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 Chock
David G. McLone
Michael Scott
Jack Walker

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PAOLO RAIMONDI LECTURERS

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

SHULMAN AWARD

Kim Manwaring 1983
Neonatal Post-hemorrhagic Ventriculomegaly:
Management with Pulsed Lumbar Cisternostomy
Amo 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 Ventriculitics

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 Nuelsen 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 Hydrocephalic Dogs"
William C. Olivero, Harold E.
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. 6. "Shunt Implantation: To Ward
Zero Infection"
Maurice Choux, Gabriel Lena,
Lorenzo Genitori, Etom Empine
Marseille, France

10:45 a.m. 6. "Doppler Cranial Sonography of
Normal and Abnormal Neonates"
Joanna J. Seibert, Charles M.
Gasier, Timothy C. McComb,
William M. Chadduck
Little Rock, AR

11:00 a.m. 7. "Cranial Doppler Ultrasonography
Correlates with Shunting
Criteria and Cerebral Perfusion
Pressure"
William M. Chadduck, 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. Dgnan
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 VentricuPeritoneal 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 LY136032 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

THURSDAY, DECEMBER 10, 1987

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, Ghans 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)

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, Takanishi 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. Luerssen, Melville R. Klauber, Lawrence F. Marshall, Howard M. Eisenberg  
San Diego, CA

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

8:30 a.m.  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. Luerssen

9:15 a.m. R  28. "Severe Head Injury in Children: The Use of 1123 HIPDM SPECT Scan and Arteriojugular Venous O-2 Difference"  
John H. 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. Buchholz, 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

Antonio Discisani, Gregory Stidham, Robert Sanford  
San Francisco, CA

10:30 a.m.  COFFEE BREAK- Foyer (4th Floor)  
View Exhibits
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, Marlon L.
Walker, Bruce B. Storrs, M. Peter
Heilbrun
Salt Lake City, UT

11:30 a.m.  34.  "Temporal Lobectomy for Complex Partial Seizures Beginning in Childhood"
Giuseppe Erba, John R. Adler,
Keesley 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

12:00 noon  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.  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 Cogentital 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 - Moderator: 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

9:15 a.m.  45. "Ganglioglioma: Prognosis and Rationale for Post-Operative Management"  
Edison P. McDaniel II, Philip H. Gogen 
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 Ploidity 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 E. 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 Myelomeningocoele 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 Lipomyelomingocoele and Tethered Cord"  
John D. Brophy, Leslie N. Sutton, Robert D. Zimmerman, Edward Bury, Luis Schut 
Silver Springs, MD
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


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. McDaniel 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 Ploidity 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 Agent for the Treatment of Medulloblastoma: Mutant Diphtheria Toxin Linked to Anti-Transferrin Receptor Monoclonal Antibody or Transferrin" Charles J. Krobel, 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

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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

12:00 p.m.  54. "Detethering - Does It Always Help?"
John R. Hawk, 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"
TakaharuFuse, 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. "Lumbar Peritoneal Shunts for Treatment of Syringomyelia"
T. S. Park, Wayne S. Call, 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. Muhlbaumer
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 Germinoma 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 \( Y = 4.0 * \ln(X) + 1.88 \), \( r = .9299 \). Periventricular edema appeared initially in the superolateral angles of the frontal horns. The extent of the edema was variable. The T1 of 98.61 msec in normal white matter increased to 195.28 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 22%. 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 saline, glaubern 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 \( P < 0.01 \) 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 \( P < 0.01 \) 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 hydrocephalus rabbits. An increase \( P < 0.05 \) in the number of subependymal astrocytes was observed in the CC and CN of 8 week hydrocephalus. Because 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 Stephanian, Martin H. Weiss (Los Angeles, CA)

The activation of membrane-bound adenylate 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 adenylate 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. Chadduck (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 BP×100%) 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 7±13 and for <32 weeks gestation 8±13. The highest RI in clinically abnormal infants were seen in cardiac ischemia 8±13 (N=7), PDA 8±15 (N=72), and infants with bloody CSF 8±13 (N=22). (All p < 0.05 compared to normals.) Sonographic abnormalities associated with increased RI included ICH (Grades 1-4) 8±23 (N=152), hydrocephalus 8±15 (N=28), and brain edema 9±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. Chadduck, 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 cranectomy. 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 386 patients who had complete records. In this cohort of children, 584 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 falx 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 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 subarachnoid space or shunting the cyst contents into 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 Children's 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, milipore 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 were taken at the initial incision were cultured in place of skin culture collection. 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 76 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 VENTRICULOOPERITONEAL 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 hyalome scarring and pseudopod extension. Pseudopod orientation suggested a chemotactic response and focal migration. Adherent macrophages were often associated with fibrin, the strands enfolding and partially obscuring the underlying macrophages. Red blood cells and platelets were seen only occasionally, indicating a predominantly monocyte-mediated 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 choroidplexus 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 BACTRACIN 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. Two sections were handled with sterile gloves and divided into 2-cm portions; each portion of this tubing was then immersed in S.U.B. (% SDS, 9M urea, 2% 2-mercaptoethanol). A second section was handled briefly without gloves and then washed three times with sterile saline and processed in a similar manner, but was immersed in bactracin 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 bactracin 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, bactracin 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 μg (n=6), intraventricular vancomycin 120 μg (n=3), or intraventricular LY146032 7.5 μg (n=6). Ventriculitis was established by inoculation of 10^7/10 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 (PVM) was bioassayed for the antibiotics postmortem.

Results of this study were: 1) S. aureus was vancomycin-resistant (MBC > 64 μg/ml) but LY146032 sensitive (MBC = 2.0 μg/ml); 2) S. aureus demonstrated static growth 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 μg/ml) did not lower S. aureus titers over 8 h (p > 0.1), whereas LY146032 (peaks-30 μg/ml) did (p < 0.01); 4) ventriculitis did not significantly alter clearance of intraventricular vancomycin; T1/2g = 1.7 h (infected) and T1/2g = 1.9 h (uninfected); 5) vancomycin was detectable in the PVM only in the presence of ventriculitis.

We conclude that: 1) drug carry-over artificially lowers the MBC of vancomycin-resistant S. aureus; 2) intraventricular antibiotics do not reduce bacterial counts; 3) intraventricular antibiotics facilitate transventricular drug clearance into the PVM; 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 T6 - 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.
18. CEREBRAL VENOUS MALFORMATIONS IN CHILDREN

Daniele Rigamonti, Harold A. Rekate, Robert F. Spetzler, Karen Hoening-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 doubtfully 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
categorized 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. Díaz,
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 encephalomyosyn-
giosis, 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 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 facilitory 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 electrophysiological 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 electrophysiological 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.

Electrophysiological 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 have electrophysiological evidence of spinal cord dysfunction preoperatively; (4) \( \frac{H_{\max}}{H_{\max}} \) 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 myotatic 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 states 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. Luerssen, 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. Galyn, 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.
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 craniofacial 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.

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) 16 of 16 (100%) 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 1-123 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 1-123 labelled HIPDM. (N.N.-trimethylen-[2-Hydroxy-3-Methyl-5-Jodobenzyl]-1,3-Probenecidamine). In addition arterio-jugular venous oxygen difference (AVDO-2) measurements were correlated with SPECT scan, CT scan, ICP, MAP & PaCO2. 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 6. 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 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. HYPERBARIIC 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 cerebrospinal 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 cerebrospinal 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 study that HBO lowers ICP during treatment of diffuse cerebrospinal 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 cranopharyngiomas, 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 STERIOTACTIC 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 stereotactic 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 2% 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 CHILDMHOOD

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 18.6±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 if 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 aphasia. 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 19F 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 133Xe, but radioactivity limits its use, especially in children. We have developed a method of determining regional CBF non-invasively using clearance of Freon 23 (CHF3), 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 16-minute period of desaturation as the Freon was abruptly discontinued. CBF was calculated for each run by the formula CBF = t1/2 where... is the partition coefficient for Freon (-1.5) and t1/2 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 100cfl/100 gm/min (R=0.78) (see graph [mflow=micrornosphere, flow-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, MAP 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 surgeon's 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 cases 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 in 18/38 patients preoperatively, 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 29/42 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. Follow-up was incomplete in 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 36/39 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 84/2, 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, 2 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 teenage 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.

<table>
<thead>
<tr>
<th>Type of Tumor</th>
<th># of pts</th>
<th>M/F</th>
<th>Micro/</th>
<th>Age range, years</th>
</tr>
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<td>Prolactin</td>
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<td>8/44</td>
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<tr>
<td>ACTH</td>
<td>26</td>
<td>13/13</td>
<td>22/4</td>
<td>7-19</td>
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<tr>
<td>GH (acromegaly)</td>
<td>9</td>
<td>3/6</td>
<td>3/6</td>
<td>15-19</td>
</tr>
<tr>
<td>Others (TSH,-SU, Null)</td>
<td>5</td>
<td>4/1</td>
<td>0/5</td>
<td>16-18</td>
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<tr>
<td>Total</td>
<td>92</td>
<td>28/64</td>
<td>50/42</td>
<td>7-19</td>
</tr>
</tbody>
</table>

Although pituitary tumors in children are uncommon,
when they do occur they are frequently large and
aggressive. The relative incidence of macroadenomas to
microadenomas in children is higher 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 follow up 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 radiosensitive 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 rational 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 torticolis. 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 myelodysplasia 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 modullic 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 transplanum-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. McDaniel, 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 patient 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, 1/2 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 ploidity, whereas the M staging correlated with the ploidity; 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 ploidity. 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 subtotaly 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 alive 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 dipheria 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 dipheria 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 x 10^-11 M and 2.6 x 10^-11 M respectively decreased protein synthesis to 50% of control cultures (IC50). CRM 107 was also covalently linked to human transferrin (Tfn); the IC50 values were 3.9 x 10^-13 M and 2.1 x 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 IC50 of one tumor was 2.5 x 10^-12 M; another tumor was less sensitive (IC50 of 1.1 x 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 IC50’s for medulloblastoma cells. Guinea pigs tolerated a Tfn-CRM 107 CSF concentration of \(1 \times 10^{-3}\) M while \(1 \times 10^{-8}\) M was uniformly fatal. Two Rhesus monkeys tolerated \(3.3 \times 10^{-10}\) M and \(1 \times 10^{-9}\) M initial CSF concentrations of Tfn-CRM 107, and one monkey tolerated \(1 \times 10^{-9}\) 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 coccgeal 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 viewpoints 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. Methylazamide 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 myelomeningoceles 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 duralchisis 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 myelomeningoceles represent a special form of the "split cord syndrome" caused by accessory neurenteric canals.

52. THE COMBINATION OF VATER ASSOCIATION AND SPINAL DYSRAPIRH

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 dysraphism is a known syndrome of the occult spinal congenital disorders, with insidious presentation and progressive neurological deterioration involving lower extremities, bladder, or both. We herein 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. 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 neuroradiology to determine that the patient does not also have the combination of these nonrandom findings with spinal dysraphism. 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. Hawk, 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 mantained 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 syringes associated with tethered cords and occult dysraphism, and 2 patients had post arachnoiditis syringes.

All patients were investigated with pre operative MRI and intraoperative ultrasound. These neuroradiographic 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 syringes 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 syringes.
57. LUMBOPERITONEAL SHUNTS FOR TREATMENT OF SYRINGOMYELIA

T.S. Park, Wayne S. Call, 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 lumbo-peritoneal shunt effects reduction of the syringleal 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 arachnoid 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.

Lumbo-peritoneal shunts were placed in 7 of the 13 patients. Postoperative MR revealed marked and sustained reductions of the syringleal 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 maneuvers was applied to the patients in the sitting position, the intraspinal pressure rose predominantly.

The reduction of the syringleal cavity following placement of lumbo-peritoneal shunts is attributed to decreases in the intraspinal pressure. Our observations support the notion of Ball and Dayan, and Amplekter 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 (12%) 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 excision or tumor resection in all cases with drainage of rostral and caudal cysts when found. All patients received adjuvant 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 within 13 months of 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 ventriculoperitoneal procedures. Eleven of the 19 (59%) developed disseminated disease, most commonly on the basis of subarachnoid spread. Extraneural 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 multimodal chemotherapy and radiation directed at the entire neuraxis.

60. SURGICAL MANAGEMENT OF LAMBDIID 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 synostomectomies 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 16%. 41% 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 noted 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 LAMBDOID (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 lambdoidal sutures. The term "lazy lambdoid" has been coined to denote similar skull base abnormalities in which the lambdoidal 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 lambdoidal suture and underwent surgical resection. Two patients had flattening due to hypertrophy of the inner surface of the lambdoidal 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 lambdoidal 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 SYNOSTOSIS

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) Trigonocephaly with severe bitemporal narrowing is treated by bifrontal craniotomy followed by removal of a supra-orbital bar of bone. The supra-orbital bar is osteotomised 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 intraventricular hemorrhage in both
infants and subsequent hydrocephalus was treated
successfully with lumbar puncture. By 30 days after
birth prominent frontal bossing and ridding 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
calvaraeotomy 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 biconoral 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 cranietomy 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 diversinomy procedure. Her hemiparesis failed to improve and further study demonstrated an unusual lip of bone extending caudal 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 craniovertebral 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.
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

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