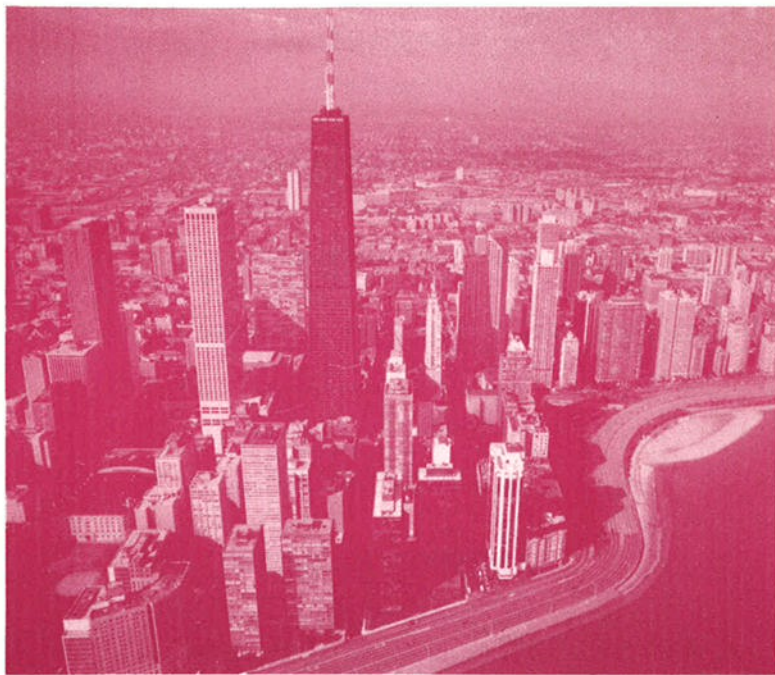


**Section of Pediatric Neurological Surgeons
of the
American Association of Neurological Surgeons**

16th WINTER MEETING



**The Palmer House
Chicago, Illinois
December 8-11, 1987**

PROGRAM SUMMARY

Paolo Raimondi Lecturers

Shulman Award

Pediatric Section Chairman

Pediatric Annual Meeting Sites

Exhibitors

16th Winter Meeting Scientific Program

16th Winter Meeting Scientific Abstracts

Pediatric Section Member Listing

This program has been approved by the
Joint Committee on Education of the
American Association of Neurological
Surgeons and Congress of Neurological
Surgeons for a maximum of 20 hours of
Category I credit toward the Continuing
Education Award in Neurosurgery.

PROGRAM COMMITTEE

William Cheek

David G. McLone

Michael Scott

Jack Walker

PAOLO RAIMONDI LECTURERS

E. Bruce Hendrick	1978	(Panel Discussion)	1982
Paul C. Bucy	1979	Derek Harwood-Nash	1983
Floyd Gilles	1980	Anthony E. Gallo, Jr.	1984
(Panel Discussion)	1981	Frank Nulsen	1985
	William F. Meacham	1986	

SHULMAN AWARD

Kim Manwaring	1983
Neonatal Post-hemorrhagic Ventriculomegaly: Management with Pulsed Lumbar Cisternostomy	
Arno Fried	1984
A Laboratory Model of Shunt Dependent Hydrocephalus	
Anne-Christine Duhaime	1985
The Shaken Baby Syndrome	
Robert E. Breeze	1986
CSF Formation in Acute Ventriculitis	

PEDIATRIC SECTION CHAIRMEN

Robert L. McLaurin	1972-73	Luis Schut	1978-79
M. Peter Sayers	1973-74	Fred Epstein	1979-81
Frank Anderson	1974-75	Joan L. Venes	1981-83
Kenneth Shulman	1975-76	Harold J. Hoffman	1983-85
E. Bruce Hendrick	1976-77	William R. Cheek	1985-87
Frank Nulsen	1977-78	David G. McLone	1987-89

PEDIATRIC ANNUAL MEETING SITES

Cincinnati	1972	New York	1980
Columbus	1973	Dallas	1981
Los Angeles	1974	San Francisco	1982
Philadelphia	1975	Toronto	1983
Toronto	1976	Salt Lake City	1984
Cleveland	1977	Houston	1985
Philadelphia	1978	Pittsburgh	1986
New York	1979	Chicago	1987

The Pediatric Section of Neurological Surgery of the American Association of Neurological Surgeons gratefully recognizes the support of the following exhibitors for the 1987 Pediatric Annual Meeting:*

Cordis Corp. - Miami, Florida
 Lone Star Medical Products - Houston, Texas
 Malliner Labs - Edmonton, Alberta, Canada
 Midas Rex Instruments - Fort Worth, Texas
 New Medico Associates, Inc. - Lynn, Massachusetts
 P.S. Medical - Goleta, California
 Wild/E. Leitz, Inc. - Rockleigh, NJ

All registrants are encouraged to visit the exhibit area frequently during the meeting.

*A complete listing of all exhibitors will be distributed to all participants at the meeting.

PROGRAM
PEDIATRIC SECTION
AMERICAN ASSOCIATION OF NEUROLOGICAL SURGEONS
The Palmer House
Chicago, Illinois
December 8-11, 1987

TUESDAY, DECEMBER 8, 1987

- 6:00 - 8:00 p.m. Registration - Grand Foyer (6th Floor)
7:00 - 9:00 p.m. Reception - Adams Room (6th Floor)

WEDNESDAY, DECEMBER 9, 1987

- 7:00 a.m. Registration - Foyer (4th Floor)
7:00 a.m. Continental Breakfast - Foyer (4th Floor)
8:00 a.m. Meeting - Red Lacquer Room (4th Floor)
Welcome - David G. McLone - Chairman
AANS Pediatric Section
8th Annual Special Lecture
"Excellence in Search of Recognition"
Dale Johnson, M.D.

HYDROCEPHALUS RESEARCH- Moderators: Howard M. Eisenberg
J. Gordon McComb

- 8:45 a.m. 1. "Characterization of a Canine Hydrocephalus Model with MRI"
James M. Drake, C. Lemaire,
D. G. Potts
Toronto, ON

*R indicates Resident paper and F indicates paper being presented by Fellow.

- 9:00 a.m. 2. "Cerebral Blood Flow Alternations in Experimental Infantile Hydrocephalus"
Ingrid Van Orden, Usha Vasthare, Ronald T. Tuma, Robert Rosenwasser, James P. McAllister Philadelphia, PA
- 9:15 a.m. R 3. "Shunt-induced Reversal of Periventricular Pathology in Experimental Hydrocephalus"
Marc R. Del Bigio Winnipeg, Manitoba CANADA
- 9:30 a.m. 4. "The Relationship Between Intracranial and Superior Sagittal Sinus Pressure in Normal and Hydrocephalus Dogs"
William C. Olivero, Harold L. ReKate, John McCormick Phoenix, AZ
- 9:45 a.m. 5. "Cyclic AMP and Adenylate Cyclase Activators on Cerebrospinal Fluid Formation"
Shigeyo Hyman, J. Gordon McComb, Erick Stephanian, Martin H. Weiss Los Angeles, CA
- 10:00 a.m. **COFFEE BREAK-** Foyer (4th Floor)
View Exhibits
- HYDROCEPHALUS CLINICAL-** Moderators: Arthur E. Marlin
Gail A. Magid
- 10:30 a.m. 67. "Shunt Implantation: To Ward Zero Infection"
Maurice Choux, Gabriel Lena, Lorenzo Genitori, Etom Empime Marseille, France
- 10:45 a.m. 6. "Doppler Cranial Sonography of Normal and Abnormal Neonates"
Joanna J. Seibert, Charles M. Glasier, Timothy C. McCowan, William M. Chaddock Little Rock, AR

- 11:00 a.m. 7. "Cranial Doppler Ultrasonography Correlates with Shunting Criteria and Cerebral Perfusion Pressure"
William M. Chaddock, James Adametz, Mark Crabtree, Cynthia Ann Stansell Little Rock, AR
- 11:15 a.m. R 8. "Seizures and Shunt Malfunction"
Brett Scott, Dennis Johnson, Joan Conry San Diego, CA
- 11:30 a.m. R 9. "X-Linked Hydrocephalus: Progress with an Enigmatic Syndrome"
Stuart M. Weil, Thomas S. Berger, Kerry R. Crone, Peter T. Dignan Cincinnati, OH
- 11:45 a.m. R 10. "Hydrocephalus Complicating Tuberculosis in Children"
Steven D. Ham, Alexa I. Canady Detroit, MI
- 12:00 noon 11. "To Fenestrate or to Shunt: Which Method is the Best Operative Treatment for Children with Arachnoid Cysts?"
Corey Raffel, J. Gordon McComb Los Angeles, CA
- 12:15 **LUNCH-** Monroe Ballroom (6th Floor)
- INFECTION -** Moderators: Mark O'Brien, Francisco Gutierrez
- 1:15 p.m. F 12. "Where Do the Bacteria Come From in Operative-Acquired Shunt Infections?"
Ann-Christine Duhaime, Luis Schut Kathleen Bonner Providence, RI

- 1:30 p.m. R 13. "Macrophage Activation and Adhesion in Proximal VentriculoPeritoneal Shunt Malfunction"
Timothy B. Garner, William O. Bell, Jon C. Lewis
Winston-Salem, NC
- 1:45 p.m. R 14. "The Effect of Bacitracin Upon the Electrostatic Properties of Silicone Plastic"
David J. Gower, K. Stuart Lee, J. M. McWhorter
Winston-Salem, NC
- 2:00 p.m. R 15. "Treatment of Staphylococcus Aureus Ventriculitis with Intraventricular Vancomycin and LY146032 in Hydrocephalic Rabbits"
Charles S. Haworth, T. S. Park, Michael W. Sobieski, W. Michael Scheld
Charlottesville, VA
- 2:15 p.m. R 16. "Salmonella Epidural Abscess in a Sickle Cell Patient"
Anthony Martino, Jeffrey A. Winfield
Syracuse, NY
- 2:30 p.m. F 17. "Cerebral Abscess in Children"
S. David Moss, David G. McLone
Chicago, IL
- 2:45 p.m. **COFFEE BREAK**- Foyer (4th Floor)
View Exhibits
- VASCULAR**- Moderators: A. Loren Amacher,
Richard A. Coulon, Jr.
- 3:15 p.m. 18. "Cerebral Venous Malformations in Children"
Daniele Rigamonti, Harold A. Rekate, Robert F. Spetzler,
Karen Hoenig-Rigamonti
Baltimore, MD

- 3:30 p.m. R 19. "Conservative Management of Thrombosed Vein of Galen Aneurysm"
Patricia A. Aronin, Edward J. Zampelle
Birmingham, AL
- 3:45 p.m. 20. "Surgical Treatment of Stroke in Children"
Balaji Sadasivan, Manuel Dujovny, Fernando G. Diaz, James I. Ausman
Detroit, MI
- 4:00 p.m. 21. "Balloon Embolization of Non-Traumatic Extra-Cranial Vertebral Arteriovenous Fistula in Children"
Balaji Sadasivan, Manuel Dujovny, Bharat Mehta, Ghaus M. Malik, James I. Ausman
Detroit, MI
- 4:15 p.m. 22. "Experience in Pediatric Neurosurgery with the Linear Accelerator as a Neurosurgical Tool"
Ken R. Winston
Boston, MA
- 4:30 p.m. Annual Business Meeting - Parlor B (6th Floor)
- 9:00 p.m. Hospitality Room - Parlor B (6th Floor)
- THURSDAY, DECEMBER 10, 1987**
- 6:30 a.m. **BREAKFAST**- Foyer (4th Floor)
- 7:00 a.m. Registration - Foyer (4th Floor)
- 7:00 a.m. **BREAKFAST SEMINAR: RHIZOTOMY**
Moderator: John W. Peacock
- 7:00 a.m. 1. "Selective Dorsal Rhizotomy for Spasticity"
Jogi V. Pattisapu, Marion L. Walker
Salt Lake City, UT

7:10 a.m. 2. "Clinical and Electrophysiologic Studies in Selective Dorsal Rhizotomy"
Leslie D. Cahan, Lauren M. Beeler
David McPherson, M. S. Kundi
Orange, CA

7:20 a.m. 3. "Selective Posterior Rhizotomy for the Treatment of Spasticity: Relationship of Intraoperative EMG Patterns to the Post-Operative Course"
Rick Abbott, Jeffrey H. Wisoff,
Alfred D. Grant, Fred Epstein
New York, NY

7:30 a.m. 4. "H-Reflex (H2/H1 Ratio) as a Standard for Electrophysiological Monitoring in Selective Posterior Rhizotomy"
Bruce B. Storrs, Takashi Nishida
Chicago, IL

7:40 a.m. Discussion

TRAUMA- Moderators: Michael J. Turner, Hal W. Pittman

8:00 a.m. 23. "Outcome of Moderate Head Injury in Childhood"
Thomas G. Luerksen, Melville R. Klauber, Lawrence F. Marshall,
Howard M. Eisenberg
San Diego, CA

8:15 a.m. R 24. "An Unusual Syndrome of Pediatric Brainstem Trauma"
Daniel D. Galyon, Jeffrey A. Winfield
Syracuse, NY

8:30 a.m. R 25. "The Infant Whiplash Shake Injury Syndrome a Clinical and Pathological Study"
Mark N. Hadley, Volker K. H. Sonntag, Harold L. Rekate
Phoenix, AZ

8:45 a.m. F 26. "The Clinical Exam, Brain Scintigraphy and EEG in the Assessment of Brain Death in Children"
Shelly Lester, Martha J. Barthel,
David G. McLone
Chicago, IL

9:00 a.m. 27. "Management of Central Nervous System Hemorrhage Associated with Hemophilia"
John C. Godersky, Arnold H. Menzes, C. Thomas Kisker,
Raymond Tannous
Iowa City, IA

TRAUMA- Moderators: Bruce Bressler, Thomas G. Luerksen

9:15 a.m. R 28. "Severe Head Injury in Children: The Use of I-123 HIPDM SPECT Scan and Arteriojugular Venous O-2 Difference"
John Ruge, Yoon S. Hahn, James J. Conway, Zehava Noah
Chicago, IL

9:30 a.m. 29. "Efficacy of Barbiturates in Head Injured Children"
Richard D. Bucholz, Thomas Pittman, Diane Williams
St. Louis, MO

10:00 a.m. 30. "Hyperbaric Oxygen in the Treatment of Elevated Intracranial Pressure After Head Injury"
Jeffrey A. Brown, Mark C. Preul, Assad M. Taha
Toledo, OH

10:15 a.m. R 31. "Cerebral Salt Wasting (CSW) Post Hypothalamic Region Surgery: Diagnosis, Management and Pathophysiology"
Antonio Disclafani, Gregory Stidham, Robert Sanford
San Francisco, CA

10:30 a.m. COFFEE BREAK- Foyer (4th Floor)
View Exhibits

**STEREOTACTIC & EPILEPSY - Moderators: Larry K. Page
William Hanigan**

- 11:00 a.m. 32. "Computer-Assisted Stereotactic Biopsy of Intracranial Lesions in Pediatric Patients"
Dudley H. Davis, Patrick J. Kelly
W. Richard Marsh, Bruce Kall
Rochester, MN
- 11:15 a.m. 33. "Use of Stereotactic Techniques in Children"
Jogi V. Pattisapu, Marion L. Walker, Bruce B. Storrs, M. Peter Heilbrun
Saly Lake City, UT
- 11:30 a.m. 34. "Temporal Lobectomy for Complex Partial Seizures Beginning in Childhood"
Giuseppe Erba, John R. Adler, Keasley Welch, Robert Ziegler, Peter McL. Black
Boston, MA
- 11:45 a.m. 35. "Intraoperative Cortical Mapping to Enhance Tumor Resection in the Pediatric Age Group"
Mitchel S. Berger, George A. Ojemann
Seattle, WA
- TECHNIQUE - Moderator: Patricia A. Aronin,
Charles C. Duncan**
- 12:00 noon R 36. "Use of 19F Magnetic Resonance Spectroscopy for Measurement of Cerebral Blood Flow"
D. Barranco, L. N. Sutton, A. McLaughlin, J. Greenberg
Philadelphia, PA
- 12:15 p.m. 37. "The Lateral Decubitus Position for the Surgical Approach to Pineal Location Tumors"
J. Gordon McComb
Los Angeles, CA

- 12:30 p.m. R 38. "Childhood Posterior Fossa Tumors Managed with Intraoperative CO-2 Laser and Postoperative MRI"
Charles L. Y. Cheng, John Ragheb, Walker Robinson, John Joslyn
Baltimore, MD

- 12:45 p.m. 39. "Three-Dimensional Display of the Ventricles and Cisterns from Routine, Non-Contrast CT Scans, Hydrocephalus and Cogenital Malformations"
Thomas P. Naidich, Sharon E. Byrd, Erin C. Prenger, Arthur Nieves, Bruce C. Teeter
Chicago, IL

FREE AFTERNOON

6:30 p.m. Reception - Empire Room (Main Lobby)

7:30 p.m. Banquet - Empire Room (Main Lobby)

FRIDAY, DECEMBER 11, 1987

7:30 a.m. Continental Breakfast - Foyer (4th Floor)

**BRAIN TUMORS - Moderators: David C. McCullough
Osamu Sato**

- 8:00 a.m. 40. "Management of Craniopharyngioma in Children"
Michael S. Edwards, James E. Baumgartner, Rodger Hudgins, Charles B. Wilson
San Francisco, CA

- 8:15 a.m. 41. "Pituitary Adenomas in Childhood and Adolescence"
Edward R. Laws, Bernd W. Scheithauer
Washington, DC

- 8:30 a.m. 42. "Choroid Plexus Tumors of Childhood: A Forty-Five Year Surgical Experience and Historical Perspective"
Richard G. Ellenbogen, Ken R. Winston, William J. Kupsky
Boston, MA
- 8:45 a.m. R 43. "Pineal Region Lesions: A Pediatric Series"
Hillel Z. Baldwin, Volker K. H. Sonntag, Harold L. Rekate, Stephen M. Bloomfield
Phoenix, AZ
- 9:00 a.m. R 44. "Pediatric Chordomas: Two Cases and Literature Review"
Lawrence Foody, Kerry R. Crone, Thomas S. Berger
Cincinnati, OH
- BRAIN TUMORS-** Moderators: Alexa Canady
Michael S. B. Edwards
- 9:15 a.m. 45. "Ganglioglioma: Prognosis and Rationale for Post-Operative Management"
Edison P. McDaniels II, Philip H. Cogen
Stanford, CA
- 9:30 a.m. R 46. "Gadolinium-DPTA Enhanced MR Imaging in Determining Treatment Options Following Surgical Resection of Pediatric Brain Tumors"
Curtis A. Dickman, Harold A. Rekate, C. Roger Bird, Marjorie Medina
Phoenix, AZ
- 9:45 a.m. R 47. "Prognosis Importance of Cellular Ploidy in Medulloblastoma of Childhood"
Masaharu Yasue, Tadanori Tomita, Herbert Engelhard, David G. McLone, Kenneth Bauer
Chicago, IL

- 10:00 a.m. R 48. "A Novel Chemotherapeutic for the Treatment of Medulloblastoma: Mutant Diphtheria Toxin Linked to Anti-Transferrin Receptor Monoclonal Antibody or Transferrin"
Charles J. Wrobel, Virginia Johnson, John Zovickian, Richard Youle
Bethesda, MD

10:15 a.m. **COFFEE BREAK-** Foyer (4th Floor)
View Exhibits

CONGENITAL SPINAL CORD LESIONS- Moderators: John Walsh
Dachling Pang

- 10:45 a.m. R 49. "A Tale of Two Tails"
Robert F. Keating, James T. Goodrich
Bronx, NY

- 11:00 a.m. R 50. "Sacral Agenesis"
Michel Malek, Anthony E. Gallo, Jr.,
Portland, OR

- 11:15 a.m. R 51. "Cervical Myelomeningocele - Description and a New Theory Regarding Embryogenesis"
Mark S. Dias, Dachling Pang, Diana Claassen
Pittsburgh, PA

- 11:30 a.m. R 52. "The Combination of Vater Association and Spinal Dysraphism"
Randall Chestnut, Hector E. James, Ken Lyons
San Diego, CA

TETHERED CORD- Moderators: Jack Walker, Arnold Menezes

- 11:45 a.m. R 53. "MR Imaging of Lipomyelomeningocele and Tethered Cord"
John D. Brophy, Leslie N. Sutton, Robert D. Zimmerman, Edward Bury, Luis Schut
Silver Springs, MD

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John D. Brophy, Leslie N. Sutton, Robert D. Zimmerman, Edward Bury, Luis Schut
Silver Springs, MD

12:00 p.m. 54. "Detethering - Does It Always Help?"
John R. Mawk, James R. McConnell
Omaha, NE

12:15 p.m. R 55. "Is the Long Tract Involved in the Experimentally Tethered Cord? Study of HRP Axonal Transport"
Takaharu Fuse, John W. Patrickson, Shokei Yamada
Loma Linda, CA

12:30 p.m. LUNCH- State Ballroom (4th Floor)

HYDROMYELIA AND SPINAL CORD TUMORS- Moderators: Michael Scott, Steven Wald

1:30 p.m. 56. "Management of Childhood Hydromyelia"
Jeffrey H. Wiseoff, Fred Epstein
New York, NY

1:45 p.m. 57. "Lumboperitoneal Shunts for Treatment of Syringomyelia"
T. S. Park, Wayne S. Cail, Melissa G. Walker
Charlottesville, VA

2:00 p.m. R 58. "Clinical and Pathological Analysis of Childhood Cord Astrocytomas"
Eugene Rossitch, Jr., Seth M. Zeidman, W. Jerry Oakes, Carter Harsh
Durham, NC

2:15 p.m. R 59. "Malignant Astrocytomas of the Spinal Cord"
Allan R. Cohen, Jeffrey Wisoff, Jeffrey Allen, Fred Epstein
New York, NY

2:30 p.m. COFFEE BREAK-Foyer (4th Floor)

CRANIAL ANOMALIES- Moderators: Hal Rekate, Barry French

3:00 p.m. 60. "Surgical Management of Lambdoid Synostosis: Review of 88 Cases"
Antonio Disclafani, Donald A. Ross, Roger Hudgins, Michael Edwards
San Francisco, CA

3:15 p.m. 61. "Lazy Lambdoid (Skull Base Abnormality - Nonsurgical Treatment)"
Robert A. Sanford, Matthew Wood, Jr., Michael S. Muhlbauer
Memphis, TN

3:30 p.m. R 62. "The Surgical Treatment of Metopic Suture Synostosis"
Tariq Javed, John E. Kalsbeck, A. Michael Sadove
Indianapolis, IN

3:45 p.m. 63. "Multi-Suture Craniosynostosis in Dizygotic Twins with Germinal Matrix Hemorrhage - Surgical Treatment by Cranial Release and Transposition"
George R. Cybulski, Martin E. Gryfinski
Chicago, IL

4:00 p.m. 64. "The Treatment of Craniosynostosis in the First Year of Life"
Concezio Di Rocco
Rome, Italy

4:15 p.m. 65. "Early Intervention in Fibrous Dysplasia of the Orbit and Frontal Vault"
Joan L. Venes
Ann Arbor, MI

4:30 p.m. R 66. "Foramen Magnum Changes in Achondroplasia"
Frederick A. Boop, Harold J. Hoffman
San Antonio, TX

SCIENTIFIC ABSTRACTS

1. CHARACTERIZATION OF A CANINE HYDROCEPHALIC MODEL WITH MRI

James M. Drake, C. Lemaire, D. G. Potts (Toronto, ON)

Magnetic Resonance Imaging (MRI) is a sensitive technique for detecting alterations in brain morphology and pathological changes associated with increased water content. MRI was used to characterize the appearance and evolution of silastic induced hydrocephalus in beagle dogs.

Ten dogs weighing 8-12 kg had 2-3 cc of Silastic mixture introduced into the basal cisterns and/or the fourth ventricle via a small posterior fossa craniotomy. The animals were imaged serially for up to 10 weeks on a 2 Tesla small bore unit. Their heads were immobilized in a cast acrylic stereotactic frame containing an external reference system. Ventricular size was calculated as % total cross sectional area of a standard anterior frontal slice. T1 and T2 relaxation times were calculated from the images.

Six animals survived the initial postoperative period. Ventricular area (Y) was related to postoperative day (X) as $Y = 4.0 \cdot \ln(X) + 1.88$, $r = .9299$. Periventricular edema appeared initially in the superolateral angles of the frontal horns. The extent of the edema was variable. The T1 of 986.81 msec in normal white matter increased to 1956.28 msec in the area of the edema, $p < .0001$. The T2 of 83.61 msec in normal white matter increased to 234.85 msec in the area of the edema, $p < .0001$.

Having established noninvasively the characteristics of this model, the effects of interventions such as new CSF shunting devices may now be determined.

2. CEREBRAL BLOOD FLOW ALTERATIONS IN EXPERIMENTAL INFANTILE HYDROCEPHALUS

Ingrid Van Orden, Usha Vasthare, Ronald F. Tuma, Robert Rossenwasser, James P. McAllister (Philadelphia, PA)

Our previous studies have shown that pyramidal cells degenerate and monoamine levels are drastically reduced in the cerebral cortex of hydrocephalic kittens. The present study was undertaken to determine the relationship between these morphologic and biochemical changes and cerebral blood flow. Hydrocephalus was induced in 4-10 day old kittens by injections of 25% kaolin into the 4th ventricle and cisterna magna. Littermate control animals received similar injections of sterile saline. Ultrasonography was used to estimate the severity of the ventriculomegaly. Fifteen to 20 days post-injection cerebral blood flow was measured on all animals by the isotope labelled microsphere technique. Significant ($p < 0.05$) decreases in blood flow were detected in frontal (Area 4), parietal (Area 22) and occipital (Area 17) regions of the cerebral cortex, as well as the thalamus, midbrain and pons, cerebellum and caudate nucleus. Throughout the brain, the percentage of blood flow reduction and the increase in vascular resistance averaged 54.4% and 54.9%, respectively. The most severe decreases in blood flow occurred in Areas 22 and 17 of the cortex, which correlates well with the neuronal degeneration and monoamine changes noted previously in these regions. The caudate nucleus, which was markedly compressed, exhibited a 58% decrease in blood flow. These alterations in cerebral blood flow were associated with statistically significant systemic changes. Heart rate was reduced by 22% and cardiac output decreased by 70%. While mean arterial pressure did not change, there was a general increase in sympathetic tone, as shown by increases in peripheral resistance in the kidney, stomach and duodenum. Total peripheral resistance increased 221%. These data suggest that reductions in cerebral blood flow may be responsible, in part, for the neurologic deficits that accompany hydrocephalus, as well as the neuronal degeneration seen in the cerebral cortex.

3. SHUNT-INDUCED REVERSAL OF PERIVENTRICULAR PATHOLOGY IN EXPERIMENTAL HYDROCEPHALUS

Marc R. Del Bigio (Winnipeg, MANITOBA)

The degree to which neuropathology associated with hydrocephalus can be reversed by cerebrospinal fluid shunting is not well documented. Hydrocephalus was induced in young adult rabbits by injection of silicone oil into the cisterna magna. After 1 and 8 weeks, hydrocephalic rabbits were shunted for 1 week. Rabbits were killed by perfusion with aldehydes, then gross and microscopic aspects of control, hydrocephalic, and shunted rabbit brains were compared. The frontal horns of the lateral ventricles were dilated at 3 days, post-injection ($P < 0.05$) and did not change in size thereafter. Shunting returned the ventricles to control size. Mitotic activity among ependymal cells was increased ($P < 0.05$) over the corpus callosum (CC) at 3 days and over the caudate nucleus (CN) at 2 weeks. This response was attenuated by shunting. Similarly, mitotic activity among subependymal astrocytes was increased over the CN ($P < 0.05$) and CC at 1 week. In both regions, the mitotic activity was further increased after shunting. The number of ependymal cells per unit distance was decreased ($P < 0.05$) over the CN, CC, and lateral septal area (LSA) at 1 and 8 weeks. Shunting increased ($P < 0.05$) the number of ependymal cells after 1 week of hydrocephalus but not after 8 weeks. The number of patent capillaries in the CN, CC, and LSA was decreased ($P < 0.05$) 1 and 8 weeks after the induction of hydrocephalus. This change was reversible by shunting only in the 1 week hydrocephalic rabbits. An increase ($P < 0.05$) in the number of subependymal astrocytes was observed in the CC and CN of 8 week hydrocephalic rabbits. It is hypothesized that the overabundant astrocytes tend to fix stretched ependymal cells and collapsed capillaries in their distorted configurations in chronically hydrocephalic animals. Thus, early shunting allows more rapid and perhaps more complete reversal of periventricular neuropathology in experimental hydrocephalus.

4. THE RELATIONSHIP BETWEEN INTRACRANIAL AND SUPERIOR SAGITTAL SINUS PRESSURE IN NORMAL AND HYDROCEPHALIC DOGS

William C. Olivero, Harold L. Rekate, John McCormick (Phoenix, AZ)

The relationship between elevations in intracranial pressure (ICP) and superior sagittal sinus pressure (SSSP) remains unclear. We studied normal adult greyhound dogs and dogs made hydrocephalic with kaolin to better define this relationship and to determine its functional significance. Intraventricular constant infusions and rapid injections were performed while ventricular and SSSP were recorded. BP and pCO₂ were monitored and maintained within the normal range. In the majority of normal dogs we observed a small elevation in SSSP with rises in intraventricular pressure in hydrocephalic dogs the elevation in SSSP was much more significant and the baseline pressure difference between the sagittal sinus and ventricle was decreased. We infused saline into the proximal sagittal sinus while recording pressure in the distal sinus. Elevations in SSSP did not occur until very high infusion rates suggesting that the rises in SSSP that accompany rises in ventricular pressure are not caused by increased flow but increased resistance in the sinus. It appears that the sagittal sinus's normal capacity to accommodate elevations in ICP is decreased in hydrocephalus probably by distorting the sinus and causing increased resistance to flow.

It seems that in the kaolin model for hydrocephalus the normal pressure gradient across the arachnoid villi into the superior sagittal sinus necessary for absorption of cerebrospinal fluid is diminished. This loss of absorptive capacity may contribute to the formation and/or maintenance of the hydrocephalic state. This can be predicted by using a mathematical model of ventricular volume regulation.

5. CYCLIC AMP AND ADENYLATE CYCLASE ACTIVATORS ON CEREBROSPINAL FLUID FORMATION

Shigeyo Hyman, J. Gordon McComb, Erick Stephanfan, Martin H. Weiss (Los Angeles, CA)

The activation of membrane-bound adenylyate cyclase or inhibition of the specific phosphodiesterase result in the increase in intracellular cAMP followed by active secretion of water and electrolytes in a number of *in vitro* preparations. The accumulation of cAMP in mammalian choroid plexus has been studied to a limited extent by using either isolated choroid plexus tissue or cultured choroid plexus epithelial cells, but these experimental conditions do not allow determination of CSF formation.

Cyclic AMP, histamine and serotonin were examined because of their known *in vitro* effect on adenylyate cyclase. Also included were dibutyryl cAMP, a functionally equivalent but more membrane-permeable analog of cAMP; adenosine, a cAMP metabolite; and aminophylline, a phosphodiesterase inhibitor. Various concentrations of these substances were added to artificial CSF and perfused through the ventricles of rabbits and cats. Alteration in the rate of CSF formation was measured by noting the change in concentration of a nondiffusible reference marker.

In most cases the CSF formation rate was unchanged. Some significant alterations were noted but were inconsistent in regard to animal species and/or concentration of the substance under consideration.

Although CSF formation may not be readily responsive to most of the substances tested in this experimental situation, the process appears to be too complex to study *in vivo* using the ventriculo-cisternal perfusion technique since such factors as choroidal arterial blood flow, cellular permeability, alterations in the blood-CSF-brain-barrier, ability of the substance to reach the appropriate receptor sites and the intracellular processes all effect CSF formation but to an unknown extent.

6. DOPPLER CRANIAL SONOGRAPHY OF NORMAL AND ABNORMAL NEONATES

Joanna J. Seibert, Charles M. Glasier, Timothy C. McCowan, William M. Chaddock (Little Rock, AR)

383 head ultrasound studies (N=383) with Doppler evaluation of the cerebral vessels were obtained in 278 normal and abnormal neonates. The Resistive Index (RI) (systolic BP-diastolic BP/systolic BPX100%) was obtained for the anterior and middle cerebral arteries and the internal carotid artery bilaterally. The overall RI was an average of these values. All neonates were assessed for a variety of clinical and sonographic abnormalities. The RI for clinically and sonographically normal infants (N=75) decreased linearly with increasing gestational age at birth. RI for term (38-42 weeks) infants was 72 ± 13 and for < 32 weeks gestation 82 ± 13 . The highest RI in clinically abnormal infants were seen in cardiac ischemia 84 ± 19 (N=7), PDA 86 ± 15 (N=72), and infants with bloody CSF 88 ± 13 (N=22). (All $p < 0.05$ compared to normals.) Sonographic abnormalities associated with increased RI included ICH (Grades 1-4) 85 ± 23 (N=152), hydrocephalus 87 ± 15 (N=28), and brain edema 96 ± 24 (N=13). (All $p < 0.05$ compared to normals.) Thirteen studies were done pre and post ventricular tap in patients with increasing hydrocephalus. RI decreased from 98 ± 18 pre-tap to 79 ± 13 following CSF aspiration ($p < 0.05$). Abnormal Doppler RI is a good predictor of intracranial pathology. It is especially helpful in differentiating ventricular dilatation secondary to increased CSF pressure from cerebral atrophy.

7. CRANIAL DOPPLER ULTRASONOGRAPHY CORRELATES WITH SHUNTING CRITERIA AND CEREBRAL PERFUSION PRESSURE

William M. Chaddock, James Adametz, Mark Crabtree, Cynthia Ann Stansell (Little Rock, AR)

Twenty-two patients ultimately requiring ventriculo-peritoneal shunts for hydrocephalus associated with intraventricular hemorrhage (IVH) or myelomeningocele (MM) were studied with serial measurements of head circumference, ventricular volume and ultrasound studies of Resistive Index (RI) (systolic BP-diastolic BP/systolic BP) in the internal carotid, anterior cerebral and middle cerebral arteries bilaterally. The decisions for shunting were based on classical criteria, (change in head circumference, change in ventricular volume, and clinical status). High RI values were found in 21 of 22 patients requiring shunts, and the RI values decreased post shunting in 20 patients to levels considered to be normal in another study of 383 normal and abnormal neonates. Resistive indices in non-shunted patients with IVH and MM also fell into the normal range.

Doppler ultrasonography is helpful in selecting patients needing ventriculo-peritoneal shunts. Further, newer transcranial Doppler equipment may be helpful in assessing shunt malfunction in older patients.

Experimentally, dogs were studied by using intracranial pressure over a wide range with cisterna magna saline infusions. Intracranial pressure was recorded from a fibre-optic brain tissue monitor, and cranial Doppler sonographic measurements were made through a craniectomy. Cerebral perfusion pressure was calculated using simultaneously recorded arterial pressure and ICP. Linear correlations between cerebral perfusion pressure and RI were uniformly found.

Transcranial evaluations of RI in pediatric head injured patients monitored with arterial pressure lines and fibre-optic ICP systems, suggest that transcranial Doppler monitoring could replace invasive intracranial pressure monitoring.

8. SEIZURES AND SHUNT MALFUNCTION

Brett Scott, Dennis Johnson, Joan Conry (San Diego, CA)

Many children with shunted hydrocephalus also have a seizure disorder. The role of the shunt in the seizure disorder has not been well-defined, and it is equally difficult to know whether the seizure disorder relates to shunt function. More specifically, what is the incidence of seizures following shunting, and how often does a seizure indicate shunt malfunction? To answer these questions the records of 3890 pediatric neurosurgical patients at CHNMC were reviewed, and 886 children with shunted hydrocephalus were extracted. The etiology of hydrocephalus is displayed in Table 1. A retrospective analysis of both shunt and seizure histories was done in 816 patients who had complete records. In this cohort of children, 544 emergency room encounters for seizures and 1831 shunt revisions were analyzed. The mean length of follow-up was 6.71 years, and the mean number of shunt revisions was 1.25 procedures per child or 0.93 per child per 5 years.

Of the patients with shunted hydrocephalus, 307 or 38% also had seizures. The first seizure occurred after shunting in 181 or 22% of children; 37 or 12% had their first seizure in the first postoperative week. The median duration of shunting prior to onset of the seizure was 1.6 years. In a subset of patients who had only one ventricular site for their entire shunt history, the incidence of seizures (18%) was the same in the parieto-occipital region as it was in the parieto-temporal region.

Fourteen patients were identified who had a seizure in the week prior to shunt revision, i.e., shunt malfunction was heralded by a seizure on 0.7% of children. The chance of a seizure encounter resulting in a shunt revision was 2.6%.

9. X-LINKED HYDROCEPHALUS: PROGRESS WITH AN ENIGMATIC SYNDROME

Stuart M. Weil, Thomas S. Berger, Kerry R. Crone, Peter T. Dignan (Cincinnati, OH)

When congenital hydrocephalus does not occur in the presence of neural tube defects, the most common familial etiology is an x-linked association of mental retardation and hydrocephalus which is present in 2% of cases of congenital hydrocephalus. Bickers and Adams introduced the concept of a sex linked hydrocephalus in 1949, and 48 families have been reported since that time. Since 1979, five additional families (11 cases) have been evaluated at our institution.

In all previous cases as well as the cases currently reported, the sine qua non is congenital hydrocephalus and mental retardation. There is controversy as to the anatomical cause of hydrocephalus but all autopsy cases to this point have demonstrated aqueductal stenosis. All of the patients at this institution are still alive but radiologic evaluation of our cases has been consistent with aqueductal stenosis.

The most common systemic abnormality is a 25-40% occurrence of adducted thumbs. This finding has been present in nine of eleven cases in our series. Electromyographic evaluation at our institution has suggested a variable etiology for this phenomenon but tends to show an agenesis of abductors and extensors of the thumb. Other systemic findings reported in the literature include plagiocephaly, as well as lower extremity spasticity, hyperreflexia, and seizures, which do not generally improve with ventricular shunting. In addition, one of our patients had epispadias, two had lateral nystagmus, four had facial hemangioma and seven had a simian crease. Although there is no previous report of x-linked hydrocephalus with neural tube defects, two of our patients had the stigmata of spina bifida occulta, including lumbosacral grooves and tufts of hair.

The specific chromosomal defect resulting in x-linked hydrocephalus is yet to be determined. At this time, ongoing analysis of carriers and probands is being conducted in our laboratories.

9. X-LINKED HYDROCEPHALUS: PROGRESS WITH AN ENIGMATIC SYNDROME (Con't)

Ultrasound has been suggested as an adjunct in prenatal diagnosis of congenital hydrocephalus. In our experience ultrasound examination prior to 26 weeks gestation has failed to demonstrate hydrocephalus. Based on this observation late second trimester examination is recommended for women at risk if previous evaluation is normal.

10. HYDROCEPHALUS COMPLICATING TUBERCULOSIS IN CHILDREN

Alexa I. Canady, Steven D. Ham (Detroit, MI)

Although central nervous system involvement of tuberculosis is common in underdeveloped countries, this manifestation in the United States is unusual and frequently considered a delayed complication. In the last two years four patients were seen with hydrocephalus present at the time of initial diagnosis of central nervous system tuberculosis. Three were the results of tubercular meningitis and one secondary to a posterior fossa tuberculoma. Three cases of tubercular meningitis presented in children aged eleven months, three and four years as lethargy and nuchal rigidity. The cerebrospinal fluid was characterized by a predominance of monocytes and elevated pressure. Initial computerized scans indicated ventriculomegaly and two required emergent ventriculostomy for control of intracranial pressure. All were eventually treated with ventricular peritoneal shunts. The child with tuberculoma was four years old and presented with headaches, vomiting and visual changes. Computerized scan demonstrated a left posterior fossa mass with hydrocephalus. The child was treated with ventricular peritoneal shunt followed by suboccipital craniectomy. None of the children were known to have previous exposure to or symptoms of tuberculosis at the time of initial treatment. All four responded to antitubercular therapy and required permanent shunting. A high index of suspicion must be maintained in treating patients with meningeal signs and acute hydrocephalus. In the case of an intracranial mass, tissue diagnosis is most important for appropriate therapy. It is of note that of these four patients, only two were inner city children with one from an affluent suburban community and one from a rural environment.

11. TO FENESTRATE OR TO SHUNT: WHICH METHOD IS THE BEST OPERATIVE TREATMENT FOR CHILDREN WITH ARACHNOID CYSTS?

Corey Raffel, J. Gordon McComb (Los Angeles, CA)

The two treatment options for intracranial arachnoid cysts are either craniotomy and fenestration of the cyst into the subarchnoid space or shunting the cyst contents to the peritoneal cavity. Successful fenestration eliminates the need to shunt, but is a major operative procedure. To determine which treatment option provides the greatest benefit with the fewest complications, the records of 31 patients with 34 arachnoid cysts treated at the Childrens Hospital of Los Angeles between 1976 and 1986 were reviewed. The mean age of the patients was 4.4 years with a range of 0.5 months to 15.5 years. The most common location was the middle fossa (14), followed by the posterior fossa (7), suprasellar region (5), and hemispheric (5). Signs and symptoms related to abnormally enlarging head size in infants and to increased intracranial pressure or seizures in older patients.

The initial treatment of 29 cysts was fenestration. Twenty-two (76%) were successful, with no additional procedure needed for the cyst. The other 7 cysts required the placement of a CP shunt. In five cases the cysts were treated by shunting initially. Of the 12 cyst-peritoneal shunts, 5 have required revisions on one or more occasions. No significant difference in morbidity was noted with either operative modality.

Based upon this series, we suggest that fenestration be the initial procedure of choice for patients with arachnoid cysts since this eliminates the need for permanent dependency upon a shunt.

12. WHERE DO THE BACTERIA COME FROM IN OPERATIVE-ACQUIRED SHUNT INFECTIONS?

Ann-Christine Duhaime, Luis Schut, Kathleen Bonner, Stanley Plotkin (Providence, RI)

In order to study the distribution of bacteria in the operating room environment, cultures were obtained during 112 unselected shunt operations at Children's Hospital of Philadelphia between November 1986 and September 1987. After routine skin preparation and drying, millipore filters for bacterial collection were placed on the patient's prepped skin underneath the drapes, on top of the drapes in the operative field, and/or on the instrument table, for the duration of the case. In 49 patients full-thickness skin biopsies taken at the initial incision were cultured in place of skin surface cultures. Perioperative CSF cultures and shunt infections were monitored.

Of the 288 environmental (skin and surfaces) cultures, 24 were positive (20 staph coag negative and 4 staph aureus). Positive cultures were found in 15 of 112 drape cultures (13.34%), 7 of 78 instrument table cultures (8.97%), and 2 of 98 skin cultures (2.04%). Positive cultures were not correlated with the surgeon, length of case, time of day, or type of shunt operation, but were more likely to occur in a room other than the designated neurosurgery OR.

Sixteen patients had positive CSF cultures, including 9 clinical shunt infections (8.04%). Positive environmental cultures were found in 16 of the 96 patients with negative CSF cultures (16.67%), and in 5 of the 16 patients with positive CSF cultures (31.25%), although the organism was not always the same.

We conclude that bacteria most often responsible for shunt infections are airborne in the operating room, rather than remaining on the patient's skin. Maintaining a designated OR in which traffic is limited as well as strict adherence to covering skin surfaces of OR personnel may help to reduce shunt infection rates.

13. MACROPHAGE ACTIVATION AND ADHESION IN PROXIMAL VENTRICULOPERITONEAL SHUNT MALFUNCTION

Timothy B. Garner, William O. Bell, Jon C. Lewis (Winston-Salem, NC)

Proximal shunt malfunction remains a serious obstacle to long-term cerebral spinal fluid diversion. Presented are ultrastructural studies of eight catheters removed for proximal malfunction; the results suggest that macrophage adhesion and activation may play an important role in subsequent obstruction.

All eight patients had clinical and/or computed tomographic evidence of shunt malfunction. All shunts were cultured, with negative results. The catheters had been indwelling for five weeks to 25 years. The catheters were examined using high-resolution scanning electron microscopy.

Luminal surface of five of the catheters were characterized by macrophage adhesion and activation, involving extensive hyalomere elaboration and pseudopod extension. Pseudopod orientation suggested a chemotactic response and focal migration. Adherent macrophages were often associated with fibrin, the strands enshrouding and partially obscuring the underlying macrophages. Red blood cells and platelets were seen only occasionally, indicating a predominantly monocytemediated event. The polymer interface of these five shunts was characterized by a proteinaceous coat. Discontinuities of the proteinaceous coat were common, and were sometimes in close association with activated macrophages. The remaining three catheters had varied characteristics: organized and acute clot in one, a portion of choroid plexus alone in one, and, in the 25-year catheter, flakes of calcification. None of these three catheters had macrophages. Some component of choroid was present in all but the 25-year catheter.

Macrophage activation on the inner surface of these catheters with evidence of chemotaxis and fibrin deposition suggests a cell-mediated immune response. Such a response may enhance the predilection for choroid plexus to occlude ventricular catheters, and offers an alternative explanation to purely mechanical obstruction.

14. THE EFFECT OF BACITRACIN UPON THE ELECTROSTATIC PROPERTIES OF SILICONE PLASTIC

David J. Gower, K. Stuart Lee, J. M. McWhorter (Winston-Salem, NC)

Silicone polymer plastic used for shunt tubing has a strong surface electrostatic charge. Similar polymers with this characteristic are highly attracted to proteins. This study evaluates the degree of attraction of silicone plastic for foreign proteins as well as methods to minimize this attraction.

Sterile silicone plastic shunt tubing (Codman) was divided into three sections 30 cm in length. One section was handled with sterile gloves and divided into 2-cm portions; each portion of this tubing was then immersed in S.U.B. (1% SDS, 9M urea, 2% 2-mercaptoethanol). A second section was handled briefly without gloves then washed three times with sterile saline and processed in a similar manner, but was immersed in bacitracin solution (200 U/cc) following gloveless handling. Equal quantities of S.U.B. were evaluated using electrophoresis (10%) at 8 w for 3 hours and stained with Serva Blue-R.

The sterile tubing had no associated proteins, but produced a smear consistent with the elution of multiple length polymers from the plastic. This was enhanced by gently heating the tubing (70° C for 15 minutes). The plastic which was handled readily attracted foreign proteins from the skin, but these proteins were almost completely removed by bacitracin treatment.

These data suggest that even sterile, protein-free shunt tubing may elute various length silicone plastic polymers while heated. In addition, sterile shunt tubing should not be handled, resterilized, and then implanted because of the high affinity of the tubing for foreign proteins. Finally, bacitracin is highly effective in removing surface adherent proteins from silicone plastic, again demonstrating the utility of this drug in shunt surgery.

15. TREATMENT OF STAPHYLOCOCCUS AUREUS VENTRICULITIS WITH INTRAVENTRICULAR VANCOMYCIN AND LY146032 IN HYDROCEPHALIC RABBITS

Charles S. Haworth, T. S. Park, Michael W. Sobieski, W. Michael Sheld (Charlottesville, VA)

Ventriculitis is a serious complication of shunted hydrocephalus. To evaluate current therapy of the ventriculitis, vancomycin and a new antibiotic, LY146032, were tested in vitro and in vivo against Staphylococcus aureus (*S. aureus*). In vivo tests were performed on rabbits with kaolin-induced hydrocephalus.

Five groups of rabbits were studied: Ventriculitis only (n=6), intraventricular vancomycin only (n=5), ventriculitis treated with intraventricular vancomycin 30 μ g. (n=6), intraventricular vancomycin 120 μ g. (n=3), or intraventricular LY146032 7.5 μ g. (n=6). Ventriculitis was established by inoculation of 10^{7-13} *S. aureus* (mean). Intraventricular antibiotics were injected 16 h after inoculation of *S. aureus*. Cerebrospinal fluid (CSF) cultures were obtained at 16 and 24 h. CSF concentrations of the antibiotics were sampled at 15 min and at 24 h. Periventricular white matter (PVWM) was bioassayed for the antibiotics postmortem.

Results of this study were: 1) *S. aureus* was vancomycin-resistant (MBC > 64 μ g/ml) but LY146032 sensitive (MBC=2.0 μ g/ml); 2) *S. aureus* demonstrated static growth in in vitro CSF and in ventriculitis at a maximum titer of 10^{5-6} cfu/ml; 3) a single dose of intraventricular vancomycin (peaks-100 and 500 μ g/ml) did not lower *S. aureus* titers over 8 h ($p > 0.1$), whereas LY146032 (peaks-30 μ g/ml) did ($p < 0.01$); 4) ventriculitis did not significantly alter clearance of intraventricular vancomycin; $T_{1/2B} = 1.7$ h (infected) and $T_{1/2B} = 1.9$ h (uninfected); 5) vancomycin was detectable in the PVWM only in the presence of ventriculitis.

We conclude that: 1) drug carry-over artificially lowers the MBC of vancomycin-resistant *S. aureus*; 2) in vitro time-kill curves in CSF correlate with in vivo results; 3) ventriculitis facilitates transepndymal drug clearance into the PVWM; 4) LY146032 is effective against vancomycin-resistant *S. aureus* ventriculitis.

16. SALMONELLA EPIDURAL ABSCESS IN A SICKLE CELL PATIENT

Anthony Martino, Jeffrey A. Winfield (Syracuse, NY)

Neurologic complications of sickle cell anemia are most commonly ischemic strokes secondary to sludging in cerebral arteries. We discuss the case of paraplegia in a 15 year old black male with sickle cell anemia.

A 15 year old black male with sickle cell disease presented to University Hospital with a two day history of progressive right leg weakness. His past medical history was remarkable for recurrent episodes of back and left leg pain. Two weeks prior to this admission, he developed severe left leg pain due to aseptic necrosis of the left hip. Two days prior to presentation he complained of a progressive weakness of his right leg and then of urinary incontinence. On initial exam he was awake, alert, and cranial nerves were intact. Peripheral motor exam revealed a diffuse weakness in his right leg. DTR's and sensation were intact throughout. The pediatricians felt the presentation represented a vascular infarct of the spinal cord and an MRI was performed from T9 to the sacrum which was negative. He received an exchange transfusion. His neurologic status deteriorated over the next 24 hours developing weakness of the left leg and a T8 sensory level. An emergency CT-myelogram was performed revealing an epidural lesion from T5 - T8 with paravertebral extension. He underwent an emergency decompressive laminectomy. At surgery a large epidural abscess was drained and salmonella cultured.

The association of sickle cell anemia and salmonella osteomyelitis has been previously described. Vaso-occlusive phenomenon of bones is common in patients with sickle cell hemoglobinopathies. These produce painful crises which must be distinguished from osteomyelitis in sicklers, a febrile course usually leads one to diagnose the latter. The literature describes only three cases of paravertebral abscess complicating salmonella osteomyelitis. We describe this unique case of salmonella epidural abscess in sickle cell anemia, the roentgenologic findings and surgical treatment as well as those roentgenologic features which will help distinguish between bone infarct and osteomyelitis.

17. CEREBRAL ABSCESS IN CHILDREN

S. David Moss, David G. McLone (Chicago, IL)

50 consecutive children have been treated for cerebral abscess at Children's Memorial Hospital from 1958 to 1987. The average age was 6.5 years, ranging from 3 days to 19 years. A very wide range of organisms and etiologies were encountered. Previous reviews have advocated many different modes of therapy. Both aspiration and craniotomy with drainage-evacuation or resection were employed in our series. The predominant mode of surgical therapy was craniotomy with resection of the abscess and capsule. Overall mortality was 16% and operative mortality was 6% (excluding post-operative cerebral abscess and medically treated abscess confirmed by diagnostic biopsy). 22% of the cases had no known cause. 24% had cyanotic heart disease, of which 3 patients had dental abscess and one had otitis media. 14% were related to hydrocephalus/shunt infections. Sinusitis and otitis accounted for 10%. One case was associated with a nasal dermal sinus and one case was congenital. 24% of the cases produced negative cultures. 26% were streptococcus of mixed types. Staphylococcus represented only 8%. Our congenital abscess was caused by salmonella. 4% were fungal, all of which died. Epilepsy occurred in 28% of our patients. 4 cases were treated medically, of which 3 died. 46 cases had surgical intervention; 11 were aspirated, 13 were evacuated without capsule resection, and 22 were resected. Mortality in the aspirated group was twice that of the evacuated or resected group (18%, 8%, and 9% respectively). The factor which correlated best with mortality was the patient's clinical status on admission. The advent of CT scan improved mortality by allowing accurate diagnosis and surgical intervention.

18. CEREBRAL VENOUS MALFORMATIONS IN CHILDREN

Daniele Rigamonti, Harold A. Rekate, Robert F. Spetzler, Karen Hoenig-Rigamonti, Marjorie K. Medina, Michael A. Epstein (Baltimore, MD)

Cerebral venous malformations consist of a radial array of small medullary veins converging into a dilated central trunk. They are usually diagnosed in the third and fourth decades. Our series includes 21 angiographically confirmed venous malformations: five patients (24%) were children. Headache was the presenting symptom in each patient; eye pain, leg pain, vertigo, "rage attacks" occurred only once (20%) and could not be undoubtedly attributed to malformation. CT scan showed in every case an enhancing linear structure after contrast administration. In two cases an associated globular hyperdensity was seen in close proximity to the linear structure. MRI was performed in four children and in each case depicted a tubular structure characterized by decreased signal intensity (S.I.) on T₂-weighted images. Only one of the two children with the globular density on CT underwent MRI: the globular density appeared on MRI as a mixed core of decreased and increased S.I. with a rim of decreased S.I. on T₂-weighted images. This patient underwent surgery: a cavernous malformation was excised, without interfering with the venous malformation. Our review indicates that venous malformations are relatively frequent in children, and headache, often severe, is the main symptom. They may be associated with cavernous malformations and MRI is useful in recognizing the two entities. Because they consist of abnormal veins draining normal brain parenchyma, surgical excision should be restricted only to those cases in which the symptoms are incapacitating and undoubtedly related to the venous anomaly.

19. CONSERVATIVE MANAGEMENT OF THROMBOSED VEIN OF GALEN ANEURYSM

Patricia A. Aronin, Edward J. Zampella (Birmingham, AL)

Aneurysms of the vein of Galen comprise less than 1% of all arteriovenous malformations. Spontaneous thrombosis of these lesions is even more rare, with only eleven cases reported in the literature. The clinical presentation of these cases include progressive cranial enlargement and nonspecific findings of increased intracranial pressure. Diagnosis of a thrombosed vein of Galen aneurysm was made in most cases by angiography or at the time of surgery; CT scanning was utilized in making a diagnosis in four cases. The management of all previously reported cases has been surgical and comprises either a ventricular shunt alone, or in tandem with excision of the lesion or removal of the clot. The post operative course in half of these has been described as "stormy", and in only three cases has the long term outcome been characterized as "remarkably improved".

We present two cases of infants referred for evaluation of increasing head circumference who were found to have thrombosed vein of Galen aneurysms. Both patients were evaluated with angiography, CT scan and MRI scan. The management of these patients has been conservative and neither shunting nor direct intervention has been performed. Followup examination has been 18 months in the first patient and three months in the second. Both are progressing from a behavioral standpoint and their head circumferences are approaching normal range for age.

We propose that the natural history of thrombosed vein of Galen aneurysms is yet to be elucidated and that, in the absence of neurologic deterioration, surgical intervention may subject the patient to an unnecessary risk of severe morbidity or mortality.

20. SURGICAL TREATMENT OF STROKE IN CHILDREN

Balaji Sadasivan, Manuel Dujovny, Fernando G. Diaz, James I. Ausman (Detroit, MI)

The incidence of pediatric stroke is higher than the incidence of pediatric brain tumors. Most cases of stroke are secondary to systemic disease, but there is a group of pediatric stroke patients with surgically treatable lesions. Patients need to be fully investigated if these lesions are to be identified and treated.

Experience with eight pediatric patients treated surgically is presented. Seven of the patients presented with transient ischemic attacks or minor strokes. One patient presented with the complaint of a "swishing" noise in the ear. All the patients had cerebral angiography done. The diagnosis was Moya-Moya disease in four cases, traumatic carotid dissection in two cases, and idiopathic carotid stenosis and idiopathic carotid disease in one case each. All the patients had surgical procedures to increase the blood supply to the brain. The surgical procedures performed include nine EC-IC bypass operations, two encephalomyoarteriosynangiosis, two encephalomyosynangiosis, and one internal carotid artery ligation. There were no complications or mortality. The management of surgically treatable lesions in cerebrovascular occlusive disease in children is discussed. The careful evaluation of all children presenting with a stroke is emphasized. If a definite medical cause for stroke is not found, selective four vessel cerebral angiography is indicated. The potential benefit of surgical intervention may depend on the promptness with which diagnosis is made.

21. BALLOON EMBOLIZATION OF NON-TRAUMATIC
EXTRA-CRANIAL VERTEBRAL ARTERIOVENOUS FISTULA IN
CHILDREN

Balaji Sadasivan, Manuel Dujovny, Bharat Mehta, Ghaus
M. Malik, James I. Ausman (Detroit, MI)

Two children, aged four and eight years, with non-traumatic vertebral AV fistula were treated by balloon embolization. The first patient presented with neck bruit, occasional headache and mild cardiomegaly. The second patient presented with increasing frequency of headache, photophobia, episodes of frequent blinking, squinting, and pursing of lips, and a neck bruit. This patient was managed as a case of migraine with benign childhood tics for several years. Both patients had an extra-cranial vertebral AV fistula on cerebral angiography. In both patients, under fluoroscopic guidance, using a percutaneous endovascular technique, a detachable silicon balloon was placed in the fistula. The auditory evoked potential was monitored during the procedure. Immediate post-embolization angiography showed occlusion of the fistula. Follow-up angiography a year later also showed occlusion of the fistula in both cases. In both patients, the bruit disappeared and symptoms improved. Pre and post-embolization MRI and CT scan studies were done in both patients. There were no complications. The management of non-traumatic vertebral AV fistula in children and the technique of embolization is described. The pre and post-embolization angiography, CT and MRI studies are discussed.

22. EXPERIENCE IN PEDIATRIC NEUROSURGERY WITH THE
LINEAR ACCELERATOR AS A NEUROSURGICAL TOOL

Ken R. Winston (Boston, MA)

Four patients with arteriovenous malformations of the brain have been treated with stereotactic radiosurgery. The details of these cases and the short-term follow-up form the basis of this report. The system used is new and was developed for stereotactically delivering high doses of radiation to precisely located sites of approximately 0.6 - 12.8 ml within the brain. This is accomplished with a Brown-Roberts-Wells (BRW) stereotactic apparatus and a 6 MeV linear accelerator equipped with a special collimator (12.5 - 30 mm diameter). The dosage outside the treatment field declines to 80% of the prescribed dose over 1.8 mm and to 50% over the next 3.4 mm. This system allows localization by either CT or cerebral angiography. Treatment was accomplished with an arcing beam of photon radiation with the head rigidly immobilized in each of four positions. The system has been extensively tested for accuracy of alignment and for distribution of radiation. Safety of operation was emphasized throughout the design and testing phase.

1. SELECTIVE DORSAL RHIZOTOMY FOR SPASTICITY

Jogi V. Pattisapu, Marion L. Walker (Salt Lake City, UT)

Spasticity results from an exaggerated and pathological spinal reflex response to muscle stretch. Various therapeutic modalities have been attempted to relieve spasticity, and of these, selective dorsal rhizotomy (SDR) appears most promising, since it permanently relieves spasticity while sparing sensation and bladder function.

Over a two year period, 31 children underwent selective dorsal rhizotomy for spasticity. Cerebral palsy was the etiology in 24 children and 4 patients had spasticity due to cerebral anoxic encephalopathy. Two children developed spasticity following severe head injury and one patient had lower extremity spasticity secondary to thoracic transverse myelitis. The mean age was 7.3 years (range 3-18 years).

The procedure is performed through a limited L2 and S2 laminectomy. The posterior rootlets are stimulated individually and the action potentials from the muscle groups are recorded and correlated with the observed motor response. The abnormal rootlets from L2 to S2 responsible for spasticity are thus identified and divided. The patient is entered into an intensive rehabilitation program for posture and gait pattern training.

There were no complications except one superficial wound infection (compl. rate of 3%) with excellent relief of spasticity in all the children. Transient hyperesthesia and numbness were seen in 75% of the children and improved within two weeks. Motor function significantly improved in 22 cases and was stable in 4 patients; 5 children are still progressing in therapy. Interestingly, 13 of 21 children with upper extremity involvement had functional improvement with decreased spasticity, better coordination and hand control. The average follow up is 10.6 months (range 4-32 months).

Based on our findings, selective dorsal rhizotomy is most beneficial in children with spasticity secondary to cerebral palsy, anoxic encephalopathy, and head injury; its efficacy in patients with spasticity due

1. SELECTIVE DORSAL RHIZOTOMY FOR SPASTICITY (Con't)

to spinal cord injury remains to be determined. Patient selection is most crucial for a successful outcome, and younger children have better results.

Selective dorsal rhizotomy corrects the fundamental abnormality within the reflex arc to balance the facilitatory and inhibitory influences on the anterior horn cell, thereby providing the most logical and permanent relief of spasticity.

2. CLINICAL AND ELECTROPHYSIOLOGIC STUDIES IN SELECTIVE DORSAL RHIZOTOMY

Leslie D. Cahan, Lauren M. Beeler, David McPherson,
M. S. Kundi (Orange, CA)

Fasano and Peacock have described selective dorsal rhizotomy for alleviation of spasticity in children with spastic diplegia (Little's disease). We here present the results of a series of 30 patients undergoing this operation. Pre and post-operatively, patients were assessed by a variety of electrophysiologic examinations (somatosensory evoked potentials of mixed nerves of upper and lower extremities (cSSEP) and F-wave and H reflex studies) as well as independent physical therapy evaluation (assessment of lower and upper extremity tone using the Ashworth scale; range of motion in arms, legs and spine; and functional mobility skills).

The patients in this report had the electrophysiologic studies performed one week after surgery and again 3 to 6 months later. The physical therapy assessments are reported at the 6 month follow up.

Significant reduction of muscle tone in lower extremities especially in adductors, hamstrings, internal rotators, quadriceps are seen without significant clinical sensory loss. Response of tone in plantar flexors of the feet is more variable; while some children experience good reduction of tone, in others some hypertonicity remains.

Range of motion has improved in lower extremities especially about the hips. Persistent limitation of motion about knee (hamstring) and ankle (plantar flexors) is seen in some children, but others experience good improvement here as well.

Functional gains are seen but are closely dependent upon preoperative status and post operative physical therapy.

Electrophysiologic studies have had a number of interesting findings: (1) many of these children have abnormal cSSEP preoperatively; (2) usually the cSSEP is unchanged in latency after surgery; (3) some children

2. CLINICAL AND ELECTROPHYSIOLOGIC STUDIES IN SELECTIVE DORSAL RHIZOTOMY (Con't)

have electrophysiologic evidence of spinal cord dysfunction preoperatively; (4) H_{max}/M_{max} ratio decreases after surgery confirming decrease in tone.

3. SELECTIVE POSTERIOR RHIZOTOMY FOR THE TREATMENT OF SPASTICITY: RELATIONSHIP OF INTRAOPERATIVE EMG PATTERNS TO THE POST-OPERATIVE COURSE

Rick Abbott, Jeffrey H. Wisoff, Neil Spielholz, Alfred D. Grant, Allan M. Strongwater, Fred J. Epstein (New York, NY)

Fifty patients with spastic cerebral palsy have been treated by selective posterior rhizotomy over the past twelve months. Intraoperative EMG was carried out with 5 or 8 channel monitoring to include both quadriceps and gastrocnemius. The fifth channel monitored either the right hamstring. The sixth seventh and eighth channels when available were used to monitor the left hamstring and the right and left adductors. Additionally, somatosensory evoked potentials within the spinal cord were monitored in some of these patients using an epidural catheter.

We observed several characteristic patterns of abnormal response when stimulating the component rootlets of the L2-S2 dorsal roots in these patients. These responses can be subdivided into two components; that which is obtained during stimulation of the rootlet and that which persists afterwards. We observed, as have others, both tetanic contraction and spread of activity to involve antagonistic muscles and muscles outside the myotactic unit. We also observed other abnormal responses within the synergistic muscle groups; specifically, a loss of fatiguing in the muscle during stimulus trains and an irregular electrical response in the musculature suggesting polysynaptic transmission.

Surgical and monitoring technique will be discussed and the intraoperative EMG pattern types and somatosensory evoked potentials will be correlated with the post operative course of the patients in an attempt to grade the degree of abnormality of these patterns thus increasing their intra-operative utility.

4. H-REFLEX (H2/H1 RATIO) AS A STANDARD FOR ELECTROPHYSIOLOGICAL MONITORING IN SELECTIVE POSTERIOR RHIZOTOMY

Bruce B. Storrs, Takashi Mishida (Chicago, IL)

The H-Reflex is a standard electrophysiological measurement that may be performed pre-, post-, and intra-operatively. The H2/H1 ratio is predictably altered in disease stated affecting the upper motor neuron. This alteration in function allows those posterior rootlets contributing to the problem to be reliably identified. Stimulator motion artifact is eliminated because of the use of a sub-maximal threshold stimulus. There is a high correlation between abnormal H2/H1 ratio and abnormal muscular performance with maximal stimulus. The use of this technique allows approximately 50% of tested rootlets to be identified as electrically abnormal.

The use of a standardized test and equipment will allow reproducibility of results and evaluation of multi-center studies.

23. OUTCOME OF MODERATE HEAD INJURY IN CHILDHOOD

Thomas G. Luerksen, Melville R. Klauber, Lawrence F. Marshall, Howard M. Eisenberg (San Diego, CA)

A longitudinal prospective study which included three metropolitan areas acquired data on 1906 head injured patients less than 15 years of age. One-hundred and thirty-eight children suffered a moderate head injury by Glasgow Coma Score (GCS) criteria, presenting with a GCS of 9, 10, 11 or 12 after nonsurgical resuscitation.

The data from these patients were studied in order to characterize the presentation and outcome of moderate head injury in childhood. Skull fractures were common, occurring in one-third of the patients, but depressed skull fractures were rare. Eighty-seven patients (63%) underwent CT scanning, and one-third of the CT scans showed abnormalities. Almost 20% of the CT scans showed cortical contusion, or swelling. Although surgical lesions were rare, epidural hematoma was seen most frequently. Mortality was only 1.4%, and a good outcome by Glasgow Outcome Scale scoring was achieved in over 90% of patients.

These results are remarkably different from those reported for the adult age group, in regard to outcome, the distribution of surgical lesions and the occurrence of abnormalities on admitting CT scans. These differences and the overall expectations of outcome for moderate head injury in childhood will be discussed.

(This study was supported by Contracts N01-NS-9-2312, 2313, and 2314B of the National Institute of Neurological and Communicative Disorders and Stroke)

24. AN UNUSUAL SYNDROME OF PEDIATRIC BRAINSTEM TRAUMA

Daniel D. Galyon, Jeffrey A. Winfield (Syracuse, NY)

Prior concepts of brainstem injury secondary to trauma are based on autopsy studies which describe these lesions in conjunction with diffuse white matter injury of the brain. The clinical hallmark of such lesions is an alteration in level of consciousness. We present two cases of apparently minor pediatric head trauma without associated changes in sensorium which resulted in a delayed (6-12 hours), profound hemiparesis that could not be explained on the basis of CT findings.

Magnetic resonance imaging is a well recognized modality for the evaluation of brain injuries not visible on routine CT examinations, particularly those involving the brainstem. Both patients we present had MRI scans shortly after presentation demonstrating a focal lesion contralateral to their hemiparesis in either the ventral pons or midbrain. These lesions were interpreted as isolated contusions of the cerebral peduncle. One patient had several small lesions in the periventricular white matter consistent with diffuse shearing injury, while the other patient's brainstem lesion was an isolated finding. In this latter patient, a follow-up MRI showed a corresponding region of encephalomalacia. Both children's hemiparesis resolved completely within several weeks of their injury.

We have identified a clinical syndrome following minor pediatric head trauma which results in a brainstem injury visualized only by MRI. These lesions can exist in isolation of associated brain injuries, produce a pure motor deficit without alteration of consciousness, and carry a favorable prognosis. The postulated mechanics of these injuries, as well as a possible explanation for the delay in appearance of a deficit will be discussed in the context of relevant literature.

25. THE INFANT WHIPLASH SHAKE INJURY SYNDROME: A CLINICAL AND PATHOLOGICAL STUDY

Mark N. Hadley, Volker K. H. Sonntag, Harold L. Rekate (Phoenix, AZ)

Infant child abuse is not uncommon. The "shaken baby syndrome" has been well described in the literature and is characterized by retinal hemorrhages, subdural and/or subarachnoid hemorrhages and minimal or absent signs of external trauma. Of dispute is whether the infant suffers a shake whiplash injury or whether actual cranial impact trauma is required to produce the injury pattern common to this syndrome.

We have had experience with 35 cases of severe nonaccidental head injuries in infants. Of this group 12 infants (age 1.5 to 13 months) had no evidence of external cranio facial trauma or skull/facial fractures as determined by standard radiographic and CT studies. All 12 patients in this isolated whiplash subgroup presented with decreased level of consciousness, seizures and retinal hemorrhages. All had a low hematocrit at the time of admission and all 12 had evidence of subarachnoid and/or subdural hemorrhages on CT studies.

Seven of 12 patients died. Complete autopsies were obtained on 5 patients. All had subdural hematomas and brain swelling with contusions. Four of 5 patients had high cervical spinal cord epidural/subdural hemorrhages and 3 of 5 patients had high cervical spinal cord contusions. This is the first study to document high spinal cord injuries in whiplash shake syndrome. These findings demonstrate that severe cranial and spinal neurological injury can occur following whiplash shake injury and suggest that direct cranial trauma is not an essential component of the injury mechanism. The clinical characteristics and outcome of these patients and the pathophysiology of the whiplash shake injury will be presented.

26. THE CLINICAL EXAM, BRAIN SCINTIGRAPHY AND EEG IN THE ASSESSMENT OF BRAIN DEATH IN CHILDREN

Ann Marie Flannery, James J. Conway, Sue Weiss, Shelly Lester, Martha J. Barthel, David G. McLone (Chicago, IL)

A retrospective review found 100 children under the age of 16 evaluated for brain death since 1973. We reviewed the clinical exam including pupillary response, caloric stimulation and respiratory effort. We determined the most probable cause of brain damage, the number of EEG's and the EEG results as well as the number of brain scintigrams and brain scintigram can result. Other factors examined include the use of barbiturates, muscle relaxant drugs and vasopressors, the admitting coma score, patient temperature, patient age, and the professional level of the physician performing the clinical exam.

Eighty-two children had brain scintigraphy diagnostic of absent cerebral perfusion. Except for two children who could not physically be examined, the clinical exam revealed no evidence of respiratory effort, pupillary response or response to caloric stimulation. Twenty-six of 82 children (31%) had evidence of EEG activity.

In 18 cases (18%) cerebral perfusion was present on brain scintigraphy. In this group, of the EEG's done (16) 15 of 16 (93.7%) showed activity. Seven of 18 (38%) survived. These seven cases all showed evidence of respiratory effort on clinical exams. Eleven died despite continued support; seven were evaluated for respiratory effort and all showed a response.

We conclude that although a clinical exam with emphasis on pupillary response, caloric response and apnea testing continues to be the cornerstone in the determination of brain death, in those cases where high dose barbiturates, muscle relaxants, or other factors confound the exam, brain scintigraphy can reliably show absent cerebral perfusion. In all age groups including neonates, a positive correlation exists between respiratory effort and evidence of cerebral perfusion on brain scintigraphy. We found no survivors when brain flow was absent despite 26 cases in which the EEG showed activity.

27. MANAGEMENT OF CENTRAL NERVOUS SYSTEM HEMORRHAGE ASSOCIATED WITH HEMOPHILIA

John C. Godersky, Arnold H. Menezes, C. Thomas Kisker, Raymond Tannous (Iowa City, IA)

The presence of coagulation defect may potentially complicate the management of central nervous system (CNS) hemorrhage. Nine male patients with hemophilia and CNS hemorrhage, have been treated over the past ten years (15 total admissions for CNS hemorrhage). Their ages ranged from 6 months to 22 years (mean 7 years) at the time of first CNS hemorrhage. The hemorrhages were intracranial in 14 instances and the remaining one a spinal epidural hemorrhage. Intracranial hemorrhages included 2 epidural hematomas, 3 subdural hematomas, 5 intracerebral hematomas, and 5 episodes of subarachnoid hemorrhage. All patients were treated with factor replacement maintained at a 100% level for at least 10 days. Operative procedures for clot removal were required during 5/15 admissions - a total of 10 operative procedures being performed (5 in one child). The trauma leading to the hemorrhage was considered trivial in 9 cases, moderate in 2, severe in 1 and in 3 instances the hemorrhage was spontaneous. The interval from trauma to hospital admission ranged from hours to 8 days with a mean of 3 days. One person was left with a moderate disability and the remainder made a good recovery. The median hospital stay was 2 weeks per event, although 2 people were hospitalized for over 10 weeks. We conclude that, 1) CNS hemorrhage associated with hemophilia is a manageable problem; 2) Surgical indications are the same as in the general population; 3) Although multiple procedures may be required to control the hemorrhage, the overall prognosis is good.

28. SEVERE HEAD INJURY IN CHILDREN: THE USE OF I-123 HIPDM SPECT SCAN AND ARTERIOJUGULAR VENOUS O-2 DIFFERENCE

John Ruge, Yoon S. Hahn, James Conway, Zehava Noah David G. McLone (Chicago, IL)

Raised intracranial pressure in children with severe head injury may be associated with either cerebral ischemia or hyperemia. Therapy should be directed to minimize ischemic complications of the brain. In order to identify those children at risk for further ischemic damage of the brain, a pilot study was performed on six children with severe head injury. Single Photon Emission Computed Tomography (SPECT) gives a tomographic image of regional cerebral blood flow as previously shown in adults with either ischemic stroke or hyperemia associated with seizure activity.

SPECT scan was performed on six children with severe head injury (GCS 3-5) within the first four days following the head injury using intravenous I-123 labelled HIPDM.

(N.N.N.-trimethye-N[2-Hydroxy-3-Methyl-5-Iodobenzy]l-1,3-Probenediamine). In addition arterio-jugular venous oxygen difference (AVDO-2) measurements were correlated with SPECT scan, CT scan, ICP, MAP & PaCO-2. For two children SPECT scans were repeated about a month later for follow-up.

A preliminary study showed the following results:

1. In certain cases, SPECT scans were more sensitive than CT scans and showed areas with deranged blood flow (hyperemia or ischemia).
2. The positive uptake of I-123 HIPDM was mostly correlated with a decrease of AVDO-2 suggesting regional hyperemia (or inhomogenous blood flow).
3. All 72 AVDO-2 measurements performed within the first five days showed a trend of regional hyperemia except for three whose AVDO-2 were over 10.
4. The combination of the regionally sensitive perfusion data generated by SPECT scan and the global perfusion data of AVDO-2 may aid in the management of the severe head injury in children.

29. EFFICACY OF BARBITURATES IN HEAD INJURED CHILDREN

Richard D. Bucholz, Thomas Pittman, Diane Williams (St. Louis, MO)

Recent studies have shown an extremely poor prognosis in head injured pediatric patients with intracranial pressures (ICP) greater than 40 and a coma scale of less than 6. High ICP reduces cerebral perfusion pressure (CPP), producing cerebral ischemia. As barbiturates reduce cerebral metabolic rate, their use may directly alter the outcome following low CPP, and detract from the prognostic value of ICP reading.

A retrospective analysis was performed on all children receiving large doses of barbiturates admitted to a pediatric hospital between 1984 and 1985. Seven children were identified who sustained head injury with a Children's Coma Scale (CCS) of less than 5. All children were maximally treated with Mannitol, hyperventilation, and drainage and were placed on barbiturates only when ICP became uncontrollable. In the 7 children treated, CPP less than 50 torr for at least 2 to 3 hours and in two CPP fell transiently to 0 during plateau waves. At 12 months following injury, 3 have had good recoveries, 2 experience moderate disability, and 2 are vegetative. Although hypotension was seen in all patients during barbiturate coma, none experienced permanent sequela.

Barbiturate therapy markedly alters the outcome in patients with high ICP and low coma scores. Although hypotension is seen with this therapy, low CPP is tolerated. Preliminary results of barbiturates in a multi-center study of uncontrolled ICP in adults have demonstrated their efficacy. This study suggests a similar trial should be conducted with children. In addition, low CPP in patients treated with barbiturates are not necessarily indicative of irreversible cerebral damage.

30. HYPERBARIC OXYGEN IN THE TREATMENT OF ELEVATED INTRACRANIAL PRESSURE AFTER HEAD INJURY

Jeffrey A. Brown, Mark C. Preul, Assad M. Taha (Toledo, OH)

Hyperbaric oxygen (HBO) has been used as an adjunct to the treatment of cerebral edema. There has been limited clinical evaluation of the extent and time course of effectiveness of HBO in treatment of cerebral edema, especially in association with head injury. HBO may be most useful in the treatment of the "diffuse cerebral swelling" seen with pediatric head trauma. We studied the effect of HBO at 2 atmospheres on ICP in 2 patients, ages 5 and 21 years, with diffuse cerebral swelling after blunt trauma and after a gun shot wound, respectively. Both required controlled hyperventilation, osmotic diuretics and ICP monitoring. ICP, mean arterial blood pressure, pulse and atmospheric pressure were recorded at one minute intervals during one-hour treatments and for 15 minutes before and after HBO therapy. Controlled hyperventilation was continued during therapy and PCO₂ was measured at intervals. Each patient underwent 4 treatments. Data was divided into 5 phases: 1) prior to therapy 2) during descent from 1-2 atmospheres 3) at 2 atmospheres 4) during ascent from 2-1 atmospheres 5) after therapy. Mean ICP during descent dropped from a mean of 13 to 8 torr, rising to 14 torr during therapy at 2 atmospheres and to 16 torr during ascent to 1 atmosphere, then returning to 12 torr after therapy. We conclude from this preliminary work that HBO lowers ICP during treatment of diffuse cerebral swelling during the descent phase of therapy. No lasting effects of treatment are seen after concluding therapy. Ambient temperature elevation during therapy may account for the loss of effectiveness of the HBO. Further careful study in patients with injury severe enough to require ICP monitoring is needed.

31. CEREBRAL SALT WASTING (CSW) POST HYPOTHALAMIC REGION SURGERY: DIAGNOSIS, MANAGEMENT AND PATHOPHYSIOLOGY.

Antonio Disclafani, Gregory Stidham, Robert Sanford (San Francisco, CA)

CSW is a condition in which hyponatremia in neurologically insulted patients results from excessive urinary sodium loss. The condition is thought to result from a failure of the CNS to modulate renal sodium handling, although proof of this mechanism was lacking. We have observed 5 children status post hypothalamic region surgery who exhibited true CSW. The recently described diuretic and natriuretic hormone, atrial natriuretic hormone (ANH) has been implicated as the responsible factor in three patients in whom detailed endocrinological studies were done, including ANH, ADH, aldosterone, renin, thyroid function and cortisol levels. Detailed studies of daily sodium and water balance were undertaken in fourteen patients undergoing surgery for hypothalamic region tumors in our institution between 1981 and 1986. Thirteen of these patients had craniopharyngiomas, one had a pituitary adenoma. Five of these children experienced significant hyponatremia (less than 125 mEq/L) which could not be explained by water retention. All five of these patients had polyuria which resembled the polyuria of diabetes insipidus expected in this patient population, but had a profound natriuresis not compatible with the diagnosis of D.I., with urine sodium concentrations of 275-325 mEq/L while hyponatremic and in documented negative water balance.

Three patients with CSW had high ANP and high normal ADH in the face of negative water and sodium balance. We believe ANH produced the negative sodium/water balance and that the high normal ADH levels were an appropriate response to volume depletion.

We feel that CSW may mimic SIADH and DI and that its pathophysiology mandates a unique therapeutic approach to avoid serious electrolyte disturbances.

32. COMPUTER-ASSISTED STEREOTACTIC BIOPSY OF INTRACRANIAL LESIONS IN PEDIATRIC PATIENTS

Dudley H. Davis, Patrick J. Kelly, W. Richard Marsh, Bruce Kall, Stephen J. Goerss (Rochester, MN)

A computer-assisted stereotactic biopsy technique has been employed in 26 pediatric patients with intracranial lesions whose ages ranged from 3 to 16 years. The computer program allows the integration of stereotactically gathered CT, MRI and digital angiographic data in the planning of a biopsy trajectory. Eleven patients had cortical or subcortical lesions, 6 patients had lesions in the thalamus or hypothalamus, 7 patients had lesions in the brain stem or cerebellum and 2 patients had lesions in the pineal region. Diagnostic tissue was obtained in 24 cases. In 2 cases therapeutic interventions, third ventriculostomy and cyst aspiration, were accomplished at the time of the biopsy. There were no major complications related to the procedure, however 2 patients had transient worsening of their preoperative neurologic deficit. The technique, histologic results and complications will be presented and discussed. Computer-assisted stereotactic biopsy is a valuable diagnostic alternative in the pediatric patient with an intracranial lesion.

33. USE OF STEREOTACTIC TECHNIQUES IN CHILDREN

Jogi V. Pattisapu, Marion L. Walker, Bruce B. Storrs, M. Peter Heilbrun (Salt Lake City, UT)

Stereotactic biopsy has been popularized over the last decade since the advent of newer and more sophisticated instrumentation and technology. However, less than 200 cases of pediatric stereotaxy are recorded in the literature, and few reports emphasize the necessary modifications required in children.

52 stereotaxic procedures were performed on 49 children using the Brown-Roberts-Wells (BRW) apparatus at our institution. The mean age was 9.6 years (range 11 mos to 18 yrs). The indication for the technique was diagnostic in 30 cases, and therapeutic in 7 cases; location was a factor in 15 children, with the lesion involving a "deficit prone" area of the brain.

The procedure is begun in the CT scanner, and performed under general endotracheal anesthesia. The base ring is applied after intubation and a localizing scan of the involved area is obtained. The patient is transported to the operating room, and the biopsy is performed using the computer generated coordinates, which are confirmed on the phantom base.

Virtually all areas of the brain were accessible using the BRW apparatus in our series including 7 biopsies of posterior fossa lesions. Eleven procedures were stereotaxy-guided craniotomies for precise localization of small or critically located lesions. Follow-up CT scans were obtained immediately after the procedure to confirm the accuracy of the biopsy site. The BRW technique was applicable in 48 cases (92%); inability to enter cysts or biopsy of necrotic tissue and tumor capsule occurred in 8% of the cases. One patient had a transient increase in cerebral edema, for an overall complication rate of 2%.

It appears that the BRW stereotactic technique is equally effective in children and adults, and will no doubt play an ever increasing role in the future of pediatric neurosurgery.

34. TEMPORAL LOBECTOMY FOR COMPLEX PARTIAL SEIZURES BEGINNING IN CHILDHOOD

Giuseppe Erba, John R. Adler, Keasley Welch, Robert Ziegler, Peter McL. Black (Boston, MA)

From 1972-84 at the Children's Hospital in Boston, 34 patients underwent temporal lobectomy for medically intractable complex partial seizures beginning in childhood. Mean age at onset was 6.4±8.0 years, and the age at surgery 18.8±1.0 years (mean±SEM). For statistical analysis, patients were divided into two groups: those completely seizure-free and those with persistent seizures. Two factors appeared important in rendering patients seizure free: the duration of seizures and the pathological findings. Mean duration of seizures for patients with successful surgery was 11.4±1.1 years and for those with persistent seizures 19.3±3.1 years (mean±SEM).

There was also a significant correlation between duration of seizures and poor outcome. Regarding pathological findings, there were 18 patients with tumors, eight with atrophic lesions, three with AVMs, three with nonspecific changes and two with hamartomas. Less satisfactory outcomes were associated with diffuse gliosis or nonspecific pathological changes. In patients with tumors, worse outcome was associated with longer duration of seizures. These data suggest that relief from intractable seizures beginning in childhood will be easier to achieve if surgery is performed early.

35. INTRAOPERATIVE CORTICAL MAPPING TO ENHANCE TUMOR RESECTION IN THE PEDIATRIC AGE GROUP

Mitchel S. Berger, George A. Ojemann, (Seattle, WA).

Cortical mapping techniques have been utilized in 8 pediatric patients with intrinsic glial brain tumors to 1) maximize resection while minimizing neurological risks by identifying eloquent areas, and 2) maximize seizure control by including epileptogenic zones in the resection.

The patients (4 female, 4 male) were aged 4.5 to 19 years. All had a seizure history and a CT/MRI lesion in temporal (5), posterior-medial temporal (1), parietal (1), or frontal (1) lobes. Intraoperative mapping in all cases (6 under general anesthesia) included electrocorticography (ECOG) to identify the epileptogenic zone and identification of Rolandic cortex by electrical stimulation evoked movements, supplemented by somatosensory evoked potentials, in one case. Language cortex was identified in 3 patients, aged 12 and 19 years by intraoperative stimulation mapping under local anesthesia, and the other patient (age 4.5 years) via a chronic subdural electrode assay placed 1 week prior to resection.

In all cases, the ECOG demonstrated epileptogenic zones which were surrounded by, but did not involve, the gross tumor nidus. All patients have been seizure-free following resection of both the gross tumor and epileptogenic zones. Intraoperative identification of Rolandic cortex allowed resection of a tumor in somatosensory cortex without a motor deficit. Identification of language areas allowed dominant temporal lobe resections of up to 100 mm from the temporal tip to be accomplished without postoperative aphasias. With the large resections that could be safely done with cortical mapping, post resection CT scans showed no evidence of disease in these cases.

36. USE OF ^{19}F MAGNETIC RESONANCE SPECTROSCOPY FOR MEASUREMENT OF CEREBRAL BLOOD FLOW

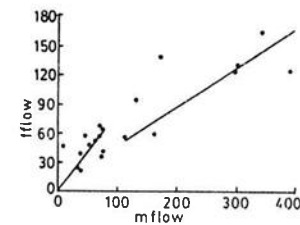
D. Barranco, L.N. Sutton, A. McLaughlin, J. Greenberg, (Philadelphia, PA)

CBF is widely determined by the clearance of some radioactive substance, such as ^{133}Xe , but radioactivity limits its use, especially in children. We have developed a method of determining regional CBF non-invasively using clearance of Freon 23 (CH_2F), a commonly used commercial refrigerant.

Eight cats were anesthetized with ketamine and placed in a 2.2T superconducting magnet with a 2 cm. surface coil over the calvarium. CBF determinations were done by allowing the decay of the fluorine peak obtained at 88.1 MHz over a 15-minute period of desaturation as the freon was abruptly discontinued. CBF was calculated for each run by the formula $\text{CBF} = \frac{1}{t_e} \cdot \frac{1}{\lambda}$, where... is the partition coefficient for freon ($\lambda=1.5$) and t_e is the time for the freon peak to fall to 1/e of its baseline value. Flows were obtained at 3 pCO₂ values: baseline normocapnic, hyperventilation (pCO₂ 10) and hypercarbia (pCO₂ 70). At each point CBF was simultaneously determined by triple labeled microsphere technique using cobalt, strontium and scandium.

CBF determined by freon inhalation showed the expected responsiveness to alteration in pCO₂ and correlated well with microsphere flows up to values of 100 ml/100 gm/min ($R=0.78$) (see graph [mflow=microsphere, fflow=freon]). At higher flows, freon underestimated microsphere flow by 1/2, as is reported with stable Xenon. Freon inhalation was well tolerated by the animals and had little effect on ICP, MAPB or pCO₂.

It is clear from our data that inhalational freon has potential as a technique for the non-invasive measurement of CBF.



37. THE LATERAL DECUBITUS POSITION FOR THE SURGICAL APPROACH TO PINEAL LOCATION TUMORS

J. Gordon McComb, (Los Angeles, CA)

Placing the patient in the lateral decubitus position provides a comfortable working distance and stance for the surgeon. The surgeons hands can be placed side by side at the waist or chest level obviating the need to work with the arms extended, one hand above the other as would be the case with the patient sitting. Another factor of considerable importance is that the operating microscope can be maneuvered over a wide arc in the horizontal and vertical planes, the latter further augmented by raising and lowering the height of the operating table.

A posterior interhemispheric retrocallosal or transcallosal approach provides the shortest and most direct access to the structures in proximity to the pineal region with minimal physiologic consequence. Excellent exposure to the region can be easily obtained by draining CSF and allowing gravity to retract the dependent hemisphere. Most lesions can be approached retrocallosally, thereby avoiding resection of the corpus callosum. The falx cerebri and tentorium can be divided to provide additional exposure.

Other advantages of placing the patient in the lateral decubitus position compared to the sitting include less set-up time (eliminating the need for a right atrial catheter), reduction of postural hypotension, decreased cardiovascular instability and negligible risk of air embolism.

38. CHILDHOOD POSTERIOR FOSSA TUMORS MANAGED WITH INTRAOPERATIVE CO₂ LASER AND POSTOPERATIVE MRI

Charles L.Y. Cheng, John Ragheb, Walker Robinson, John Joslyn, (Baltimore, MD)

The advent of the CO₂ laser has brought "no touch technique" to the already delicate approach to posterior fossa surgery in childhood. Using the CO₂ laser, vital and eloquent areas can be dissected precisely under the microscope without excessive retraction, vibration, manipulation, or exploration of the cerebellar hemispheres, cerebellar peduncles, 4th ventricle, or brainstem. Sixty consecutive cases of pediatric brain tumors treated with microscope mounted CO₂ laser were reviewed, including 24 astrocytomas, 22 medulloblastomas, 8 ependymomas, 3 ependymoblastomas, and 3 hemangioblastomas. Ninety-seven percent were gross total resections, 3% were near total. Operative morbidity was only 7%, with no mortality. Recurrence rate was 7% at 4.5 years average followup. In addition, more recent case were followed post-operatively with magnetic resonance scans which yielded good correlation with clinical course. Combining intraoperative CO₂ laser and postoperative MRI has greatly enhanced the safety and efficacy of pediatric posterior fossa surgery.

39. THREE-DIMENSIONAL DISPLAY OF THE VENTRICLES AND CISTERNS FROM ROUTINE, NON-CONTRAST CT SCANS. HYDROCEPHALUS AND CONGENITAL MALFORMATIONS

Thomas P Naidich, Sharon E. Byrd, Erin C. Prenger, Arthur Nieves, Bruce C. Teeter

Standard image data from routine head CTs can now be reprocessed in minutes to display cerebrospinal fluid free from surrounding brain and bone without need for any contrast agent(s).

The 3-D image processing is performed on-line using standard commercially available hardware (GE 9800) and a software package that is to be released by GE in the first quarter of 1988. Linear and angular measurements of the ventricles and cisterns can be made from the 3-D image. Volumetric measurements will be available soon. Thus, this technique can be utilized for patient management throughout North America.

The 3-D images are generated from the routine image data, not the raw data, so patients may be studied routinely in a busy department and the 3-D images can be generated later, overnight or at other convenient times. The 3-D images may also be obtained on any old case from the magnetic tape archives.

Practically, the 3-D images help the radiologist and neurosurgeon to assess ventricular sizes, desired sites and trajectories for shunting, size changes post shunting, loculations, arachnoid cysts, relationships of the ventricles to masses and the complex anatomy of congenital malformations.

40. MANAGEMENT OF CRANIOPHARYNGIOMA IN CHILDREN

Michael S. Edwards, James E. Baumgartner, Rodger Hudgins, Charles B. Wilson, (San Francisco, CA)

A series of 42 children (age less than 18 yr.) with craniopharyngioma were treated over a 20 year period 1966-86. Of these, 21 were male and 21 female with a mean age of 9.2 yr. (range 3-17 yr.). The most common presenting symptoms were headache 30/42, decreased linear growth 19/42 and visual dysfunction 19/42. Hydrocephalus was found on 18/38 patients preoperative studies, but only 2 patients required V/P shunting before definitive surgery. Endocrinopathies were found in 17 of 38 patients preoperatively.

Ten tumors were approached transphenoidally, 29 fronto-temporally and 3 via a bifrontal route. There were 10/42 total resections, 5/42 presumed total resections, 14/42 aggressive subtotal resections and 13/42 biopsies +/-cyst aspiration. A second operation was required in 10/42 patients while 6/42 required more than 2 operations for tumor recurrence. Radiation (approx. 5000 rad. XRT) was given to 22/42 patients following their initial surgery, 7/42 following their second surgery and 13/42 received no radiation. Follow-up averaged 7.2 yr. (range 1.5-21 yr.), 3 patients died and 3 were lost to follow-up.

Of 10/42 patients undergoing total resection, none recurred and none received XRT. The 5/42 presumed total resections received no XRT following their first operation. All 5 recurred with 4 requiring a second operation and 1 more than 2 operations. All 5 received XRT following operation #2. 12/14 patients undergoing aggressive subtotal resection received XRT, 4/12 recurred with 3 requiring a second operation and 1 more than 2 surgeries. Followup was incomplete on 1 patient and 1 patient died. Of the patients undergoing biopsy +/-cyst aspiration, 10/13 received XRT, 6 recurred, 1 required a second operation and 5 more than 2 operations. There were 2 deaths and 2 patients were lost to follow-up. All deaths occurred in patients who refused XRT. Endocrinopathy was found in 40/42 patients postoperatively and in 38/39 at last follow-up. Worsening endocrine status was most frequently associated with repeat operation. Vision remained normal in 17/42 patients, improved from

40. MANAGEMENT OF CRANIOPHARYNGIOMA IN CHILDREN (Con't)

abnormal to normal in 8/42, improved from abnormal to better in 1/42, deteriorated from normal to abnormal in 3/42 and changed from abnormal to worse in 10/42 patients. Of the 13 patients with decreased vision at last follow-up, 8 had tumor recurrence requiring reoperation. The results of this study suggest that total resection and aggressive subtotal resection with XRT are the most acceptable forms of treatment of craniopharyngioma in children.

41. PITUITARY ADENOMAS IN CHILDHOOD AND ADOLESCENCE

Between 1972 and 1987 more than 1,600 pituitary adenomas were treated by transsphenoidal microsurgery. Approximately 6% of these tumors occurred in patients less than 20 years of age. These patients are the subject of this report.

A retrospective analysis of 92 patients under 20 years of age was completed (Table). There were 28 boys (29%) and 65 girls (71%) ranging in age from 5 to 19 years. The female preponderance is mainly the result of the relative large number of postpubertal teenaged girls with prolactinomas. The mean age was 16 and the majority of patients were 12 years of age or older. Only 6 patients were less than 12 years old, 5 boys and 1 girl.

Hyperfunctioning pituitary tumors and their clinical manifestations were present in 90% of the patients, but it was interesting to note that some 40% of the patients had large and often invasive tumors. ACTH secreting tumors accounted for more than half of those seen in patients less than 12 years old. Technical aspects of transsphenoidal surgery in children will be discussed. All patients with visual loss improved and there was no operative mortality.

Type of Tumor	# of pts	Table M/F	Micro/ macro	Age range, years
Prolactin	52	8/44	25/27	12-19
ACTH	26	13/13	22/4	7-19
GH (acromegaly)	9	3/6	3/6	15-19
Others (TSH,-SU, Null)	5	4/1	0/5	15-18
Total	92	28/64	50/42	7-19

Although pituitary tumors in children are uncommon, when they do occur they are frequently large and aggressive. The relative incidence of macroadenomas to microadenomas is higher in children than adults, the incidence of ACTH-secreting tumors is higher, and the incidence of invasive tumors is also somewhat higher. Transsphenoidal microsurgery, occasionally followed by radiation therapy, provides excellent control in the majority of cases.

42. CHOROID PLEXUS TUMORS OF CHILDHOOD: A FORTY-FIVE YEAR SURGICAL EXPERIENCE AND HISTORICAL PERSPECTIVE

Richard G. Ellenbogen, Ken R. Winston, William J. Kupsky (Boston, MA)

A series of 40 consecutive surgically treated choroid plexus tumors from the Children's Hospital in Boston between the years 1941 and 1986, inclusive is reviewed. This is the largest pediatric series of this rare tumor described in the literature. All medical records, operative reports and histopathologic specimens were reviewed by the authors. The clinical features and pathology of the choroid plexus tumors, benign and malignant, were analyzed with respect to long term neurologic outcome. The mean followup for the survivors was 12 years, with the range from 6 months to 30 years. Evolving management issues including surgical approaches to these tumors and treatment of their sequelae are discussed.

The primary goal in all cases was total excision which was accomplished in 33 of the patients. Review of the pathologic specimens revealed that fifteen were choroid plexus carcinomas and 25 were papillomas based on strict adherence to Russell and Rubinstein's and Lewis' criteria for malignancy. The overall mortality rate for this operative series was 25%. Of the 10 fatalities, 7 patients had carcinoma and 3 papilloma. The neurologic outcome of the surviving children was excellent or good for 15 with papilloma and 7 with carcinoma. It was fair or poor for 6 children with papilloma and 2 with carcinoma. The major operative morbidity consisted of a seizure disorder (23%), visual field defect (23%) and mild hemiparesis (23%). Three carcinomas previously believed to have been completely excised recurred. Although CSF overproduction was documented in several of our patients, complete resection of choroid plexus tumors did not invariably alleviate the associated hydrocephalus.

The histological appearance of these tumors did not always correlate with biological behavior or prognosis. For, some tumors with relatively benign histologic features were invasive, while some anaplastic tumors had well circumscribed surgical borders and were thus curable. The characteristics

43. PINEAL REGION LESIONS: A PEDIATRIC SERIES

Hillel Z. Baldwin, Volker K. H. Sonntag, Harold L. Rekate (Phoenix, AZ)

The treatment of pineal region masses remains controversial. At issue is whether it is in the patient's best interest to explore the lesion at the time of their diagnosis, or whether the obstructive hydrocephalus should be shunted, and the posterior third ventricular tumor irradiated without a tumor diagnosis. The authors' experience with the clinical presentation, differential diagnosis, and therapeutic intervention of 11 children with pineal region lesions is discussed. Mean age at diagnosis was 7.1 years, with a range of 3 weeks to 12 years old. With the safety of modern neurosurgical techniques, a therapeutic protocol was implemented, making possible: 1) accurate histological diagnosis in all patients, indicating the necessity of adjuvant radiation and/or chemotherapy, 2) gross total resection of tumor in 64% of cases, 3) subtotal resection in 36%, 4) avoidance of subjecting patients with radioresistant tumors (45%) to the unnecessary morbidity of radiotherapy, 5) 0% surgical mortality rate, and 6) 91% survival with a mean follow-up of over 2 years. The anatomic rationale for the various approaches to pineal region lesions is discussed, and in consideration of present operative expertise, the traditional approach of empiric radiation to these lesions may no longer be warranted.

44. PEDIATRIC CHORDOMAS: TWO CASES AND LITERATURE REVIEW

Lawrence Foody, Kerry R. Crone, Thomas S. Berger (Cincinnati, OH)

Chordomas are uncommon tumors and rarely present early in life. During the past 17 years two patients with chordomas have been treated at Children's Hospital Medical Center.

A four month old male presented in 1970 with torticollis. Diagnostic studies including skull films, cervical spine series, and myelography were considered normal. Transsection of the sternal cleidomastoid provided temporary relief. One year later, a cervical myeloradiculopathy developed resulting in a laminectomy which revealed a presumed cervical arteriovenous malformation. The child progressively deteriorated and died one month later. Autopsy revealed a clival chordoma with medullary intraparenchymal extension and multiple systemic metastases.

An 11 year old female complained of diplopia and progressive lethargy. Computed tomography and magnetic resonance imaging demonstrated a destructive rostral clival mass resulting in hypophyseal elevation and pontine compression. Following a translabial-transphenoidal excision, the patient remains asymptomatic.

Our review of the literature reveals 86 chordomas in patients less than 20 years of age. Cranial chordomas were most common (63.7%) followed by spinal (18.7%) and sacrococcygeal (17.6%). This is in contrast to the general literature where the sacrococcygeal tumors account for 50-55%.

Thirty-six cranial cases survived for at least one month and provided some follow-up data. Patients who received both surgery and radiation appeared to have a better prognosis. 4/19 died (mean 39 months) as compared to 5/9 (mean 34 months), treated solely with surgery. Nine of 19 patients treated with combination therapy have survived for significantly longer periods 60-252 months (mean 113.5).

45. GANGLIOGLIOMA: PROGNOSIS AND RATIONALE FOR POST-OPERATIVE MANAGEMENT

Edison P. McDaniels, II, Philip H. Cogen (Stanford, CA)

Ganglioglioma is a rare neoplasm of the central nervous system characterized histologically by the presence of neuronal elements in a glial matrix. This tumor commonly presents in children and adolescents, and adult patients are often able to date the onset of their symptoms to an early age. Despite the usually benign pathology of the lesion, most series to date contain patients that received post-operative radiation therapy, particularly when the surgery consisted of biopsy or partial resection only. In light of the long-term side effects of such treatment, especially in young adults and children, we have reviewed our series of patients who underwent excision of gangliogliomas, to address the question of appropriate post-operative management. In addition, we have been able to follow a small group of patients with malignant elements in their tumors, to better define their outlook.

The records of eleven patients were available for study. While the mean age at presentation was 20.3 years, the majority dated their symptoms to childhood or adolescence, with one patient symptomatic for 23 years. Seven of the patients underwent resection of a histologically benign lesion, and none received radiation therapy post-operatively. One infant died in the immediate post-operative period of sepsis. Four of the patients in this group had either a biopsy or subtotal resection only. Of these patients, 3 have long-term follow-up (19, 16, and 5 years), and there has been no evidence of progression in either their symptoms or tumors. Three of the patients with benign lesions underwent total resection, and there has been no recurrence to date, although 1 patient died of unrelated causes 3 years post-operatively. The remaining 4 patients in the series had evidence of malignant glial elements in the tumor, and all received post-operative irradiation. One patient was lost to follow-up. The remaining 3 patients are all alive and stable without evidence of recurrence at 4 and 5 years, despite the glioblastoma-like histology in 2 of the lesions.

45. GANGLIOGLIOMA: PROGNOSIS AND RATIONALE FOR POST-OPERATIVE MANAGEMENT (Con't)

In conclusion, we feel that it is justified to withhold post-operative radiation therapy in patients with benign gangliogliomas, even if surgical resection is partial. Our long follow-up period, the longest to date for non-irradiated patients, supports this recommendation. There is, however, a clear role for irradiation if the tumor is malignant, and preliminary evidence suggests that these patients may fare better than those without ganglion cells in their lesions.

46. GADOLINIUM-DPTA ENHANCED MR IMAGING IN DETERMINING TREATMENT OPTIONS FOLLOWING SURGICAL RESECTION OF PEDIATRIC BRAIN TUMORS

Curtis A. Dickman, Harold L. Rekate, C. Roger Bird, Marjorie Medina (Phoenix, AZ)

Gadolinium-DPTA is a chelated paramagnetic contrast agent available experimentally for use in MR imaging. Following administration, an increased signal intensity in tumor and improved tumor delineation may result. We have examined the utility of enhanced MR imaging in the pediatric population following resection of brain tumors. An analysis of 15 pediatric patients with a history of previous brain tumors is presented. All patients underwent postoperative Gadolinium-DPTA enhanced MR imaging, which was used to guide further therapy. Residual tumor was demonstrated in 9 of the 15 patients. Four patients had additional surgery, 2 had radiation therapy, and 1 had immunotherapy based on the findings of the enhanced MR studies. Surveillance was recommended for the remaining 8 patients. Gadolinium-DPTA enhanced MR imaging appears to be a safe and effective means of providing an accurate postoperative assessment in pediatric brain tumor patients. It is as effective as contrast enhanced computerized tomography and has the advantages of the sensitivity and anatomic resolution provided by MR imaging.

47. PROGNOSTIC IMPORTANCE OF CELLULAR PLOIDITY IN MEDULLOBLASTOMA OF CHILDHOOD

Masaharu Yasue, Tadanori Tomita, Herbert Engelhard, David G. McLone, Kenneth Bauer (Chicago, IL)

The DNA content of 53 medulloblastomas was analyzed by means of flow cytometry and compared with the clinical and histological findings. DNA analysis showed about half of the cases were diploid and another half were aneuploid. The patients of the young age group had more diploid tumors than aneuploid tumors. The rate of undifferentiated tumors to differentiated tumors among the aneuploid group was higher than that among the diploid group. No correlation was found between Chang's T staging system and the DNA ploidy, whereas the M staging correlated with the ploidy; diploid medulloblastomas had a greater tendency to metastasize than aneuploid medulloblastomas ($P < 0.01$). Four year survival was compared with the extent of resectability, dose of radiation, and ploidy. The patients with total resection, appropriate post operative radiation, and aneuploid medulloblastoma had a better prognosis than those with subtotal, inappropriate radiation, and diploid group ($P < 0.025$, and $P < 0.25$, respectively). Only 1 out of 8 patients with diploid medulloblastomas which was resected subtotally survived while all of the 7 patients with aneuploid medulloblastomas resected totally survived. Although the G0/G1 phase fraction in the alive group and the S phase fraction in the dead group were a little higher than the G0/G1 phase fraction in the dead group and the S phase fraction in the alive group, these cell cycle analyses did not offer significant information.

48. A NOVEL CHEMOTHERAPEUTIC AGENT FOR THE TREATMENT OF MEDULLOBLASTOMA: MUTANT DIPHTHERIA TOXIN LINKED TO ANTI-TRANSFERRIN RECEPTOR MONOCLONAL ANTIBODY OR TRANSFERRIN

Charles J. Wrobel, Virginia Johnson, John Zovickian, Richard Youle (Bethesda, MD)

Immunotoxins are hybrid proteins composed of a monoclonal antibody linked to a peptide toxin. Peptide toxins, such as diphtheria toxin, consist of an A chain that can inhibit cellular protein synthesis, and a B chain that binds cells and then facilitates A chain translocation across the cell membrane. Most immunotoxins are conjugates of an A chain and a monoclonal antibody that targets the A chain to cells bearing the appropriate antigen. However, antibody mediated membrane translocation of the A chain is much less efficient than B chain mediated entry, so that A chain conjugates are relatively nontoxic. We report the development of a highly potent immunotoxin for medulloblastoma therapy that utilizes a full length diphtheria toxin, CRM 107, bearing a critical point mutation that conserves B chain entry function but greatly diminishes cell binding.

Since medulloblastoma cells express increased levels of transferrin receptor relative to normal brain, CRM 107 was linked to 454-A12, an anti-transferrin receptor monoclonal antibody. Cytotoxicity was assayed using the permanent medulloblastoma cell lines SNB 40 and TE 671. Concentrations of 3.5×10^{-11} M and 2.6×10^{-11} M respectively decreased protein synthesis to 50% of control cultures (IC_{50}). CRM 107 was also covalently linked to human transferrin (Tfn); the IC_{50} values were 3.9×10^{-13} M and 2.1×10^{-12} M.

Because cell lines adapted to culture conditions may possess transferrin receptor levels different than the tumors of origin, we established two primary medulloblastoma cultures and determined sensitivity to Tfn-CRM 107. The IC_{50} of one tumor was 2.5×10^{-12} M; another tumor was less sensitive (IC_{50} of 1.1×10^{-10} M).

48. A NOVEL CHEMOTHERAPEUTIC AGENT FOR THE TREATMENT OF MEDULLOBLASTOMA: MUTANT DIPHTHERIA TOXIN LINKED TO ANTI-TRANSFERRIN RECEPTOR MONOCLONAL ANTIBODY OR TRANSFERRIN (Con't)

To assess the feasibility of intrathecal therapy, single dose toxicity testing was performed in the guinea pig and Rhesus monkey. Both species tolerated intrathecal concentrations of Tfn-CRM 107 one to three logs greater than the IC_{50} 's for medulloblastoma cells. Guinea pigs tolerated a Tfn-CRM 107 CSF concentration of 1×10^{-9} M while 1×10^{-8} M was uniformly fatal. Two Rhesus monkeys tolerated 3.3×10^{-10} M and 1×10^{-9} M initial CSF concentrations of Tfn-CRM 107, and one monkey tolerated 1×10^{-9} M 454-A12-CRM107. These results suggest that intrathecal immunotoxin therapy with CRM107 conjugates may be useful for treatment of medulloblastoma or prophylaxis of the craniospinal axis.

49. A TALE OF TWO TAILS

Robert F. Keating, James T. Goodrich, (Bronx, NY)

The human tail has long been a medical curiosity and while examples are less frequently cited today, past literature documented many strange and unusual tales regarding lumbosacral dermal appendages. Often these appendages (pseudotails) are anomalous prolongations of coccygeal vertebrae, as well as lipomas and/or teratomas. Nevertheless, in humans, true tails may contain adipose and connective tissue, muscle or blood vessels, and are covered with skin. Bone, cartilage, notochord and neural elements are lacking.

Two newborn infants were recently encountered at our institution, both possessing true tails. Both infants were born full term after normal prenatal periods, to healthy mothers and were seen to have midline lumbosacral dermal appendages. Neither child had any neurological deficit; however, one did have other associated congenital anomalies. In addition, both individuals had normal radiological studies.

After surgical excision, both tails were studied histologically and seen to consist of adipose and connective tissue as well as having a fibrous stalk. Neither appendage had any neural, muscular or bony elements.

For the past two centuries these clinical curiosities have generated considerable discussion regarding the premise that ontogeny recapitulates phylogeny. Today, this concept remains popular, although others believe these circumstances are merely pathological entities.

50. SACRAL AGENESIS

Michel Malek, Anthony E. Gallo, Jr., (Portland, OR)

Sacral agenesis is a rare congenital condition affecting multi organ systems and therefore is of interest to neurosurgeons, urologists, orthopedists and others. Because of the multiple view points from which various specialists view this condition, the literature concerning sacral agenesis has been characterized by a lack of uniformity, and the entity has been ill defined. This paper attempts to redefine sacral agenesis by analyzing all 436 cases reported between 1852 and 1986, reviewing their epidemiology, diagnosis, clinical manifestations, associations, possible etiologies, and various classifications. A pathogenic model is proposed; vertebral primordia laid down along faulty intervertebral planes or a non-functional T-flow results in failure of production of cell surface components required for cellular interactions resulting in defective interaction between neural tube and notochord, ultimately accounting for each of the clinical manifestations. Treatment of associated hydrocephalus, spina bifida, and unstable spine is conventional. Surgical exploration of the cord leads to uniformly poor results in the absence of compressive lesions. Prognosis can be improved by early diagnosis and treatment.

51. CERVICAL MYELOMENINGOCELES - DESCRIPTION AND A NEW THEORY REGARDING EMBRYOGENESIS

Mark S. Dias, Dachling Pang, Diana Claassen (Pittsburgh, PA)

Seven children with cervical myelomeningocele were seen at the Children's Hospital of Pittsburgh over the last eight years. All seven children, at birth, had normal or nearly normal sensorimotor function. Cervical spine roentgenograms at the involved levels demonstrated widened interpedicular distances, bifid laminae and, in three instances, abnormally formed vertebral bodies. Magnetic Resonance Imaging (MRI) in several cases strongly suggested a split spinal cord within the spinal canal opposite the myelomeningocele sac; in one case, a ventral band of fibrous tissue was seen arising from the dorsal aspect of the subjacent vertebral body and forming a midline keel which bisected the spinal cord. Metrizamide myelography and computed tomography performed in three cases clearly demonstrated a split spinal cord underlying the myelomeningocele sac, in all seven cases, a normal appearing spinal cord was visualized, either by MRI or by myelography/CT, within the spinal canal caudal to the myelomeningocele.

Histological examination of the excised stalk of neural tissue from the myelomeningocele sac demonstrated, in every case, elements of a malformed spinal cord. Numerous mature neurons were present in a disorganized, gliotic neuropil; many neurons were large and resembled anterior motoneurons. Ependyma was also observed; in three cases seemingly complete, ependyma-lined central canals were seen.

In two patients, delayed neurological deterioration from spinal cord tethering led to a re-exploration of the myelomeningocele. In each case, a split spinal cord was found. In one case, the operative findings confirmed the pre-operative MRI image of a midline keel extending from between the two "hemicords" to the dorsal aspect of the vertebral body.

The preserved sensorimotor function in these patients implies the presence of an intact, functioning spinal cord caudal to the myelomeningocele. Yet histologic examination of the tissue within the myelomeningocele sac suggests that at least part of the spinal cord ends

51. CERVICAL MYELOMENINGOCELES - DESCRIPTION AND A NEW THEORY REGARDING EMBRYOGENESIS (Con't)

blindly within the placode. We feel that cervical myelomeningocele arise, not from simple myeloschisis of a single neural plate, but from partial or complete spinal cord duplication, in which one or part of both "hemicords" leaves the bony spinal canal through a dorsal duralschisis to end blindly in the myelomeningocele sac, while the other, more normal "hemicord", or the major portion of both "hemicords", remains within the spinal canal to innervate the body segments below the lesion. Theories of spinal cord duplication are reviewed. One theory, proposed by Bremer, postulates that abnormal accessory neurenteric canals may develop embryologically and are responsible for many forms of occult spinal dysraphism, including diastematomyelia, neurenteric cysts, and anterior spina bifida. In light of the above findings, we propose that cervical myelomeningocele represent a special form of the "split cord syndrome" caused by accessory neurenteric canals.

52. THE COMBINATION OF VATER ASSOCIATION AND SPINAL DYSRAPHISM

Randall Chesnut, Hector E. James (San Diego, CA)

The Vater Association is a recognized nonrandom presentation of vertebral defects, imperforate anus, esophageal atresia with tracheo-esophageal fistula, radial and renal dysplasia, and other anomalies which have been documented in the literature. Spinal dysraphia is a known syndrome of the occult spinal congenital disorders, with insidious presentation and progressive neurological deterioration involving lower extremities, bladder, or both. We here report 2 children with the previously unrecognized combination of both the Vater Association and Spinal Dysraphism, in the same patients. Both had the spectrum of the Vater Association. One had in the first year of life repeated urinary tract infections and subsequent deterioration of bladder function. At 2 years of age she was referred to neurological surgery because of these findings and the presence of progressive distal weakness of the lower extremities. She was noted to have a cavus deformity of both feet. On spinal CT she demonstrated a tethered filum and conus to a spinal and subcutaneous lipoma. She underwent operation, release of tethered cord and radical resection of the spinal lipoma. A postoperative SCF fistula was treated successfully with a subcutaneous peritoneal shunt. The second child was noted at birth to have the Vater Association and a deformed right foot, with cavus deformity. On pelvic and subsequently spinal high resolution ultrasonography, she was noted to have a tethered spinal cord and a thick filum terminale. This was confirmed at operation at 4 months of age where a thickened filum and tethered conus was encountered. This was sectioned and released.

These 2 patients demonstrate that upon recognizing the presence of the Vater Association in a given patient, the spinal canal should be studied with the appropriate neuroimaging to determine that the patient does not also have the combination of these nonrandom findings with spinal dysraphia. In reference to the possible causative mechanisms, they will be analyzed.

53. MR IMAGING OF LIPOMYELOMENINGOCELE AND TETHERED CORD

John D. Brophy, Leslie N. Sutton, Robert D. Zimmerman, Edward Bury, Luis Schut (Silver Spring, MD)

The operative and MR findings of 25 patients with the diagnosis of lipomyelomeningocele and/or tethered cord at the Children's Hospital Philadelphia were compared. Post-operative MR's on eight patients, five of whom were asymptomatic, were also compared to the pre-operative studies. In this review there was one false negative MR and four MR's in which the relationship of the lipoma to the conus or filum was not demonstrated accurately. In six cases incidental intramedullary cystic lesions at the conus were identified by MR. All post-operative MR's (one month - two years) demonstrated no change in the level of the conus. MR is an accurate screening modality in the initial diagnosis of occult spinal dysraphism. MR was not useful in the post-operative evaluation of lipomyelomeningocele and the tethered cord since the caudal, posterior displacement of the conus was unchanged in all studies.

54. DETETHERING - DOES IT ALWAYS HELP?

John R. Mawk, James R. McConnell (Omaha, NE)

Over the past two years, twenty children have undergone detethering for a variety of progressive symptoms. Detethering clearly improves function in most children with simple thickened filum, lipomyelomeningocele or other more complex variants of myelodysplasia. Children with classic myelodysplasia or myelodysplasia complicated by diastematomyelia, prior detethering or prior suboptimal repair do not uniformly benefit from release of the spinal cord. Some children are objectively improved but are actually functionally worse - a unique but definite finding in two of our patients. Mechanical injury of already dysplastic cord tissue, ischemia or venous stasis within the cord, and transfer of tethering force from cord to roots are all discussed as reasons for failure to improve or actual worsening. Detethering in classic myelodysplasia is a hazardous procedure with unpredictable outcome.

55. IS THE LONG TRACT INVOLVED IN THE EXPERIMENTALLY TETHERED CORD? STUDY OF HRP AXONAL TRANSPORT

Takaharu Fuse, John W. Patrickson, Shokei Yamada (Loma Linda, CA)

Various studies have indicated that the lesions of the tethered cord syndrome are located in the gray matter of the lumbo-sacral segments. However, some cases with long tract signs may involve the interruption of the neuronal fibers in the long tract. The present study was designed to determine if the continuity of these fibers are preserved in the tethered cord. The axonal transport of the neural tracer, horseradish peroxidase (HRP), was used to determine the neural continuity in the posterior column which represents a distinct component that can be easily analyzed.

Thirteen cats were used; four acute tethered, four chronic tethered, and five control. In the acute tethered group, laminectomy was performed under anesthesia and a 5 gram-weighted ligature was attached to the filum terminale. During traction the filum was securely sutured to the dura. HRP was then injected into nucleus gracilis. In the chronic group, HRP injection was performed 2-13 months after tethering. Control animals underwent HRP injection only. All animals were sacrificed 72 hours post-injection, and L7 and S2 dorsal root ganglia were removed. Frozen sections were made and prepared for identification of HRP positive neurons using the chromogen, tetramethyl benzidine.

The number of labeled ganglion cells was not significantly different between any of the groups.

In conclusion, the axonal continuity of the long tract is maintained in the experimentally tethered cord. This supports the concept that tethered cord syndrome is primarily a gray matter disease as previously proposed.

56. MANAGEMENT OF CHILDHOOD HYDROMYELIA

Jeffrey H. Wisoff, Fred Epstein (New York, NY)

The authors review their experience in the management of 22 patients with hydromyelia over a 26 month period. Ten children had a Chiari I malformation and hydromyelia, 4 children had myelomeningoceles: 3 with large thoracic cavities and 1 had a cervical hydromyelia, 6 children had distal syrinxes associated with tethered cords and occult dysraphism, and 2 patients had post arachnoiditis syrinxes.

All patients were investigated with pre-operative MRI and intraoperative ultrasound. These neurodiagnostic examinations dictated the type of surgical intervention. Patients with Chiari I, Arnold-Chiari II with cervical hydromyelia, or basal arachnoiditis had decompression of the hindbrain malformations, myelotomy with drainage of the cyst and placement of a stent. When the syrinx extended to the obex, as demonstrated by intraoperative ultrasound, the obex was plugged. The children with myelomeningocele and thoracic hydromyelia had cyst-pleural shunts. Patients with distal syrinxes underwent modified terminal ventriculostomy.

The classical presentation of brachial amyotrophy and dissociated sensory loss was present in only 3 patients. Progressive scoliosis without neurological deficit, pain, and Lhermitte's phenomena were common presentations. The patients with tethered cords were generally asymptomatic from their syrinx.

57. LUMBOPERITONEAL SHUNTS FOR TREATMENT OF SYRINGOMYELIA

T.S. Park, Wayne S. Cail, Melissa G. Walker (Charlottesville, VA)

Gardner's hydrodynamic theory to explain the pathogenesis of syringomyelia is based on an assumption that the central canal between the 4th ventricle and the syrinx is patent. We examined 1) whether the patent central canal is essential for maintenance of the syrinx; 2) whether drainage of CSF via a lumboperitoneal shunt effects reduction of the syringeal cavity.

We reviewed 13 patients in whom syringomyelia was diagnosed by MR. All patients except one were younger than 16 years. Associated pathological abnormalities of the patients were Chiari II anomaly in 9, Chiari I anomaly in 2, posterior fossa arachnoidal adhesion in 1 and lipomyelomeningocele in 1. In 12 patients, MR showed no evidence of the patent central canal rostral to the upper end of the syrinx. The upper end of syrinx was located between C1 and the mid-thoracic level. In only one patient who had Chiari II anomaly, MR suggested the possible existence of a CSF-filled channel between the 4th ventricle and the syrinx. These observations corroborate recent MR studies which failed to visualize the direct communication.

Lumboperitoneal shunts were placed in 7 of the 13 patients. Postoperative MR revealed marked and sustained reductions of the syringeal cavities in 4 patients. Repeat MR has not been obtained in the remaining 3 patients. In 3 patients with Chiari I anomaly, the intracranial and intraspinal pressures were concomitantly measured before the shunting procedure. In the sitting position, the resting state intraspinal pressure always exceeded the intracranial pressure. When abdominal compression or Valsalva maneuver was applied to the patients in the sitting position, the intraspinal pressure rose predominantly.

The reduction of the syringeal cavity following placement of lumboperitoneal shunts is attributed to decreases in the intraspinal pressure. Our observations support the notion of Ball and Dayan, and Aboulker that the syrinx is formed and maintained by CSF tracking under pressure into the spinal cord from the spinal theca.

58. CLINICAL AND PATHOLOGIC ANALYSIS OF CHILDHOOD SPINAL CORD ASTROCYTOMAS

Eugene Rossitch, Jr., Seth M. Zeidman, W. Jerry Oakes, Carter Harsh (Durham, NC)

Spinal cord astrocytomas are rare, accounting for only 4% of CNS tumors in childhood. We present eleven children with pathologically confirmed spinal cord astrocytomas and look at possible correlations between degree of resection, pathologic characteristics, and recurrence.

In our series, the most common presenting complaints were weakness, scoliosis and gait disturbance, regardless of tumor location. Six tumors were primarily located in the cervical cord, while five were in the thoracic region. The most common radiographic abnormality was cord widening. Eight tumors were biopsied or subtotally resected while three were felt to be grossly resected. Seven patients received postoperative radiation.

Pathologic material was analyzed with respect to histologic grade, degree of cellularity, and presence or absence of microcysts, necrosis, vascular proliferation, calcium, nucleoli, mitotic figures, Rosenthal fibers, or dark nuclei. All eleven tumors were fibrillary astrocytomas with a low or moderate degree of cellularity. No necrosis, vascular proliferation or mitotic figures were seen in any specimen. Microcysts and calcium were found in one tumor. Nucleoli were seen in five tumors, with Rosenthal fibers and dark nuclei seen in four respectively.

Recurrence was seen in 4/11 patients, two of whom died of their disease at 96 and 84 months following surgery. Seven children remain symptom free with followup ranging from 2 years to 35 years (mean = 12 years). Patients undergoing biopsy or subtotal resection had recurrence in 3/8 cases with 2 deaths. Patients who received a gross total resection had recurrence in 1/3 cases with no deaths. The uniformly well differentiated lesions seen on pathologic examination correlate with the relatively benign clinical course seen in this series. The histology, however, was not useful in predicting recurrence.

59. MALIGNANT ASTROCYTOMAS OF THE SPINAL CORD

Alan R. Cohen, Jeffrey Wisoff, Jeffrey Allen, Fred Epstein (New York, NY)

Malignant glial tumors, which so commonly affect the brain, occur only rarely in the spinal cord and have been described infrequently in the neurosurgical literature. During the past 8 years we have cared for 160 patients with astrocytomas of the spinal cord, 19 of whom (8%) harbored malignant tumors. This report details the biological activity of this subset of malignant astrocytomas.

There were 10 male patients and 9 females whose ages ranged from 1.5 to 32 years with a median age of 14. Twelve tumors were cervical and 7 were thoracic. The median duration of symptoms was 1 month prior to diagnosis. Three patients, however, had a subacute course, initially presenting with low grade astrocytomas which appeared to undergo malignant transformation.

Surgery consisted of radical cytoreductive tumor resection in all cases with drainage of rostral and caudal cysts when found. All patients received adjunctive radiotherapy and 10/19 received chemotherapy as well. The pathologic diagnosis was grade III in 7 cases and grade IV in 12.

To date 13/19 patients (68%) in this series are dead with a median survival of 6 months following surgery. Of the 6 survivors, only 3 have lived longer than 1 year, and 2 of these 3 are paraplegic with loss of sphincter function.

Hydrocephalus was present in 10 patients, 7 of whom underwent ventricular shunting procedures. Eleven of the 19 (59%) developed disseminated disease, most commonly on the basis of subarachnoid spread. Extranatural metastases did not occur in the absence of a ventricular shunt.

We conclude that malignant astrocytomas of the spinal cord are characterized by a short prodrome, rapid and relentless clinical deterioration, with a propensity to produce hydrocephalus and craniospinal dissemination. The authors' discuss their rationale for operation and encourage an aggressive approach which includes multi-agent chemotherapy and radiation directed at the entire neuraxis.

60. SURGICAL MANAGEMENT OF LAMBDROID SYNOSTOSIS: REVIEW OF 88 CASES

Antonio Disclafani, Donald A. Ross, Roger Hudgins, Michael Edwards (San Francisco, CA)

Isolated lambdoid synostosis is unusual with only a few surgical series published since 1960. We report a retrospective series of 88 consecutive lambdoid synostectomies performed by wide craniectomy. 64% of operations were performed prior to six months of age. 51% were right sided, 34% left, and 15% bilateral. Sex distribution was nearly equal. 24% had at least one other associated condition, often a chromosomal anomaly. Clinical presentation consisted of flattened occiput, prominent forehead, ipsilateral prominence of the maxilla and mandible and anterior displacement of the ipsilateral ear. CT scan and/or plain skull films were positive in 80% and negative in 7%. 14% of the patients were operated on based on clinical findings alone. Surgical indications are progressive deformity in a child that is not responsive to positioning changes or neck exercises.

80% had an excellent result (surgeon and parents pleased), 16% a good result (surgeon noted mild residual deformity), and 4% a fair result (surgeon and parents note residual deformity). The three fair results were obtained in patients with Apert Syndrome, severe fetal alcohol syndrome and microcephaly. The syndrome mean hospital stay was 3.8 days. The mean blood loss was 40cc.

Postoperative complications (30 days post surgery) included two cases of SIDS in children with Apert Syndrome and upper airway difficulty. One patient developed pneumonia and one a superficial wound infection. 13% required transfusion.

Wide synostectomy for lambdoid synostosis achieves good or excellent cosmetic results in greater than 95% of all cases with minimal morbidity.

61. LAZY LAMBDROID (SKULL BASE ABNORMALITY -
NONSURGICAL TREATMENT)

Robert A. Sanford, Matthew Wood, Jr., Michael S. Muhlbauer (Memphis, TN)

Various terms have appeared in the literature to describe the skull base asymmetry in which there is occipital flattening. In some cases, this asymmetry results from premature closure of the lambdoid sutures. The term "lazy lambdoid" has been coined to denote similar skull base abnormalities in which the lambdoid sutures remain radiologically open.

Fifty consecutive children with skull based asymmetry and occipital flattening will be described. Eight of these had premature closure of the lambdoid suture and underwent surgical resection. Two patients had flattening due to hypertrophy of the inner surface of the lambdoid sutures, which prevented outward expansion of the skull at the suture. The other 42 children were characterized as having lazy lambdoid; their sutures were radiologically patent and CT revealed no evidence of bony hypertrophy along the inner surface of the suture. These 42 patients were treated nonoperatively and had excellent cosmetic results.

Lazy lambdoid sutures seem to comprise a clear clinical syndrome, being only associated with head tilt, and asymmetric facies with maxillary flattening on the same side of the occipital flattening. The majority of children with lazy lambdoid have a parent with similar findings. Parents exhibiting the syndrome had each been noted by their parents to have had asymmetric skulls during childhood, and had gone untreated. CT scans were obtained on 3 of these, and each demonstrated the same skull base abnormality.

In summary, lazy lambdoid is a descriptive term which refers to children who have the triad of skull base abnormality with occipital flattening, facial asymmetry and head tilt. Their appearance will greatly resemble that of the parent with similar clinical features.

62. THE SURGICAL TREATMENT OF METOPIC SUTURE
SYNSTOSIS

Tariq Javed, John E. Kalsbeck, A. Michael Sadove
(Indianapolis, IN)

Sixteen patients with metopic suture synostosis underwent seventeen surgical corrective procedures, between March 1983 and July 1987, using a modification of previous surgical techniques. The extent of surgical intervention is individually tailored to the degree of cosmetic deformity present. Our approach to this condition is as follows: a) Midline ridging and moderate cosmetic defects are treated by bifrontal craniotomy and recontouring. b) Trigonoccephaly with severe bitemporal narrowing is treated by bifrontal craniotomy followed by removal of a supra-orbital bar of bone. The supra-orbital bar is osteotomized in the midline and recontoured to the desired shape. This is secured back in place using an L shaped Luhr mini compression plate screwed into the fronto-nasal area. The bifrontal bone flap is cut along the metopic suture, rotated 180 degrees, recontoured and attached to the supra-orbital bar using stainless steel wire. c) The hypotelorism is corrected by performing an osteotomy in the nasal bone and wedging a small piece of bone into the osteotomy site.

Two patients underwent bifrontal craniotomy with recontouring, one bifrontal craniotomy with lateral canthal advancement, two bifrontal craniotomies with removal of supraorbital bar and 12 bifrontal craniotomies, removal of supra-orbital bar, osteotomy of nasal bone, and fixation of supra-orbital bar with compression plate. Mean follow-up is 24 months with no deaths, infections, CSF leaks, or damage to the visual or cerebral function. We believe this is a safe and effective approach to the surgical management of metopic suture synostosis, particularly in cases of severe deformity and the older child.

63. MULTI-SUTURE CRANIOSYNOSTOSIS IN DIZYGOTIC TWINS WITH GERMINAL MATRIX HEMORRHAGE - SURGICAL TREATMENT BY CRANIAL RELEASE AND TRANSPOSITION

George R. Cybulski, Martin E. Gryfinski (Chicago, IL)

Intraventricular germinal matrix hemorrhage causes morbidity in premature infants mainly on the basis of development of posthemorrhagic hydrocephalus. However, abnormal head shape also is observed in survivors and usually takes the form of dolichocephaly. The etiology of this skull configuration is unknown and craniosynostosis has not been reported with germinal matrix hemorrhage.

We have observed multi-suture craniosynostosis in dizygotic twins with germinal matrix hemorrhage. Prenatal history was unremarkable and family history was negative for craniosynostosis. Neurosonograms performed on the third day of life demonstrated subependymal and intraentricular hemorrhage in both infants and subsequent hydrocephalus was treated successfully with lumbar puncture. By 30 days after birth prominent frontal bossing and ridging of the sagittal sutures were noted in both infants, and skull x-rays and CT scan confirmed synostosis of the sagittal, lambdoidal, and coronal sutures.

The infants were followed until one year of age when each underwent cranial release via subtotal calvariectomy from the supraorbital ridge posterior to just above the foramen magnum. The excised cranial bones were separated and replaced in a configuration to achieve more appropriate contour. Each infant tolerated the procedure as well.

Although the etiology of craniosynostosis remains unknown the association of craniosynostosis with conditions affecting bone metabolism (vitamin D deficiency) or brain size (shunted hydrocephalus) has been noted. It is postulated that the infants presented here developed posthemorrhagic hydrocephalus and secondary craniosynostosis after the hydrocephalus was treated. A technique using cranial release and transposition was successful in treating this form of secondary craniosynostosis.

64. THE TREATMENT OF CRANIOSYNOSTOSIS IN THE FIRST YEAR OF LIFE

Concezio Di Rocco (Rome, Italy)

During the period October 1981-October 1986, 150 children less than 1 year of age have been operated on at the Catholic University, Institute of Neurosurgery, because of craniosynostosis. The diagnosis has been obtained by integrating clinical data and the findings provided by skull x-ray examination, calvarial bone scanning and computed tomography. In 20 children also the intracranial dynamics have been evaluated.

Linear craniectomies extending down to the base of the skull have been utilized for treating sagittal craniosynostosis, while more complex surgical procedures, involving the opening of the roof of the orbit and the anterior displacement of the superior orbital ridge and of the lateral canthus have been used for the correction of the anterior craniosynostoses (metopic, unicoronal and bicoronal craniosynostoses). One surgical death has been recorded (sagittal craniosynostosis).

The recurrence of the process, necessitating a second operation, has been observed in only 4% of cases, even though foreign material (to be inserted on the edges of the cut calvarial bone in order to prevent bone formation) has not been employed.

On the grounds of the obtained results and clinical follow-up, the correction of the scaphocephaly and trigonocephaly has been proved to be the most rewarding operation; on the other hand, unicoronal craniosynostosis has appeared the most challenging form of early fusion of cranial sutures.

65. EARLY INTERVENTION IN FIBROUS DYSPLASIA OF THE ORBIT AND FRONTAL VAULT

Joan L. Venes (Ann Arbor, MI)

Fibrous dysplasia is a poorly understood condition of bone, often classed among the hamartomas. It is generally agreed that the progression of bone involvement ceases at about the same time as closure of the iliac crest epiphysis occurs. Craniofacial reconstructive surgery for cosmesis in fibrous dysplasia is often delayed until that time to avoid the necessity for reoperation. Earlier intervention has been limited to cases of visual dysfunction (usually oculomotor imbalance, but occasionally decreased visual acuity).

On occasion children have been referred to the Craniofacial Center at the University of Michigan in whom either mild vertical diplopia was accepted or psychosocial implications were discounted because the surgeon did not think it appropriate to operate for this condition in the preadolescent. We have performed extensive craniectomy of the frontal and middle fossae in three children between five and eight years of age. Over a two year period the disease has not progressed, facial growth has not been impaired and the cosmetic results are excellent. The value of early cosmetic surgery in terms of body image and self-esteem is well accepted. We would like to present our operative approach to fibrous dysplasia and suggest that our results indicate that radical craniectomy and reconstruction should be considered sooner rather than later in the management of these children.

66. FORAMEN MAGNUM CHANGES IN ACHONDROPLASIA

Frederick A. Boop, Harold J. Hoffman (San Antonio, TX)

Achondroplasia occurs in 2.5/100,000 live births with most cases being sporadic. The disease is associated with a high infant mortality, with nearly half of those living longer presenting to hospital with neurologic afflictions.

We present the unusual case of an achondroplastic infant referred for evaluation of a right hemiparesis first detected at eight months of age. Investigation revealed hydrocephalus which was treated with a diversionary procedure. Her hemiparesis failed to improve and further study demonstrated an unusual lip of bone extending caudad from the posterior rim of a congenitally narrowed foramen magnum. The inferior extent of this lip of bone lay within the arch of the atlas. Following suboccipital decompression this child made a dramatic recovery.

Neurologic sequelae are commonplace in achondroplastics. The interpretation of findings in these infants is difficult with hydrocephalus, craniovertebral junction anomalies and spinal stenosis being well-recognized problems. These infants are generally hypotonic in infancy and manifest delay in motor development. Indeed, the surprisingly high infant mortality is associated with chronic aspiration and respiratory distress. Early recognition of craniocervical compression related to a congenitally deformed foramen magnum may lead to prompt surgical correction, thus preventing neurologic deterioration and, in some cases, avoiding sudden death.

The developmental pathophysiology, radiographic findings and operative results are to be reviewed.

67. SHUNT IMPLANTATION: TOWARD ZERO INFECTION

Maurice Choux, Gabriel Lena, Lorenzo Genitori, Etom Empime (Marseille, France)

Infection remains the main problem in Shunt implantation in children, even if the infection rate has significantly decreased to 3 to 5% in most of the series in the recent years.

Since we support the idea that Shunt infection is exclusively an operative complication, we have studied the factors which may be, possibly, the cause of infection.

These factors are described in the preoperative and immediate post operative period of shunt procedure.

Since January 1983 to December 1986, we have initiated a new protocol including technical modifications and preoperative antibiotics.-

Among 234 new shunts in children under 15 years old, we have had no infection. During the same period, 333 shunts revisions were performed and only one infection was detected. In this case, the patient has been operated on three times in one month.

We are convinced that 0% infection rate is not an impossible goal in Shunt implantation. We demonstrated too that late neurological and intellectual results, in cases of hydrocephalus, are closely related with the number of procedures and, especially, with Shunt infections.

Our conclusion is that Shunt implantation must be considered as a complex surgical procedure which must be done by a neurosurgeon familiar with Shunt implantation.

SECTION OF

PEDIATRIC NEUROLOGICAL SURGERY



MEMBERSHIP ROSTER

1987

Alphabetical by Name

Albright, A Leland, M.D.
 Childrens Hosp Of Pgh
 Pittsburgh PA 15213
 412/647-5090
 Active

Altenau, Lance, M.D.
 4060 Fourth Avenue
 Suite #610
 San Diego CA 92103
 Active

Amacher, A Loren, M.D.
 Dept Of Neurosurgery
 Gelsingner Clinic
 Danville PA 17822
 717/271-6437
 Active

Anderson, Frank M, M.D.
 2161 Redcliff St
 Los Angeles CA 90039
 213/661-7845
 Lifetime Active

Anderson, Jim D, M.D.
 Redwood City Offices
 1150 Veterans Blvd
 Redwood City CA 94063
 415/780-2287
 Active

Arkins, Thomas J, M.D.
 Neurosurgical Associates
 60 Temple Street
 New Haven CT 06510
 203/789-2030
 Active

Aronin, Patricia A, M.D.
 1600 Seventh Avenue, S.
 Birmingham AL 35233
 205/939-9653
 Candidate

Arpin, Elaine Joy, M.D.
 1328 Navajo Court
 Louisville KY 40207
 502/588-5433
 Active

Bailey, Walter L, M.D.
 Doctors Professional Bldg
 280 Smith Avenue, N.
 St Paul MN 55102
 Active

Bartkowiak, Henry M, M.D.
 Neurological Associates
 931 Chatham Ln
 Columbus OH 43221
 614/457-4880
 Active

Becker, Donald Paul, M.D.
 UCLA Med Ctr D/Neurosurg
 Rm/74-140 Chs 405 Hilgard
 Los Angeles CA 90024
 213/825-5111
 Active

Barger, Thomas S, M.D.
 Mayfield Neurological Inst
 506 Oak Street
 Cincinnati OH 45219
 513/221-1100
 Active

Bering Jr, Edgar A, M.D.
 Creek House
 Rt #1, Box #94
 Oxford MD 21654
 301/822-5480
 Lifetime Active

Bressler, Bruce C, M.D.
 704 S Webster Ave
 Green Bay WI 54301
 414/465-1900
 Active

Bruce, Derek A, M.D.
 7777 Forest Lane
 Suite #C-703
 Dallas TX 75230
 214/788-6660
 Active

Buchheit, William A, M.D.
 Temple Univ Health Ctr
 3401 N Broad St
 Philadelphia PA 19140
 215/221-4068
 Active

Cahan, Leslie D, M.D.
 U.C.I. Medical Center
 101 City Drive South
 Orange CA 92668
 714/634-6336
 Active

Canady, Alexa Irene, M.D.
 4160 John Rd
 Suite #1031
 Detroit MI 48201
 313/833-4490
 Active

Carmel, Peter W, M.D.
 Neurological Institute
 710 W 168th St
 New York NY 10032
 212/305-5208
 Active

Chadduck, William M, M.D.
 718 North Ash
 Little Rock AR 72205
 501/370-1448
 Active

Chapman, Paul H, M.D.
 Massachusetts Gen Hosp
 Fruit Street
 Boston MA 02114
 617/726-3887
 Active

Cheek, William R, M.D.
 Texas Childrens Hospital
 Room #0-202
 Houston TX 77030
 713/790-9053
 Active

Chour, Maurice, M.D.
 Hopital Des Enfants
 De Ia Timone
 Marseille FRANCE
 Corresponding

Coulon Jr, Richard A, M.D.
 311 Midway Dr
 River Ridge LA 70123
 504/834-7070
 Active

Alphabetical by Name

Crosby, Robert M N, M.D.
 1205 York Rd
 U.C.I. Medical Center
 Lutherville MD 21093
 301/823-1300
 Active

Davidson, Robin Ian, M.D.
 Univ Of Mass Medical Center
 55 Lake Avenue, N
 Worcester MA 01605
 617/856-3079
 Active

Dorsen, Michael, M.D.
 711 W 38th St
 Building B/Suite #B4-A
 Austin TX 78705
 512/453-0093
 Active

Duncan, Charles Cecil, M.D.
 Yale Univ Sch Of Med
 Neurosurgery Section
 New Haven CT 06511
 203/785-2805
 Active

Edwards, Michael, M.D.
 Univ Of California
 Dept Of Neurosurgery/M787
 San Francisco CA 94143
 415/476-1087
 Active

Eisenberg, Howard M, M.D.
 Univ Of Texas Medical Branch
 Division Of Neurosurgery
 Galveston TX 77550
 409/761-1500
 Active

Eisenbrey, Arthur B, M.D.
 4160 John Rd
 Suite #1031
 Detroit MI 48201
 313/833-4490
 Active

Epstein, Fred J, M.D.
 New York Univ Medical Ctr
 550 First Avenue
 New York NY 10016
 212/340-6419
 Active

Epstein, Mel H, M.D.
 Rhode Island Hosp/Dept Neuros
 110 Lockwood Street
 Providence RI 02903
 Active

Fell, David A, M.D.
 6767 S Yale
 Tulsa OK 74136
 918/492-7587
 Active

Fischer, Edwin G, M.D.
 Childrens Hosp Medical Center
 Boston MA 02115
 617/735-6009
 Active

Folts, Eldon L, M.D.
 U.C.I. Medical Center
 101 City Drive South
 Orange CA 92668
 714/634-5775
 Active

French, Barry Norman, M.D.
 2801 K Street
 Suite #300
 Sacramento CA 95816
 916/452-4811
 Active

Gahn, Norman Henry, M.D.
 100 Retreat Ave
 Suite #705
 Hartford CT 06106
 203/278-0070
 Active

Gainaburg, Duane B, M.D.
 Nicolet Clinic
 411 Lincoln Street
 Neenah WI 54956
 414/727-4210
 Active

Galicich, Joseph H, M.D.
 Memorial Hosp
 1275 York Ave
 New York NY 10021
 212/988-1629
 Active

Gallo Jr, Anthony E, M.D.
 3181 Sw Sam Jackson Park Road
 Portland OR 97201
 503/279-7736
 Active

Gamache Jr, Francis W, M.D.
 525 E 68 St
 Room #K-6
 New York NY 10021
 212/472-4909
 Active

Guido, Laurance Jacobius, M.D.
 340 Capitol Ave
 Bridgeport CT 06606
 Active

Gutierrez, Francisco A, M.D.
 1210 Broadmeadow Road
 Winnetka IL 60093
 312/256-5039
 Active

Hahn, Joseph F, M.D.
 Cleveland Clinic
 9500 Euclid Ave
 Cleveland OH 44106
 216/444-5753
 Active

Haines, Stephen J, M.D.
 Box #96 Mayo
 420 Delaware St, S.E.
 Minneapolis MN 55455
 612/636-3337
 Active

Hammargren, Lonnie, M.D.
 3196 S Maryland Pkwy
 Las Vegas NV 89109
 702/735-7272
 Active

Hammock, Mary Kathryn, M.D.
 111 Michigan N W
 Washington DC 20010
 202/745-3020
 Active

Alphabetical by Name

Hanigan, William C, M.D.
214 N E Glen Oak Avenue
Suite #500
Peoria IL 61603
309/676-0766
Active

Harwood-Nash, Derek C, M.D.
Hosp For Sick Children
Neuro Radiologist
Toronto ON M5GLX8
416/598-6026
Associate

Hawk, Thomas J, M.D.
3545 Olentangy River Rd
Columbus OH 43214
614/268-5655
Active

Hawkins III, John C, M.D.
1871 Montgomery Place
Jacksonville FL 32205
904/388-6516
Active

Hellbusch, Leslie C, M.D.
8905 Douglas Ct
Omaha NE 68114
Active

Hemmy, David C, M.D.
Medical College Of Wisconsin
8700 W Wisconsin/Neurosurgery
Milwaukee WI 53226
414/257-4846
Active

Hendee Jr, Robert W, M.D.
1010 E 19th Ave
Tammen Hall/Suite #605
Denver CO 80218
303/861-4985
Active

Hendrick, E Bruce, M.D.
Hosp For Sick Children
555 University Ave/#1502
Toronto ON M5GLX8
416/597-0808
Active

Hoffman, Harold J, M.D.
Hosp For Sick Children
555 University Ave
Toronto ON M5GLX8
416/598-1210
Active

Hollenberg, Robert D, M.D.
Mc Master Univ Medical Ct
Dept Of Surgery / Rm 4U4
Hamilton ON L8N3Z5
416/521-2100
Active

Rudson, Alan Roy, M.D.
Division Of Neurosurgery
38 Shuter St
Toronto ON M5B1A6
416/864-5588
Active

Humphreys, Robin P, M.D.
Hosp For Sick Children
555 Univ Ave/Ste 1504
Toronto ON M5GLX8
416/598-6427
Active

James, Hector E, M.D.
225 Dickinson St H-893
San Diego CA 92103
619/543-5540
Active

Jerva, Michael Joseph, M.D.
55 E Washington St
Suite #3005
Chicago IL 60602
312/236-0668
Active

Johnson, Dennis L, M.D.
Childrens Hosp Ntl M C
111 Michigan Avenue
Washington DC 20010
202/745-3020
Active

Johnson, Martin, M.D.
2800 N Vancouver
Suite #106
Portland OR 97227
503/287-2646
Active

Johnson, Mary M, M.D.
5555 Peachtree Dunwoody
Suite #337
Atlanta GA 30342
404/256-4510
Candidate

Joseph, Allen S, M.D.
2237 S Acadian Thruway
Suite #400
Baton Rouge LA 70808
504/928-5972
Active

Kalsbeck, John E, M.D.
Indiana Univ Med Ctr
545 Barnhill/Emerson #139
Indianapolis IN 46223
317/264-8910
Active

Kasoff, Samuel S, M.D.
New York Med Col/Neuros
Muger Pavillion/Rm #329
Valhalla NY 10595
Active

Keener, Ellis B, M.D.
434 Academy Street,N.E.
Gainesville GA 30501
404/532-6333
Active

Kelly Jr, David L, M.D.
Bowman Gray Sch Of Med
Section Of Neurosurgery
Winston-Salem NC 27103
919/748-4049
Active

Klein, David Mendel, M.D.
Dept Of Neurosurgery
219 Bryant St
Buffalo NY 14222
716/878-7386
Active

Kosnik, Edward J, M.D.
Chatham Vlg Prof Bldg
931 Chatham Ln
Columbus OH 43221
614/457-4880
Active

Alphabetical by Name

Kramer, Paul William, M.D.
4640 N Federal Hwy
Ft Lauderdale FL 33308
305/772-0100
Active

Laurent, John P, M.D.
Texas Childrens Hosp
Suite #0-202
Houston TX 77030
713/790-9053
Active

Laws, Edward R, M.D.
George Wash. Univ. Med. Center
2150 Pennsylvania Nw
Washington DC 20037
202/994-4035
Active

Loefer, John D, M.D.
Univ Of Washington
Dept Of Neuro RL-20
Seattle WA 98195
206/543-3574
Active

Longo-Cordero, Rafael, M.D.
Ashford Medical Bldg
Suite #604
Santurce PR 00907
809/723-9075
Active

Luerssen, Thomas G, M.D.
University Hospital H893
225 Dickinson St
San Diego CA 92103
Active

Magid, Gail A, M.D.
1661 Soquel Dr
Santa Cruz CA 95065
408/476-8900
Active

Mapstone, Timothy, M.D.
University Hospitals
2074 Abington Road
Cleveland OH 44106
Active

Marlin, Arthur E, M.D.
343 W Houston
San Antonio TX 78205
512/224-1631
Active

Mawk, John Robert, M.D.
Univ Of Nebraska Med Ctr
42ND St & Dewey Ave
Omaha NE 68105
402/559-4301
Active

McCallum, Jack E, M.D.
1522 Cooper St
Fort Worth TX 76104
Active

McComb, J Gordon, M.D.
1300 N Vermont Ave
Suite #903
Los Angeles CA 90027
213/663-8128
Active

McCullough, David C, M.D.
111 Michigan Ave,N.W.
Washington DC 20010
202/745-3020
Active

McLanahan, C Scott, M.D.
Charlotte Neuros Assoc
1010 Edgehill Road N
Charlotte NC 28207
704/376-1605
Active

McLaurin, Robert L, M.D.
111 Wellington Pl
Cincinnati OH 45219
513/381-6111
Lifetime Active

McLone, David Gordon, M.D.
Childrens Memorial Hospital
2300 Childrens Plaza
Chicago IL 60614
312/880-4373
Active

Meacham, William F, M.D.
Vanderbilt Univ Hosp
Dept Of Neuro Surgery
Nashville TN 37232
615/322-3343
Senior

Mealey Jr, John, M.D.
Indiana Univ Med Ctr
545 Barnhill/Emerson #139
Indianapolis IN 46223
317/264-8549
Active

Menezes, Arnold H, M.D.
Univ Of Iowa Hospital
Dept Of Neurosurgery
Iowa City IA 52242
319/356-2768
Active

Meyer, Glenn A, M.D.
Medical College Of Wisconsin
8700 W Wisconsin/Neurosurgery
Milwaukee WI 53226
414/257-6465
Active

Michelsen, W Jost, M.D.
161 Fort Washington Ave
New York NY 10032
212/305-5451
Active

Milhorat, Thomas H, M.D.
Health Science Ctr @Bklyn
450 Clarkson Ave/Box#1189
Brooklyn NY 11203
718/270-2111
Active

Miller, Clinton F, M.D.
330 Borthwick Avenue
Suite #108
Portsmouth NH 03801
603/433-4666
Active

Minella, Philip Arthur, M.D.
146 Wycoring St
Dayton OH 45409
513/228-5261
Active

Alphabetical by Name

Moliel, Richard H, M.D.
7400 Fannin St
1160 One Fannin Bldg
Houston TX 77054
713/797-1160
Active

Morrison, Glenn, M.D.
C/O Medical Arts Bldg
3200 S.W. 60 Court/Suite #301
Miami FL 33155
305/661-5318
Active

Moyes, Peter D, M.D.
6581 Pinehurst Drive
Vancouver BC V5K4N9
604/266-4881
Lifetime Active

Murtagh, Frederick, M.D.
Univ Of Pennsylvania Hosp
3400 Spruce St
Philadelphia PA 19104
215/662-3490
Active

Myles, Terence, M.D.
1820 Richmond Rd S W
Calgary AB T2T5C7
403/229-7805
Active

Nadell, Joseph M, M.D.
L S U Medical Center
1430 Tulane Ave/Dept Neurosurg
New Orleans LA 70112
504/588-5565
Active

Nijensohn, Daniel E, M.D.
340 Capitol Ave
Bridgeport CT 06606
203/367-4433
Active

Nilsen, Frank E, M.D.
P.O. Box #841
Manchester VT 05255
813/262-0809
Lifetime Active

O'Brien, Mark Stephen, M.D.
Emory Univ Clinic
1365 Clifton Rd, N.E.
Atlanta GA 30322
813/321-0111
Active

Oakes, W Jerry, M.D.
Box 3272
Duke Univ Med Ctr
Durham NC 27710
919/684-5013
Active

Page, Larry Keith, M.D.
1501 N W Ninth Ave
Miami FL 33136
305/547-6946
Active

Pang, Dachling, M.D.
Child Hosp Of Pittsburgh
125 Desoto St
Pittsburgh PA 15213
412/647-5090
Active

Parent, Andrew D, M.D.
Univ Of Miss Med Ctr
2500 N State Street
Jackson MS 39216
601/984-5703
Active

Park, Tae Sung, M.D.
Univ Of Virginia
Neurosurgery, Box 212
Charlottesville VA 22908
804/924-2203
Active

Parkinson, Dwight, M.D.
5111-750 Bannatyne Ave
Winnipeg MB R3E0W3
204/943-1627
Lifetime Active

Penix, Jerry O'Don, M.D.
607 Medical Tower
Norfolk VA 23507
804/622-5325
Active

Pevhouse, Byron C, M.D.
2351 Clay St
Suite #314
San Francisco CA 94115
415/923-3616
Active

Pittman, Hal Watson, M.D.
Barrow Neurological Institute
2910 N 3rd Ave
Phoenix AZ 85013
602/285-3468
Active

Pitts, Frederick W, M.D.
1245 Wilshire Blvd
Ste 305
Los Angeles CA 90017
213/977-1102
Active

Portnoy, Harold D, M.D.
1431 Woodward Ave
Bloomfield HI MI 48013
313/334-2568
Active

Pugetz, Robert H, M.D.
574 Garfield Ave
So Pasadena CA 91030
Senior

Raimondi, Anthony J, M.D.
C.P.#401
Verona ITALY
045/404-0700
Active

Ransohoff, Joseph, M.D.
New York Univ Medical Ctr
550 First Avenue
New York NY 10016
212/340-6414
Active

Reigel, Donald H, M.D.
4815 Liberty Ave
Suite #158
Pittsburgh PA 15224
412/682-0400
Active

Alphabetical by Name

Rekate, Harold Louis, M.D.
Barrow Neurological Institute
2910 N 3rd Ave
Phoenix AZ 85013
602/285-3632
Active

Reynolds Jr, Arden F, M.D.
Magan Medical Clinic
420 West Rowland St
Covina CA 91723
Active

Rhoton Jr, Albert L, M.D.
Univ Of Florida Hlth Ctr
Dept Of Neuros / Box J265
Gainesville FL 32610
904/392-4331
Active

Robinson, Walker L, M.D.
22 S Green Street
Baltimore MD 21202
301/823-1300
Active

Salmon, James Henry, M.D.
220 Anderson Drive
Erie PA 16509
814/453-6858
Lifetime Active

Sanford, Robert Alexander, M.D
Semmes-Murphy Clinic
920 Madison Ave/Suite #201
Memphis TN 38103
Active

Sato, Osamu, M.D.
Dept Of Neurosurgery
Bo Sei D Ai Isehara
Kanagawa JAPAN
046/393-1121
Corresponding

Saunders, Richard, M.D.
Dartmouth-Hitchcock M C
Section Of Neurosurgery
Hanover NH 03756
Active

Scarff, Timothy B, M.D.
6584 Professional Pl
Suite #B
Riverdale GA 30274
404/991-0150
Active

Schut, Luis, M.D.
Child Hosp Of Phila
34th & Civic Ctr Blvd
Philadelphia PA 19104
215/596-9368
Active

Scott, R Michael, M.D.
New England Medical Center
Boston MA 02111
617/956-5860
Active

Seljeskog, Edward L, M.D.
Box #479 U.M.H.C.
420 Delaware St, S.E.
Minneapolis MN 55455
612/624-6666
Active

Selker, Robert G, M.D.
Montefiore Hospital
3459 Fifth Ave
Pittsburgh PA 15213
412/648-6753
Active

Shallat, Ronald F, M.D.
3000 Colby
Berkeley CA 94705
415/843-0261
Active

Shapiro, Kenneth N, M.D.
7777 Forest Lane
Suite #703
Dallas TX 75230
212/920-4162
Active

Shillito Jr, John, M.D.
19 Denny Road
Chestnut Hill MA 02167
617/735-6012
Active

Simmons, James C H, M.D.
920 Madison Ave
Suite #201
Memphis TN 38103
901/525-8431
Active

Sklar, Frederick H, M.D.
7777 Forest Lane
Suite #C-703
Dallas TX 75230
214/361-2233
Active

Smith, Frank P, M.D.
880 Cass St
Boston MA 02111
617/956-5860
Active

Smith, Harold, M.D.
2011 Church St
Suite #800
Nashville TN 37203
615/329-7840
Active

Stein, Sherman Charles, M.D.
80 Congress Street
Springfield MA 01104
403/733-4153
Active

Steinbok, Paul, M.D.
B.C. Children's Hospital
4480 Oak Street
Vancouver BC V6H3V4
604/875-2094
Active

Storrs, Bruce B, M.D.
Childrens Memorial Hospital
2300 Childrens Plaza
Chicago IL 60614
312/880-4373
Active

Sukoff, Michael, M.D.
801 N Tustin
Suite #406
Santa Ana CA 92705
714/834-1303
Active

Alphabetical by Name

Susan, Anthony F, M.D.
3471 Fifth Avenue
Suite #811
Pittsburgh PA 15213
412/682-5900
Active

Sutton, Leslie N, M.D.
Childrens Hosp Of Phila
Dept Of Neurosurgery
Philadelphia PA 19104
215/387-6059
Active

Taekman, Michael S, M.D.
3000 Colby
Berkeley CA 94705
415/843-0261
Active

Tew Jr, John M, M.D.
506 Oak St
Cincinnati OH 45219
513/221-1100
Active

Tomita, Tadanori, M.D.
Childrens Memorial Hospital
2300 Childrens Plaza
Chicago IL 60614
312/880-4373
Active

Turner, Michael S, M.D.
1801 N Senate Blvd
Suite #535
Indianapolis IN 46202
Active

Venes, Joan, M.D.
2128 Taubman Center
1500 East Medical Ctr Dr
Ann Arbor MI 48109
313/936-5016
Active

Ventureyra, Enr Cls Garcia, M.
401 Smyth Rd
Ottawa ON K1H8L1
613/737-2316
Active

Vries, John Kenric, M.D.
Child Hosp Of Pittsburgh
125 Desoto St
Pittsburgh PA 15213
412/647-5090
Active

Wald, Steven L, M.D.
7536 North 10Th Street
Phoenix AZ 85020
Active

Waldman, John B, M.D.
Albany Medical College
Div Of Neurosurgery
Albany NY 12208
Active

Walker, Marion L, M.D.
Primary Childrens
320 12Th Avenue
Salt Lake City UT 84103
801/521-1209
Active

Walsh, John Willson, M.D.
Univ Of Kentucky Med Ctr
Div Of Neurosurgery
Lexington KY 40536
606/233-5974
Active

Waltz, Thomas A, M.D.
Scripps Clinic
10666 N Torrey Pines
La Jolla CA 92037
619/455-9100
Active

Ward, John D, M.D.
1504 Harborough Rd
Richmond VA 23233
804/786-9659
Active

Weiss, Martin H, M.D.
U S C Medical Center
1200 N State St/Box #786
Los Angeles CA 90033
213/226-7421
Active

Welch, W Keasley, M.D.
Childrens Hosp Medical Center
300 Longwood Avenue
Boston MA 02115
617/735-6008
Active

White, Robert Joseph, M.D.
Metropolitan Gen Hosp
3395 Scranton Rd
Cleveland OH 44109
216/459-4383
Active

Winston, Ken Rose, M.D.
300 Longwood Avenue
Boston MA 02115
617/735-6011
Active

Yamada, Shokai, M.D.
Loma Linda U Schl Of Med
Section Of Neurosurgery
Loma Linda CA 92350
714/824-4417
Active