as case reports only but no large series has previously been compiled to assess their incidence and attempt to clarify appropriate treatment. Our series reveals that complete excision is the treatment of choice, with cryotherapy being the next best treatment modality. Curettage is associated with up to a 71% recurrence rate and XRT has been associated with radiation-induced sarcoma.

Conclusions: Therefore, we feel that ABCs are a lesion that the neurosurgeon must consider in the differential diagnosis of bony lesions of the skull and spine and that complete excision is the optimal treatment.
54 Upper Cervical Spine Fusion in the Pediatric Population*

David W. Lowry, Ian E. Pollack, Brent Clyde, P. David Adelson, A. Leland Albright (Pittsburgh, PA)

We review the outcome of 25 pediatric patients who underwent upper cervical or occipito-cervical fusion at the authors' institution since 1983. At a mean age of nine years, the patients presented with instability associated with odontoid in 11 cases, with rotatory subluxation in 5 cases, with odontoid fracture in 2 cases, with atlanto-occipital dislocation in 2 cases, and with congenital atlantoaxial instability in 5 patients, 4 of whom had Down's syndrome (Trisomy 21). Ten children had abnormal neurologic examinations preoperatively, but nine experienced improvement or resolution of deficits by latest follow-up (mean 14 months). Fusion was achieved with the first operation in 21 of 25 patients, and all except one eventually fused. Four patients exhibited persistent instability after an initial procedure. In two cases this was due to erosion of a multistranded cable through the intact arch of C2, in one case to pin site infection necessitating early halo removal, and in one case to slippage in a halo following a Gallie procedure, which was revised with a Brooks fusion. This series, the largest yet published, shows that with appropriate surgical management posterior upper cervical fusion in the pediatric population is highly successful. Careful attention to halo pin site care and caution in using multistranded cable in young patients may improve results.

55 Pediatric C1-2 Transarticular Screw Fixation for Atlanto-Occipital and Atlanto-Axial Instability

Douglas Brockmeyer, MD, Ronald Apfelbaum, MD (Salt Lake City, UT)

Atlanto-occipital and atlanto-axial instability in pediatric patients continues to be a challenging problem in neurosurgery. Fusion failure, growth potential, anatomical size constraints and congenital anomalies are just a few of the problems encountered by the pediatric spinal surgeon. From December 1991 to August 1996, twenty-five C1-2 transarticular screws were placed in thirteen patients ranging from 5 to 16 years of age for instability at the atlanto-occipital level, atlanto-axial level, or both. Ten of the thirteen patients were unstable at C1-2, two were unstable at O-C1, and one was unstable at both levels. Reasons for translational or vertical instability included: odontoid fracture—5, Cotarsal fracture—3, chronic type II odontoid fracture—2, transoral odontoid resection—2, and cervicomедullary tumor resection—1. After the procedure with rigid screw fixation, all patients achieved satisfactory bony union at the appropriate level, including four patients with prior bone and wire attempts at fusion. The occipital-cervical constructs included titanium "U" bars affixed to the occiput with screws and incorporated into the lateral mass screws with rigid couplers. Surgical morbidity included one wound infection not requiring hardware removal and one vocal cord palsy from patient positioning. There were no vertebral artery injuries and no mortalities. We conclude that C1-2 transarticular screw fixation is a valuable technique in managing a wide variety of difficult craniovertebral problems in the pediatric population.
56 Virchow Among the Craniologists: His Contribution to the
Problem of Craniosynostosis

Cary D. Alberstone, MD (Albuquerque, New Mexico)

When Rudolph Virchow (1821-1902) published his study, "On Cretinism, Especially in Franconia, and on Pathological Conformations of the Skull" (1851), his morphometric analysis and classification of deformed skulls both borrowed and departed from a long and well developed tradition of craniometry that was primarily concerned with the establishment of a scientific basis for racism. The inheritors of this tradition applied the methods of craniometry to identify racial types, which invariably "confirmed" the superiority of the author's race.

Applying these anthropological tools to a more respectable end, Virchow attempted to articulate a biological explanation for the pathologic skull shapes that were observed in individuals with cretinism. Virchow's rejection of the racial agenda that tainted the efforts of many of his predecessors was characteristic of his methods, and an important ingredient in his scientific successes.

From his earlier studies on osseous growth, Virchow understood that the sutures of the skull were the growth zones of the cranial bones. He reasoned from this knowledge that premature closure of the sutures results in abnormal skull growth, and that closure of one suture is compensated by growth in another direction (thus maintaining normal cranial volume), whereas closure of many sutures results in microcephaly.

Based on a study of twenty-nine deformed skulls, and the contemporary work of a racial craniologist, Anders Retzius, Virchow formulated a general classification of pathologic skull types, including (1) macrocephaly, (2) microcephaly, (3) pathological dolichocephaly (subtypes: sheno-, lept-, clinocephaly), and (4) pathological brachycephaly (subtypes: pachy-, oxy-, platy-, trocho-, plagioccephaly). Although the methods and conclusions of Virchow's study of craniosynostosis had been utilized or suggested by previous investigators, Virchow's comprehensive, unbiased, biological approach to the problem rightly earns him the credit for the first description of craniosynostosis.

57 Occipital Plagiocephaly: Standards for Treatment

Benjamin S. Carson, Carol S. James, Craig A. VanderKolk (Baltimore, MD)

Referrals for occipital plagiocephaly have significantly increased over the past decade, yet there is little evidence that the cranial asymmetry is caused by an increased incidence in primary lambdoidal craniosynostosis. Concerns have been raised about the role of neurosurgery in the treatment of occipital plagiocephaly. Several neurosurgeons have advanced clinical decision rules for surgical intervention based on radiographic detection of the lambdoidal sutures. Although such rules are valuable, we believe they may be difficult to evaluate because premature fusion can be difficult to detect. More importantly, our experience suggests that children can have abnormal appearing sutures with normal neurological status, and normal appearing sutures with neurological deficits or marked cerebral compression.

To guide treatment decisions and to quantitate the results, we describe a new set of rules based on radiographic indicators of the brain rather than the bone, and clinical assessment of the child. These rules are simple to apply and they offer a powerful tool for the crucial early evaluation and follow-up of children with suspected craniosynostosis.
58 Occipital Flattening: Indications for Intervention and Techniques for Repair

Richard Polin, John Jane, Kant Lin (Charlottesville, VA)

Introduction: Lambdoid synostosis, functional lambdoid synostosis, lazy lambdoid occipital plagiocephaly all refer to flattening at the back of the head. True lambdoid synostosis is exceedingly rare. By true, we mean actual fusion of the lambdoid suture. On the other hand, unilateral/bilateral flattening is becoming more common and these may or may not be related to the prohibition of the prone position for babies because of the Sudden Infant Death Syndrome. The question is as follows: should occipital flattening be treated in the absence of true synostosis?

Materials and methods: During a period from 1992 to 1995 we saw 73 patients that we evaluated for unilateral or bilateral occipital flattening. There was only one case of true lambdoid synostosis and this patient was not operated on. Six patients were operated on after a trial of conservative management of at least six months duration encompassing positional therapy and skull molding cap therapy.

Results: All patients who underwent therapy also underwent a skull remodeling procedure with use of miniplates for rigid fixation to correct the occipital flattening. Excellent cosmetic results were seen in all patients.

Conclusions: Should occipital flattening be treated in the absence of true synostosis? Our answer is yes. Any alternative including skull molding has been tried by us for 20 years and is only modestly effective. The indication for surgery really is the same as it has always been. If the deformity is aesthetically unacceptable and the parents understand the risks involved in the correction, then surgery is warranted.

59 Surgical Management of Sagittal Synostosis: A Comparative Outcome Analysis

Todd A. Maugans, MD, J. Gordon McComb, MD, Michael Levy, MD (Los Angeles, CA)

A ten year (1986-95) retrospective review examined the surgical management of sagittal synostosis at CHLA. Strip craniectomy (SC) was performed in 72 cases (61%) and biparietal-widening, occipital-reducing calvarial vault remodeling (CVR) in 46 cases (39%). There was a 3.5:1 male predominance in both groups. Patients undergoing CVR were older (x=5.3 vs. 3.0 months, p<01) and more likely to manifest severe keel formation and occipital protuberance than the SC group. In the CVR group, surgical procedure time was longer (x= 2.1 vs. 1.4 hours, p<.01), intraoperative blood loss was greater (x=252 vs. 55 cc, p<.01), and the frequency of intraoperative blood transfusion was greater (96% vs. 24%, p<.01; mean volume, 328 vs. 108 cc, p<.01). No intraoperative complications occurred in either group. Total time of hospitalization was similar (x= 4.4 vs. 3.8 days, p=.02). No neurological, hematological or transfusion-related complications or deaths were encountered.

Cosmetic outcomes were significantly better in the CVR group. Residual frontal bossing was less frequent in the CVR patients (7% vs. 13%, p<.01). 62% were rated as excellent, compared with only 33% in the SC group. 58% of the SC patients manifested palpable bony defects at last follow-up visit, with one patient requiring reoperation.

In conclusion, CVR is a surgical option that affords superior immediate correction of skull deformities compared to SC, especially in older infants with occipital protuberance enlargement and/or severe keel formation.
Endoscopic Cranietomy for Release of Stenosed Cranial Sutures

David E. Jimenez, MD, Constance Barone, MD (Columbia, MO), Anthony Nobles (Fountain Valley, CA)

Three patients underwent release of six stenotic cranial sutures using fiberoptic and rigid endoscopes. The sutures affected included three sagittal, two coronal and one metopic. Two patients had single isolated sagittal synostosis and one patient had Pfeiffer's syndrome with one sagittal, two coronal and a metopic suture stenosis. A 2.0 cm incision was made over the anterior fontanel which was released from the surrounding bone. A 2.0 cm incision was made over the lambda and a 7 mm burr hole was placed over the midline. Using S-shaped refractors along with 30K, 2.7mm, and 4mm rigid endoscopes, subgaleal and epidural dissection was carried out between these two points. Two parasagittal osteotomies were performed with bone cutters and an entire 2cm by 1cm calvarial strip was removed without complications. Barrel stave osteotomies were easily performed in the parietal bones bilaterally. For resection of the coronal and metopic sutures, a similar approach was used through the anterior fontanel and dissection was carried out laterally and anteriorly respectively. In all cases the dura was easily and successfully freed without injury to the sagittal sinus or underlying brain. Mean operative time was 1.1 hours, blood loss was less than 5cc's per case and all patients were discharged the following day. Postoperative follow-up of 8½ months, with molding helmets, indicate that all patients have obtained excellent results with correction of severe scaphocephaly in two of patients. Progressive cranial deformity was halted and reversed in the case of Pfeiffer's syndrome patient.

61 The Transpalatal Approach for the Treatment of Infantile Transsphenoidal Encephaloceles in Four Patients

Kerry R. Crone, David P. Gruber, Erin M. Kennedy, David A. Billmire (Cincinnati, OH)

The surgical treatment of transsphenoidal encephaloceles in infants has been controversial because of the high postoperative rates of hypothalamic dysfunction, infection, and death. These rare anomalies are often associated with facial and ocular abnormalities, median cleft face syndrome, or endocrine disturbances that can cause postoperative morbidity. We present 4 patients, aged 2 weeks to 10 months, who underwent the successful transpalatal repair of three encephaloceles and one cephalocele; surgery was undertaken for progressive apnea due to nasal obstruction. Two patients had a concomitant diagnoses of median cleft face syndrome; one had an associated Chiari I malformation and one was without other cranial abnormalities. All infants underwent preoperative auditory, ophthalmologic, and endocrine evaluations. Each lesion was approached through the median Rafe of the hard and soft palates; the bone was harvested to serve as a graft. The encephalocele was then dissected free from the surrounding bone and coagulated, and the defect was closed with the autogenous graft. The sac was not opened and anomalous neural elements were not resected. At follow-up, patients demonstrated resolution of their preoperative symptoms without evidence of infection or worsening endocrine dysfunction. We conclude that this approach for transpalatal repair is safe and reliable for the treatment of infantile transsphenoidal encephaloceles.
62 Frameless Stereotactic Applications in Cranial Neuroendoscopy

Allen S. Waitze, MD, Joseph Petronio, MD, Nell Paris, RN (Atlanta, GA)

Previous experience has shown that many complications of ventriculoscopic procedures in children result from anatomic confusion, especially in cases with extensive distortion of normal anatomy. The application of stereotactic technology has helped to localize target structures for endoscopic procedures, but conventional framed systems have proven less than ideal. In this series we review our preliminary experience with the application of image-guided endoscopy and frameless stereotaxy to intracranial endoscopy in a largely pediatric population. We have performed 15 procedures on 11 patients, ranging in age from 5 days to 43 years at the time of initial surgical treatment. Indications for frameless stereotactic localization during neuroendoscopic procedures included cyst (arachnoid or tumor) fenestration in four cases, septum pellucidum fenestration in four cases, and catheter placement (cyst or ventricular) in seven cases. Stereotactic localization was done utilizing MRI in five cases and CT in ten; registration was done using scalp fiducial markers (two cases) or fitting of a computer-generated skin surface mask (thirteen cases). Target localization was successful in all but two instances. There were no operative mortalities and the three operative morbidities, including one aborted procedure due to inaccurate registration, one missed target, and one case of a postoperative gran-negative ventriculitis. The present study demonstrates that frameless stereotactic guidance for neuroendoscopic procedures is technically feasible in the pediatric population, including infants, and can be performed in all three cranial fossae. Frameless stereotactic localization is usually complementary to current neuroendoscopic techniques and should be utilized for target localization in challenging cases.

63 Treatment of Non-Communicating Hydrocephalus with Ventriculoscopic Third Ventriculocisternostomy: Outcome and CSF Flow Patterns

Liliana Goumnerova, MD (Boston, MA), David Prim, MD, PhD (Chicago, IL), Sandra Rufo (Boston, MA)

Traditionally, non-communicating hydrocephalus has been treated with insertion of a ventriculo-peritoneal (VP) shunt. With the technological development of ventriculoscopic instruments, third ventriculocisternostomy has become a method gaining popularity in the initial management of this problem. However, questions remain as to the proper selection of patients, as well as to its effectiveness in the long-term management of hydrocephalus and appropriate follow-up. We performed 22 ventriculoscopic third ventriculocisternostomies in 22 patients, ranging in age from one day to 33 years, median 8.7 years. All of the procedures were performed by the senior author via the same technique. There were 13 males and nine females.

Primary aqueductal stenosis were present in 16 patients and tectal masses with secondary aqueductal stenosis were present in the remaining six. Follow-up consisted of routine surgical evaluation and a cine MRI to evaluate flow through the ventriculocisternostomy. There were five patients who required the subsequent insertion of a VP shunt. Three patients had this procedure performed at time of a shunt malfunction without revision of the existing shunt and have done well with no subsequent need for the shunt. Outcome with respect to control of the symptoms of hydrocephalus correlated well with the results of the cine MRI, even if there was persistent ventriculomegaly. Third ventriculocisternostomy has a role in the management of non-communicating hydrocephalus in a select group of patients. Adequate follow-up includes flow studies to evaluate the patency of the ventriculocisternostomy and this correlates well with outcome.
Complications of Neuroendoscopic Third Ventriculostomy

Steven J. Schneider, MD (New Hyde Park, NY)

Neuroendoscopic third ventriculostomy continues to increase in popularity for the treatment of select forms of hydrocephalus. Between 1991-1996, third ventriculostomy procedures were attempted in 89 patients. In 73 patients, the initial procedure appeared to be effective. However, there was late failure in 7 cases including two patients with stenotic foramen of Monro, and one herniation of the basilar artery corrected by a secondary procedure. Four additional late failures occurred despite what appeared to be a patent third ventriculostomy. In 14 patients, the initial procedure revealed anatomy which was not conducive to third ventriculostomy. In 4 patients, the initial procedure was complicated by bleeding. Six patients underwent re-exploration with a successful fenestration accomplished in 4 cases. One case of late re-hemorrhage and pseudoaneurysm was seen. In our series, there were two permanent neurologic deficits including a third nerve palsy and a brain stem dysfunction. Several transient deficits occurred and resolved. There was no mortality. Our report focuses on the prognostic indicators and the occurrence of complications both intraoperative and perioperative and a stratagem we have developed to help avoid them.

Scopeless Endoscopy: The Use of Virtual Operating Environments in the Ventricular Chamber

Terri L. Harphold, MD, Michael L.J. Apuzzo, MD, J. Gordon McComb, MD, Ann Amar, MD, SooHo Chei, MD, Michael L. Levy, MD (Los Angeles, CA)

Introduction: With increasing technologies available for the use in the operating room, the integration of advanced imaging and computerized anatomic environments remains undocumented. Since increases in the availability of anatomic information contributes to the surgical approach and outcome, we have integrated a virtual imaging system into our operating practice.

Methods: Over the past 18 months we have developed an integrated computerized visualization system for use in surgery of the ventricular chambers. The interface is via an LCD head-up display incorporated into the optics of the operating microscope. In addition the observer is allowed to visualize all images in three dimensions through the use of a stereoscopic headset which allows for unlimited viewing of the operative field and the ability to move around the operating room and assist the primary surgeon from any number of positions. Digital images are incorporated from the operative site using a 90° endoscope rotated along an axis of 360°. The image is stored in a mainframe directly attached to the operating microscope and observer headsets. Using voice control the image plane can then be viewed in a 360° cylindrical image.

Results: High density allows for the superimposition of structures and the appreciation of 3D spatial relationships. The input is updatable and provides endoscopic views for the operating surgeon while working under the operating microscope without the need for the continuous use of an endoscope during surgery. Benefits include the ability to view the endoscopically generated image while simultaneously working within the ventricular compartment.

Conclusions: Scopeless endoscopy allows for better appreciation of anatomic structures and their relationships, increased concurrent visualization, and a more accurate and thorough undertaking of operative intervention within the ventricular chamber.
Endoscopic Third Ventriculostomy and the Slit Ventricle Syndrome

Jonathan J. Baskin, MD, Kim H. Manwaring, MD, Harold L. Rekate, MD (Phoenix, AZ)

Introduction: The slit ventricle syndrome (SVS) is defined as the triad in shunted patients of intermittent headaches lasting 10 to 90 minutes, small ventricles, and a flushing reservoir that slowly refills. The pathophysiology of this condition relates to transient proximal catheter occlusion, and symptomatic patients typically respond to shunt revision that incorporates an anti-siphon device. This treatment is imperfect, however, with clinical improvement sometimes failing to occur or occurring only transiently. This study illustrates the utility of endoscopic intervention as a means of sometimes attaining a symptom-free and shunt independent state in patients with SVS.

Methods: Twenty-one patients previously diagnosed with SVS and who presented with headaches refractory to shunt revision as described above underwent distal shunt externalization and occlusion with concomitant fiberoptic intracranial pressure monitoring. Those patients demonstrating symptomatic hydrocephalus underwent endoscopic third ventriculostomy and shunt removal with postoperative intensive care unit observation.

Results: Five patients (23.8%) were seen to have intracranial pressures consistently within a normal range and headache complaints that were unrelated to intracranial pressure. These patients had their shunts removed. The remaining 16 patients (76.2%) underwent endoscopic third ventriculostomy for treatment of hydrocephalus with 11 patients (68.8%) tolerating shunt removal and reporting continued headache relief at their one year follow-up evaluation.

Discussion and conclusions: Endoscopic, third ventriculostomy has successfully treated a significant percentage of patients with SVS, both from the standpoint of attaining symptomatic relief and shunt independence. This study also emphasizes the capacity for patients previously requiring a CSF diversion procedure to regain an ability to intrinsically manage their own CSF dynamics and tolerate shunt removal, thereby simplifying their medical follow-up.

Stereolithography and the Evaluation of Computer Image Based Modeling in the Peri-Operative Management of Patients with Cerebral Vascular Anomalies and Neoplasms*

Michael L. Levy, MD, Terry Harpold, MD, Joseph Chen, MD, J. Gordon McComb, MD (Los Angeles, CA)

Currently a number of modalities are being integrated into the care of neurosurgical patients. These include the use of virtual immersive and semi-immersive headsets, in addition to computerized reconstructions of MRI and CT images which allow for further evaluation of patients in the peri-operative period. We will describe the use of our own system, which has allowed us to not only increase our understanding of the appropriate surgical anatomy in our pre-operative evaluation of patients with CNS disorders, but also as used as a teaching tool for the training of neurosurgical residents. Initial images for each patient are constructed from either data obtained from MRI and/or CT imaging. The data is subsequently transferred via disk or an ethernet connection for stereolithographic construction. The data is then integrated into SLA-250 stereolithography apparatus. Multiple passes of yttrium vanadate sulfate laser over the photo isomer resin allows for the completion of the three dimensional CAD.

We will discuss our applications in these models in three scenarios. These will include application and approach to vascular abnormalities of the CNS approaches to complex tumors in the CNS in children and finally in the pre-operative assessment and management of children with complex craniofacial abnormalities. We believe that the use of these technologies will be indispensable not only in the future peri-operative planning and management of patients with CNS disorders, but also in the future teaching of those individuals in neurosurgical training.
**Surgery of Arachnoid Cysts Involving the Third Ventricle**

SooHo Choi, MD (Los Angeles, CA)

Introduction: Despite the prevalence of studies regarding arachnoid, cysts in children and adolescents, little is known about the presentation, treatment, outcome, and complications of patients harboring third ventricular region arachnoid cysts.

Methods: We reviewed our series of thirty-one patients with third ventricular arachnoid cysts presenting over a fifteen-year period. There were twenty males and eleven females in the study with a mean follow-up of seventy-eight months. A total of thirty-nine arachnoid cysts were present in thirty-one patients, with six individuals having multiple cysts. The majority of patients (17/31) had intraventricular lesions. Thirty patients were diagnosed by radiographic imaging following the progression of a neurological deficit. All cysts were found to be increasing in size on serial scans.

Results: Eighty-seven percent of patients (27/31) had hydrocephalus on their initial imaging studies; while, four patients had normal ventricular size. Five patients were treated with shunting and aspiration of the cyst, with two of the cyst catheters being placed stereotactically. Thirteen patients underwent fenestration alone; whereas, another thirteen patients had craniotomy along with temporary external drainage. Post-operatively, an additional sixteen patients eventually required shunting. Hyponatremia or hypernatremia occurred in nineteen percent of patients; while, less common complications included transient hemiparesis, subdural hygroma, seizure and progressive visual loss. Two patients had recurrence of their arachnoid cysts.

Conclusion: Third ventricular arachnoid cysts can be approached safely with a minimal amount of morbidity or mortality. Even after shunt placement for hydrocephalus, the cyst can progressively enlarge and cause neurologic deterioration. Therefore, fenestration is important in the long term treatment of these cysts. With fenestration, thirty-nine percent (10/26) of the patients were able to avoid shunting.

**Benefits of Aggressive Fenestration Versus Limited Fenestration or Shunting of Enlarging Temporal Region Arachnoid Cysts**

Arun Amar (Los Angeles, CA)

The natural history of temporal region arachnoid cysts in children is variable. While some remain static, incidental findings, others may enlarge and exert local mass effects or contribute to hydrocephalus. The extent of communication between the cyst and the ventricles, which is similarly variable, is often difficult to assess preoperatively. This explains, in part, the poor results in some patients who merely undergo CSF diversion or limited fenestration into the Sylvian fissure. Recently, we have been aggressively treating these cysts by exposing the entire cyst wall and fenestrating them into the suprasellar and basilar cisterns. We compared the results with this approach to those with smaller fenestrations or initial attempts at shunting the cyst and/or ventricles.

Methods: Over the past 15 years, 42 children (31 males and 11 females) presented with enlarging temporal region arachnoid cysts. Six had associated hydrocephalus at presentation. The mean age was 65 months, and mean duration of follow-up was 56 months. 24 patients (group 1) were initially managed with shunting (5 patients) or limited fenestration (19 patients). The remaining 18 patients (group 2) underwent aggressive fenestration.

Results: Of the 5 patients undergoing initial shunting, 3 (60%) required subsequent fenestration. Of the 19 patients initially treated with limited fenestration, 10 (53%) required subsequent shunting, compared with only 2 of the 18 patients (11%) in group 2. Operative complications group 1 included 2 patients with transient hemiparesis and 1 patient requiring a subdural-peritoneal shunt. Those for group 2 included 2 patients with transient hemiparesis, 2 patients with transient CN III compromise, 1 post-operative seizure and 1 patient requiring a subdural-peritoneal shunt.

Discussion and conclusions: Aggressive fenestration of temporal arachnoid cysts minimizes the chance of needing a shunt and its attendant morbidity. Less aggressive attempts with initial shunting in the absence of hydrocephalus do not result in significant reduction of cyst size and may require subsequent craniotomy for fenestration. Patients with hydrocephalus at presentation, however, will likely need both a shunt and a fenestration.
One-Stage Aggressive Surgical Management of Pediatric Arachnoid Cysts

Paul Jensen (Columbus, OH)

The goal of surgical management of pediatric arachnoid cysts is to accomplish decompression by means of a single operative procedure that will not require additional revision and can be performed without morbidity. While it has been proposed that ideal management would leave a child treated for this disorder shunt independent, we feel that placement of cystoperitoneal shunt during initial cyst decompression is an important adjunct to the achievement of this goal.

We present a retrospective analysis of a large series of pediatric arachnoid cysts operated on by one neurosurgeon and describe the surgical strategy that has resulted in a near 100% long-term cyst decompression without need for subsequent reoperation. Between 1980 and 1996 over one hundred children with an arachnoid cyst were operated on by Dr. Edward Kosnik at Columbus Children’s Hospital. All patients underwent craniotomy to gain access to the cyst. Excision of as much cyst wall as possible under microscopic vision was performed in most patients. Finally, cystoperitoneal shunt was performed under direct vision and magnification to assure ideal location that would not be subject to occlusion by residual cyst membrane or collapsing cyst. A smaller number of patients primarily underwent excision alone, such as suprasellar and CP angle cysts, where shunt placement was not deemed optimal. All patients were followed up by CT scan analysis.

In our total series of pure arachnoid cyst patients, none required secondary surgery for shunt revision. In a few patients with other additional problems, a few required subsequent revision. There was no significant morbidity and no mortality.

We conclude that concomitant cystoperitoneal shunting with open arachnoid cyst decompression provides the optimal approach for treatment of pediatric arachnoid cysts.

Arachnoid Cysts in Children: How Do Postsurgical Cyst Size and Seizure Outcome Correlate?

Christian A. Koch (Worthington, OH), J. Layne Moore (Columbus, OH), Dieter Voth (Mainz, Germany)

Introduction: Arachnoid cysts (ACs) are congenital cystic brain malformations associated with epilepsy. The purpose of this study was to determine the effect of surgical intervention of ACs on cyst size and seizure outcome. Efficacy and safety of surgical treatment of ACs for seizures may depend not only on the mode of operation, cyst location, and patients’ age, but also on the postoperative AC size.

Methods: We reviewed the world’s medical literature dealing with surgically treated ACs in epilepsy patients. Our study only included children, in which the relationship between pre- and postoperative CT size of the AC and seizure outcome was described. We also included 3 of our own children with ACs and epilepsy treated surgically at the University of Mainz. Postoperative AC size and seizure outcome were analyzed with respect to mode of operation and cyst location.

Results: A total of 30 children was reviewed. Twenty (67%) had a smaller AC postoperatively. Sixteen of these 20 children (80%) experienced seizure improvement. Four patients (20%) remained unchanged. In 10 of the 30 children (33%) the AC size on postoperative CT was unchanged. Seven of these 10 (70%) improved. One child remained unchanged and 2 (20%) worsened. A positive correlation between postoperative AC size and seizure outcome was well demonstrated among children treated with cyst fenestration or needle aspiration. Among patients treated with cystoperitoneal shunting this direct correlation was less clear.

Conclusions: Seizure outcome correlates directly with postoperative AC size. Seizure reduction is associated with a decreased AC size postoperatively and depends on the mode of operation. Based on these data we would expect that children with epilepsy secondary to ACs would demonstrate improved seizure control with lower AC volume. Conversely, we might expect increasing AC size to correlate with worse seizure control. This relationship may guide physicians in efficacious and timely patient management.
72 Long-Term Outcome in Myelomeningocele—These Kids Are Doing Better Than You Think*

Michael Seiff, MD, Robert Keating, MD, Leonard Seimon, MD, Robert Marion, MD, James Goodrich, MD, PhD (Bronx and Valhalla, NY)

The consensus amongst clinicians is that myelomeningocele carries a poor prognosis. Many patients in fact have higher cognitive capacity and fewer delayed complications than believed. A multidisciplinary review of 123 patients with spinal dysraphism followed in an outpatient clinic of a large pediatric rehabilitation hospital was conducted, including 118 myelomeningocele patients. The study period ranged from 1969 to present, average follow-up was 10.03 years (7 months to 27 years).

There were 61 males and 62 females. Most defects were in the lumbosacral region, with 20.3% lumbar, 35.8% lumbosacral, and 12.2% sacral. Thoracic defects numbered 15.4%, and thoracoacetabular 16.3%. Fifty-one patients or 41.4% are independent community ambulators. Household ambulators number 14.6%, nonfunctional ambulators 12.2% and non-ambulators 28.5%. As expected, lower level correlated with higher grade of ambulation. Details on hydrocephalus were available for 99 patients, 14 of which required no shunting. Of the rest, 28 required no revision, and 32 were revised only once, nearly half being within the first 12 months. Tethered cords were released in 17 patients, and Chiari crisis with subsequent decompression occurred in 4 patients. Of 75 patients for which data on cognitive function was available, nearly 40% had normal intellectual function, 20% were mildly impaired, 35% were moderately delayed, and only 7% had profound MR. There were only three deaths; one secondary to Chiari crisis, one patient who died in her sleep following a headache, and one trauma.

Orthopaedic procedures to correct deformities and facilitate bracing or ambulation included soft tissue releases in 31 patients (25.2%) and osteotomies/hip repair in 39 patients (32%). Spinal Fusions were performed for kyphosis or scoliosis in 14 patients (11.3%). Urologic diversionary or augmentation procedures were performed in 17 patients (13.8%).

These results challenge the prevailing sentiment of poor outcome in myelomeningocele. As a result of the multidisciplinary approach in a clinic focusing on their unique problems, these patients can do quite well, living productive and meaningful lives.

73 The Variability of Familial Neural Tube Defects

Timothy M. George, MD, Marcy C. Speer, PhD, Chantelle M. Wolpert, BA, Joanne F. Mackey, RN, Herbert E. Fuchs, MD, PhD, Gordon Worley, MD (Durham, NC)

A major challenge in the study of genes causing neural tube defects (NTDs) is defining whether a mutant gene can result in a spectrum of developmental neural anomalies. The hereditary basis for the most frequent phenotypic expression of NTDs (myelomeningocele and anencephaly) is well documented by an increased risk to relatives of probands; however, the occurrence of NTD in families with myelomeningocele and/or other types of neural anomalies (e.g., fatty filum terminale, lipomyelomeningocele, encephalocele, Chiari malformation, spina bifida occulta and others) not commonly grouped together has rarely been reported. We collected family histories and imaging studies on four families with a spectrum of NTDs. In these families, we found an association between myelomeningocele, spina bifida occulta, Chiari I malformation, and fatty filum terminale. As in other reported series, the genetic factor followed maternal lines. Not all family members were affected indicating incomplete penetrance or a non-dominant inheritance pattern. These families provide supporting evidence in favor of a genetic predisposition to a spectrum of NTDs, although environmental causes or chance cannot be ruled out. Based on these cases, we conclude that the range of NTDs may be broad and familial studies should be aimed at targeting a spectrum of neural defects to determine their genetic significance. This hypothesis will be tested as molecular probes for candidate genes become available. A detailed description of each family will be presented.
74 Pathogenetic Classification of Spinal Cord Anomalies: A Systematic Approach

Michael D. Parrington, MD (Denver, CO), Mark S. Dias, MD (Buffalo, NY), David G. McLone, MD, PhD (Chicago, IL)

For more than a century, spinal cord anomalies have been classified primarily on the basis of their final gross morphology. The best-known example of this is the open vs closed classification for spina bifida. In recent years, a wealth of new information on the pathogenesis of these lesions has been developed from both clinical and laboratory research and has provided new insights into the inter-relationships between these lesions. For this reason, we propose that these anomalies should be classified according to the point during embryogenesis from which they are derived. The major divisions of spinal embryogenesis (i.e., gastrulation, neurulation, differentiation, and post-neurulation) can then be used as a framework for this classification.

Lesions of gastrulation include those lesions associated with the formation of the notochord (i.e., lumbosacral agenesis, split notochord, split cord malformation, neurenteric cyst and others). Primary neurulation would include the defects of the upper neural tube, such as myelomeningocele, lipomyelomeningocele and dermal sinus tract. Secondary neurulation defects are primarily those involving derivatives of the caudal cell mass, such as sacral agenesis or fatty filum. Post-neurulation defects are rare in the spinal cord, but possible examples would include meningocele and terminal myelocystocele.

A classification scheme of this type should prove useful both in the design of future experimental efforts and in clinical work.

75 Surgical Decision Making in Tethered Cord Syndrome: A Role for Thoracolumbar Phase Motion MRI

J. Michael Desalons, MD, Zeev T. Feldman, MD, Charles W. McCluggage, MD, Robert C. Dauser, MD, John P. Laurent, MD (Houston, TX)

The purpose of this retrospective study is to evaluate the role of phase motion MRI in deciding whether to untether patients with tethered cord syndrome. In addition to standard stationery MR imaging, cine gradient-echo examinations (phase motion MRI) were performed on the thoracolumbar spinal cord in 34 children with spinal dysraphism. In 12 patients with previously closed myelomeningoceles, 4 showed significant motion on cine MRI while 8 showed lack of motion. Of 22 patients with other forms of dysraphism, mostly lipomas, 11 showed motion and 11 did not. Overall, only 2 of 15 patients with preserved motion underwent untethering procedures as opposed to 14 of 19 without motion (p=0.005, Fisher Exact Test).

Of the myelomeningocele group, 8 patients were symptomatic on presentation; 2 showed motion and 6 did not. Surgery was avoided in the 2 patients with motion while all 6 without motion underwent untethering (p=0.03). In the symptomatic lipoma group (n=15), 1 of 6 with motion and 7 of 9 without motion underwent untethering (p=0.04). Of significant note, the symptomatic patients foregining surgery had generally mild symptoms, most experiencing paresthesias. Clinical outcome of the patients treated conservatively (n=7) showed improvement in 4 and non-progression in 2 (follow-up 4 months to 2 years). One patient was lost to follow-up. In case by case review, phase motion MRI directly influenced surgical decision making in 9 of 34 patients (26.5%).

Phase motion MRI of the thoracolumbar spinal cord influences surgical decision making in spinal dysraphic patients at our institution. Patients with lipomas and those with mild symptoms are more likely to demonstrate residual motion.
SCIENTIFIC POSTERS*

*Considered for Shulman Award.
1. A New Model of Human Medulloblastoma in the Nude Mouse

Daniel R. LeMay, MD, PhD, Toby MacDonald, PhD, Ronnie I. Mimran, BS, Keith A. Harvitz, BS, J. Gordon McComb, MD, Martin H. Weiss, MD, Berislav V. Zlokovic, MD, PhD (Los Angeles, CA)

Introduction: Animal models of CNS tumors are tools which have been extremely useful in allowing researchers to explore the genetics, pathophysiology and treatment of brain tumors. We describe a new model of the human medulloblastoma in the nude mouse which will provide an additional tool for these investigations.

Methods: A human medulloblastoma cell line was isolated and grown in vitro. The immunodeficient nude mouse was the animal model chosen because of the need to avoid a foreign tissue immune response to human cells. Day 1 mice (n=3) were stereotactically implanted with $10^4$ human medulloblastoma cells in 10 uliters into the right frontal lobe 4mm lateral to the sagittal suture along the coronal suture at 5mm depth. On day 21 mice were sacrificed and the brain was removed and immersion fixated in formalin. The brains were sectioned with a vibratome into 100 micron thick coronal slices, stained with H+E, and area analysis of the tumor was performed using an image analyzer. In each rat the slice containing the largest area was compared.

Results: At 18 days post-implantation tumor grew to a mean area of 1.16 mm$^2$ ± .27 SE.

Discussion and Conclusion: These results demonstrate that human medulloblastoma cells grow in the nude mouse brain and therefore a new animal model of human brain tumor is available for the study of medulloblastoma.
2 Cerebellar Astrocytoma in Pediatric vs. Adult Patients

Vittorio Morone, Michael J. Ebersold, Lynn M. Quast, Joseph E. Parisi
(Rochester, MN)

In this paper, we compare the tumor characteristics, treatment, and outcome of pediatric and adult patients with cerebellar astrocytomas. We specifically used objective CT and/or MRI data to assess the degree of surgical resection and tumor recurrence.

A retrospective analysis was done on the clinical records of 54 patients who were surgically treated for cerebellar astrocytoma from 1978-1990. All patients had all their surgery done at Mayo Rochester. Follow-up was done through a combination of phone calls and mailed questionnaires.

Patients' ages ranged from 1 to 80 years old with a mean age of 26. One-half of the patients were under 15 years old. Forty-eight percent were males and 52 percent were females. Seventy-two percent of tumors were pilocytic astrocytomas, 17% were grade 4 astrocytomas, 6% were grade 3 astrocytomas, 4% were low grade non-pilocytic astrocytomas, and 1% were gliosarcomas. Eighty-five percent of pilocytic astrocytomas were cystic. Most patients presented with headache, nausea, vomiting, and ataxia. Gross total resection was obtained in 35 out of 54 patients. Nineteen patients had residual tumor and 4 patients had tumor recurrence as documented by postoperative CT or MRI scans.

Patient survival is strongly dependent on tumor pathology. Pilocytic astrocytoma had the longest 5- and 10-year survival rates. Long symptom-free postoperative survival can be had with subtotal removal of tumor when the tumor is of low grade. No benefit from postoperative radiation was observed. We recommend gross total resection when possible.

3 Should Age Be the Deciding Factor in Determining the Need for Arteriography to Exclude Aneurysm in Children with Oculomotor Palsy?

Mark S. Dias, MD, Imtiaz A. Mehkri, MD, Steven Awner, MD, Scott E. Olitsky, MD (Buffalo, NY)

A posterior communicating artery aneurysm as a cause of isolated oculomotor palsy in children younger than 14 years is extremely rare; to date, only one such child, aged 11 years, has been described in the literature. Several references in the ophthalmology literature have suggested that arteriography to exclude an aneurysm is unnecessary in this population. We present a 10-year-old boy with an isolated complete oculomotor palsy caused by a posterior communicating artery aneurysm. At presentation, the child had a three day history of headache, followed by diplopia and unilateral ptosis. Examination disclosed a complete unilateral third nerve palsy with proptosis, mydriasis, and extraocular palsies. MRI and magnetic resonance angiography suggested a lesion in the region of the posterior communicating artery. A cerebral arteriogram confirmed a 5 mm posterior communicating artery aneurysm directed posterolaterally. At surgery, there was no evidence of subarachnoid hemorrhage and the aneurysm was successfully clipped. The child made a complete recovery.

Although rare, the potentially devastating consequences of aneurysm rupture and the low complication rate of cerebral angiography in children mandate that an intracranial aneurysm be excluded as a source of the oculomotor palsy regardless of age. We recommend that young children with unilateral oculomotor palsy involving the pupil in whom another cause is not apparent undergo cerebral angiography to exclude an aneurysm.
4 Subdural Fluid as an Indication of Hydrocephalus

Michael H. Handler, MD (Denver, CO)

Ventricular enlargement is the hallmark of impaired CSF dynamics and elevated pressure in hydrocephalus. We treated five patients in whom the ventricular system had been opened to the subdural space, three by a transcortical resection of an intraventricular lesion, and two by placement of a ventriculoperitoneal shunt. When they developed signs and symptoms of elevated intracranial pressure, they also developed progressive enlargement of subdural collections with evidence of mass effect, not necessarily accompanied by progressive ventricular enlargement. All had resolution of symptoms, and four had complete resolution of the subdural, with a correctly functioning VP shunt. If the ventricles can drain to the subdural space, symptomatic hydrocephalus may manifest itself by subdural fluid collections on imaging, rather than by ventriculomegaly.

5 Cerebral Gnathostomiasis

John R. Mawk, MD (Portland, OR)

Only two parasites are known to move after their larval stages embolize the central nervous system - *Angiostrongylus*, which migrates in the subarachnoid space, and *Gnathostoma*, which has a propensity to traverse the parenchyma. The latter parasite is the subject of Victorian era horror literature.

An eight-year-old boy visited the South Pacific and ate a meal probably containing partially cooked fish. Four months later, he developed severe, lancinating headaches with partial visual loss. These headaches came on very suddenly every few weeks, lasted for days, and terminated suddenly. A CT scan demonstrated no tumor. After a period of many months they ceased without recurrence. Ten years later, the boy presented to a trauma service after a fall, and CT scanning demonstrated a small temporal lobe cyst. Magnetic resonance scanning demonstrated a serpiginous tract winding through the basal ganglia and terminating in the cyst. This is believed to represent a case of cerebral gnathostomiasis.

The popular literature of a century ago, the pertinent medical literature and treatment options are outlined. To have one's brain consumed by a worm from within remains without question the horrifying notion it was at the time the disease was first described.
6 Intraventricular Urokinase for the Treatment of Posthemorrhagic Hydrocephalus: Does a Fibrinolytic State Prevent Need for Shunting?

Joseph Madsen (Boston, MA)

Posthemorrhagic hydrocephalus is a common problem in premature infants, so a medical intervention which would avoid the need for shunting could considerably decrease shunt-related morbidity. Intraventricular fibrinolytic therapy has been proposed as means to this end, but major questions have not been addressed: is a fibrinolytic state actually achieved in neonates treated with these drugs, does fibrinolysis still occur if intermittent drainage is used with the infusion, and, if fibrinolysis is achieved, does this prevent need for a shunt? To approach these questions with respect to intraventricular urokinase, we conducted a prospective clinical trial conducted at three academic hospitals. Six prematurely born infants with medically refractory progressive hydrocephalus requiring treatment with ventricular drains received alternating infusion of intraventricular urokinase with drainage of CSF for three days. Of the six treated patients, median gestational age at birth was 26.5 weeks and the median age at treatment was 30 days (range 26 to 37). No patient had evidence of secondary bleeding, systemic absorption, or side effects. Fibrinolysis was achieved in the cerebrospinal fluid, as documented by markedly elevated D-dimer levels (median 1841 µg/l, range 83 to 9528 µg/l, normal plasma range 4-78 µg/l). Clot size appeared to diminish by ultrasound evaluation in at least one patient. In spite of the fibrinolysis, all six patients eventually required a VP shunt based on conservative prospective clinical criteria. We conclude that intermittent infusion of intraventricular urokinase alternating with periods of CSF drainage is a safe way to effectively achieve a fibrinolytic state. However, when administered at the relatively late point when a ventricular drain is required, this fibrinolytic state is not sufficient to decrease the rate of VP shunt.

7 A Novel Method of Cranioplasty Using Coralline Hydroxyapatite

SooHo Choi, MD, Michael L. Levy, MD, J. Gordon McComb, MD (Los Angeles, CA)

Current materials used for cranioplasty include autologous or homologous bone grafts, wire mesh, and methyl methacrylate, either alone or in combination. However, each material has its own unique disadvantages.

Although coralline hydroxyapatite has been used extensively for orthopedic, ophthalmologic, craniofacial and dental procedures as a bone substitute, it has not been used to repair cranial bony defects. Coralline hydroxyapatite, a substance similar to that found in autologous or homologous bone grafts provides a matrix in which to form a living structure. The hydroxyapatite comes as granules which are mixed with Avitene, thrombin and autologous blood to form a paste which can be shaped as needed.

Over the past few years we have used hydroxyapatite either alone or applied to the surface of tantalum mesh in 17 neurosurgical patients, ranging in age from 4 months to 19 years, for a variety of conditions. The cosmetic results were good in all. Other than one patient extruding granules over several months, there were no complications.

In neurosurgical procedures when a bone substitute is needed, hydroxyapatite is an effective alternative to materials currently being used.
8 Prevention of Spinal Cord Retethering Using Free Dermal Fat Grafts

Michael R. Egnor, MD, Mark D. Epstein, MD (Stony Brook, NY)

Retethering of the spinal cord following repair of myelomeningocele or other spinal surgery in children is a common problem, and a variety of operations have been used to minimize this problem. Closure with fascia lata or cadaver dura, while providing good CSF leak prevention, is associated with frequent retethering. Some pediatric neurosurgeons have described reconstruction of the spinal canal to create a large CSF sac, and while this technique is promising, it is technically demanding and the results are mixed at best. Synthetic grafts may reduce scar tissue formation, but infection and CSF leak have been limited to this approach.

In an effort to provide good tissue coverage and minimize scar formation, plastic surgeons have used free dermal fat grafts, which helps the fat graft develop a blood supply and minimizes graft resorption. Eighteen months ago, we began using autologous free dermal fat grafts for children with recurrent spinal cord retethering. Nine children have undergone the procedure, and no child has had further retethering, CSF leak or infection. One child developed scar tissue formation above the graft site which required extension of the graft.

The potential long term problems associated with our procedure are graft resorption, epithelial cyst formation, and growth of the iatrogenic “lipoma”, although we have not seen these problems thus far. Our early results are encouraging, and we suggest this method of dural repair be considered for children with particularly difficult recurrent tethering or CSF leaks.

9 The Relationship between Intraoperative Hypothermia and Ventricular Shunt Infections

Peter C. Gerszten, MD, A. Leland Albright, MD, Ian F. Pollack, MD, P. David Adelson, MD (Pittsburgh, PA)

Ventricular shunt infections continue to be a major problem in the management of pediatric neurosurgery patients. Several recent studies have demonstrated a positive relationship between intraoperative hypothermia and postoperative infection. Perioperative hypothermia may promote infection by triggering thermoregulatory vasoconstriction, thus decreasing subcutaneous oxygen tension and impairing oxidative killing by neutrophils. In addition, hypothermia directly impairs immune function.

Between 1990 and 1996, 68 children who underwent ventricular shunt placement at our institution subsequently developed a shunt infection (overall shunt infection rate of 4%). Mean age was 8 years (range, neonate to 20 years). Shunt infections related to a primary abdominal process or wound dehiscence were excluded. The last 74 children who underwent ventricular shunt placement without subsequent infection served as a control group. The anesthetic record of these children were reviewed for lowest core temperature recorded during the surgical procedure. The lowest core temperature varied from 33.9°C to 37.7°C (mean 36.0°C). Hypothermia was defined as a temperature less than 35.1°C. Ten patients each in both the infected group and the control group were hypothermic during surgery.

No relationship was found between hypothermia and the subsequent occurrence of a shunt infection (p=0.45). Given that infants are more prone to hypothermia during surgical procedures, those children less than 2 years old were then excluded from analysis. Among children over 2 years old (n=88), there was a trend towards the occurrence of shunt infections following intraoperative hypothermia which did not reach statistical significance (p=0.07). Therefore, in this series we found no clear relationship between intraoperative hypothermia and the development of shunt infections. Perhaps a larger series of patients would demonstrate that such a relationship indeed exists.
Use of the ISG Wand in the Posterior Fossa

Rick Abbott, MD (New York, NY)

Traditionally, use of frameless stereotaxis in the posterior fossa has been discouraged due to the high degree of inaccuracy in referencing of that region of the patient's anatomy to the imaging studies using fiducials placed about the forehead and ears. We have found that using ISGs surface fit algorithm the accuracy of the referencing is more than acceptable and has allowed us to perform more aggressive surgeries within the brain stem as well as develop new treatments for trapped CSF spaces within the posterior fossa. After correlating five fixed anatomical points on the patient to the same points on their imaging studies, 50 randomly selected points about the scalp were touched with the system's pointer. These points included sites in the suboccipital and retromastoid regions. The system then generated a topographical cloud made up of these points and performed a least square fit with the 3-dimensional reconstruction of the imagery data set to register the patient to the image set. We found that this gives a very accurate registration with the brain stem which is maintained during tumor resections within the stem. We have also found the registration to be accurate enough to allow us to place catheters via a coronal burr hole into fluid spaces within the posterior fossa which are greater than 2 cm in diameter. This technique will be discussed and case examples shown.
Abbassy, Munir H., MD
80 Congress St.
Springfield, MA 01104
Active Member

Andrews, Brian T., MD
UCSF/Dept. of Neurosurgery
2100 Webster St., Suite #521
San Francisco, CA 94115
Active Member

Beatty, Robert M., MD
8919 Parallel Pkwy.
Suite #455
Kansas City, KS 66112
Active Member

Abbott, Ira R., III MD
550 First Ave./Ped. Neuro.
New York, NY 10016
Active Member

Akins, Thomas J., MD
Neurosurgical Associates
60 Temple St.
New Haven, CT 06510
Active Member

Bell, William O., MD
Carolina Neuro Assoc., PA
2810 Maplewood Ave.
Winston-Salem, NC 27103
Active Member

Adelson, P. David, MD
Children's Hosp. of Pittsburgh
3705 Fifth Ave./Neurosurgery
Pittsburgh, PA 15213
Active Member

Aronin, Patricia A., MD
521 Dubuque St.
Northville, MI 48177
Active Member

Benzel, Edward C., MD
Univ. of New Mexico
Div. of Neurosurgery
Albuquerque, NM 87131
Active Member

Albright, A. Leland, MD
Children's Hosp., Pittsburgh
3705 Fifth Ave./Neurosurgery
Pittsburgh, PA 15213
Active Member

Arpin, Elaine J., MD
S.W. Florida Neuro. Assoc.
413 Del Prado Blvd., Ste. #102
Cape Coral, FL 33990-5710
Active Member

Berger, Mitchel S., MD
Univ. of Washington Med. Ctr.
1859 N.E. Pacific St., #359470
Seattle, WA 98195
Active Member

Altenau, Lance, MD
501 Washington
Suite #700
San Diego, CA 92103
Active Member

Bailey, Walter L., MD
Doctors' Professional Bldg.
280 N. Smith Ave., Suite #234
St. Paul, MN 55102
Active Member

Berger, Thomas S., MD
Mayfield Neurological Clinic
590 Oak St.
Cincinnati, OH 45219
Active Member

Amacher, A. Loren, MD
Gelinsger Clinic
Dept. of Neurosurgery
Danville, PA 17822
Active Member

Ball, Gene A., MD
3003 E. Fletcher Ave.
Suite #340
Tampa, FL 33613-4645
Active Member

Bierbrauer, Karin S., MD
St. Christopher's Hosp.
Erie & Front St./Ped. Neuro.
Philadelphia, PA 19134
Active Member

Amador, Luis V., MD
1440 Veteran Ave., #336
Los Angeles, CA 90024
Lifetime Member

Barrer, Steven J., MD
1584 Old York Rd.
Abington, PA 19001
Active Member

Black, Peter McL., MD PhD
Children's Hosp./Neuro.
300 Longwood Ave.
Boston, MA 02115
Active Member

Anderson, Jim D., MD
Redwood City Offices
1150 Veterans Blvd.
Redwood City, CA 94063
Active Member

Bartkowski, Henry M., MD PhD
Neurological Assoc.
931 Chatham Ln.
Columbus, OH 43221
Active Member

Boggs, John Scott, MD
1820 Barry St.
Suite #104
Jacksonville, FL 32204
Active Member
Boop, Frederick A., MD
Arkansas Children's Hosp.
800 Marshall St., Neurosurgery
Little Rock, AR 72202
Active Member

Bressler, Bruce C., MD
720 S. Van Buren
Green Bay, WI 54301
Active Member

Brockmeyer, Douglas L., MD
Primary Children's Med. Ctr.
100 N. Medical Dr.
Salt Lake City, UT 84113-1100
Active Member

Chapman, Paul H., MD
Massachusetts Gen. Hosp.
Fruit St., GB8502
Boston, MA 02114
Active Member

Cheek, William R., MD
2809 Robinhood
Houston, TX 77005
Lifetime Member

Choux, Maurice, MD
Hopital Des Enfants
De La Timone, Cedex 5
Marseille 13005
France
International Member

Cogen, Philip Harry, MD PhD
Children's National Med. Ctr.
111 Michigan Ave., N.W./Neuro.
Washington, DC 20010
Active Member

Cohen, Alan R., MD
Case Western Reserve Univ.
11100 Euclid Ave./Neuro.
Cleveland, OH 44106
Active Member

Constantini, Shiomi, MD
Dana Children's Hosp.
6 Wolman St./Fed. Neurosurg.
Tel-Aviv, 64239
Israel
International Member

Coulson, Richard A., Jr., MD
OrthoClinicNeuro.
1514 Jefferson Hwy.
New Orleans, LA 70121
Active Member

Cron, Kerry R., MD
3333 Burnet Ave.
Cincinnati, OH 45229-3039
Active Member

Dagi, T. Forcht, MD
933 F. Johnson Ferry Rd.
Suite #100
Atlanta, GA 30342
Active Member

Davidson, Robin I., MD
Univ. Massachusetts Med. Ctr.
55 Lake Ave., N./Neuro.
Worcester, MA 01655
Active Member

Di Rocco, Concezio, MD
Univ Collochia/Neurochirurgia
Largo Gemelli 8
Rome, 00168
Italy
International Member

Dias, Mark S., MD
Children's Hosp. of Buffalo
219 Bryant St.
Buffalo, NY 14222
Active Member

Doreen, Michael, MD
The Neurog. Group of Austin
2005 San Gabriel, #310
Austin, TX 78705
Active Member

Drake, James M., MD
Hosp. for Sick Children
555 University Ave., #1504-D
Toronto, ON M5G 1X8
Canada
Active Member

Duhamel, Ann-Christine, MD
Child. Hosp. of Philadelphia
34th & Civic Ctr. Blvd./Neuro.
Philadelphia, PA 19104
Active Member

Edwards, Michael S. B., MD
2100 Webster St.
Suite #420
San Francisco, CA 94115
Active Member

Eisenberg, Howard M., MD
Div. of Neurosurgery
22 S. Greene St., #1101
Baltimore, MD 21201-1734
Active Member

Epstein, Fred J., MD
550 First Ave.
New York, NY 10016
Active Member

Epstein, Mel H., MD
2 Dudley St.
Suite #505
Providence, RI 02905
Active Member

Erasmus, Mark D., MD
502 Lomas Blvd., N.E.
Albuquerque, NM 87102
Active Member

Fallace, Walter J., MD
Univ. Med. Ctr.
653 W. Eighth St.
Jacksonville, FL 32209
Active Member

Fallon, Robert J., MD
8707 S. Yale
Tulsa, OK 74136-3303
Active Member

Fell, David A., MD
Neurological Surg. Inc.
6767 S. Yale
Active Member

Fischer, Edwin G., MD
110 Francis St.
Suite #3B
Boston, MA 02215
Active Member

Flannery, Ann Marie, MD
Med. Coll. of Georgia
1120 15th St./Neurosurgery
Augusta, GA 30912-4010
Active Member

Foltz, Eldon L., MD
UCI Med. Ctr./Neurosurgery
PO. Box 14091
Orange, CA 92613-4091
Active Member

Freed, Michael H., MD
825 Washington St.
Suite #100
Norwood, MA 02062
Active Member

French, Kathleen B., MD
3020 Hamaker Ctr.
Suite B104
Fairfax, VA 22031
Active Member

Gahm, Norman H., MD
100 Retreat Ave.
Suite #705
Hartford, CT 06116
Active Member

Galitsch, Joseph H., MD
P.O. Box 276
Alpine, NJ 07820
Lifetime Member

Gamache, Francis W., Jr. MD
Alten Neurosci. Inst.
553 E. 72nd St.
New York, NY 10021
Active Member

Gennuso, Rosemaria, MD
35 Calvert Ave., E.
Edison, NJ 08820
Active Member

Gerszten, John C., MD
1015 Chestnut St.
Suite #1400
Philadelphia, PA 19107
Active Member

Buchheit, William A., MD
1015 Chestnut St.
Suite #1400
Philadelphia, PA 19107
Active Member

Cahan, Leslie D., MD
1505 N. Edgermont St.
Neurosurgery/Room #4141
Los Angeles, CA 90027
Active Member

Canady, Alexa Irene, MD
Children's Hosp. of Michigan
3001 Beaubien
Detroit, MI 48201
Active Member
Hamilton, Mark G., MD  
Alberta Children's Hosp.  
1020 Richardson Rd., S.W.  
Calgary, AB T2T 5C7  
CANADA  
Active Member

Hammargren, Lonnie L., MD  
3196 S. Maryland Pkwy.  
Las Vegas, NV 89109  
Active Member

Hammock, Mary Kathryn, MD  
8680 Sudley Rd.  
Suite #200  
Manassas, VA 20110-4416  
Active Member

Handler, Michael H., MD  
1010 E. 19th St.  
Suite #405, Tamman Hall  
Denver, CO 80218  
Active Member

Hanigan, William C., MD PhD  
214 N.E. Glen Oak Ave.  
Suite #500  
Peoria, IL 61603  
Active Member

Hawkins, John C., III MD  
2545 Rivercliff Ave.  
Jacksonville, FL 32204  
Active Member

Hofbrook, Thomas J., Jr. MD  
3 Medical Park Rd.  
Suite #310  
Columbia, SC 29203  
Active Member

Hofmann, Harold J., MD  
Hosp. for Sick Children  
555 University Ave., #1504  
Toronto, ON M5G 1X8  
CANADA  
Active Member

Holstein, Robert D., MD  
McMaster Univ. Med. Ctr.  
Dept. of Surgery/Roentgen #404  
Hamilton, ON L8N 3C5  
CANADA  
Active Member

Hudgens, Roger, MD  
5455 Meridian Mark Rd.  
Suite #540  
Atlanta, GA 30342  
Active Member

Hutcheson, D.A., MD  
642 West 8th St.  
Daytona Beach, FL 32114  
Active Member

Humphreys, Robin P., MD  
Hosp. for Sick Children  
555 University Ave., #1504  
Toronto, ON M5G 1X8  
CANADA  
Active Member

Jones, Robert F. C., MD  
21 Norfolk St.  
Paddington, NSW 2021  
Australia  
International Member

James, Hector E., MD  
7790 Frost St.  
Suite #304  
San Diego, CA 92123  
Active Member

Jenneck, John A., MD PhD  
Univ. of Virginia  
Neuro./Hosp. Box 212  
Charlottesville, VA 22908  
Active Member

Johnson, Dennis L., MD  
Milton Hershey Med. Ctr.  
PO. Box 850Dx of Neuro.  
Hershey, PA 17033  
Active Member

Johnson, John K., MD  
Greenville Neuro. Group  
27 Memorial Medical Dr.  
Greenville, SC 29605  
Active Member

Johnson, Mary M., MD  
4516 Chambers Dunwoody Rd.  
#347  
Atlanta, GA 30338-6232  
Lifetime Member

Jones, Robert F. C., MD  
21 Norfolk St.  
Paddington, NSW 2021  
Australia  
International Member

Jones, Robert F. C., MD  
21 Norfolk St.  
Paddington, NSW 2021  
Australia  
International Member

Joseph, Allen S., MD  
7777 Hannessy Blvd.  
Suite #1000  
Baton Rouge, LA 70808  
Active Member

Kalsbeck, John E., MD  
Riley Children's Hosp.  
702 Barnhill Dr.  
Indianapolis, IN 46202-5200  
Active Member

Kanvey, Paul M., MD  
2793 W. Grand Blvd.  
Detroit, MI 48202  
Active Member

Kasper, Samuel S., MD  
Munger Pavilion, Room #329  
Vailhalla, NY 10595  
Active Member

Kaufman, Bruce A., MD  
St. Louis Children's Hosp.  
1 Children's Pl./Neuro.  
St. Louis, MO 63110  
Active Member

Kelly, David L., Jr. MD  
Boman Gray Sch. of Med.  
Medical Center Blvd./Neuro.  
Winston-Salem, NC 27157-1229  
Active Member

Kleiher, Laurence I., MD  
5304 Indian Grave Rd.  
Suite A  
Roanoke, VA 24014  
Active Member

Kosnik, Edward J., MD  
Chatham Village Prof. Brig.  
931 Chatham Ln.  
Columbus, OH 43221  
Active Member

Krisil, David S., MD  
Dr. Pod. Neuro./Room 5700H  
Loma Linda Univ Childrens Hosp  
Loma Linda, CA 92354  
Active Member

Krin, Merrin R., MD  
168 Oakdale St.  
Redwood City, CA 94062  
Active Member

Kunl, Joseph S., MD  
210 Farnsworth St.  
Stoughton, MA 02072  
Active Member

Lawrent, John R., MD  
Clinical Care Ctr.  
3–3435  
6621 Fannin, Suite 1950  
Houston, TX 77030  
Active Member

Lawler, Robert F., Jr. MD  
Univ. of Virginia/Neuro.  
Health Sciences Ctr., Box 212  
Charlottesville, VA 22908  
Active Member

Lipow, Kenneth L., MD  
287 Grant St.  
Bridgetown, CT 06610  
Active Member

Lipow, Kenneth L., MD  
287 Grant St.  
Bridgetown, CT 06610  
Active Member
<table>
<thead>
<tr>
<th>Name</th>
<th>Address/Location</th>
<th>Phone/Other Info</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tomita, Tad, MD</td>
<td>Children's Mchn. Hosp. 3300 Children's Plaza</td>
<td></td>
</tr>
<tr>
<td>Tullpan, Noel, MD</td>
<td>8533 McCreary Ln. Nashville, TN 37229</td>
<td></td>
</tr>
<tr>
<td>Turner, Michael S.,</td>
<td>1831 N. Senate Blvd. Suite #535 Indianapolis, IN 46202</td>
<td></td>
</tr>
<tr>
<td>Uscinski, Ronald H.,</td>
<td>3301 Wootburn Rd., #209 Annandale, VA 22003</td>
<td></td>
</tr>
<tr>
<td>Veres, Joan L., MD</td>
<td>27517 Via Montoya San Juan Capistrano, CA 92675</td>
<td></td>
</tr>
<tr>
<td>Ventureyra, Enrique C, MD</td>
<td>401 Smyth Rd. Ottawa, ON K1H 8L1 CANADA</td>
<td></td>
</tr>
<tr>
<td>Vries, John Kanrie, MD</td>
<td>Unio of Pittsburgh 217 Victoria Blvd. Pittsburgh, PA 15261</td>
<td></td>
</tr>
<tr>
<td>Wald, Steven L., MD</td>
<td>10 Grove Ln. Shelburne, VT 05482</td>
<td></td>
</tr>
<tr>
<td>Waldman, John B., MD</td>
<td>Albany Med. Coll. Div of Neurosurgery/A-61-NE Albany, NY 12208 Active Member</td>
<td></td>
</tr>
<tr>
<td>Walker, Marion L.,</td>
<td>Primary Children's Med. Ctr. 100 N. Medical Dr. Salt Lake City, UT 84113-1100</td>
<td></td>
</tr>
<tr>
<td>Winfield, Jeffrey A., MD PhD</td>
<td>Hill Med. Ctr. 1000 E. Genesee St., Ste. #202 Syracuse, NY 13210 Active Member</td>
<td></td>
</tr>
<tr>
<td>Werd, John D., MD</td>
<td>Virginia Commonwealth Univ. Neuro., MCV Station, Box 831 Richmond, VA 23298</td>
<td></td>
</tr>
<tr>
<td>Weiss, Martin H., MD</td>
<td>LAC/USC Med. Ctr., Box 786 1200 N. State St., Room 5046 Los Angeles, CA 90033</td>
<td></td>
</tr>
<tr>
<td>Wernick, Shelley, MD</td>
<td>2550 W. Villard Ave. Room #101 Milwaukee, WI 53209 Active Member</td>
<td></td>
</tr>
<tr>
<td>White, Robert Joseph, MD PhD</td>
<td>MetroHealth Med Ctr. 2500 MetroHealth Dr. Cleveland, OH 44109-1988 Active Member</td>
<td></td>
</tr>
<tr>
<td>Yamada, Shokei, MD</td>
<td>Loma Linda Univ. Sch. of Med. Room #2033/Neurosurgery Loma Linda, CA 92840 Active Member</td>
<td></td>
</tr>
<tr>
<td>Zakalik, Karol, MD</td>
<td>Wm. Beaumont Hosp. 3535 W. 13 Mile Rd., Suite 504 Royal Oak, MI 48073 Active Member</td>
<td></td>
</tr>
<tr>
<td>Zampella, Edward J., MD</td>
<td>10 Parrott Mill Rd. PO. Box 808 Chatham, NJ 07928 Active Member</td>
<td></td>
</tr>
<tr>
<td>Zavala, L. Manuel, MD</td>
<td>2147 Mowry Ave. Suite D-1 Fremont, CA 94538 Active Member</td>
<td></td>
</tr>
<tr>
<td>Zaretti, Paul Henry, MD</td>
<td>5920 Saratoga Blvd. Suite #440 Corpus Christi, TX 78414-4108 Active Member</td>
<td></td>
</tr>
<tr>
<td>Zovickian, John G., MD</td>
<td>3000 Colby St. Suite #101 Berkeley, CA 94705 Active Member</td>
<td></td>
</tr>
</tbody>
</table>
NOTES

Health Maintenance Scores

Class III Outcome Assessment
Level Spec Ed - Respite
Self Care ED
Days School Missed

John Kristin's Skills

Review Complications This Week