Pediatric Section
American Association of Neurological Surgeons

21st Winter Meeting Program

Vancouver, B.C.
December 6 - 9, 1992
Pediatric Section
American Association of Neurological Surgeons

21st Annual Meeting

Westin Bayshore Hotel
Vancouver, B.C.
December 6 - 9, 1992

The Joint Committee on Education of the American Association of Neurological Surgeons (AANS) and the Congress of Neurological Surgeons (CNS) designates this continuing medical education activity for 25.50 neurosurgical credit hours in Category I toward the Continuing Education Award in Neurosurgery and the Physician's Recognition Award of the American Medical Association. The Joint Committee on Education of the AANS/CNS is accredited by the Accreditation Council for Continuing Medical Education to sponsor continuing medical education for Physicians.
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Paolo Raimondi Lecturers

1976—E. Bruce Hendrick 1985—Frank Nelsen
1979—Paul C. Bucy 1986—William F. Meacham
1980—Floyd Giles 1987—Dale Johnson
1982—Panel Discussion 1989—Martin Eichelberger
1983—Derek Harwood-Nash 1990—George R. Leopold
1984—Anthony E. Gallo, Jr. 1991—Judah Folkman
1992—Olof Flodmark

Kenneth Shulman Award Recipients:


1984 — Arno Fried: A Laboratory Model of Shunt Dependent Hydrocephalus

1985 — Anne-Christine Duhaime: The Shaken Baby Syndrome

1986 — Robert E. Breeze: CSF Formation in Acute Ventriculitis

1987 — Marc R. DelBigio: Shunt-induced Reversal of Periventricular Pathology in Experimental Hydrocephalus

1988 — Scott Falci: Rear Seatlap belts. Are They Really “Safe” for Children?

1989 — James M. Herman: Tethered Cord as a Cause of Scoliosis in Children with a Myelomeningocele

1990 — Christopher D. Heffner: Basilar Pons Attracts Its Cortical Innervation by Chemotropic Induction of Collateral Branch Formation

1991 — P. David Adelson: Reorganization of the Cortical-tectal Pathway Following Neonatal Cerebral Hemispherectomy in Cats
Hydrocephalus Association Prize Recipients:

1989 — Eric Altschuler: Management of Persistent Venticulomegaly Due to Altered Brain Compliance
1991 — Nesher G. Asner: Venous Sinus Occlusion and Venticulomegaly in Craniectomized Rabbits

Pediatric Section Chairman:

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

Annual Winter Meeting Sites:

1972 Cincinnati
1973 Columbus
1974 Los Angeles
1975 Philadelphia
1976 Toronto
1977 Cleveland
1978 Philadelphia
1979 New York
1980 New York
1981 Dallas
1982 San Francisco
1983 Toronto
1984 Salt Lake City
1985 Houston
1986 Pittsburgh
1987 Chicago
1988 Scottsdale
1989 Washington, D.C.
1990 San Diego & Pebble Beach
1991 Boston
1992 Vancouver, B.C.
1993 San Antonio, TX

Acknowledgements

The Section on Pediatric Neurological Surgery of the American Association of Neurological Surgeons gratefully recognizes the support of the following exhibitors for the 1992 Pediatric Annual Meeting.

Acra-Cut, Inc. - Acton, MA
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Howmedica - Rutherford, NJ
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Journal of Pediatric Neurosurgery - New York, NY
Laserscope - San Jose, CA
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Redmond Neurotechnologies Corporation - Lake Zurich, IL
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Steritek, Inc. - Paterson, NJ
Valleylab, Inc. - Boulder, CO
Wild Microscopes, Division of Leica, Inc. - Rockleigh, NJ
Officers of the Pediatric Section of the American Association of Neurological Surgeons

Chairman: R. Michael Scott (term expires April 1993)

Secretary-Treasurer: Harold L. Rekate (1994)

Executive Council: Donald H. Reigel (immediate past president, 1993)
William Chadduck (1993)
Tae Sung Park (1993)
Dennis Johnson (1994)
Gordon McComb (1994)

Membership Committee: Walker

Rules and Regulations: Leslie Sutton (Chairman) (1994)

Ad hoc Committees:
Committee on Utilization of Treasury Surplus
Ken Winston, Chairman
William Bell
Bruce Storrs
Timothy Mapstone

Committee for Continuing Education (SANS V):
Joseph Platt
Corey Raffel
Michael Turner
John Waldman
Ken Winston

Program of the Pediatric Section 21st Winter Meeting

SUNDAY, DECEMBER 6, 1992
4:00 pm - 8:30 pm
Registration - Main Foyer
6:30 pm - 8:30 pm
Welcoming Cocktail Reception, Garden Lounge

MONDAY, DECEMBER 7, 1992
7:00 am - 7:45 am
Continental Breakfast and Exhibits - Stanley/Park
7:45 am - 8:00 am
Welcome and Introductory Remarks — Paul Steinbok, Local Meeting Chairman, R. Michael Scott, Section and Program Committee Chairman

SCIENTIFIC SESSION I — Fraser/Mackenzie
SPINAL DYSPHURISM:
Moderators: Donald Reigel, Shokei Yamada

8:00 am - 8:15 am

8:15 am - 8:30 am
2. "Radiation Exposure in the Myelomeningocele Population", S.J. Gaskill, Durham, NC

*Indicates Resident Paper

8:45 am - 9:00 am


SCIENTIFIC SESSION II — Fraser/Mackenzie

SPINAL DYSRAPHISM (continued)

Moderators: David McLone, Ben Warf

9:00 am - 9:15 am


9:15 am - 9:30 am


9:30 am - 9:45 am


9:45 am - 10:00 am

8. "Outcome After Operative Intervention in Patients with Meningocele Manque", D. Warder, W.J. Oakes, Durham, NC

10:00 am - 10:30 am

COFFEE BREAK & EXHIBITS — Stanley Park

EVALUATION FORM

SCIENTIFIC SESSION III — Fraser/Mackenzie

BRAIN TUMORS

Moderators: Tadano Tomita, Leland Albright

10:30 am - 10:45 am


10:45 am - 11:00 am


11:00 am - 11:15 am

11. "Expression of Insulin-Like Growth Factors in Medulloblastoma", L.S. Chin, C. Raffel, Los Angeles, CA

11:15 am - 11:30 am

12. "Efficacy of Transferrin Receptor-Targeted Immunotoxins in Brain Tumor Cell Lines and Pediatric Brain Tumors", K.M. Muraszko, L. Martell, A. Agrawal, D. Ross, Ann Arbor, MI

SCIENTIFIC SESSION IV — Fraser/Mackenzie

BRAIN TUMORS (continued)

Moderators: Robert A. Sanford, Doug Cochrane

11:30 am - 11:45 am


11:45 am - 12:00 pm

14. "Use of MoAb to hTGF-alpha as a Immunocojugate for Tumor Targeting", A.P. Bowles, Jr., Augusta, GA

12:00 pm - 12:15 pm


12:15 pm - 12:30 pm

16. "Pediatric Oligodendroglioma: Natural History, Treatment and Outcome. Experience at Boston Children's Hospital", L.C. Goumnerova, P. Barnes, R.M. Scott, Boston, MA

12:30 pm - 1:30 pm

LUNCH AND EXHIBITS — Stanley/Park

SCIENTIFIC SESSION V — Fraser/Mackenzie

BRAIN TUMORS (continued)

Moderators: Enrique Ventureyra, Jack Walker

1:30 pm - 1:45 pm

17. "Childhood Brain Tumors Presenting with Seizures: Results of Tumor Excision", R. Munn, D. Cochrane, P. Steinbok, K. Farrell, Vancouver, BC

1:45 pm - 2:00 pm

18. "Prospective Endocrinologic Function in Children with Brain Tumors", P.M. Kanev, K.S. Bierbrauer, P.M. Thomas, C. Gtech, Philadelphia, PA
2:00 pm - 2:15 pm

2:15 pm - 2:30 pm

2:30 pm - 2:45 pm

2:45 pm - 3:00 pm

3:00 pm - 3:30 pm
COFFEE BREAK AND EXHIBITS — Stanley/Park

SCIENTIFIC SESSION VI — Fraser/Mackenzie
THE CHIARI MALFORMATIONS
Moderators: Jerry Oakes, Timothy Mapstone

3:30 pm - 3:45 pm
23. "The Experimental Model: Chiari II-Like Change in Surgically Induced Spina Bifida Aperta in Chick", T. Inagaki, D.G. McLone, Y. Yamanouchi, Salt Lake City, UT

3:45 pm - 4:00 pm

4:00 pm - 4:15 pm

4:15 pm - 4:30 pm
26. "Symptomatic Chiari I Malformations: Clinical Features, Surgical Management, and Outcome", M. Herman, D.G. McLone, M.S. Diaz, Chicago, IL

4:30 pm - 4:45 pm

4:45 pm - 5:00 pm

5:00 pm - 5:30 pm
BUSINESS MEETING FOR MEMBERS OF THE PEDIATRIC SECTION

TUESDAY, DECEMBER 8, 1992

7:00 am - 8:00 am
CONTINENTAL BREAKFAST AND EXHIBITS — Stanley/Park

7:30 am - 8:00 am
Update on Status of National Tumor Protocols
CCG: Leland Albright
POG: Robert A. Sanford

SCIENTIFIC SESSION VII — Fraser/Mackenzie
VASCULAR DISORDERS
Moderators: Robin Humphreyes, Arthur Marlin

8:00 am - 8:15 am

8:15 am - 9:30 am
30. "Diagnosis and Treatment of Dural Sinus Thrombosis (DST) with Thrombolysis Therapy", K. Zakalik, A.M. Wand, M.G. Goetting, Royal Oak, MI

8:30 am - 8:45 am

9:00 am - 9:30 am

Paolo Raimondi Lecture. Olof Flodmark, MD, Ph.D., FRCP, Stockholm, Sweden: "Intracranial Hemorrhage in the Newborn." To be introduced by Paul Steinbock, Vancouver, BC

9:30 am - 10:00 am

Special Lecture: Dr. Ken Poskitt and Dr. Margaret Norman, Vancouver, BC: Cerebral Dysplasias and Hamartomas.

10:00 am - 10:30 am

COFFEE BREAK - VIEW EXHIBITS — Stanley/Park

SCIENTIFIC SESSION VIII — Fraser/Mackenzie

LABORATORY RESEARCH

Moderators: Parker Mickle, Charles Duncan

10:30 am - 10:45 am


10:45 am - 11:00 am


11:00 am - 11:15 am


11:15 am - 11:30 am


SCIENTIFIC SESSION IX — Fraser/Mackenzie

SPASTICITY

Moderators: Bruce Storr, Joseph Madsen

11:30 am - 11:45 am


11:45 am - 12:00 pm


12:00 pm - 12:15 pm


12:15 pm - 12:30 pm

40. "Effects of Selective Dorsal Rhizotomy for Spastic Diplegia on Hip Migration in Cerebral Palsy", T.S. Park, G.P. Vogler, B.A. Kaufman, M. Ortman, St. Louis, MO

Afternoon Free

6:30 pm - 7:30 pm

RECEPTION — Fraser/Mackenzie

7:30 pm - 10:00 pm

ANNUAL BANQUET — Fraser/Mackenzie

WEDNESDAY, DECEMBER 9, 1992

7:15 am - 8:00 am

SCIENTIFIC SESSION X — Fraser/Mackenzie

TRAUMA

Moderators: Thomas Luerssen, Anne-Christine Duhaime

8:00 am - 8:15 pm

*41. "Child Abuse Head Injury Study", J.K. Levitt, H. Bartowski, Columbus, OH

8:15 am - 8:30 am

10:45 am - 11:00 am
50. "Late Decompression Does Not Restore Protein Synthetic Capacity in Neonatal Hydrocephalic Kittens", J.P. McAllister II, C.L. Wolfgang, K. Bierbrauer, Philadelphia, PA

11:00 am - 11:15 am

11:15 am - 11:30 am
52. "MR Visualization of the Olfactory Pathway of Cerebral Spinal Fluid Flow in the Dog", M.J. Burke, D. Symonds, Denver, CO

11:30 am - 11:45 am
53. "Current Applications of Endoscopy to Ventricular Shunt", K.R. Crane, Cincinnati, OH

11:45 am - 12:00 pm

12:00 pm - 12:15 pm
55. "The Diagnosis and Treatment of Trapped Fourth Ventricle", J.R.B. Nashold, W.J. Oakes, Durham, NC

12:15 pm - 12:30 pm
56. "Initial Management of Congenital and Acquired Hydrocephalus with a High Pressure Valve", J. Holsapple, L. Hochhauser, J.A. Winfield, Syracuse, NY

12:30 pm - 1:30 pm
LUNCH AND EXHIBITS — Stanley/Park

SCIENTIFIC SESSION XIV — Fraser/Mackenzie
HYDROCEPHALUS (continued)
Moderators: Walker Robinson, William Chaddock

1:30 pm - 1:45 pm
1:45 pm - 2:00 pm

2:00 pm - 2:15 pm

2:15 pm - 2:30 pm
60. "Significance of Broth Only or Anaerobic Positive Ventricular CSF Cultures", J. Chen, J.G. McComb, C. Raffel, Los Angeles, CA

2:30 pm - 2:45 pm

2:45 pm - 3:00 pm

3:00 pm - 3:30 pm
COFFEE BREAK AND EXHIBITS — Stanley/Park

SCIENTIFIC SESSION XV — Fraser/Mackenzie
SEIZURE DISORDERS
Moderator: Glenn Morrison

3:30 pm - 3:45 pm

3:45 pm - 4:00 pm

SCIENTIFIC SESSION XVI — Fraser/Mackenzie
THE SPINAL CORD
Moderators: John Laurent, Andrew Parent

4:00 pm - 4:15 pm
65. "Cervicomedullary Compression in Achondroplasia", T. Ryken, A.H. Menezes, Iowa City, IA

4:15 pm - 4:30 pm

4:30 pm - 4:45 pm
67. "Treatment of Pediatric Spinal Aneurysmal Bone Cysts with Selective Arterial Embolization", J.D. Callahan, J.E. Kalsbeck, T.G. Luersen, J.C. Boaz, G.P. DeRosa, Indianapolis, IN

4:45 pm - 5:00 pm

Closing remarks and adjournment — Paul Steinbok, Michael Scott
1. DISBANDING A MULTIDISCIPLINARY CLINIC — EFFECTS ON HEALTH CARE

Bruce A. Kaufman, MD, Amy Terbrock, RN, Arlene Klosterman, RN, Nan Winters, MEd, T.S. Park, MD (St. Louis, MO)

The multidisciplinary clinic is an accepted method of health care delivery for children with spinal dysraphism. Such a clinic was established at our institution in 1963, but disbanded in 1988. Each service was still available to the patients individually. Myelomeningocele patients in this clinic (n=60) were surveyed by telephone to determine the effects of clinic closing on their health and use of health care. During the four years after the clinic closed, 53% had no neurosurgical care, 47% no orthopedic care, 65% no physical therapy, and 28% had no urologic care. General pediatric care was absent in 30% and episodic in 30%. Patients with private insurance (versus government programs) were twice as likely to have an identified pediatrician and regular pediatric visits. Insurance had no effect on whether the patients had neurosurgical, orthopedic, or urological caregivers, but those on government programs had infrequent neurosurgical or orthopedic visits. Forty-six percent had undergone operations (67% neurosurgical, 11% orthopedic, 11% urologic, 11% other). Posterior fossa decompression and untethering were performed only on patients who had regular follow-up by all specialties. The patients with perhaps preventable operations (foot amputation, nephrectomy, osteomyelitis, decubitus repair) had received only urologic or no specialty care.

Most myelomeningocele patients will fail to obtain appropriate specialty care when a multidisciplinary clinic disbands, despite continued availability of these services individually. Private insurance does not affect access, but does affect the frequency of specialty care use. The lack of specialty follow-up may prevent the identification of conditions that are potentially treatable.
2. RADIATION EXPOSURE IN THE MYELOMENINGOCELE POPULATION

Sarah J. Gaskill, MD (Durham, NC)

Radiation induced carcinoma is now recognized to occur with lower doses of exposure than previously thought. The myelomeningocele population, because of their complex medical problems, is exposed to relatively high doses of diagnostic x-ray examination. Presented is the calculated radiographic exposure for patients in a myelomeningocele clinic. Of the 243 patients in the clinic, 112 patients had been in the clinic from birth. These patients comprise the study group. For each patient, the radiographic history was reviewed, and calculations of radiation exposure were made.

The total skin dose ranged from 3.96 to 171.29 rads with a mean of 42.99 (SD 33.58). The total skin dose per year of life ranged from .79 to 38.61 rads/year of life with a mean of 6.38 rads/year of life (SD 5.72). The total red marrow dose ranged from .3 to 29.04 rads with a mean of 7.02 rads/year (SD 6.24). The total red marrow dose per year of life ranged from .1 to 5.35 rads/year with a mean of 1.01 rads/year (SD 1.06). Neurosurgical examinations contributed between 0 and 92.95 total skin dose rads with a mean of 21.65 (SD 17.34) and 0 to 9.29 total red marrow rads with a mean of 1.92 (SD 1.89). Because of lost x-rays, studies done outside the clinic, repeat x-rays, fluoroscopic examinations, and examinations without a known exposure value, these numbers are clearly an underestimate.

These data suggest that radiation exposure within the myelomeningocele population is within the range to raise concern for carcinogenesis. Recommendations and guidelines for limitations of radiographic examination, and the implementation of cancer screening in this population are discussed.

3. LONG TERM OUTCOME IN HYDROCEPHALIC MYELOMENINGOCELE PATIENTS

John R.W. Kestle, MD, FRCS (Vancouver, B.C.), Harold J. Hoffman, MD, FRCS, R.P. Humphreys, MD, FRCS, E. Bruce Hendrick, MD, FRCS (Toronto, Ontario)

In order to assess the long term outcome of patients who have had a myelomenigocele closed and a CSF shunt inserted at The Hospital for Sick Children a retrospective review of such patients born between January 1, 1970 and December 31, 1975, has been performed.

There were 89 children with a median followup of 16.3 years. Six patients were lost to follow-up and 18 have died leaving 65 available for longterm follow-up. Fifty-six of the 65 survivors are independent (26/65) or are living at home, partially dependent on others in their activities of daily living (30/65). Thirty-four (of 65) attended school at a level appropriate for their age (31) or advanced (3). Forty-nine (of 65) patients enrolled in the regular school system. Six patients are in the work force. Thirty-three (of 65) patients walk with assistive devices (29) or independently (4). Neuropsychological test results were available for 26 patients. Mean scores were: verbal 86, performance 77, full scale 80.

Lobter's selection criteria were retrospectively applied and the results for the patients who would not have been treated were compared with the results for those who would have been treated if those criteria had been used. The two groups differed significantly in their scores on independent living (p = 0.0004), ambulatory status (p = 0.001), mean IQ (p = 0.01) and school level achieved (p = 0.03) but there were notable individual exceptions. Some patients who would not have been treated did very well and vice versa. As a result we continue to offer treatment to all patients with this disorder.
4. NEONATAL KYPHECTOMY: ITS ROLE IN MYELOMENINGOCELE
MANAGEMENT

Thomas S. Berger, MD, Alvin Crawford, MD, Kerry R. Crone, MD
(Cincinnati, OH)

It is estimated that 10-15% of children born with myelomeningocele will have a significant kyphotic deformity. The typical lesion is in the thoracolumbar area with the apex of the kyphosis being at lumbar two or three. This deformity makes it difficult to obtain an initial skin closure and allows early breakdown of the subsequently closed skin because of the bony prominence. Both problems may lead to meningitis with its attendant complications of deterioration of neurologic function and mental retardation. If the kyphosis is left uncorrected there is a steady increase in the angle of deformity which increases after the first year when the child starts to sit frequently leading to an operative procedure by age three. The uncorrected deformity makes it difficult to fit appliances for standing or to be supine.

Sharrod introduced neonatal kyphectomy in 1968. The procedure did not become popular because of complications, particularly blood loss and a high rate of failure of fusion. With improved technique, both surgical and anesthetic, we feel this operation can be performed safely and successfully.

We have performed this procedure seven times with a neurosurgical-orthopedic team using a modification of a technique described by Reigel. The follow-up is six months to 9.5 years (average 4.4 years). There have been no deaths or significant complications. None of the children have progressed to a severe enough kyphoscoliosis to require a second procedure.

In conclusion, we feel that newborns with a myelomeningocele associated with a significant kyphotic deformity should be considered for a kyphectomy at the time of initial repair.

EXPERIENCE

Charles F. Harvey, Mark S. Dias, David G. McLone (Chicago, IL)

Two hundred forty-four consecutive operations for the release of congenitally tethered spinal cords were performed on children with spinal lipomas over the twenty year period from 1971-1991. A retrospective analysis demonstrates that patients with these lesions suffer an insidious decline in neurologic function from an early age due to spinal cord tethering. Operative release of the tethered cord can be performed safely, with minimal risk of neurologic worsening. In this series, only one patient (0.4%) was permanently worsened by operation, and no patient who was neurologically normal preoperatively was rendered worse by surgery.

Retethering occurred in 36 patients (18.2%). Factors associated with a lower incidence of retethering included operation after 1980, and terminal attachment of a combined intra and extradural lipoma to the conus medullaris below the last intact nerve root (the "terminal" lipoma). Most importantly, infants less than one year and neurologically intact patients had a significantly lower incidence of retethering. Moreover, these children required fewer orthopedic and urological procedures during the follow-up period than did older children and those with preoperative neurologic deficits.

Follow-up in this series averaged 6.3 years. At last evaluation, 92% of patients have either stabilized or improved their neurologic function; 95% of patients are ambulatory and 65% are continent. Our data strongly support an early and aggressive approach to the management of children with spinal lipomas. We recommend an early evaluation with spinal magnetic resonance imaging (MRI) for all patients with suspected spinal lipomas, and untethering of all lesions regardless of neurologic status. Untethering should be performed as soon as possible, preferably before one year of age, and certainly before neurologic deterioration ensues.
6. THE VALUE OF INTRAOPERATIVE URODYNAMIC MONITORING FOR THE RELEASE OF THE TETHERED SPINAL CORD

Kenneth W. Reichert, Enrique C.G. Ventureyra, Michael J. Higgins, Ginette Potvin (Ottawa, Ontario)

The tethered spinal cord is an entity with potentially grave neurological implications. Presenting symptoms include progressive spinal deformity, back and leg pain, motor and sensory deficits of the lower and upper limbs, and increasing urinary and bowel incontinence. The value of intraoperative urodynamic monitoring in assisting the neurosurgeon to untether a spinal cord and preserve urologic function is presently under debate. We have used intraoperative urodynamic monitoring for tethered chord release since January of 1981. The urologic history was correlated with preoperative and postoperative urodynamics on 94 consecutive myelodysplastic children. A total of 104 intraoperative urodynamic monitoring procedures were performed on these children. This report does not include 39 operations which were done for the initial closure of open neural tube defects at birth. Three additional children in whom urodynamic monitoring was performed, have not reached training age and three (2.8%) were lost to follow-up. These are excluded from this report. In this study, we noted that clinical symptomatology and urodynamic parameters improved in 88 (87.1%) of those children who successfully underwent spinal cord untethering. Thirty-three (32.7%) who were incontinent preoperatively have a normal voiding pattern postoperatively. A total of 54 (53.5%) are able to void without catheterization. One (1.0%) showed deterioration of urodynamic parameters. In 2 (2%) children urodynamic parameters reverted a complete untethering, surprisingly without a further decrease in their urologic function so far. We feel this data supports the benefit of intraoperative urodynamic monitoring for the release of tethered spinal cords.

7. TERMINAL SYRINGOHYDROMYELIA

Bermans Iskandar, MD, W. Jerry Oakes, MD, Colleen McLaughlin, RN, BSN, Alan K. Osumi, MD, Robert D. Tien, MD, MPH (Durham, NC)

Syringohydromyelia, or cystic dilatation of the spinal cord may be caused by trauma, arachnoiditis, and neoplasms, or it may be associated with a hindbrain hernia (Chiari malformation). Terminal syringohydromyelia, which refers to a syrinx located only in the lower third of the spinal cord, is frequently associated with other congenital spinal anomalies. The association of a syrinx with other forms of occult spinal dysraphism (OSD) has gained special significance since the advent of Magnetic Resonance Imaging (MRI).

In this study, we have reviewed all OSD cases (143) seen at Duke University Medical Center between 1972 and 1992. A terminal syrinx was found in 24 of the 90 cases which were evaluated by MRI scans (27%). In contrast, 3 out of 48 (6.2%) cases, evaluated by myelograms and postmyelographic CT scans had syrinx documented.

The relative radiographic severity of the different syringes was estimated using measurements of the syrinx and spinal cord on the MRI scans. Common presenting symptoms and signs, which were thought to be due to the syrinx, included pain, motor and sensory deficits of the lower extremities, scoliosis and rarely bowel or bladder dysfunction. The change in symptoms following surgery will be discussed. The association of terminal syringohydromyelia with other manifestations OSD defects will be described, demonstrating a strong association with anorectal anomalies, tethered spinal cord, meningocoele manque, and diastematomyelia. An infrequent association was seen with other spinal anomalies.

Finally, results of the surgical management of syringohydromyelia in our patient population were analyzed, highlighting the necessity of shunting the large syringes, in addition to repairing the OSD defects.
8. OUTCOME AFTER OPERATIVE INTERVENTION IN PATIENTS WITH MENINGOCELE MANQUE

Daryl Warder, MD, W. Jerry Oakes, MD (Durham, NC)

The purpose of this review is to describe the neurologic presentation and follow up of patients with meningocele manque, and to describe the associated dysraphic features. We have reviewed our series of nineteen patients with meningocele manque collected over a fifteen year period. We noted the frequency at presentation of neurologic symptoms and compared these symptoms with those at follow up to assess neurologic outcome. We also noted other features of occult spinal dysraphism including, diastematomyelia, tethered cord syndrome from a thickened filum terminale intraspinal lipomas, meningocele, myelomeningocele presence of a median septum associated with diastematomyelia, presence of cutaneous abnormalities, syringohydromelia and vertebral anomalies. 72% of the patients presented with abnormal neurologic examination; the most common symptoms were: 1) lower extremity weakness; 2) radicular leg pain; 3) spasticity of the lower extremities and 4) urinary incontinence and infection.

At the time of follow up after surgery, 37% had no change in their symptoms; 37% had improvement and 26% had progression of their symptoms. The mean period of follow up was 2.9 years. 83% of the patients had the tethered cord syndrome from a thickened filum terminale; 76% had diastematomyelia; 78% had cutaneous stigmata of occult spinal dysraphism, 72% had major vertebral anomalies; 56% had a median septum and 33% had syringohydromelia.

We have reviewed our series of nineteen patients with meningocele manque. We examined their neurologic symptoms at presentation and follow up after division of the dorsal tethering bands. We also identify associated dysraphic features in these patients. The five patients whose neurologic status appeared to worsen despite operative intervention will be discussed.

9. MULTIDRUG RESISTANCE GENE EXPRESSION IN PEDIATRIC BRAIN TUMOR

Frederick A. Boop, William M. Chadduck, MD, Gary Roloson, MD, Jeff Sawyer, PhD (Little Rock, AR)

The multi-drug resistance gene (MDR-1) resides on human chromosome 7 and codes for a 170-Kd membrane glycoprotein (pgy). Pgy functions as a selective efflux pump which protects cells from damage by certain toxins. This phosphorylating glycoprotein has also been shown to be inducible in tumors, and its amplification is a likely mechanism by which neoplasms develop resistance to adjuvant chemotherapy. This study was undertaken to demonstrate the presence of the MDR-1 gene product in pediatric brain tumor as well as in primary pediatric brain tumors. Thirty-three specimens, (four brain biopsies with normal histopathology, five temporal lobe specimens removed for intractable epilepsy, five cerebellar astrocytomas, five gangliogliomas, six primitive neuroectodermal tumors and eight malignant astrocytomas) underwent immunohistochemical staining for presence of pgy (Biogenex, California). All normal tissues and benign neoplasms were negative to trace positive as compared to controls. Four of five medulloblastomas had minimal expression, whereas one was positive. All malignant astrocytomas stained positively. Interestingly, the reactive glia surrounding rapidly growing neoplasms also stained positively. Since experimental evidence suggests pgy can be phenotypically reversed by the addition of calcium entry blockers, we have now begun a randomized prospective study of children with malignant brain tumors adding Verapamil to their adjuvant chemotherapy regimen, should their tumors show positivity for MDR-1 initially or upon re-sampling following failure to respond to adjuvant therapy. An update of our findings will be presented.
DNA content or ploidy was measured by flow cytometry in fresh tumor specimens obtained from 67 children with newly diagnosed supratentorial and infratentorial glial neoplasms. Each patient had a complete evaluation with post operative staging to ascertain extent of surgical resection and metastatic disease. Other parameters evaluated included histology, age at diagnosis, sex, treatment, disease control interval and overall survival length. The DNA content was correlated with the various factors, particularly survival and disease-free interval, in the hopes of finding an independent prognostic factor for this group of brain tumor patients.

Our study group included 33 low-grade astrocytomas, 14 glioblastoma multiformes, 11 anaplastic astrocytomas, 5 oligodendrogliomas, and 2 giant cell astrocytomas of tuberous sclerosis and 2 xanthoastrocytomas. In the low-grade astrocytomas, 29 of 33 tumors had a DNA index (DI) of 1.00 (diploid), and all 33 are alive today. Two patients had disease progression, 1 with aneuploid tumor and the other diploid. Ten of 14 patients with glioblastoma multiforme had hypodiploid tumors (DI greater than 1.00), and 11 of 14 patients are dead from disease. Five of 11 patients with anaplastic astrocytoma had hypodiploid tumors, and only 3 of 11 are alive today. The remaining tumors had variable DNA indices and survival rates, but the numbers are too low for correlation.

In conclusion, tumor DNA content does not have prognostic significance beyond the association between DNA content and histology. Tumor histology remains the best predictive factor for survival. Other parameters, specifically the relationship between disease control interval and DNA content, will be discussed in detail.

**Expression of Insulin-like Growth Factors in Medulloblastoma**

Lawrence S. Chin, MD, Corey Raffel, MD, PhD (Los Angeles, CA)

Insulin-like growth factors (IGF) (somatomedins) are polypeptide hormones related to proinsulin in structure. These growth factors stimulate proliferation of cells from many tissues in vitro. The presence of IGF-I, IGF-II, and IGF-I receptor in a variety of tumors suggest that they may be involved in an autocrine stimulation loop. Their presence in the adult and fetal central nervous system (CNS) implicates a role in the development of the CNS. Indeed, expression of IGF-I has been reported in astrocytomas. We examined expression of IGF-I in 13 medulloblastoma specimens obtained at the time of surgery. Total RNA was extracted from each specimen and RNA blotting with specific probes for IGF-I, IGF-II, and IGF-I receptor was performed. Expression of IGF-II was seen in 7 out of 13 tumors. Transcripts of 5.0 kb and 3.8 kb were identified in each instance. No expression of IGF-I nor IGF-I receptor was seen. This suggests that IGF-II may be involved in an autocrine mechanism of tumor growth in PNET's. Confirmation of this awaits analysis of IGF-I receptor expression.
12. Efficacy of Transferrin Receptor-Targeted Immunotoxins in Brain Tumor Cell Lines and Pediatric Brain Tumors

Karin M. Muraszko, MD, Lori Martell, PhD, Alka Agrawal, Donald Ross, MD (Ann Arbor, MI)

The efficacy and cytotoxic properties of immunotoxin conjugates directed toward the transferrin receptor were examined in cell lines and pediatric brain tumor specimens. Dose-response relationships were assessed for immunotoxin-mediated inhibition of protein synthesis for two immunotoxins, 454A12-IRA (anti-transferrin receptor antibody [454A12] conjugated to a recombinant ricin A chain toxin [IRA]) and anti-thnR-CRM 107 (antitransferrin receptor antibody [anti-thnR] conjugated to a diphtheria toxin mutant [CRM 107]). Protein synthesis inhibition assays were used to test the efficacy of these immunotoxins. Three target medulloblastoma cell lines (DAOY, D283 MED, D341 MED), a glioblastoma cell line (U373) and two neuroblastoma cell lines (SKNSH, SHSY5Y) exhibited similar sensitivity to both immunotoxins with IC50's in the 10-9M to 10-10M range. The time course of protein synthesis inhibition by the immunotoxins in DAOY cells showed that inhibition by anti-thnR-CRM 107 was rapid and apparent by 6 hours of incubation. In contrast a response to 454A12-IRA was not observed until 16 hours. Cell viability was decreased 30-40% by 24 hours after removing 454A12-IRA (1 x 10-9M) and was maximally decreased 70-80% after 3 days.

The efficacy of these immunotoxins in a variety of pediatric brain tumor specimens (N=20) was also examined. The more aggressive and malignant tumor types such as glioblastoma multiforme, ependymoma and medulloblastoma had low IC50 values (10-11M to 10-12M) when tested with each of the immunotoxins. The rate of protein synthesis in slow growing and more benign tumors (pilocytic astrocytoma and craniopharyngioma) was not affected by immunotoxins until very high levels of immunotoxin was applied (10-7M to 10-8M). Immunoblots showed expression of transferrin receptors on cell lines and tumors which correlated with in vitro sensitivity to immunotoxin.

Previous studies have shown that the toxic in vivo concentration of immunotoxin in the CSF is 10-9M to 10-4M. This has been confirmed in several species. Using infant rats (16-20 days old) and slightly older rats (21-27 days old) we have shown that there is a higher tolerance to immunotoxin within the younger brain. Animals require a higher amount of immunotoxin (5 x 105M versus 1 x 105M) to display toxicity. Animal tumor models using medulloblastoma cell lines implanted in nude rats have documented efficacy of immunotoxin therapy at concentrations of 10-9M to 10-6M.

The data presented suggests that immunotoxin therapy is a viable therapy option for children with CNS tumors. There is a window of therapy in which efficacy can be achieved and toxicity avoided.

13. A Human Medulloblastoma Xenograft in a Rodent Model of Leptomeningeal Neoplasia

Dennis Wen, Walter Hall, Oystein Fodstad (Minneapolis, MN)

Prognosis for patients with medulloblastoma remains poor despite recent advances in chemotherapy and radiation therapy which is often contraindicated in young children. New treatment modalities are clearly indicated and the use of targeted-toxin therapies (immunotoxins) may prove beneficial. Immunotoxins have a mechanism of action different from that of radiation therapy and chemotherapy. The compartmentalized nature of the subarachnoid space and the predilection for medulloblastoma to disseminate throughout the CSF makes immunotoxins an appropriate mode of therapy. We developed a rodent model of human leptomeningeal neoplasia with medulloblastoma for testing immunotoxins.

Nude rats (nu/nu) weighing 200-220 gm were anesthetized with ketamine/xylazine and an intrathecal catheter was placed into the subarachnoid space of the spinal canal via the cisterna magna. Forty-eight hours after catheter placement, those animals which remained neurologically intact were inoculated with a single cell suspension of a human medulloblastoma-derived cell line (DAOY). Animals were examined daily for the development of neurologic injury such as paraplegia.

Animals received 5 x 105 (n=5) to 1 x 106 (n=5) viable DAOY cells in 50 µl. Paraplegia developed at 31 and 34 days in 2 animals (40%) given 5 x 105 cells. All 5 animals that received 1 x 106 DAOY cells became paraplegic between 25-55 days, mean 34 days, mean 40 ± 14 (S.D.) days. Necropsy demonstrated focal deposits of tumor along the spinal neuraxis. This reproducible model of medulloblastoma provides a means of testing immunotoxins and other intrathecal therapies for potential therapeutic effect. Experiments to test the efficacy of a transferrin-Pseudomonas exotoxin immunotoxin in this model are currently in progress.
14. USE OF MoAb-rhTGF-alpha AS AN IMMUNOCONJUGATE FOR TUMOR TARGETING

Alfred P. Bowles, Jr., MD (Augusta, GA)

A recent consideration for the treatment of brain tumors has been the use of antitumor antibodies as carriers of drugs and toxins, however therapy has been inadequate because of the inaccessibility of distinctive tumor antigens in the CNS. TGF-alpha has been distinctly identified in a number of brain tumors, as defined in our laboratory and others, especially of a more embryonic origin, and therefore may yield a more favorable malignant determinant.

In previous and separate studies, we have demonstrated daunomycin induced cytolysis activity and also antiproliferative enhancement with MoAb-TGF-alpha in a number of glioma cell lines. We therefore conjugated daunomycin to MoAb to TGF-alpha (MoAb-daunomycin) by reacting peroxidase-treated daunomycin with antibody for form Schiff bases, followed by sodium borohydride reduction. In vitro conjugates were assessed, and the therapeutic efficacy was determined from an xenogenic model. Two established cell lines were used in this study. Both U 87 (glioblastoma multiforme) and TE 671 produce significant quantities TGF-alpha as defined by Western and Northern blot analysis.

The in vitro cytolytic activity of MoAb-daunomycin was determined from the incubation of tumor cells with conjugates and control. Cells were labeled with propidium iodide and fluorescence assessed by flow cytometry. Exposed DNA may bind to propidium iodide, with the fluorescence emitted representing the percentage of cell death. The relative fluorescence of albumen conjugated to daunomycin and free daunomycin was not significantly different from control. However, significant cytolysis was achieved with MoAb-daunomycin with 48% cell death (p<0.001).

Age matched athymic homozygous (nu/nu) mice, with subcutaneous tumors of either U 87 or TE 671, were treated intratumorally with drug-antibody conjugates and control. The relative tumor weights were determined and variability standardized. MoAb-daunomycin was effective in limiting tumor growth (p<0.001), with regression in tumor size.

Although some success has been achieved with drug antibody conjugates in tumor bearing animals, therapy with antibody conjugates in malignant gliomas has proven unsuccessful predominately because of the inaccessibility of distinctive tumor antigens. Although previous studies have suggested a favorable correlation of malignant determination in gliomas with TGF-alpha, the current study more readily defines the importance of TGF-alpha in glioma cell growth, with effective treatment demonstrated with drug conjugates to MoAb-TGF-alpha.

15. MULTICENTRIC JUVENILE PILOCYTIC ASTROCYTOMAS

Adam N. Mamelak, MD, Michael Prados, MD, Phillip Cogen, MD, PhD, Samuel F. Citello, MD, Michael S.B. Edwards, MD (San Francisco, CA)

Juvenile Pilocytic Astrocytoma (JPA) is a low grade glioma that is felt to carry an excellent prognosis for long-term survival or cure, even in cases of subtotal resection. Although JPAs rarely disseminate, multicentric and metastatic cases have been described, but risk factors for the development of multicentric JPA have not been identified, and treatment course, prognosis, and longterm survival are not known. Between 1987 and 1991 we treated 44 patients with a diagnosis of JPA following subtotal resection or biopsy, 8 of which (18%) have developed multicentric disease. There were 6 children and 2 adults. Age at initial diagnosis of JPA ranged from 5 months to 43 years. All the children were less than 4 years at presentation. All patients had primary tumors located in the hypothalamic-suprasellar region, with a volume of 9 cm³ or more. Histological descriptions were typical of JPA in all cases, and mitoses were absent in all cases. Bromodeoxyuridine (BrdU) labeling was performed in one patient, with a labelling index of 1.1%. Four patients were treated with chemotherapy and 2 underwent focal radiation therapy. Multicentric disease was noted in 7 of 8 patients within 1.5 years of initial diagnosis, but only 1 patient was symptomatic. Extensive ependymal spread was noted in the two adult patients, while in the children metastatic lesions to the posterior fossa (n=2), hemispheres (n=2), and spine (n=4) were noted, with leptomeningeal involvement noted in every case. CSF cytology was positive in 50% of patients tested (n=4). Histological confirmation of multicentric disease was performed in 4 cases, and all were consistent with "benign" JPA. All patients received adjuvant chemotherapy after multicentric disease was identified, and two patients received craniospinal XRT. Followup ranged from 1.25 to 12 years, with stabilization of disease in all cases. One patient died from a cerebral infarction with stable disease on scan and positive cytology. These observations suggest that multicentric JPA may not be as rare as previously assumed. Children less than 4 years old with large, subtotaly resected, suprasellar JPAs appear to be at highest risk for dissemination. Multicentric spread typically occurs within 1.5 years of initial diagnosis, but patients are usually asymptomatic when distant disease is discovered. Leptomeningeal and subarachnoid spread are the most likely mechanisms of dissemination. Although followup is short, outcome in these patients does not correlate with extent of disease, particularly when compared to patients with disseminated malignant astrocytoma. Chemotherapy appears useful in stabilizing disease, although long-term outcome is still undefined.
16. PEDIATRIC OLIGODENDROGLIOMA: NATURAL HISTORY, TREATMENT AND OUTCOME. EXPERIENCE AT BOSTON CHILDREN'S HOSPITAL

Liliana C. Gounnerova, MD, FRCSC, Patrick Barnes, MD, R. Michael Scott, MD (Boston, MA)

We have reviewed the records of all patients treated at Children's Hospital in Boston with the diagnosis of oligodendroglioma since 1956. There were 43 confirmed cases of pure oligodendrogliomas (23 males, 20 females). The majority of patients presented with seizures (88%) and often had had a long history of medically intractable seizures requiring surgical treatment. A minority of the patients presented with headaches (19%) and/or increased intracranial pressure (9%). Patients in the pre-CT scan era were investigated with pneumoencephalograms, angiograms and skull X-rays. CT scans and MRIs were reviewed and demonstrated calcification in 40%, tumor enhancement in 25% and 55% had well-defined margins. All patients have undergone surgery for biopsy and excision. Complete resections were performed in 27 patients, subtotal in 10 and biopsy only in 6, two of whom presented with diffuse leptomeningeal oligodendrogliomatosis. The patients with medically intractable seizures underwent inoperative mapping and resection of epileptogenic foci in addition to the tumor resection. In all 40 cases postoperatively where followup examinations were available, no progression/recurrence of tumor was demonstrated. There have been no deaths in this series and the long term followup has been from 1-20 years. The long term outcome will be presented and discussed with regards to survival, seizure control and the role of adjuvant therapy (radiotherapy and chemotherapy) for residual disease.

17. CHILDHOOD BRAIN TUMORS PRESENTING WITH SEIZURES: RESULTS OF TUMOR EXCISION

Robert Munn, MD, Doug Cochrane, MD, Paul Steinbok, MD, Kevin Farrell, MD (Vancouver, BC)

The surgical management of children with brain tumors, who present with seizures, is controversial. Surgery may include corticography and the excision of epileptogenic foci in addition to removal of the tumor, although it is not known what procedure(s) will optimize seizure control.

This study reviews the clinical features, pathologic findings, and seizure control following tumor excision in 28 children diagnosed with a supratentorial tumor who presented with seizures as the primary manifestation of their tumor (April 1983-September 1991) and who underwent tumor excision.

The average age at first seizure was 5.8 years (3 months to 16.3 years); the duration of seizures prior to operation averaged 3.7 years (3 months to 14.7 years). Partial seizures occurred in 22 patients. Prior to operation, seizure control was attained in three patients.

Computed tomography demonstrated the tumor in 25 patients at seizure onset. Three with normal CT scans were followed (mean 114 months) before the tumor was identified.

Thirteen tumors were temporal and 15 extra temporal. Gangliogliomas occurred in 14 (4 associated with cortical dysplasia), low grade astrocytoma in 8 and malignant tumors in 4 patients.

At followup (average 31.3 months, range 6-105 months), 19 of the 25 survivors were seizure-free. Six patients reported occasional seizures. Nineteen patients remained on anticonvulsants.

This study confirms that most tumors presenting with seizures in childhood are benign and that seizure control can be obtained after tumor excision. Patients with intractable seizures should be reinvestigated regardless of prior studies.
18. PROSPECTIVE ENDOCRINOLOGIC FUNCTION IN CHILDREN WITH BRAIN TUMORS

Paul M. Kanav, MD, Karin S. Bierzabauer, MD, Patrick M. Thomas, MD, Christopher Geth, MD (Philadelphia, PA)

Endocrinologic complications following radiation therapy of brain tumors in children are very common. Growth hormone (GH) is the most sensitive endocrine axis to the effects of radiation, independent of tumor histology. We have instituted prospective endocrinologic testing following surgery and adjuvant therapies to assess the patterns of hormone failure. 45 children with brain tumors received care from July 1990 - December 1991. Pathology was confirmed at craniotomy in 39 patients and presumptive MRI diagnosis was made in 6 children with brain stem or optic pathway tumors. 26 children completed radiation therapy and chemotherapy was used in 9 patients. No postoperative therapy followed tumor resection in 9 children. Prior to surgery and steroid use, heights, TSH, T4, and somatomedin-C levels were obtained and repeated quarterly. Every 6 months provocative stimulation tested GH function. All children maintained normal thyroid function during follow-up. 23/26 patients who received radiation therapy had depressed somatomedin-C levels and GH deficiency. The onset of hormone deficiency was progressive following completion of radiation therapy. Endocrine failure was not seen in patients who only received chemotherapy. Growth velocity declined 3 to 9 months following confirmation of GH deficiency. 9 children with GH deficiency elected recombinant hormone replacement. These prospective data suggest GH deficiency begins earlier than previously recognized, preceding changes in height velocity. Serial thyroid and somatomedin-C levels were and effective screen for hormone deficiency. We recommend careful endocrinologic studies following radiation therapy and upon confirmation of GH deficiency, beginning hormone replacement therapy.

19. STEREOTACTIC SURGERY FOR CYSTIC OR SOLID CRANIOPHARYNGIOMAS IN CHILDREN

D. Kondziolka, MD, FRSCC (C), L. Dade Lunsford, MD (Pittsburgh, PA)

To determine the efficacy of stereotactic surgery in the management of craniopharyngioma in children, we reviewed our 10-year experience in 16 patients. Eleven children had stereotactic intracavitary irradiation (colloidal phosphorus-32) for cystic tumors, three had stereotactic biopsy only, and two had gamma knife radiosurgery for solid recurrent tumor after prior craniotomy. The mean age of patients that had intracavitary irradiation was 14 years. Cystic tumor volumes varied from 2.4 to 90 ml (mean, 13.6). Cyst puncture and injection was performed using a 0.9 mm stereotactic needle. Injected radiation dose varied from 17,000 to 32,600 cGy (mean, 23,800). Seven of 11 patients underwent prior craniotomy and resection of solid and/or cystic tumors. Six patients had shunts. Four had received prior fractionated radiotherapy. Six patients had hypothalamic damage. Mean followup for this group was 3 years (range, 2 to 84 months). Of 10 evaluable patients, 7 had a significant decrease in tumor size and 3 had no further change. There was no treatment related morbidity. One patient had reaspiration of cyst fluid because of symptomatic chiasmatic compression. One patient died from continued growth of solid tumor. Three patients had stereotactic biopsy only of solid tumors thought to be cystic on preoperative CT scan. MRI has eliminated this diagnostic problem. Two children (ages 9 and 12) had stereotactic radiosurgery for recurrent solid tumor. One had complete disappearance of residual tumor in the posterior third ventricle one year after radiosurgery; she then developed a new recurrence in the sella and had a second radiosurgical procedure. She remains well now at three years. The second child had radiosurgery for a small solid recurrence in the wall of the cavernous sinus, after two craniotomies and a transphenoidal resection.

Stereotactic techniques are valuable in the diagnosis and treatment of children with craniopharyngioma. The safety and efficacy Of stereotactic intracavitary irradiation warrant its consideration as primary management in patients with cystic tumors, as well as for recurrence.

Stereotactic radiosurgery will have an increasing role in the management of small volume solid tumors.
20. RADICAL SURGICAL RESECTION OF EXOPHYTIC CHIASMATIC-HYPOTHALAMIC TUMORS

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

33 children underwent 35 operations for radical resection of exophytic chiasmatic-hypothalamic tumors. There were 26 gliomas, 2 ependymomas, 2 hamartomas, 2 germ cell tumors, and 1 PNET. Only 5 children had neurofibromatosis. Clinical presentation correlated with age: <1 year - macrocephaly, failure to thrive, and severe visual loss, 1-5 years - precocious puberty with mild visual loss, and >5 years - slowly progressive loss of vision. Surgical approach was dictated by the direction of maximal growth: 21 children with primarily suprasellar neoplasms had a pterional craniotomy, 11 children with tumor extending superiorly into the 3rd ventricle had a transcallosal resection, and 1 child had a combined approach. There was no operative mortality. Significant permanent morbidity (neurological deficits, visual loss, memory difficulties, and permanent DI) occurred in 5 patients with gliomas and both children with germ cell tumors. Outcome was related to age: 8/9 infants have had biologically aggressive tumors in spite of benign histology and have progressive disease (3 children) or have died (5 children). In contrast, 75% of the older children have remained in continuous remission following radical resection without adjuvant therapy. The authors conclude: 1) exophytic chiasmatic-hypothalamic gliomas and ependymomas are amenable to radical surgical resection with minimal morbidity, 2) radical surgery for germ cell tumors is hazardous and should not be performed, 3) 75% of children over 1 year of age will have prolonged progression free survival after radical surgery, 4) chiasmatic-hypothalamic tumors of infancy are biologically aggressive neoplasms that require multimodality therapy.

21. DETECTION OF TUMOR RECURRENCE BY SURVEILLANCE SCANNING IN CHILDREN WITH MEDULLOBLASTOMA

Leslie N. Sutton, MD, C. Torres, S. Rebeamen, P. Phillips, L. Bilaniuk, J. Goldwein, B. Lange (Philadelphia, PA)

It has been the practice at our institution to follow children with medulloblastoma (posterior fossa primitive neuroectodermal tumor) by surveillance CT or MRI scans of brain every 3 months for 1 year after surgery, at least every 6 months for the next 2 years and yearly thereafter. Surveillance spine myelography or MRI is performed in those with cord disease or infants treated with 1800 cGy craniospinal axis irradiation. Between January 1980 and March 1992, 702 surveillance head and 75 spine scans or myelograms have been performed on 90 children 5 mos to 21 years of age (mean 7.1 years) followed for 2-158 months (mean 47.3 mos) from surgery. Sixty-three children have had no recurrences; 27 have had recurrence or progression, 26 of which were in the CNS. Twenty-two of 42 patients with either residual tumor after surgery or disseminated disease at initial staging had recurrence compared to 5 of 48 with no detectable tumor. Recurrences developed 2-64 mos (mean, 16.9 mos; median, 13 mos) from diagnosis. Clinical symptoms heralded 23 of 27 recurrences at an average of 4.7 mos from the previous scan; surveillance scans detected only 4 of 27 recurrences. Three patients with clinically detected disease survive 1, 14 and 17 mos; one with relapse detected by scan survives 17 mos. Death occurred 1 to 38 months from recurrence (mean, 11.0 mos) in those with clinically detected disease and at 8, 24 and 35 mos in those with disease detected by scan. No spinal cord relapses occurred without symptoms or without concurrent brain disease; thus, surveillance spine scans did not yield useful information. Surveillance brain scans failed to detect recurrent disease in most patients and had virtually no impact on outcome. We conclude that surveillance scans may not be necessary in those with no residual disease after surgery or no evidence of dissemination. In these patients, at least one post-treatment scan is necessary as a baseline. During treatment and thereafter, followup should emphasize parent education and neurologic evaluation to detect relapse and to elicit and remediate late effects of therapy.
22. THE ROLE OF 201-THALLIUM CHLORIDE SPECT IMAGING IN
CHILDHOOD BRAIN TUMORS
Bernard L. Maria, MD, J. Parker Mickle, MD, Ronald G. Quisling, MD,
Walter E. Drain, MD (Gainesville, FL)

The potential utility of 201-Thallium Chloride single photon emission computer
tomography scanning was investigated as a diagnostic tool in 38 children with
brain tumors. A total of 75 spect scans were performed on these individuals. The
goal was to define the specificity of the spect scan in a wide range of histologies
and to define its relationship to magnetic resonance imaging volumetric assess-
ments and gadolinium enhancement of tumor and necrosis. Also, the usefulness
of the 201-Thallium spect scan was looked at in terms of postoperative assess-
ment and at the time of clinical progression. Thallium uptake was detected in 23
of 27 neoplasms with MRI measurable residual disease including 14 of 18
posterior fossa tumors. The spect scan demonstrated uptake in tumors with
Thallium ranging from .03 cc to 60 cc. Spect imaging did not correlate with MRI
gadolinium enhancement, necrosis, or exocytic and multi/unicentric character-
istics. Significantly, spect imaging was negative in patients with tumors of
mal-developmental origin and radionecrosis. 201-Thallium spect scanning was
found to be helpful postoperatively, and at time of recurrence. It is concluded
from this survey that 201-Thallium spect scanning is an important imaging adjunct in
the assessment of children with brain tumors.

23. THE EXPERIMENTAL MODEL: CHIARI II-LIKE CHANGE IN
SURGICALLY INDUCED SPINA BIFIDA APERTA IN CHICK
Takayuki Inagaki, MD, David G. McLone, MD, Yasuo Yamanouchi, MD
(Salt Lake City, UT)

It is well known clinically that the patient with myelomeningocele usually has an
accompanying Chiari II malformation. Also reported in some drug induced and
congenital animal models with spina bifida aperta were small posterior fossas with
downward herniation of their contents.

We hypothesize that the leakage of cerebrospinal fluid through unclosed or
reopened neural tubes at the early stage of vertebrate embryos may be a cause
of Chiari II malformation in animals with spina bifida aperta. To determine the
possibility of this hypothesis, the closed neural tube was surgically opened at the
early stage of embryo and the posterior fossa was examined histologically. The
chick embryo was chosen for this study. The eggs were incubated in a humidified
incubator at 38 degrees celsius.

After opening a window, embryos were staged under a dissecting microscope.
These embryos were operated at stages between 12 to 22 (Hamburger and
Hamilton). Surgery was repeated at least three times every 6 to 8 hours. The
window sites were sealed with plastic tape and the eggs were reincubated.
The embryos were harvested after 14 to 16 days of incubation and immersed in fresh
4% PFA fixative.

Midsagittal section of the head revealed the downward movement of the choroid
plexus of the fourth ventricle and narrowing of the subarachnoid space at the high
cervical portion in the embryos with spina bifida aperta. Our data may indicate the
possible role of cerebrospinal fluid leakage in the development of Chiari II
malformations in mechanically induced spina bifida in chick.
We have studied a group of patients with craniovertebral junction anomalies to include Chiari I and II malformations as well as basilar impression with Cine-mode MRI. Dynamic flow velocity images were obtained by cardiac-gated phase contrast scanning, in which the average CSF velocity and direction of flow were compiled and video-recorded. We observed physiologic CSF velocity differences as compared to normal controls. Pre- and postoperative images were also obtained in all patients who underwent surgery. Specifically, in Chiari I patients there was a decrease in CSF flow as compared with normal controls dorsal to the cerebellar tonsils and valleculae and through the foramen magendie. Post-operatively, after posterior fossa decompression with dural grafting and lysis of adhesions there was a significant increase in the flow through the foramen Magendie and increase in the flow at the foramen Magnum. In patients who had decrease in the size of their syrinx-postoperatively there was also an increase in the CSF flow in the craniovertebral junction. There was no flow appreciated between the obex and the central canal in patients with a syrinx. Additionally, this technique demonstrated the patency of a dorsal cyst in a patient with a Chiari II malformation and hypoplastic cerebellum. The Cine-mode MRI also corroborated decreased CSF flow ventral to the craniovertebral junction.

Using Cine-mode MRI, the patterns and dynamics of CSF flow are clearly demonstrated in craniovertebral junction anomalies. Information gathered from this technique has implications in understanding the pathophysiology of these disorders as well as being useful in assessing the relative merits of the various surgical therapies for these entities.
26. SYMPTOMATIC CHIARI I MALFORMATIONS IN CHILDREN: CLINICAL FEATURES, SURGICAL MANAGEMENT, AND OUTCOME

Martin Herman, D.G. McLone, Mark S. Dias, M. Alonzo (Chicago, IL)

We describe the presenting symptoms and signs, magnetic resonance image (MRI) features, and clinical outcome of twenty-one children (all without myelodysplasia) undergoing surgical decompression for symptomatic Chiari I malformations. Headache and neck pain were predominant symptoms, occurring in 80% of patients. Pain characteristically began with exertion or Valsalva's maneuver, and the child's response was, in many instances, stereotypical. The child would immediately cease all activity, grab the neck and become perfectly still (often not even crying) until the pain resolved, then resume normal activity. Other signs and symptoms included nausea and vomiting in 50%, upper and lower extremity motor signs in 30%, sensory disturbance in 25%, and cerebellar or cranial nerve dysfunction each in <10%.

Preoperative MRI in all patients demonstrated caudal displacement of the cerebellar tonsils to the level of the 1st cervical vertebra in 90% of patients, and to the C2 or C3 vertebra in 10%. Hydrocephalus was absent in all patients, but hydromyelia was found in 5 children (24%). Posterior fossa bony decompression and cervical laminectomy were performed in all patients; 14 patients (67%) underwent C1 laminectomy, and 7 patients (33%) underwent C1 and C2 laminectomy. Sixteen patients (76%) also underwent duraplasty. Postoperatively, 85% of patients experienced complete resolution of signs and symptoms, whereas 10% had minimal or no improvement, and one patient (5%) died of respiratory complications. Postoperative MRI in all patients demonstrated no change in the position of the cerebellar tonsils. Hydromyelia was significantly improved in 2 of 4 patients (50%) who had postoperative MRI evaluations.

We emphasize the characteristic behavioral response to the headache and/or cervical pain associated with a symptomatic Chiari I malformation. Our results suggest that surgical decompression is an effective treatment for symptomatic Chiari I malformations in children with and without hydromyelia.

27. TREATMENT OF HYDROMYELIA IN CHILDREN PRESENTING WITH SCOLIOSIS

Mary Elizabeth Dunn, MD, Walter L. Bailey, MD (St. Paul, MN)

Hydromyelia in ten children with Chiari I malformation ages three to fifteen have been identified on MRIs ordered as a screening procedure for progressive scoliosis. The hydromyelia on MRI scan was dramatic and measured at least 50% of the cord diameter preoperatively over at least ten segments. Scoliosis was the presenting complaint in each case and was of the idiopathic variety when initially evaluated.

Surgery in each case, cervicomedullary decompression with patch graft or without tonsillar tack up sutures, assured communication of CSF flow from the fourth ventricle to the cervical subarachnoid space. The obex was noted to be open in two cases. No muscle plugging was done. There was no significant morbidity; hospitalization was four to six days. Postoperative MRI sagittal films in all cases show dramatic collapse of the hydromyelic cavity with follow up to four years in some cases.

MRI pre and postoperative films will be shown as well as specifics of the operative techniques. Cervicomedullary decompression seems to be a safe, adequate treatment for whole cord hydromyelia with scoliosis as the presenting symptoms. Scoliosis in eight of ten patients was significantly improved in long term follow up. Two cases of the teenage group had their scoliosis advance, requiring orthopaedic fusion of the spine despite continued demonstration of collapse of the hydromyelic cavity. Examples of adult patients with scoliosis during childhood presenting with neurological deficit as adults emphasizes the beneficial potential of early treatment of hydromyelia in children without neurologic deficit, but with scoliosis.
28. MANAGEMENT OF SYMPTOMATIC CHIARI II MALFORMATIONS IN CHILDREN WITH MYELOMENINGOCELE

Mark S. Dias, David G. McLone, Mario Alonzo (Chicago, IL)

Between 1983 and 1991, 43 patients with myelomeningoceles and symptomatic Chiari malformations underwent surgical treatment; their presenting features, radiographic characteristics, and surgical outcomes were analyzed retrospectively. The average age at the onset of symptoms was 3.7 years; 13 patients (30%) became symptomatic within the first year of life. The average duration of symptoms was 5.8 months. Presenting signs and symptoms included disorders of swallowing in 32 patients (74%), oculomotor dysfunction in 7 patients (16%), disordered upper extremity motor function in 23 patients (53%), cerebellar dysfunction in 15 patients (35%), and pain (headache and/or neck pain) in 19 patients (44%). Disorders of swallowing occurred most frequently in infancy and declined with advancing age; in contrast, pain was uncommon in infancy and increased with advancing age. Other abnormalities were evenly distributed across all age groups.

Thirty patients (70%) underwent cervical decompression (CD), 12 patients underwent both cervical and posterior fossa decompression (PFD), and one patient underwent only PFD. Improvement was seen in 80% of patients following CD, and in 62% of patients undergoing CD plus PFD (χ² = 1.6; not significant). The fourth ventricle was opened in 17 patients, 8 of whom also underwent shunting to the subarachnoid space, and 2 of whom additionally underwent plugging of the obex: 14 patients (69%) were improved postoperatively. The remaining 26 patients underwent only bony opening and duraplasty; 14 patients (54%) were improved postoperatively. The difference in outcomes is not statistically significant (χ² = 0.93). The age at onset of symptoms was a significant predictor of outcome following decompression. The average age for those patients who improved post-operatively was 4.51 years, versus 1.48 years for those who did not (P < 0.005). Only 17 (14%) patients whose symptoms began immediately after birth improved postoperatively. Pain, cerebellar dysfunction, and upper extremity motor problems improved in 16/17 (94%), 5/5 (100%), and 19/23 (83%) patients respectively. In contrast, swallowing dysfunction was least likely to improve post-operatively, occurring in only 16 of 30 patients (53%).

Our results suggest that 1) surgical treatment for symptomatic Chiari II malformations in children with myelodysplasia need not include a posterior fossa decompression, as cervical decompression with duraplasty alone is just as effective, 2) no additional benefit was derived from opening, shunting or shunting the fourth ventricle, 3) infants whose symptoms begin at or shortly after birth are unlikely to benefit from operation, and 4) although some signs and symptoms respond readily to decompression, swallowing dysfunction improves much less frequently.

29. MR ARTERIOGRAPHY AND VENOGRAPHY IN CHILDREN

Benjamin C.P. Lee, MD, T.S. Park, MD, Bruce A. Kaufman, MD (St. Louis, MO)

PURPOSE:

To evaluate the clinical usefulness of MR arteriography (MRA) and venography (MRV) in pediatric patients.

MATERIAL AND METHODS:

100 patients from newborn to 16 years of age were studied: 50 by MRA and 50 MRV. The indications for MRA included vascular malformations, dural fistula, aneurysms, arteritis, suspected internal, major cerebral arterial occlusions and tumors. Indications for MRV included congenital venous anomalies, dural sinus, deep venous thrombosis and compressions, tumor displacement, cortical venous occlusions secondary to meningiitis and subdural hematoma. Conventional cerebral angiography were performed in selected cases.

RESULTS:

The internal carotid artery and proximal branches of the circle of Willis were routinely visualized with MRA. Abnormalities were demonstrated in 25 patients: 2 arteriovenous malformations, 2 vein of Galen aneurysms, one dural fistula, one aneurysm, one vasculitis, 11 internal carotid, and one middle cerebral artery occlusion, 5 tumor displacements and one vascular tumor. The findings were confirmed by conventional angiography in 10 cases. The deep sinuses and deep cerebral veins were visualized in all cases with MRV and abnormalities were demonstrated in 20 patients: 4 congenital venous anomalies, one deep venous occlusion, 2 deep venous and dural sinus occlusions, one dural sinus occlusion, 6 cortical vein occlusions, 2 compressions of dural sinuses by extrinsic masses, and 4 deep venous displacement by tumors.

CONCLUSIONS:

In our institution MRA has become part of routine MR scanning. Conventional angiography is reserved for evaluation of subtle abnormalities such as arteritis and for examination of the peripheral branches of the cerebral arteries. MRA is the preferred method and is superior to conventional cerebral angiography in evaluation of deep veins and deep cerebral veins.
30. DIAGNOSIS AND TREATMENT OF DURAL SINUS THROMBOSIS (DST) WITH THROMBOLYSIS THERAPY
Karol Zakalik, MD, Ay-Ming Wand, MD, Mark G. Goetting, MD (Royal Oak, MI)

Prior to advancement of MRI/MRA DST was difficult to diagnose and potentially fatal. We present 4 children with DST that were diagnosed by MRI and 3-D MRA and were successfully treated with local thrombolysis with the use of microcatheters. Follow up MRA showed continued patency in all cases.

Case 1. A 6 d.o. presented with seizures, r. hemiparesis and 1. occipitoparietal brain swelling by CT. MRI/MRA confirmed brain edema and showed a fresh clot in the 1. transverse sinus without flow. The clot was dislodged by microcatheter. Seizures and hemiparesis resolved.

Case 2. A 10 m.o. had a 1. 6th nerve palsy and a bulging fontanel. MRI/MRA revealed thrombosis of the basal vein, 1. cavernous, sagittal, straight and right transverse and sigmoid sinuses. Local urokinase restored patency. His fontanelle was soft the next day and his 6th nerve palsy resolved.

Case 3. A 13 y.o. with protein losing enteropathy and dehydration developed headaches, 6th nerve palsy. MRI/MRA revealed complete thrombosis of the sagittal, r. transverse, torcular and part of the 1. transverse and cavernous sinuses. Urokinase dissolved the clot. The headache and diplopia improved.

Case 4. A 3 w.o. with dehydration developed apneas. CT suggested sagittal sinus thrombosis, edema of occipital lobes, left cerebellum, and right thalamus. MRI/MRA showed bilateral transverse and partial sagittal sinus thrombosis as well as basilar artery occlusion. The venous sinuses were cleared with urokinase.

MRA facilitates the diagnosis of DST. Prompt local thrombolysis with microcatheters followed by heparin can rapidly achieve and maintain sinus patency.

31. TRAUMATIC ANEURYSMS IN CHILDHOOD
Phillip A. Yazbak, J. Gordon McComb, Corey Raffel (Los Angeles, CA)

From a group of eighteen children treated for an intracranial aneurysm between 1975 & 1992, seven were traumatic in origin. The mean age was nine years (117), with a 5:2 male:female ratio. The nature of the trauma was as follows: motor vehicle accidents-3, nonaccidental trauma-2, fall-1, stabbing-1. Five patients presented with hemorrhage, one with a growing fracture, and one with behavioral change. The time from injury to presentation varied from one week to two years. The location of the aneurysm was as follows: anterior cerebral artery (two pericallosal and one distal)-3, distal middle cerebral artery-3, and distal posterior inferior cerebellar artery-1. Surgical treatment consisted of excision in five, clipping in one, and wrapping/gluing in one. Neurologic outcome in six of the seven was unchanged or improved.

Traumatic aneurysms as a percentage of the total number of aneurysms is considerably higher in children compared with adults. Angiography is indicated in those patients with a high index of suspicion. All aneurysms should be surgically treated. The outcome is usually quite favorable but depends upon the severity of the initial head injury, location of the aneurysm, and surgical treatment before rupture.
32. CHILDHOOD CEREBRAL CAVERNOUS ANGIOMAS: CLINICAL, RADIOLOGIC, PATHOLOGIC CORRELATION, SURGICAL MANAGEMENT AND OUTCOME

Spyros S. Kollias, MD, Erin C. Prenger, DO, William S. Ball, MD, Kerry R. Crone, MD, Bradbury Skidmore, MD (Cincinnati, OH)

Cerebral cavernous angiomas are uncommon lesions, being more rare in childhood. We describe the clinical, pathologic and radiologic (angiographic, CT, MRI) features and illustrate the surgical management and outcome in 15 pediatric patients (mean age 8.4, M:F ratio 9:6). MRI was available in all cases. Pre and postcontrast was 13, and angiography in 10. Twelve were surgically treated and had histologic correlation.

The most common clinical presentation was seizures (11/15). Multiple lesions were present in 5 cases, 2 of which proved to be familial. MRI detected 31 lesions varying in size from punctate to 4.5 cm and was more sensitive than CT in detecting additional small lesions. Location was supratentorial (30/31) and infratentorial (1/31). Two lesions were intraventricular. Typical appearance was that of well defined, nonenhancing lesions with marked T2 shortening consistent with hemosiderin, with or without central high signal foci consistent with methemoglobin. Intense enhancement and edema was present in 2 lesions complicating the differential diagnosis. Angiography was normal in 8/10 cases and showed only mass effect in 2/10. Pathologically consisted of single lined endothelial vascular spaces with no intervening neural tissue. Thrombosis, calcification, surrounding gliosis, and hemosiderin deposition were also present.

Total surgical removal was possible in all cases. The outcome over a mean followup period of 1.5 years was excellent (seizurefree without medication in 9/12 cases). We conclude that MRI is the investigation of choice and usually helps to establish the correct diagnosis. Surgical treatment is safe and offers an excellent prognosis for seizure control. It is always indicated in symptomatic patients because of their tendency to enlarge and the risk of hemorrhage.

33. EFFECTS OF BIOLOGICALLY DELIVERED NEUROTROPHINS IN ANIMAL MODELS OF NEURAL DEGENERATION

David M. Frim, MD, PhD, Tara Uhler, Christina Fleet, Priscilla Short, Xandra Brakefield, Ole Isacson (Boston, MA)

Neurotrophic molecules, such as the members of the Nerve Growth Factor (NGF) related Neurotrophin family, are known to promote neuronal survival in vitro and prevent cell death during development in vivo. The effect of the neurotrophins on cell death in the pediatric or adult brain is unknown; however, NGF axonogenously delivered by either mechanical or biological means potently protects against a variety of neuronal insults. To further explore the protective potential of the neurotrophins, we have used retroviral gene transfer to construct immortalized rat fibroblast cell lines capable of secreting high levels of NGF, or the related neurotrophin, Brain Derived Neurotrophic Factor (BDNF). NGF-, BDNF-, and non-secreting cell lines were implanted intracerebrally near the striatum in rats 7 days before striatal infusion of an excitotoxin, mimicking the toxicity of a variety of neuronal insults. Analysis of subsequent striatal neuronal death revealed that NGF, but not BDNF, was able to ameliorate the excitotoxicity. This finding suggests that neurotrophin protection is likely specific to neuronal subpopulation, predicting that NGF, but not BDNF, may be of potential value in the study and treatment of striatal degenerative disorders. The relevance to pediatric neurosurgery of this experimental approach will be in the identification and chronic biological delivery of neurotrophic factors capable of supporting the neuronal subpopulations affected by pediatric neurodegenerative disorders.
34. HISTOPATHOLOGIC DIFFERENCES IN TISSUE REACTIONS TO SILASTIC VS. SILASTIC-DACRON DURAL SUBSTITUTE

Anthony E. Russell, MD, William M. Chadduck, MD (Little Rock, AR)

Previously we reported the use of Silastic duraplasty for pediatric patients with spinal abnormalities having a predilection for tethering of neural structures to overlying tissues. We emphasized using non-reinforced Silastic sheeting because Dacron reinforced Silastic had been observed to cause marked fibrioblastic responses. In December, 1991, Dow-Corning advised neurosurgeons that Silastic should not be used for dural repair; however, they did not differentiate between non-reinforced Silastic sheeting and that reinforced with Dacron. Because our favorable results using the non-reinforced Silastic sheeting differed from reports in the literature using Dacron reinforced material, we performed chronic subcutaneous implantations of the two varieties of Silastic sheeting in rabbits. After three months, the animals were sacrificed; the Silastic and surrounding tissues were excised en bloc. Histopathologic examination of the six non-reinforced Silastic samples showed them to be covered with a thin membrane of fibrous tissue, lined by a single layer of epithelial cells. There was no inflammatory reaction, and no evidence of breakdown of the material. In contrast, the six samples of Dacron reinforced Silastic showed intense fibrotic reactions, lymphocytic and eosinophilic inflammatory responses, and giant cell formation with phagocytosis of Dacron particles. These animal experiments demonstrate a significant difference in tissue reaction between non-reinforced and Dacron reinforced Silastic. The results further support our previous conclusions, that (1) the use of non-reinforced Silastic material is an excellent dural substitute when prevention of tissue adherence to neural structures is important, and (2) that Dacron reinforced Silastic should not be used for duraplasty.

35. METHYL PREDNISOLONE UPTAKE AT THE BLOOD-BRAIN INTERFACE

Thomas C. Chen, J. Gordon McComb, Jasmina B. Mackic, Berislav V. Zlokovic (Los Angeles, CA)

Our understanding of the pharmacokinetics of methylprednisolone (MP) uptake at the blood-brain interface, and its mode of action in the brain is limited.

An in-situ vascular brain perfusion technique in guinea-pigs was used to determine the pharmacokinetics of [3H]-MP transport at the blood-brain interface. A capillary depletion step was used to distinguish between blood-brain interface transport versus capillary sequestration. [14C]-Sucrose was used as a cerebrovascular space marker. After ten minutes of vascular brain perfusion, [3H]-MP uptake was progressive, time dependent, and 5-6 times higher relative to [14C]-sucrose. MP blood-brain interface in-situ binding and transport exhibit saturation kinetics. Brain compartmental analysis indicated a significant capillary sequestration of [3H]-MP prior to its entry into brain tissue.

It is concluded that MP crosses the blood-brain interface in a saturable manner, and that a significant portion of steroid remains sequestered at the level of endothelial cells at the blood-brain interface.
36. PRENATAL CORTICAL INJURY: METABOLIC AND ANATOMICAL EFFECTS IN THE CAT

P. David Adelson, MD, David A. Hovda, PhD, Louise D. Loopuit, PhD, Jaime R. Villablanca, MD (Los Angeles, CA)

Improved neurosurgical techniques have made prenatal surgery possible but little is known of the effects of intrauterine Central Nervous System damage. We have previously shown that cerebral lesions sustained early in life resulted in an improved recovery of function as demonstrated behaviorally, metabolically, and anatomically. It has been proposed that the earlier the injury, the better opportunity for complete recovery. Recently we have shown that prenatal injured animals were more impaired neurologically than those that had been injured during the neonatal period. Anatomically, they were also found to have thalamic and cortical atrophy but caudate hypertrophy. To define the metabolic changes that occur, a comparison of the oxidative metabolism and the local cerebral metabolic rates for glucose (LCMRglc) were determined for these regions in intact adult, neonatal-hemisphereonized, adult hemispherectomized, and prenatal (E43-50) unilateral frontoparietally lesioned cats. The brains were processed using cytochrome oxidase histochemistry and the LCMRglc rates were determined using 2-deoxyglucose. Optical densitometry was used to compare the lesioned side to the nonlesioned side as well as to intact controls. Preliminary, the difference between sides in the thalamus was most profound in the adult lesioned animals. The neonatal and prenatal lesioned animals had only minor differences and not to the extent of the adult. This data will be discussed. Though the metabolic differences between those animals that were lesioned prenatally compared to neonates was not significant, there was less recovery of neurologic function. This study suggests that the developing brain during the prenatal period is more vulnerable to injury.

37. SELECTIVE DORSAL RHIZOTOMY: IS S2 LESIONING NECESSARY?

Prasanna Jayakar, MD, PhD, Glenn Morrison, MD, Antonio Prats, MD, Chester Tylkowski, MD (Miami, FL)

Selective dorsal rhizotomy is conventionally carried out between segment levels L2 and S1. Rootlets which demonstrate abnormal electrophysiologic responses to threshold stimuli are resected. Inclusion of S2 has recently been advocated to minimize residual spasticity in the leg musculature but is associated with significant postoperative urinary retention and risk of sexual dysfunction. We, therefore, evaluated whether selectively increasing the degree of S1 lesioning would adequately relieve spasticity even when S2 was left intact. Eleven patients (8M, 3F) aged 4 to 14 years (mean = 6.9) were subjected to lesioning of at least 80% of the S1 root. All had preoperatively demonstrated greater than 3+ passive tone (Modified Ashworth scale) in the plantar flexors of the ankle in the supine posture. Lesioning was initially carried out at threshold intensity. If the proportion of rootlets responding abnormally were less than 80%, the stimulus intensity was increased to suprathreshold values in order to determine the most involved of the remaining rootlets. S2 segment was left intact. Ten of the 11 patients derived significant relief of spasticity in the plantar flexors (0 to 1+), with 1 showing only marginal benefit. Bladder involvement was not observed in any of the patients post-operatively except one who required catheterization once for relief of transient retention.

Our findings lead us to believe that spasticity in the leg musculature can be adequately ameliorated by increasing the degree of S1 lesioning without incurring the risk of significant bladder complications.
38. SELECTIVE DORSAL RHIZOTOMY — A FOLLOW UP STUDY OF AMBULATORS AND NONAMBULATORS
Amo Fried, MD, Jennifer Ahl, RN, Allan Gurd, MD, Janet Komisarz, RN, Diane Molinger, PT, Bonnie Boenig, MEd, OTRL, Kerri Levin, MD (Providence, RI)

The ability to predict functional improvement in children undergoing selective dorsal rhizotomy (SDR) is especially important, as the procedure is now being closely scrutinized as to its ability to improve the lifestyle of children with Cerebral Palsy. In a large CP clinic, 60 children were selected for selective dorsal rhizotomy with 190 being screened. The follow up of the children undergoing rhizotomy ranged from 6 months to 3 years with a mean of 2 years. Children were categorized as ambulators (N=29) and nonambulators (N=31). Prognosis was determined using objective evaluations as well as blinded videotapes of function, after their course of therapy.

Preoperatively, among children who were ambulators, 13/29 were independent (Group I) and 16/29 used assistive devices to walk (Group II). Among nonambulators, 11/31 could take one or two steps with maximal assistance (Group III) and 20/31 were completely nonambulatory (Group IV). The range of sensory rootlets cut was 15-40% of those tested from S1-L2.

Among ambulators, 13/29 (45%) had improved gait using assistive devices and 15/29 (52%) had improved independent gait. One was unchanged. Among nonambulators, 14/31 (45%) showed improved sitting ability and posture and 16/31 (52%) were able to walk to some degree as well, using assistive devices. Among nonambulators who became walkers, 7/20 were in Group IV preoperatively and 9/11 (82%) were in Group III preoperatively. There were 8 minor transient complications in 6 children.

Selective dorsal rhizotomy is an effective option for children with CP with predictable outcomes in ambulator and nonambulator groups. Nearly all ambulators could walk better (96%). In nonambulators all but one had posture and sitting improvement (97%) while 50% achieved some degree of walking ability, especially in Group III. This result could be due to the cutting of only 15-40% (mean 30%) of sensory rootlets.

39. THE ISOLATION OF HUMAN SCHWANN CELLS FROM SELECTIVE DORSAL RHIZOTOMY SPECIMENS
Gerald F. Tuile, MD, P. Boyer, R.C. Dauser, K.M. Muraszko, L. Rutkowski (Ann Arbor, MI)

We have developed a method for the isolation of human Schwann cells from selective dorsal rhizotomy (SDR) specimens, based on a technique we developed for Schwann cell isolation from human sural nerve biopsies. (Ann Neurol 1992;31:580) We found that pediatric SDR tissue can generate a much higher yield of Schwann cells than human sural nerve biopsies; the isolation procedure described for adult tissue produced a cell yield five times that obtained with adult tissue (5-11 x 104 cells/mg of nerve tissue). However, these same cultures were rapidly overrun by fibroblasts in a short period of time. We found that pediatric fibroblasts from SDR specimens grew faster (doubling time 48 hours) and to a higher density (greater than 103 cells/mm2) than the fibroblasts from adult nerves (doubling time >7 days and saturated at 102 cells/mm2).

We have modified our isolation protocol to counteract the vigorous proliferative capacity of pediatric fibroblasts from SDR specimens using three principle techniques. First, the SDR tissue is grown in L-valine deficient medium; this selectively inhibits fibroblast growth since they are unable to convert this essential amino acid to the usable D isomer. Secondly, a preplating technique is employed to preferentially remove the more rapidly attaching fibroblasts. Finally, the tissue is treated with a combination of agents that elevate cyclic AMP levels, thus providing selective inhibition of fibroblast mitosis and stimulation of Schwann cell growth.

The ability to culture large numbers of human Schwann cells from SDR specimens may facilitate the eventual understanding of the molecular and biochemical mechanisms of peripheral nervous system diseases. Similarly, isolated cultured Schwann cells may play an important role in the study of both peripheral and central nervous system regeneration.
40. EFFECTS OF SELECTIVE DORSAL RHIZOTOMY FOR SPASTIC DIPLEGIA ON HIP MIGRATION IN CEREBRAL PALSY
T.S. Park, MD, George P. Vogler, PhD, Bruce A. Kaufman, MD, Madeleine Ortmann, RN (St. Louis, MO)

In spastic cerebral palsy, lateral hip migration is common; reported incidence of hip subluxation or dislocation ranges from 3 to 60 percent. While a selective dorsal rhizotomy is employed for spastic diplegia, its effect on hip stability after the operation is unknown. We examined whether the dorsal rhizotomy halts or exacerbates lateral hip migration in 67 children (134 hips) with spastic diplegia. The migration percentage of Reimers (MP) computed from pre- and post-rhizotomy hip radiographs was used as an index of the severity of lateral hip migration. Age at the time of the rhizotomy was between 2 and 11 years. Twenty and 47 patients were followed for 6-10 months and 15-46 months, respectively. Postoperative MP values were unchanged in 75 percent, decreased in 18 percent and increased in 7 percent of the hips examined. Thus 93 percent of the hips were radiographically stable after the rhizotomy. Age of patients or gait function at surgery did not affect the outcome. Pre-rhizotomy hip migration significantly influenced the outcome (P<0.001, Fisher's exact test) and was highly significant as a predictor of the outcome (stepwise multiple regression analysis, P<0.001, R²=0.34). 89 percent of hips with MP of <15 before the operation were unchanged and 11% worsened; of hips with MP of 15-33, 80 percent were unchanged and 14% improved and 6 worsened; of hips with MP of 33, 55 percent improved and 45 percent were unchanged. The data indicate that selective dorsal rhizotomy offers an effective method for management of hip deformities in spastic diplegia.

41. CHILD ABUSE HEAD INJURY STUDY
Jodie K. Levitt, MD, Henry Bartkowski, MD (Columbus, OH)

The trauma registry from Columbus Children's Hospital was reviewed retrospectively from 1985 through 1992. All patients admitted to the hospital for child abuse during this period were reviewed with respect to age, sex, type of injury (head vs. spine vs. other), and severity. Of the total of sixty-six patients, thirty-four (52%), suffered head trauma and only one (2%), sustained spinal cord injury (central cord injury). Males sustained 62% of head injuries. Thirty-two of the head injured patients (94%) were under the age of two years, while eighteen (53%), were under six months. Pathology in the head trauma group included twenty-five (74%) patients with intracranial lesions, and nine with concussive symptoms and skull fracture. The closed head group included fourteen (56%) patients with severe trauma (subarachnoid/subdural hematomas or multiple contusions/hemorrhages) all requiring surgery or intracranial pressure monitoring, four (16%) with chronic pathology (chronic subdural or hygromas), one (4%) with a documented brain stem injury by MRI as well as other intracranial pathology, five (20%) with no visible pathology by CAT scan but with clinically diagnosed concussions, and one (4%) with acute hydrocephalus related to previous abuse trauma. All deaths occurred in the closed head group, one from acute hemaition secondary to hydrocephalus and four from severe intracranial hemorrhages.

Head trauma is a common form of injury in child abuse. More than half the cases hospitalized in Columbus Children's Hospital over the last four years suffered some form of head trauma, with 15% fatality. Most patients in this group are under two years old. Spinal injury is uncommon and represented only 2% of all abuse trauma. Fractures, burns, and bruises were the next most common types of child abuse.
42. MAGNETIC RESONANCE IMAGING IN THE EVALUATION OF CHILDREN WITH SPINAL CORD INJURY WITHOUT RADIOGRAPHIC ABNORMALLY

Paul A. Grabb, MD, Dachling Pang, MD, FRCS(C), FACS, William Hirsch, MD (Pittsburgh, PA)

Over a 36-month period, we evaluated 18 children with spinal cord injury without radiographic abnormality (SCIWORA). We obtained magnetic resonance images (MRI) in seven children because of 1) findings indicative of severe cord injury or 2) persistent neurologic signs. These seven children ranged in age from newborn to 17 years. Mechanisms of injury varied. There were six cervical and one thoracic cord injuries. MRIs were obtained three hours to 16 days after injury. Two MRIs obtained acutely (4 hours after injury) in one child with paraplegia, and one with hemisensory disturbance and monoparesis, were normal. The remaining MRIs showed cord hemorrhage in two children with quadriparesis and quadriplegia, respectively; cord disruption in a child with quadriplegia; anterior longitudinal ligament rupture and endplate separation in a child with hemisensory disturbance; and cord edema and posterior longitudinal ligament injury in a child with Brown-Séquard syndrome. Follow up MRI revealed chronic ligamentous instability in a child whose initial MRI did not show ligamentous injury.

We found abnormalities in 71% of MRIs in children with SCIWORA and severe or persistent neurologic signs. MRI results did not influence immobilization methods or reveal operative lesions, but did correlate with outcome (cord disruption + hemorrhage + edema = abnormality). We, however, recommend emergency MRI in children with SCIWORA with progressive or severe incomplete cord syndromes to rule out compressive lesions. We also recommend semi-elective MRI for SCIWORA patients with persistent deficits for prognostic purposes. MRI obtained less than six hours after injury may be too early to display traumatic lesions. Gradient echo images and short TR images with fat suppression algorithms should be employed to idealize the imaging of the acutely injured spinal cord and surrounding soft tissues, respectively. Though not found in every case of SCIWORA, evidence of ligamentous injury on three MRIs (acute and followup) supports the hypothesis of ligamentous strain which allows sufficient intersegmental motion to cause neural injury. More experience is necessary to assess the ability of MRI to predict ligamentous instability.
44. CONTROLLED EXTERNAL LUMBAR DRAINAGE IN DIFFUSE PEDIATRIC HEAD INJURY

David I. Levy, MD, W. Bruce Chemy, MD, Kim Manwaring, MD, S. David Mass, MD, Hillel Z. Baldwin, MD, Harold L. Rekate, MD (Phoenix, AZ)

Our experience with the use of external lumbar subarachnoid drainage in 10 children with severe diffuse head injuries is presented. All patients had Glasgow Coma Scale scores of 8 or less at 24 hours after injury and were initially treated with ventriculostomies. Four children required surgical evacuation of focal mass lesions. Within 72 hours of admission, all children manifested high intracranial pressures (ICP) refractory to maximal therapy, including hyperventilation, furosemide, mannitol, and barbiturates. After the institution of lumbar drainage, eight children had an abrupt and lasting decrease in ICP, obviating the need for continued barbiturates and hyperventilation. Eight children survived, five of whom made good recoveries. Three children are functional with disability. ICP varied passively with the height of the drainage bag in these surviving patients. Two patients died, most likely from uncontrolled ICP before the lumbar drain was placed. We conclude that controlled external lumbar subarachnoid drainage has proven useful treatment option for severe diffuse pediatric head injury when maximal medical therapy and ventricular cerebrospinal fluid (CSF) evacuation have failed to control high ICP. Posttraumatic CSF circulation disruption, white matter cerebral edema, and intracranial venous hypertension can be treated with this modality in the absence of focal mass lesions.

45. EFFICACY OF HELMET MODELING FOR DEFORMATIONAL PLAGIOCEPHALY

John M. Whitley, MD, PhD, Ann Marie Flannery, MD, Steve Smith, MD (Augusta, GA)

Cranial asymmetry in children results from different etiologies. Common etiologies include synostoses and deformation (positional molding). Associated with and thought to be secondary to deformational forces, are torticollis and unilateral hip dislocation. The majority of cranial asymmetry in the pediatric population is positional molding. We report the efficacy of helmet modeling in 11 children who presented for craniofacial evaluation at MCG with mild to moderate positional molding. All children were evaluated with skull measurements and radiographs and 3-D CAT scans of the head and neck to rule out synostoses and rotatory subluxation. Motor strength was assessed by physical therapy (PT). Of the 11 patients, 4 had varying degrees of torticollis which contributed to their plagiocephaly. Only one of the four required release of the sternocleidomastoid muscle. PT was carried out in all patients with emphasis on those with torticollis. Modeling helmets were fashioned by a local orthotics company and all children were fitted prior to 9 months of age with the average wear time of 3 months. Follow-up evaluations included photo analysis, skull measurements and 3-D CAT scans. All cases resulted in marked improvement in cranial symmetry. None of the cases progressed or required surgery. Rotatory subluxation was not a contributing factor in any of the cited cases. Based on the results to date, helmet modeling appears to be an effective treatment for mild to moderate positional molding when diagnosed and initiated early.
46. THE ROLE OF THE ASTERION IN LAMBDOIDAL SYNOSTOSIS
James T. Goodrich, MD, PhD, David Jimenez, MD, Ravelo Argamaso, MD, Robert Shprintzen, PhD

With advent of 3-D reformatting of thin slice CT reconstructions it has become evident the important role that the asterion plays in the deformational patterns seen in lambdoidal synostosis. We recently reviewed our series of lambdoidal synostosis (N=21) and found eleven cases with asterion involvement. In five of these cases 3-D reformatting showed partial synostotic patterns. These patients typically had more severe deformational changes in the calvarium including changes in the ear positions and angle of the petrous ridge. In severe cases the TMJ, orbits and forehead shifted following Virchow's Law. These asterion stenosis patterns are not always evident on plain skull x-rays, which are invariably read as normal by the radiologists. It is only on thin slice CT cuts that this asterion pattern of premature suture closure can be determined. Craniofacial reconstruction in these patients requires involving the asterion in the reconstruction. If operated on early, i.e., before one year of age, the esthetic outcomes are excellent including correction of the forehead and ear positions as the child grows. We will present the 3-D CT reformatting images along with reconstruction techniques and long-term outcomes. Lack of appreciation of the asterion's contribution to lambdoidal synostosis can lead to an unsatisfactory outcome in reconstruction.

47. OPERATIVE PREPARATION AND POSITIONING FOR PATIENTS UNDERGOING VARIOUS CRANIOSYNOSTOSIS REPAIRS
Paul C. Francel, MD, PhD, John A. Jane, MD, PhD, John A. Persing, MD, Alonzo Bell (Charlottesville, VA)

Initially, we began our operative procedures with all patients in the supine position. The modified prone procedure was then introduced in order to enable the surgeon to have a full exposure to the entire cranial vault and orbital region at a single setting which is often required in patients that have extensive craniosynostosis. However, this procedure, though being an extremely useful positioning technique for these operations, does have drawbacks; namely, it requires evaluation of the stability of the cervical spine for safety of positioning and also places the patient in a fairly hyperextended position. This is a problem in patients who have syndromic craniosynostoses in which craniovertebral anomalies are sometimes present. We have recently added two new methods for positioning of these patients which has obviated the need for the use of the modified prone positioning because it avoids exposing the patient to extremes of cervical extension. For patients that require supine positioning because of more extensive involvement anteriorly, we have been able to utilize the gelfoam collar technique which enables the entire facial cranial vault and posterior neck regions to be exposed without placing the patient in any position of instability. This procedure does not require the use of external skull fixation. For patients that require more prone positioning for approaching more posteriorly located synostoses, the angled horseshoe technique appears to work extremely well, the patient being placed in a neutral to slightly flexed position during the operation. This position enables a full exposure of the entire posterior cervical and cranial vaults and exposure anteriorly to the orbital rims.

We feel that these new positioning techniques will be quite useful for neurosurgeons performing craniosynostosis repairs, because these techniques enable the physician to have a very large area of exposure for aonestage operation without placing the patient in any risk secondary to craniovertebral instability.
48. THE ROLE OF MICROPLATES IN CRANIOFACIAL RECONSTRUCTION: A FOUR YEAR, MULTICENTER EXPERIENCE

LCDR Robert F. Keating, MC, USN, Bryant A. Toth, MD, Lawrence Shuer, MD (Oakland, CA)

Techniques and principles in craniofacial surgery continue to advance at breathtaking speed since the advent of this discipline in the 1950's. With the introduction of high-speed drills, sophisticated pediatric neuroanesthesia, as well as 3-dimensional imaging, quantum leaps have been made during the past decade. Recently the introduction of microplating techniques have allowed new horizons to unfold.

During the past four years we (Oakland Naval Hospital, Oakland Children's Hospital, and Stanford Medical Center) have had the opportunity to use a microplating system on 131 pediatric patients for craniofacial reconstruction. We have employed the use of low-profile, vitallium as well as titanium plates in the setting of craniosynostosis repair, craniofacial reconstruction for congenital lesions, cranial 1 orbital tumors, as well as facial trauma. The most significant area of use has been with craniofacial and craniosynostosis repair in the pediatric population. These plates and screws have been used effectively at an age less than 2 years (82 children) as well as in the older pediatric patient (49 individuals), especially those that had failed previous craniofacial reconstruction.

The microplates not only provide immediate and firm fixation of the calvarial vault, allowing an unlimited choice of cranial configurations, but more importantly permit significantly shorter surgical times as well as decreased intraoperative morbidity. Despite the use of different plating systems, the long-term results have been consistently favorable with no significant advantages or disadvantages noted between hardware. To date there has been a single infection (easily treated) and a minimal number of complications with the use of the microplates. In addition these plates have not demonstrated any long-term difficulties with respect to skull growth.

Although significant reservations may still exist, the use of microplating techniques in the arena of craniofacial surgery represents a paramount technological advance. In addition to allowing the craniofacial surgeon to proceed in new directions with respect to calvarial reconfiguration, they also permit a safer surgical approach to the patient with a minimum of postoperative morbidity.

49. REVERSAL OF HIGH ENERGY PHOSPHATE METABOLISM CHANGES IN EXPERIMENTAL HYDROCEPHALUS AFTER CSF SHUNTING

M.C. Da Silva, J.M. Drake, C. Lemaire, A. Cross, U.I. Tuor, P. Chumnas, G. Potts (Toronto, Ontario)

We studied the effects of hydrocephalus on the high energy phosphate metabolism (HEPM) of the brain in an experimental model of hydrocephalus and the impact of CSF shunting on these changes. HEPM was analyzed using in vivo 31 phosphorus magnetic resonance spectroscopy (MRS). Hydrocephalus was produced in 1-week-old kittens by cisternal injection of 0.05 ml of kaolin solution, 25% (n=33). Littermates were used as controls (n=16). A ventriculoperitoneal shunt with a distal silt valve was implanted in 16 hydrocephalic animals 10 days after hydrocephalus induction. MRS were obtained 3 weeks after the injection of kaolin.

Nontreated hydrocephalic animals presented a marked dilatation of the lateral ventricles and periventricular edema. MRS showed a significant decrease of the energetic index ratio phosphocreatine/inorganic phosphate (PCR/PI) (p=0.004) and an increase of the relative concentration of inorganic phosphate (Pi/ATP) (p=0.01).

Animals with a functioning shunt had a decrease of their ventricular size, compared to before shunt implantation, with no periventricular edema. PCR/PI and Pi/ATP ratios were similar to those of control animals (p=0.3, p=0.9, respectively) and different from nontreated hydrocephalics (p=0.04 , p=0.03, respectively). Some hydrocephalic animals did not show an improvement of the PCR/PI and Pi/ATP ratios after shunting, even though the shunt system was functioning properly and the ventricles were markedly decreased.

This study demonstrates CSF shunting is able to reverse the changes in HEPM caused by hydrocephalus in this experimental model. It also suggests there may be a cutoff point, after which the damage is irreversible.
50. LATE DECOMPRESSION DOES NOT RESTORE PROTEIN SYNTHETIC CAPACITY IN NEONATAL HYDROCEPHALIC KITTENS

James P. McAllister II, PhD, Christopher L. Wolfgang, MS, Karin Bierbrauer, MD (Philadelphia, PA)

While several studies have described the morphologic characteristics of cortical neurons in hydrocephalic animals, evaluations of the cellular functions of these neurons are limited. Therefore, the present study has utilized cytophotometric techniques to assess the protein synthetic capacity of cerebrocortical neurons in hydrocephalic kittens, as well as animals that underwent "early" or "late" decompression via ventriculoperitoneal shunts. Two groups of kaolin-induced hydrocephalic kittens were allowed to progress to moderate and severe stages of hydrocephaly, after which surgical decompression was performed to yield the early and late shunt groups. Hydrocephalic, shunted and normal age-matched controls were sacrificed to correspond to both the preshunt condition and a 7 day postshunt survival period. Histologic sections of primary visual, parietal association and primary motor cortices were stained with azure B for the stoichiometric binding of perikaryal RNA and nucleolar volume measurements. Cytophotometric analysis of superficial and deep layers from these cortical regions revealed a significant depletion in cytoplasmic RNA (6-48% decrease) during early stages of hydrocephalus, and a more consistent decrease (19-48%) with more severe hydrocephalus. A concomitant decrease in nucleolar volume occurred during early (19-34%) and late (16-74%) hydrocephalus. The general pattern of diminished protein synthetic capacity was relatively uniform, and thus may correlate more with a global decrease in cortical blood flow than neuronal damage. Early decompression resulted in a return of protein synthetic capacity to values that were up to 37% higher than control levels. However, the late shunted counterparts remained 26-52% below controls in neuronal RNA levels and 6-45% low in nucleolar volume. Since the normal function of neurons is dependant on the continual supply of perikaryal RNA, it follows that cerebrocortical neuronal function is adversely affected by hydrocephaly, and may not be restored by late decompression.

51. EFFECTS OF EXPERIMENTAL INFANTILE HYDROCEPHALUS ON BASAL FOREBRAIN AND HIPPOCAMPUS

Richard M. Kriebel, PhD, James P. McAllister II, PhD (Philadelphia, PA)

The neurological deficits found in infantile hydrocephalus have most often been explained by pathologic changes in cerebral neocortex. Since alterations in the basal forebrain and hippocampus could cause cognitive deficits, we have extended our ongoing work to include these subcortical areas. Kaolin injection induced obstructive hydrocephalus in a neonatal kitten model. The basal forebrain was divided according to standard atlas designations and planimetrically analyzed with a Biocuant system. Decrease in volume was observed in all areas of the basal forebrain; these changes were only partially normalized in decompressed brains. There was a decrease in ChAT labelled neurons and severe neuropil degeneration of the basal forebrain region, especially septal nuclei. These studies suggest that basal forebrain as well as cortex should be considered in mechanistic explanations of the neurological deficits seen with this disorder. Additional studies were done to examine the structural changes in the hippocampal cortex, a primary target of septal projections. Preliminary cell counts show 70% of hippocampal pyramidal neurons in varying stages of degeneration, from densification of cytoplasm to fully pyknotic. The pyramidal laminae showed hydroptic cellular degeneration, although there was no indication of edematous extracellular space. The number of synaptic contacts was decreased. In contrast, the thickness of the hippocampus was not decreased, and the ependymal covering of the hippocampus was intact. The apparent structural integrity of the hippocampus has led to the suggestion that the neuronal degeneration seen in the hippocampus may have resulted from the deficiency in basal forebrain innervation.
52. MR VISUALIZATION OF THE OLFATORY PATHWAY OF CEREBRAL SPINAL FLUID FLOW IN THE DOG.

Michael J. Burke, DVM, MD, David Symonds, MD (Denver, CO)

Past investigators have demonstrated the presence of cerebral spinal fluid around olfactory nerve fila and its passage into sub mucosal perineural lymphatics. This was demonstrated in animal models by histologic examination of the cribriform region after subarachnoid injection of particulate matter. The anatomic detail and lack of bone artifact afforded by MRI prompted the current investigation in an attempt to demonstrate this alternate pathway of cerebral spinal fluid flow in vivo.

Adult healthy dogs were anesthetized routinely. Pre-injection T1 and T2 weighted MR cranial images were obtained. 10 mmol of gadolinium DTPA was then injected into the cisterna magna. T1 and proton density MR cranial images were obtained one and three hours post injection. Linear extensions of contrast were seen to extend beyond the cribriform plate. After three hours a diffuse enhancement of the sinus mucosa was noted.

The dogs tolerated the procedure without sequelae and remained neurologically normal. We believe these contrast images demonstrate not only the subarachnoid space around olfactory nerve fila rostral to the cribriform plate but also cerebral spinal fluid absorption (enhancement) via sinus mucosa.

Alternate pathways of cerebrospinal fluid absorption could be studied on spinal nerve using this technique. Future studies might be directed in determining alternate pathway flow rates.

53. CURRENT APPLICATIONS OF ENDOSCOPY TO VENTRICULAR SHUNT

Kerry R. Crone, MD (Cincinnati, OH)

Renewed interest in neuroendoscopy has resulted from recent advances in scope miniaturization, illumination and video image processing. To date, 250 endoscopic procedures have been performed including 65 initial shunt procedures, 25 proximal shunt revisions, 140 membrane fenestrations, and 20 tumor biopsies/excisions.

Preliminary results demonstrate a 3% yearly shunt malfunction rate following endoscopic ventricular catheter placement, 0% shunt malfunction rate in proximal shunt revision, membrane patency of 96% following fenestration and histologic diagnosis of ventricular tumors in all cases.

Complications have included four patients with ventricular hemorrhage requiring external ventricular drainage, two patients with single cranial nerve palsies, one patient with a thalamic contusion and one patient with a cerebral vascular accident. All problems resolved within three months of the procedure except one patient with a persistent third nerve palsy.

Our preliminary experience suggests that endoscopy warrants continued investigation to establish its application to ventricular surgery. Discussion will include endoscopic instrumentation, energy delivery for hemostasis and examples of each procedure.
54. INITIAL CLINICAL EXPERIENCE WITH VENTRICULOSCOPY IN THE PEDIATRIC POPULATION

   Joseph Petronio, MD, Marion L. Walker, MD, Lyn C. Wright, MD (Salt Lake City, UT)

In order to better delineate the clinical applicability of ventriculoscopic surgery in the pediatric population, we conducted a retrospective review of all children undergoing ventricular endoscopy at Primary Children's Medical Center of the University of Utah. Since July, 1987 we have performed 56 ventriculoscopic procedures on 41 children, ranging in age from 3 days to 18 (median, 2.68) years at the time of initial intervention.

The largest group of patients treated included those with unilateral or bilateral multi-compartmental hydrocephalus, usually secondary to severe intraventricular hemorrhage of prematurity or recurrent ventriculitis.

In this group of patients, 30 ventriculoscopic procedures were performed in 18 patients, usually for fenestration of multiple synechiae or septations. This group had the highest failure rate when measured in terms of the radiographic recurrence of loculated collections and the need for additional ventricular catheters and complex shunts. While unilateral or focal compartmentalizations were associated with a more favorable outcome following ventriculoscopic fenestration (p), only 6 out of 18 patients were felt to have had a "satisfactory" or "good" outcome when measured in terms of radiographically demonstrable reduction in ventricular size and clinical stability.

A more favorable result with ventriculoscopic fenestration was seen with the 14 procedures performed on 13 patients with focal arachnoid cysts, including cysts of the septum pellucidum (n=5), suprasellar cistern (n=3), and interhemispheric fissure (n=2). In this group, a "satisfactory" or "good" outcome was obtained in 9/13 patients treated endoscopically, including complete shunt independence in 9/13 patients.

Lastly, in a heterogeneous group of 8 shunt-dependent patients treated with ventriculoscopic fenestration of the lamina terminalis or floor of the third ventricle using the neodymium:YAG laser, 3 have remained shunt-independent postoperatively.

This preliminary experience suggests a useful therapeutic role for ventricular endoscopy with currently available technical adjuncts, including CT- or MR-guided stereotactic targeting and fiberoptic laser delivery systems in managing certain hydrocephalic afflictions of children.

55. THE DIAGNOSIS AND TREATMENT OF TRAPPED FOURTH VENTRICLE

   James R. B. Nashold, MD, W. Jerry Oakes, MD (Durham, NC)

The trapped fourth ventricle is an unusual but potentially fatal complication of hydrocephalus. Of twenty-four patients diagnosed with a trapped fourth ventricle, twenty received shunting of the fourth ventricle with nineteen patients (95%) showing improvement or remaining asymptomatic. The highest risk factor for the development of a trapped fourth ventricle was intracranial hemorrhage in premature infants 14/24 (58%). Other etiologic risk factors included meningitis (46%) and Chiari II malformation 4/24 (17%). The symptoms attributable to the trapped fourth ventricle included headaches (25%), nausea and vomiting (25%), dysconjugate gaze (20%), dysphagia (20%), ataxia (15%), extremity paresthesias (10%), opisthotonus (10%), increased spasticity (5%), bradycardia (5%), apnea (5%) and cyanosis (5%). Treatment included shunting of the fourth ventricle by inclusion with the existing lateral ventricular shunt system or in the case of Chiari II malformations shunting into the cervical subarachnoid space. Twenty-two revisions were required in twelve patients with an average follow-up time of 18.3 months. Shunt revision was more likely in the patients with a post-meningitic trapped fourth ventricle 11/6. Six revisions were necessary in four patients with Chiari II malformations. Three post-operative complications occurred in thirty shunt procedures of the fourth ventricle; one cerebellar hemorrhage, one infection (Staphylococcus Epidermidis) and one shunt tubing disconnection. No fatalities occurred. Follow up ranged from three months to twelve years (mean 3.8 years). Because of the potential deterioration of patients with a significant trapped fourth ventricle, recognition and treatment is recommended to reverse the signs of localized increased intracranial pressure and brain stem compression. The technique of posterior fossa catheter placement will be discussed.
56. INITIAL MANAGEMENT OF CONGENITAL AND ACQUIRED HYDROCEPHALUS WITH A HIGH PRESSURE VALVE

James Holsapple, MD, Leo Hochhauser, MD, Jeffery A. Winfield, MD, PhD (Syracuse, NY)

Slit ventricle syndrome and intermittent proximal shunt obstruction are frustrating problems associated with ventriculoperitoneal shunts in children with congenital or acquired hydrocephalus. These problems are likely to result from "overshunting" and "collapse" of the ventricular system which might be avoided if the ventricles could remain expanded at physiological pressures. One approach to preventing these problems would be to use relatively high pressure valve systems at initial shunting. In this report 60 consecutive children with hydrocephalus were shunted by the senior author using a high pressure PS MED valve. All etiologies of congenital hydrocephalus were represented, and in those infants with Dandy-Walker Syndrome, the fourth ventricular cyst was also shunted with a separate high pressure system. The most frequent etiology of acquired hydrocephalus was intraventricular hemorrhage in the premature infant. At a mean follow-up period of 3 years the following were determined: 1) the radiographic appearance of the ventricular system and skull (presence or absence of slit ventricles and ventricular asymmetry, biparietal and AP ratio of the head), 2) the incidence of slit ventricle syndrome, 3) the failure rate of shunts (Kaplan-Meyer survival curves), 4) the relationship of observed to expected head circumferences, and 5) overall outcome (developmental progress). The results suggest that initial placement of a high pressure valve prevents the development of slit ventricles and their complications (slit ventricle syndrome, shunt malfunction and multiple revisions). Furthermore, the use of a high pressure does not appear to induce abnormal head growth/shape or superimposed developmental delays.

57. A SEARCH FOR DETERMINANTS OF CSF SHUNT SURVIVAL: RETROSPECTIVE ANALYSIS OF A 14-YEAR INSTITUTIONAL EXPERIENCE

Joseph H. Piatt, Jr., MD (Portland, OR)

All CSF shunt operations performed at Oregon Health Sciences University from 1976 through 1989 were reviewed retrospectively. There were 671 insertions or revisions of simple, linear CSF shunt systems. Outcomes from these operations were studied using statistical techniques for analysis of survival data. The overall estimated median survival of CSF shunts was 73 mo.

The only significant determinant of risk of obstruction was the age of the patient: Children less than 2 years old were at higher risk than older patients (p<0.00005). The following factors had no significant influence on risk of obstruction: attending surgeon, etiology of hydrocephalus, duration of operation, time of day of operation, and whether the shunt was new or revised. Totally revised shunts lasted no longer than partially revised shunts. A very slight trend favoring parietal sites over frontal sites was nullified by stratification by age group. In children less than 2 years old, trends were observed favoring new shunts over revised shunts (p=0.0355) and flat bottom valves over Hakim valves (p=0.0255).

The risk of shunt infection by 1 year was 8.5%. The following factors had no significant influence on the risk of infection: patient age, attending surgeon, etiology of hydrocephalus, duration of operation, time of day of operation, and whether the shunt was new or revised.

Duration of CSF shunt function is strongly related to the age of the patient at the time of surgery. Studies of surgical technique and shunt design must incorporate stratification of patients by age.
58. FACTORS EFFECTING THE NEED FOR VENTRICULOOPERITONEAL SHUNTS FOLLOWING POSTERIOR FOSSA TUMOR SURGERY: A RETROSPECTIVE ANALYSIS OF 117 PATIENTS

Mitchel S. Berger, MD, Deborah Culley, Russell Geyer (Seattle, WA)

This study reviewed 117 pediatric patients with posterior fossa tumors treated at Children's Hospital Medical Center, Seattle, from 1976-1990, in an attempt to determine what perioperative and intraoperative factors influenced the need for postoperative shunts. Ages in our patient population ranged between 4 months and 16 9/12 years. Factors evaluated for statistical significance include: age at diagnosis, duration of symptoms, extent of hydrocephalus, tumor location, extent of tumor resection, presence and duration of external ventricular drain (EVD), flow of cerebrospinal fluid (CSF) through the fourth ventricle at the conclusion of tumor resection, presence of hemostatic cavity linings, method of dural closure, tumor type, wound/CSF infections and leaks, as well as pseudomeningocele formation. Of these variables, young age at diagnosis, tumors affecting midline structures, subtotal tumor resection as determined by immediate postoperative scans, prolonged requirement of an EVD, cadaveric dural grafts, pseudomeningocele formation and wound/CSF infections were statistically significant predictors for postoperative shunt placement. Postoperative shunts were required in 36% of all patients regardless of histology.

Therefore, in considering the pertinent factors that may influence the need for postoperative shunt placement in children with posterior fossa tumors, the surgeon should strive for a radical tumor resection, especially with midline tumors, verified by immediate postoperative scanning, meticulous closure of the "dura" and wound to avoid a significant pseudomeningocele formation, as well as the avoidance of foreign body dural substitutes and other potential causes of an enhanced CSF inflammatory response, e.g. wound infection, to minimize the need for shunt placement following posterior fossa surgery.

59. ANAEROBIC GLYCOLYSIS — THE PRECURSOR OF PORENCEPHALIC CYST FORMATION IN HYDROCEPHALUS?


Marked hydrocephalus is associated with periventricular white matter oedema and porencephalic cyst formation but with relative sparing of the grey matter structures. In an attempt to further elucidate these white matter changes and determine their reversibility, we examined cerebral glucose utilisation using 2-deoxy glucose autoradiography in a neonatal kitten model of hydrocephalus (kaolin). In addition, sections were also taken and stained for cytochrome oxidase and H&E.

The mean cross-sectional brain area of the hydrocephalic kittens was 140% that of the controls. The mean ventricular area in the frontal region was 24% and 34% in the occipital region. There was increased white matter damage and porencephalic cyst formation in the occipital region compared to the frontal region. The porencephalic cysts were situated in the middle of the white matter.

There was no significant difference between the hydrocephalic kittens and controls in glucose utilisation in the 12 grey matter regions measured. In contrast, there were marked changes in the white matter regions and the porencephalic cysts had a significantly lower glucose utilisation (mean 4.6μmol/100g/ml) than normal white matter (mean 17.5μmol/100g/ml). Surrounding the cysts was a halo of increased glucose utilisation. In some animals it was apparent that this increase in white matter glucose utilisation actually preceded cyst formation.

Density studies from sections stained for cytochrome oxidase revealed no difference in grey matter values but a significant decrease in density in the white matter of the hydrocephalic kittens, indicating a decrease in oxidative metabolism. Interestingly, there was no halo of increased activity in the white matter to correspond to the findings from the glucose utilisation suggesting that the latter may represent anaerobic glycolysis. In fact, the pattern of white matter glucose utilisation seen in this study is remarkably similar to that seen after middle cerebral artery occlusion in cats and may be further evidence of chronic ischaemia in hydrocephalus. Preliminary studies with VP shunting after 10 days of hydrocephalus shows preservation of the white matter and a normal glucose utilisation pattern suggesting that this white matter damage is preventable at least at an early stage.
60. SIGNIFICANCE OF BROTH ONLY OR ANAEROBIC POSITIVE VENTRICULAR CSF CULTURES
Jeff Chen, MD, J. Gordon McComb, MD, Corey Raffel, MD, PhD (Los Angeles, CA)

To determine the significance of either broth only or anaerobic positive ventricular CSF cultures, a retrospective analysis of the CSF obtained at shunt related operative procedures from 1988 to 1990 was conducted. Six hundred nine samples of ventricular CSF were obtained and analyzed. There were 15 shunt infections for an infection rate of 2.5% per procedure. Of 44 cultures that were positive for growth in the broth media only, 26 (59%) patients remained asymptomatic postoperatively. Fourteen (32%) patients required subsequent revisions but clinically did not manifest any evidence of a shunt infection. In each case, subsequent CSF samples were sterile. The mean number of shunt revisions for these 14 patients did not vary significantly from the group as a whole. Four (9%) broth only positive culture patients did develop clinical evidence of a shunt infection. Although the CSF WBC counts were not or only slightly elevated, a WBC differential found marked eosinophilia (27%, 50%, 70%, 82%).

Only 2 ventricular CSF samples grew an anaerobic organism. Neither patient demonstrated clinical evidence of shunt infection, nor did either shunt need to be revised.

In conclusion, it is not cost effective to send ventricular CSF obtained at the time of shunt insertion or revision for routine anaerobic cultures. The vast majority of broth only positive cultures appear to be contaminants. Only in the presence of CSF eosinophilia should there be a high index of suspicion for shunt infection.

61. CONTRALATERAL TRANS-SEPTAL PLACEMENT OF VENTRICULAR CATHETERS TO PREVENT POST-SHUNTING VENTRICULAR ASYMMETRY: A RANDOMIZED STUDY
Paul Steinbok, MD, Kenneth Poskitt, MD, D. Douglas Cochrane, MD (Vancouver, BC)

Ventricular asymmetry is common after ventricular shunting, and occasionally isolation of the contralateral ventricle necessitates a second shunt. The following study was performed to determine whether contralateral placement of the ventricular catheter via ultrasound guided perforation of the septum pellucidum would decrease the incidence of post-shunting ventricular asymmetry.

58 children with hydrocephalus and an open fontanel, who were having a ventriculo-peritoneal shunt via a parieto-occipital approach, were randomized to have the ventricular catheter tip placed into either the ipsilateral or contralateral frontal horn. Of 33 patients randomized to contralateral placement, in only 25 was the catheter tip successfully located contralaterally, and in 6 of 25 randomized to ipsilateral placement, the catheter ended up in the contralateral ventricle.

Ventricular asymmetry occurred in 27% of patients with contralateral versus 50% with ipsilateral placement. In 7 patients with contralateral placement, in whom a special catheter with 2 sets of holes was used, such that holes were located in both ventricles, ventricular asymmetry occurred in 1 patient, and this was related to pre-existing cerebral tissue loss. In 2 children with ipsilateral placement, a second shunt was required for a symptomatic isolated lateral ventricle. 15 of 33 patients randomized to contralateral placement had shunt revisions, compared to 9 of 25 in the ipsilateral group.

Perforation of the septum pellucidum with placement of the ventricular catheter in the contralateral ventricle decreases the incidence of post-shunting ventricular asymmetry, but there may be a higher incidence of shunt revision associated with actual or attempted septal perforation.
62. AN ADJUSTABLE ENCAPSULATED ANTI-SIPHON DEVICE

J.M. Drake, R. Fox, M.C. da Silva (Toronto, Ontario)

Anti-siphon devices were designed to address the problems created by CSF over drainage in shunted hydrocephalic patients in the upright position. Both the original Anti-siphon Device and more recently modifications of this design marketed as the Siphon Control Device and Delta Valve have been demonstrated to eliminate negative pressures in shunt systems during laboratory testing. Limited testing in patients with Anti-siphon devices has also demonstrated reduction of negative intracranial pressure in the upright position.

However, common to all these devices is the requirement that the pressure external to the device be atmospheric in order for the device to function properly. Increase in the local external pressure by placement in a barometric chamber leads to a linear increase in the resistance to flow. Subcutaneous implantation in experimental animals leads similarly to increased resistance from local tissue capsule formation. Functional obstruction from tissue capsule formation has been demonstrated in a single patient and may explain the high incidence of functional obstruction of anti-siphon devices in some series.

We report the design of an anti-siphon device which is encapsulated with a gas filled rigid container. This allows normal anti-siphon device function unaffected by capsule formation, overlying scar or muscle tissue, etc. The volume of the gas can be manipulated to increase or decrease the internal pressure so that the resistance of the device can be correspondingly increased or decreased over a wide range. This manipulation can be carried out without transgressing the skin so that the device is externally adjustable.

Initial prototype construction has confirmed the feasibility of the design. It has also demonstrated the problems related to construction of gas-impermeable flexible membranes. An adjustable encapsulated anti-siphon device is a possible solution to the problems encountered by current anti-siphon devices.

63. MDR1 GENE EXPRESSION IN THE BRAINS OF MEDICALLY INTRACTABLE EPILEPTICS

Corey Raffel, MD, PhD, David M. Tishler, Kenneth I. Weinberg, David R. Hinton, Nicholas Barbaro (Los Angeles, CA)

Why some patients with seizures are successfully treated with anticonvulsant drugs and others prove medically intractable is not known. The multiple drug resistance gene (MDR1) encodes P-glycoprotein, an energy-dependent efflux pump that exports planar, hydrophobic molecules from the cell. Normal brain parenchyma does not express MDR1, but brain capillary endothelium does. Some anticonvulsant drugs have chemical structures consistent with a substrate suitable for export by P-glycoprotein. In this presentation, phenytoin is shown to be a substrate for P-glycoprotein in vitro. We examined MDR1 expression using polymerase chain reaction for MDR1 mRNA in 19 specimens of brain removed during operations for medically intractable seizures. Eleven of the 19 specimens had MDR1 levels more than 10 times that found in normal brain. Increased expression of P-glycoprotein in capillary endothelium and, in specimens with high MDR1 mRNA levels, in astrocytes was identified immunohistochemically. These results suggest lack of response to medication in some epileptics may be caused by inadequate accumulation of anticonvulsant in the brain.
64. LESIONECTOMY IN THE MANAGEMENT OF CHILDREN WITH INTRACTABLE EPILEPSY

J.L. Montes, MD, B. Rosenblatt, MD, J.P. Farmer, MD, A. O'Gorman, MD, F. Andermann, MD, G. Watters, MD (Montreal)

The results of radical excision of lesions detected by M.R.I. in children presenting with epilepsy are analyzed. There were 14 boys and 4 girls with a mean age of 9.2 years. The average age of onset of seizures was 6.8 years. The average time between onset of seizures and surgery was 2.3 years. Often C.T. scans suggested that the lesions were indolent. M.R.I. was more helpful in differentiating between neoplastic and developmental lesions. Angiography was noncontributory. The interictal E.E.G. showed epileptiform activity correlating with radiological studies in 54% of the cases.

The lesion was radically, surgically removed in all but one, of these patients. This was confirmed by intra-operative ultrasound and post-operative imaging. The average post-operative follow-up is 4.3 years. Five patients had astrocytomas, all low grade. Five others had gangliogliomas. One each mixed astrocytoma—oligodendroglioma, infantile desmoplastic ganglioglioma and cavernous angioma. Three had cortical dysplasias and two hamartomas.

Fifteen patients have been seizure-free since surgery. Two others had only auras. The patient with incomplete resection continues to have seizures. Essentially, all patients benefited from the seizure point of view. In the group with temporal lobe lesions, improvement in I.Q. was seen post-operatively. Early consideration of surgery in patients with lesions demonstrated by M.R.I. and difficult to control epilepsy is suggested.

65. CERVICOMEDULLARY COMPRESSION IN ACHERONDEPLASIA

Timothy Ryken, MD, Arnold H. Menezes, MD (Iowa City, IA)

Six patients with achondroplasia and symptoms suggestive of cervicomedullary compression are reviewed. Four females and two males with an average age of nine years were evaluated with a mean duration of symptoms prior to intervention of 1.9 years. Symptoms included occipitocervical pain, ataxia, incontinence, apnea and respiratory arrest. Radiological evaluation included plain films with flexion extension views, pluridirectional tomography, thin section computed tomography and magnetic resonance imaging. Typical findings included marked foramen magnum stenosis, ventral cervicomedullary junction compression secondary to basilar invagination and dorsal cervicomedullary junction compression secondary to ligamentous hypertrophy and invagination of the posterior atlantal arch. All patients underwent posterior fossa decompression and atlantal laminectomy. Operative findings consistently revealed marked hypertrophy of the posterior rim of the foramen magnum with thickening and invagination of the atlantal posterior arch and a dense fibrotic epidural band resulting in dorsal cervicomedullary compression. Intraoperative ultrasound was used to determine the extent of decompression required. Four patients required duroplasty. Two patients had concurrent hydrocephalus, one of which required ventriculoperitoneal shunting. One patient developed a pseudomeningoecele postoperatively requiring serial lumbar punctures with resolution. No patient developed craniovertebral instability following decompression. Improvement or resolution of symptoms was noted in all patients with an average follow-up of 5.6 years. Thus, cervicomedullary compression in patients with achondroplasia can be successfully treated with dorsal decompression of the craniovertebral junction. Dense epidural fibrotic bands are frequently noted and must be aggressively released to ensure satisfactory decompression.
66. TRAUMATIC ATLANTO-OCcipital DISLOCATION

David J. Donahue, MD, William Wamer, MD, Michael Muhlauer, MD, Robert A. Sanford, MD, Robert A. Kautman, MD (Memphis, TN)

Traumatic atlanto-occipital dislocation (AOD) is a rare and frequently fatal disorder. Death is usually due to immediate respiratory arrest from pontomedullary tearing, vascular compromise and brain stem ischemia or associated cranial injuries. The mode of injury is usually vehicular trauma but other presentations have been described. The apparent high incidence in children may be due to biochemical properties peculiar to the pediatric spine.

Upon reviewing the literature, we found only 11 children who survived traumatic AOD greater than 24 hours. The improvement of emergency medical services may account for the increasing reports of traumatic AOD survivors arriving at emergency departments.

We present 3 children (10 years of age or less) with AOD who presented to the Neurosurgical service over the past 18 months. Two were struck by motor vehicles; the third sustained a hyperextension "clothesline injury" as she rode a fourwheeler under a steel cable. All 3 required immediate treatment for a concomitant closed head injury and eventually underwent occipit-C1 fusion. Two developed post hemorrhagic communicating hydrocephalus necessitating a ventriculoperitoneal shunt. Outcome has been good with 2 children having mild deficits and 1 child has no neurologic sequelae.

The authors will discuss the mechanism of injury, review radiologic criteria for diagnosis, survey previously reported cases and describe their management and complications of traumatic AOD.

67. TREATMENT OF PEDIATRIC SPINAL ANEURYSMAL BONE CYSTS WITH SELECTIVE ARTERIAL EMBOLIZATION

James D. Callahan, MD, John E. Kalscheek, MD, Thomas G. Luerssen, MD, Joel C. Boaz, MD, G. Paul DeRosa, MD (Indianapolis, IN)

Aneurysmal bone cysts are relatively uncommon, benign, locally aggressive tumors of the bone that arise primarily in the pediatric population. Conventional treatment has been resection or curettage and bone grafting, and possibly radiotherapy. Arterial embolization has been described as a preoperative adjunct to surgical resection, and isolated treatment in select cases. The incidence of spinal lesions ranges from 3% to 20% in published series.

We are presenting a series of 3 complex aneurysmal bone cysts of the spine treated with selective arterial embolization. Two lesions involved the lumbar spine and one the cervical spine. All the patients were children ages 8, 13, and 14. They all presented with complaints of pain. Two out of the three patients had neurologic deficits consistent with cauda equina compression and myelopathy, respectively. All 3 lesions were biopsied for diagnostic purposes. Due to the size of the lesions, involvement of multiple levels, and involvement of both anterior and posterior elements, complete surgical resection was felt not to be possible. All underwent selective arteriography and embolization under general anesthesia. Post embolization all 3 patients had relief of their pain and resolution of existing neurologic deficits. Follow-up ranges from 2 1/2 to 4 1/2 years with all of the patients remaining neurologically intact and pain free. There has been progressive shrinkage and mineralization of the cysts seen on computed tomography. There has been no evidence of spinal instability. We feel embolization offers a safe effective treatment modality for those lesions not amenable to surgical resection.
THE IMPACT OF OSTEOPlastic LAMINOTOMY ON THE DEVELOPMENT OF POSTOPERATIVE KYPHOsCOLIOSIS

Fred J. Epstein, MD, Diana Gruia-Leahu, Jeffrey H. Wisoff, MD, Rick Abbott, MD (New York, NY)

This is a retrospective clinical study of the impact of osteoplastic laminotomy on the development of postoperative kyphoscoliosis. We reviewed 30 patients that underwent one surgery at NYU, for intraaxial spinal cord tumors. The patients were under 16 years of age at the time of their surgery and none of them received adjunctive therapy. Two study groups were contrasted; one which underwent osteoplastic laminotomy versus the group that had a classical laminectomy. The presence of pre-operative and post-operative spinal deformity was recorded from the patients' charts and by phone inquiry of their family and/or primary physician. We attempted to correlate the occurrence of post-operative spinal deformity with several parameters such as: the patients' age, sex, duration of symptoms, extent and site of laminectomy and motor score before and after the surgery. Our preliminary results show no significant correlation between the two groups of patients in terms of development and/or aggravation of post-operative kyphoscoliosis and the type of surgical technique involved. Our opinion is that the major risk factor for progressive post-operative kyphoscoliosis is the severity of pre-operative kyphoscoliosis. Special attention will be given to very young age patients that underwent extensive laminectomy and presented post-operatively with severe spinal deformity.
1. THE INVESTIGATIONAL USE OF POWER SPECTRAL ANALYSIS TO QUANTITATE CERVICOMEDULLARY COMPRESSION IN CHIARI MALFORMATIONS

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It is well known that children with Chiari Malformations exhibit autonomic dysfunction when compression of the cervicomedullary junction occurs. Power Spectral Analysis of heart rate variability is a method of directly measuring autonomic efferent cardiac activity. Ultimately, we believe this method can be employed to assess children with Chiari Malformations. The results are, in theory, a quantitative measure of the degree of compression on the cervicomedullary junction.

Power Spectral Analysis of heart rate variability demonstrates low-frequency (0.01 - 0.15 Hz) and high-frequency (0.15 - 1.00 Hz) heart rate power which are measures of parasympathetic and sympathetic efferent cardiac activity during stress, respectively. Preliminary data in adults with Chiari Type I Malformations and cough headache/syncope reveal preoperatively a low resting sympathetic activity and a high resting parasympathetic activity when compared to normal controls. Postoperatively, after decompressive procedures, they show a return to more normal levels of activity.

We speculate that future data collected by this method will be relevant for children and be useful for both clinical assessment and further research endeavors. Baseline data will be available for following patients clinically, as well as for comparison after decompressive procedures.

2. A MORPHOLOGIC AND HISTOCHEMICAL STUDY OF CALVARIAL BONE IN THE GROWING RABBIT: IMPLICATIONS IN MULTIPLE SUTURE CRANIOSYNOSTOSIS

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The authors have previously reported their observations on normal adult calvarial bone stained histochemically to identify osteoclasts and osteoblasts. In that work we found osteoclasts only within the diploic space. This led us to question the mechanisms involved in the formation of craniolacunia and internal calvarial scalloping. We present our findings herein.

Calvarial bone from patients with multiple synostosis removed during craniotomy for craniofacial procedures was processed for acid and alkaline phosphatase activity. An internal bony lamina was absent in areas of craniolacunia. That is the diploic space communicates with the dural surface. When these sections were stained for acid phosphatase, osteoclasts were found on the dural surface. Thus craniolacunia form via dural surface resorption of bone and this morphology may be recognized as pathologic.

These observations prompted an evaluation of fetal and neonatal rabbit calvarial bone. Similar histochemical staining was performed on these tissues. Fetal calvarial plates are initially composed of a single periosteal bony lamina and dural surface lined with osteoblasts. With further development the internal (dural) bony lamina forms thus enclosing osteoclasts within the diploic space.

Calvarial bone in multiple synostosis has a "fetal morphology" in contrast to normal bone or bone in patients with single suture synostosis. Multiple synostosis represents more than multiple single sutural synostosis.
3. ORBITOCRANIAL OPERATIVE TECHNIQUE IN CHILDREN
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Skull lesions in the parasellar area have been approached by many techniques, including the bicoronal subfrontal approach, the unilateral peritoneal approach, and even the transsphenoidal approach. With the introduction of the orbitocranial approaches the removal of the supraorbital bone and the frontozygomatic margins have allowed minimal brain retraction and shortened the distance to the targeted lesions. Most of the previous technical descriptions of these procedures have stressed the importance of preserving the facial nerve, the supraorbital nerve, and the vasculature of the pericranial and scalp flap. The anatomy of the child's skull and soft tissues, however, differs significantly from that seen in the adult. Recognizing the absence or immaturity of cranial sinuses, the pediatric neurosurgeon must weigh several considerations specific for children when choosing an orbitocranial approach. These considerations include the preservation of bony growth plates and the protection of connective tissue-fascial planes. The presence of vascular conchal sinuses bones may further modify the standard technique.

We are presenting our experience in a combined total of 30 cases of orbitocranial approaches in children less than 15 years of age. Specific recommendations regarding preoperative evaluation, operative positioning, and anatomic surgical considerations will be provided in this presentation.

4. CRANIOFACIAL HOLOGRAPHY WITH PULSED RUBY LASER
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The next technological challenge in imaging will be to capture subjects as they exist in three dimensions. As different imaging technologies are evolving toward this end, one such area of development in three dimensionality is holography. Although holographic principles were introduced in 1947, practical applications within medicine have never been realized. Recent refinements in the original concept of the pulsed wave laser makes it now possible to image previously unsuitable objects including those that are living and not movement free.

Holograms of infants with craniofacial anomalies provide information that even the best photographs can only infer. Preoperative and postoperative holograms record soft tissue in a manner that no presently employed technique can. True, three dimensionality can only be depicted in holography because it uniquely images volume. Although CT and MRIs are mandatory in planning the surgical approach, holography assists and also evaluates the outcome of this plan. No matter how well the internal anatomy is realigned, the end result of the procedure in craniofacial surgery is evaluated based on the patients final external physical features. Holography provides a permanent three dimensional record of this, long after such features are changed by the maturation process.

The pulsed ruby laser produces high resolution images of biological material in seconds that are viewable using a domestic spot lamp. The holograms can be utilized for pre-operative preparation, post-operative assessment and a teaching tool.
5. CURRENT NEUROSURGICAL TECHNIQUES FOR REPAIR OF SAGITTAL CRANIOSYNOSTOSIS

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Complete synostosis of the sagittal suture results in biparietal narrowing without abnormal squamous temporal convexity and a compensatory bulging in the frontal and occipital regions in a bilateral fashion. Patients can also have restricted sagittal synostosis, i.e., either anterior or posterior aspects of the sagittal suture being involved. How one intends to correct the sagittal synostosis depends importantly on the age of the patient, for different techniques need to be used whether the patient is less than one year of age (particularly in the first few months of life), one to three years of age, or greater than three years of age. In addition, which major morphologic features are accentuated in each individual case of sagittal synostosis determines the particular operative technique that is employed.

In this report, we present several novel operative techniques for the repair of sagittal synostosis. For complete sagittal synostosis, we have in the recent past utilized a bifrontal/biparieto-occipital/bilateral temporal cranietomies with radial remodeling and barrel stave osteotomies. More limited versions of this technique, including the Y shaped frontal technique have been commonly employed for anterior sagittal synostosis, whereas for posterior sagittal synostosis a more limited biparietal-occipital cranietomy and adjacent biparietal craniotomies are performed with radial remodeling and barrel stave osteotomies. More recently, we have utilized several new approaches for the repair of sagittal synostosis which create a more cosmetically acceptable surgical result in our patients. In particular, we have added a "double figure of 8" procedure which is utilized in patients who have total sagittal synostosis. In addition, for anterior sagittal synostosis, we have continued to use the Y technique or the pi (π) technique, but have recently added the T technique as a variation on the previous pi procedure. The inverted Y technique can be used for repair of posterior sagittal craniosynostosis.

We feel that these various operative procedures provide the necessary corrections for the repair of this craniosynostosis abnormality and enables the surgeon to tailor his repair to the morphologic abnormalities that are more accentuated in a particular patient in order to achieve the best operative result.

6. TREATMENT OF NONSYNOSTOTIC PLASIOCEPHALY UTILIZING THE CRANIAL REMODELING ORTHOSIS

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Increased concern with craniofacial dysmorphic conditions is noted because of multidisciplinary interest in reconstructive surgery and the interest of geneticists in their complex etiology. Generally, abnormal head shape can be divided into three categories: 1) syndromes (Apert's or Crouzon's), 2) cranial synostosis, and 3) nonsynostosis conditions of a positional nature. Our primary concern is with positional plagiocephaly which may result from premature birth, intrauterine environment, torticollis, cervical abnormalities, sleeping position preference, or the interaction of any of the aforementioned factors. Children with non-synostotic plagiocephaly can exhibit complex cranial asymmetries with resultant multistructural involvement of the head, face and spine. Until recently, treatment for severe plagiocephaly was reconstructive surgery. Difficulty in achieving permanent correction of some complex head shapes has lead to the development of an external cranial remodeling band that can be modified to the individual's growth specifications. In positional plagiocephaly, our technique succeeds in producing significant normalization, has a distinct advantage over other passive helmets and provides an alternative to surgical intervention. Nonsurgical means of altering skull shape are most effective during infancy as, during the first 12 months of life, the majority of skull growth occurs. Head circumference increases by approximately 8.3 cm during the first 6 months, slowing to increments of 0.5 cm each month thereafter until the child is 18 months of age. To further substantiate the need for early recognition, objective anthropometric measurements are taken to document quantitative change in response to treatment. Our goals are threefold: a) case studies illustrating the effectiveness of external orthosis, b) to illustrate the importance of early recognition, and c) demonstrate frequency of occurrence.
7. MICROPLATE FIXATION FOR PRIMARY REPAIR OF CRANIOSYNOSTOSIS

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Many surgical techniques are utilized for primary repair of craniosynostosis including bone grafts, floating segments and osteotomies with wire fixation. Microplates offer a rapid technique for immediate rigid bone fixation without impairing long term craniofacial growth. We have utilized titanium microplate fixation for primary repair of metopic, lambdoid and uni and bilateral coronal craniosynostosis. We will describe our surgical techniques of plate fixation in 19 children who underwent 21 cranietomies for synostosis. There were 6 children with metopic synostosis, 5 patients each with unilateral or bilateral coronal synostosis, 2 patients with lambdoid and one child with multiple suture craniosynostosis. Patient ages ranged from 2 months to 14 years. There were no complications of plate use and screw migration was not demonstrated on postoperative CT. It was not necessary to remove the microplates for any reason. The cosmetic results have been very satisfactory with no relapse. Miniplates avoid the complications of wire and offer superior rigid fixation. Their use in patients of all ages facilitates one stage complex cranial remodeling with rigid bone fixation. Our studies suggest there is no long term limitation of craniofacial skeletal growth.

8. ASTERION REGION SYNOSTOSIS

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Presented are four patients between the ages of 6 to 15 months with clinical features of unilateral lambdoid synostosis: unilateral posterior flattening, ear shifting, and anterior compensational plagiocephaly. Three-dimensional CT scans revealed that both lambdoid and both coronal sutures were open, and that the region of the Asterion was fused. The Asterion or the junction of the lambdoidal occipitomastoid and parietomastoid sutures were areas of involved pathology. The parietomastoid suture was involved in all four cases for approximately 4cm distance from the Asterion, and only the distal 1cm of the lambdoid suture was involved, with the remainder of the lambdoid being open. Three-D CT findings were confirmed intraoperatively, where the area of the Asterion was sclerosed and thickened compared to the opposite uninvolved side. The lambdoid sutures were found to be open. Histologic studies were performed in the region of the Asterion and lambdoid sutures: The occipitomastoid, parietomastoid and proximal squamosal sutures were fused. The lambdoid sutures were found to be open except for the area closest to the Asterion. The patients were treated by releasing the involved region and reconstructed with a posterior Bandau procedure. The term functional synostosis has been used to describe patients with posterior plagiocephaly and open lambdoid sutures. Synostosis of this area should be considered in patients with posterior plagiocephaly. It is possible that failure to do so may be the cause of poor results or recurrences that have been noted using conventional methods. Clinical, radiographic and histologic correlations will be presented.
9. MRI BEHAVIORAL AND ANATOMICAL FEATURES OF A RABBIT MODEL FOR INFANTILE HYDROCEPHALUS

Charles E. Olmstead, PhD, Warwick J. Peacock, MD, Christopher P.E. Outram, Michael, De Rosa, Robin S. Fisher, PhD, Richard Gayek, DVM (Los Angeles, CA)

A rabbit model of kaolin induced chronic infantile hydrocephalus was studied using magnetic resonance imaging (MRI), a standardized neurological examination, and basic and immunohistochemical staining to document the anatomical basis for functional deficits accompanying hydrocephalus.

Longitudinal data has been obtained from 26 experimental and 12 control male and female rabbits. Between 2 and 7 days of age kaolin was injected into the cisterna magna. Two sequelae were observed. In a small proportion of the animals, hydrocephalus developed quickly, killing the animal within 5 to 8 days. In the majority of animals, however, the hydrocephalus developed less catastrophically, even though it was readily evident by MRI within 2 days of injection. Ventricle enlargement and cranial enlargement progressed throughout the survival period (up to 7 months). At 78 days postinjection the volumes of the injected ventricles ranged from 174% to 1600% of the control littersmates. Most notable was massive dilation of the olfactory bulb ventricles (300 to 700%). This is thought to be related to increased flux through CSF drainage routes at the olfactory lymphatics, which results in partial alleviation of the raised intracranial pressure in association with a subsequent chronicity of the condition.

At no time were any substantial behavioral differences between hydrocephalic animals and littersmates detectable. Animals with chronic hydrocephalus have life spans comparable to controls.

Microscopically, cresyl violet staining revealed severe dysplasia of the cortex and dyslamination of the olfactory bulbs. Golgi staining showed convolution and shortening of apical dendrites of pyramidal neurons. Immunostaining for GFAP showed aberrant differentiation of cortical elements. Cells with the morphological properties of neurons expressed an astrocytic intermediate filament protein in hydrocephalic rabbits but not in controls.

10. HISTOLOGICAL DAMAGES TO THE SPINAL CORD BY NEW EXPERIMENTAL MODEL OF TETHERED CORD

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Tethered cord syndrome is manifested by motor and sensory deficits in lower limbs and incontinence. An elongated spinal cord attached to a thick filum terminale is often found stretched and oxidative metabolism is impaired in cat model and human tethered cord. In this report laminectomy and ligation of the filum terminale allowed traction of the lumbosacral cord as previously described. A ligature was passed over the pulley and connected to one of weights (3 g, 5 g, and 8 g) and dropped 1.0 cm. The animals were sacrificed 24 hours 2 weeks after surgery. The L2, L7, S3, and coccygeal cord segments were removed and examined under light and electron microscopy (LM and EM). Only high grade (8g) drop traction resulted in urinary incontinence and tail anesthesia. Edema and lymphocytic infiltration in the gray matter were noted by LM; and numerous neuronal membrane breaks and loss of mitochondrial cristae were found by EM. This is the first report to describe histological damage to the gray matter in cat cord tethering model and to correlate these findings with permanent incontinence in tethered cord patients.
11. TETHERED CORD DURAL REPAIR WITH INTRAOPERATIVE AUTOLOGOUS FIBRIN GLUE

LCDR Robert F. Keating, MC, USN, LCDR Paul Potter, MC, USN (Oakland, CA)

Although management and timing of surgery for tethered cord in the setting of myelomeningocele remains somewhat controversial even today, few would deny the attendant difficulties in repair of the attenuated dura as well as the dreaded complication of CSF leaks. The use of fibrin glue in this arena has been embraced by many pediatric neurosurgeons of late. Yet this can require significant labor and expense in the blood bank as well as expose the patient to viral infection. We have now used autologous fibrin glue, obtained at the time of surgery, in four patients with prior myelomeningocele repairs and subsequent tethered cords as well as one patient status post a gunshot wound to the spine with a CSF leak.

This represents a simple, inexpensive and completely safe means of providing a watertight seal to previously compromised dura mater. In addition, this platelet and plasma matrix also provides a tremendous array of growth factors, in particular PDGF and FGF, to assist in healing of the dura. Intraoperatively this was performed with an AT1000 Electromedics Cell Saver with minimal difficulty and may be accomplished safely in patients who weigh as little as 22 kg. The only difficulty encountered was a patient with an unexpected low fibrinogen level, who required a longer time for clot activation. Nevertheless, the use of intraoperative autologous fibrin glue serves to reduce the incidence of postoperative CSF leaks and can be accomplished in a safe and simple manner.

12. THE EFFECT OF VENTRICULAR DRAINAGE AND DURAL CLOSURE ON CEREBROSPINAL FLUID LEAKS AND INFECTION

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Acute complications following suboccipital craniectomy include hydrocephalus, cerebrospinal fluid (CSF) leak, and infection. The purpose of this retrospective review is to evaluate the effects of pre or intraoperative ventriculostomy and/or duraplasty on the incidence of these complications. Over a 3 year period, 50 pediatric patients underwent suboccipital craniectomy for posterior fossa tumor biopsy and/or resection. Surgery was performed for 20 astrocytomas, 23 primitive neuroectodermal tumors, 2 dermoid cysts, 1 pontine sarcoma, 3 pineal germ cell tumors, and 1 ganglioglioma. The dura was either left open (n=16), loosely reapproximated (n=6), tightly closed (n=11), or reconstructed with human cadaveric dura (n=17). All patients received 48 hours of prophylactic, intravenous antibiotics. Mean follow-up period was 6 months.

Twenty-one patients (42.0 %) had hydrocephalus. Twelve patients (24.0 %) underwent pre or intraoperative ventriculostomy placement. Three of these 12 patients (25.0%) later developed a CSF leak. Two of these 3 patients (66.0%) thereafter required a ventriculoperitoneal shunt. In contrast, only 4 of 38 patients (10.5%) without pre or intraoperative ventricular drainage subsequently developed a CSF leak. Three of these 4 (75.0 %) required placement of a ventriculoperitoneal shunt. Four of 7 patients (57.1 %) with postoperative CSF leaks developed meningitis or ventriculitis.

It is postulated that watertight dural closure or grafts help prevent CSF leakage and thereby decrease the incidence of wound infection. We conclude, however, that neither tight dural closure, duraplasty, nor precedent ventricular drainage significantly changes overall complication rate following suboccipital craniectomy.
13. STEREOTACTIC TRANSTENTORIAL VENTRICULO-PERITONEAL SHUNTING FOR THE TREATMENT OF THE SEQUESTERED FOURTH VENTRICLE

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The sequestered fourth ventricle is uncommon but when encountered is a challenge which demands considerable attention on the part of the pediatric neurosurgeon. The shunting techniques traditionally used to relieve pressure caused by an isolated fourth ventricle yielded clinical and radiographic results which were often unsatisfactory.

The authors believe that the effectiveness of shunting can be improved and the complications minimized with precise insertion of the ventricular catheter into the fourth ventricle at an angle approximating, as much as possible, the orientation of the long axis of the fourth ventricle. To this end, the authors have developed a technique of stereotactic transtentorial insertion of the ventricular shunt for the treatment of the sequestered fourth ventricle. The authors report on their series of five patients, aged 4 months to 17 years, with a followup period of 3-20 months. All patients had existing lateral ventriculo-peritoneal shunts and isolated fourth ventricles, the etiologies of which were meningitis in three patients, Chiari malformation in one patient, and intraventricular hemorrhage in one patient. All patients underwent stereotactic transtentorial insertion of ventriculoperitoneal shunt using the BRW stereotactic system. This technique is described and problems which may arise, as well as solutions leading to avoidance of such difficulties, are presented. It is hoped that the utilization of this technique will result in better success in the treatment of this difficult patient population.

14. HYDROCEPHALUS MAY CAUSE IRREVERSIBLE PATHOLOGY IN THE RETINA OF NEONATAL KITTENS

James P. McAllister II, PhD, Eileen C. Williamson, Helen E. Pearson, PhD (Philadelphia, PA)

Although perturbations of the visual system have been implicated as causes of the cognitive deficits that can accompany infantile hydrocephalus, examinations of the retina following ventriculomegaly have been lacking. Therefore, the present study was designed to analyze the retinae of normal, mildly hydrocephalic, severely hydrocephalic and surgically decompressed kittens to determine if changes occur in ganglion and glial cells. Hydrocephalus was induced in 10 day old kittens by intracisternal injection of kaolin. Kittens were allowed to survive from 7-28 days after injection. Animals that were decompressed received ventriculoperitoneal shunts 10 to 15 days after the induction of hydrocephalus and were sacrificed 10-14 days after shunt placement. The density and area of neuronal and glial cells were determined within a sample area in peripheral nasal retina. Total cell density was significantly increased 48% in mildly and 52% in severely hydrocephalic animals, but returned to normal following decompression. These changes are due to a significant increase in the glial population. More importantly, there was a significant loss of ganglion cells in both the severely hydrocephalic (45% decrease) and the shunted (53% decrease) groups, but not in the mildly hydrocephalic group. Recovery was selective in that the density of large ganglion cells returned to normal values, but small and medium-sized ganglion cells remained 47% and 58% below controls, respectively. Based on these findings, we conclude that gliosis and ganglion cell death occur in the retina following severe hydrocephalus, and relatively late decompression is unable to reverse these effects. Furthermore, gliosis alone occurs in mild cases of hydrocephalus, and may be an early indication that cellular degeneration will follow.
15. A LABORATORY MODEL FOR PERCUTANEOUS UNCLOGGING OF PROXIMAL SHUNT CATHETERS

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The majority of shunt malfunctions involve the proximal ventricular catheter, which is usually occluded with tissue debris or proteinaceous material. Recent advances in technology have resulted in angioscopic catheters of 0.5mm in diameter and laser fibers as small as 200um in diameter.

A method for visually inspecting the lumen of ventricular catheters has been developed with attempts at unclogging the catheter occlusion. Specimens from patients, and catheters imbedded in agarose/fibroblast tissue cultures were utilized for this technique.

Under direct visualization, the lumen was cleared of the occluding debris and the catheter was recanalized using NdYag laser and electrocautery probes. The proximal catheter was occluded to allow for distal perfusion and evaluation of the valve mechanism.

It is yet unclear if this model can be translated to clinical practice with permanent percutaneous recanalization of shunt malfunction. However, the technique appears promising for direct visual inspection of shunt malfunction and applying a minimally invasive technique to maintain proper shunt function.

16. DOES AN EXCITOTOXIC COMPOUND CONTRIBUTE TO DECREASED INTELLIGENCE FOLLOWING SHUNT INFECTION?

Steven J. Schiff, MD, PhD, Melvyn P. Heyes, PhD (Washington, DC)

Infections associated with cerebrospinal fluid (CSF) shunts are correlated with significant decrements in intelligence. To better account for the neurologic and cognitive deficits observed following shunt infections, this study explored the hypothesis that the endogenous excitotoxic compound, Quinolinic acid, may be elevated during such infections. In patients with an indwelling CSF shunt or temporary drainage system, Quinolinic acid concentration was measured in 49 samples of CSF from 39 patients with hydrocephalus, tumors, intraventricular hemorrhage, and bacterial infections. Quinolinic acid concentrations were not significantly elevated in the CSF of patients who had a shunt alone, with or without raised intracranial pressure, without an accompanying inflammatory or neoplastic condition. In the presence of tumors and intraventricular hemorrhage, significantly elevated CSF levels were found. Extremely high levels were detected in patients with bacterial shunt infections. Such elevated levels in infected patients are high enough to cause direct excitotoxic damage to the brain. These findings suggest that it may be possible to lessen the damage to the brain from shunt infections, and other inflammatory and neoplastic diseases, with suitable excitatory amino acid blockade.
17. DELAYED HYDROCEPHALUS AFTER POSTERIOR FOSSA TUMOR SURGERY IN CHILDREN

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Posterior fossa tumors in children are frequently associated with obstructive hydrocephalus. Upon presentation the decision to shunt is often made perioperatively and in a few situations on delayed basis. Factors predicting persistent symptomatic hydrocephalus requiring cerebrospinal fluid (CSF) shunting are unexplained.

We reviewed our surgical series of 90 children with posterior fossa tumors from 1985-1992. Thirty-nine (43%) required shunting to control symptomatic hydrocephalus. Many factors including age, extent of surgical resection, histology, tumor location, management of hydrocephalus by ventriculostomy and lumbar punctures and interval between resection and shunt placement were studied. The most important predictor for persistent symptomatic hydrocephalus requiring shunting was patient age (5 years or less). The highest percentage of delayed shunts were required in children with cerebellar astrocytoma. Other factors appear to be inconsistent with outcome.

Clinical, radiographic and periooperative features of pediatric posterior fossa tumors requiring CSF shunting will be discussed. Understanding these features will assist the neurosurgeon to predict outcome and treat hydrocephalus on a more timely basis.

18. MORPHOLOGICAL CHANGES AFTER EARLY AND LATE SHUNTING IN A RAT MODEL OF INFANTILE HYDROCEPHALUS: IMPLICATIONS ON CSF DYNAMICS

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In a companion experimental study, it has been shown that, the changes in CSF dynamics caused by infantile hydrocephalus would be reversible after early but not after late shunting. The aim of the present study is to investigate whether the failure of late shunting to recover the CSF dynamics can be explained by the changes in the brain tissue as a result of a longstanding hydrocephalic process. Morphological and CSF dynamic studies were performed before and after shunting in a kaolin model of infantile hydrocephalus in rats. Hydrocephalic rats were studied two (n = 11) or six (n = 16) weeks after kaolin injection. Two corresponding control groups were also included.

Early in the course of hydrocephalus (after two weeks), subependymal edema was pronounced, however, the axons looked morphologically intact. After shunting, this edema almost completely resolved. The morphologically intact white matter and the rapid edema resolution explain the rapid postshunting recovery of the low PVI (p0.01). In the second group, six weeks after inducing hydrocephalus, the subependymal edema was more extensive and groups of degenerated axons were frequently observed. After shunting, edema resolution was partial and degenerated axons could still be observed. The white matter degeneration and the incomplete edema resolution help clarify the failure of CSF diversion to alter the CSF dynamics in this stage.

We conclude the neural axis buffering capacity after shunting depends upon the degree of the hydrocephalic morphological changes present at the time of shunt insertion.
19. SIGNIFICANCE OF SYMPTOMATIC UNILATERAL VENTRICULOMEGALY IN SHUNTED PATIENTS

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Isolated contralateral ventricular enlargement is an uncommon but well documented finding that occurs after shunting myelodysplastic and non-myelodysplastic children. The pathophysiology is unclear although most have blamed distortion of the anatomy of the foramen of Monro resulting from shunt placement. Three children developed unilateral ventriculomegaly after shunting. The pathophysiology involved abnormal cerebral venous outflow that resulted in hydrocephalus. Treatment with ventriculoperitoneal shunting caused marked ventricular asymmetry. In all three cases, the patients returned with severe symptoms of raised intracranial pressure, despite functioning shunts. All patients improved after addition of a second shunt on the enlarged side. The etiology of the venous outflow obstruction and hydrocephalus varied among the three patients: one had Crouzon's disease with abnormal venous structures at the skull base, the second underwent repair of a fourth ventriculocoele, the third had a congenital asymmetry of the cerebellum and venous sinuses. Two of the three patients developed clinical pictures consistent with pseudotumor cerebri after shunting including elevated monitored intracranial pressures, papilledema, and chronic tonsillar herniation. Abnormal venous drainage after shunting may lead to the development of turgid brains, causing sufficient distortion to result in unilateral ventricular enlargement. This sequence of events may also underlie the development of hydrocephalus in myelodysplastic children.

20. SUBCUTANEOUS PALISADING GRANULOMA OF THE SCALP IN CHILDHOOD

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The child presenting with multiple, immobile, painless scalp lesions can present a diagnostic dilemma. Over a 17 year period, 109 operations for scalp and/or calvarial masses have been performed at Children's Hospital of Los Angeles. In three cases, a diagnosis of subcutaneous granuloma annulare was established. All three patients were 3 years of age or less, and one patient had juvenile rheumatoid arthritis. Multiple hard, painless, immobile lesions were noted; with observation some of the original masses disappeared and new lesions developed. Skull radiographs and radionuclide bone scanning were normal and CT scanning revealed only the subcutaneous masses without involvement of the underlying skull. A diagnostic biopsy was performed in each case. Histopathologic examination revealed fibrinoid necrosis, palisading histiocytes, and lymphocytes. With follow-up, all scalp masses regressed.

Subcutaneous granuloma annulare usually presents as multiple, firm, nontender, fixed scalp lesions in young children. The lesions typically vary in size and location over time; they are most common in the posterior scalp. They may be associated with rheumatoid disease, but are unlikely to herald its future onset. Imaging studies are not helpful. Histopathologically the lesions show fibrinoid necrosis and chronic inflammatory cells. Immunohistochemistry reveals markers consistent with a cell mediated immune response. A diagnostic biopsy is not indicated in the face of known collagen vascular disease. Fine needle aspiration should be avoided, as an erroneous diagnosis of malignancy may result. Spontaneous regression of the lesions can be expected.
21. CORTICAL GLUCOSE METABOLISM AND PEDIATRIC BRAIN DEATH

Michael D. Medlock, MD, Robert Cruse, DO, William C. Hanigan, MD, PhD (Peoria, IL)

The specificity of confirmatory tests for the diagnosis of pediatric brain death varies considerably. A case is reported of an infant with clinical evidence of brain death, isoelectric EEG and cortical glucose metabolism demonstrated by PET.

A 2-month-old child was brought to the emergency room after a prolonged period of anoxia. Ophthalmoscopy revealed bilateral retinal hemorrhages while a CT scan showed a parietal skull fracture, small subdural hemorrhage, and diffuse subarachnoid blood. On the second day of hospitalization, the patient demonstrated clinical brain death; five days later, an EEG showed minimal alpha activity while cerebral blood flow using Tc 99m was normal. On the 11th hospital day, the EEG was isoelectric; a PET scan with 5 PDG showed significant cortical glucose metabolism. Following a second isoelectric EEG, the ventilator was discontinued. Autopsy revealed diffuse cortical necrosis with extensive mononuclear cell infiltrates throughout all cortical layers.

Clinical evidence of brain death, isoelectric EEG, and normal cerebral blood flow has been reported in 8 infants. The presence of an expandable cranial vault allowing increased intracranial volume may prevent the cessation of blood flow. In this patient, both blood flow and cortical glucose metabolism were intact despite clinical evidence of brain death. The histological findings suggested that the glucose metabolism could not be accounted for by neuronal uptake, but may be explained by metabolic activation of microglia associated with neuronal loss. This hypothesis is strengthened by evidence that PET has shown increases in regional glucose metabolism within 48 hours of ischemic stroke.

In summary, this report indicates that cortical glucose metabolism in infants may be associated with clinical and EEG evidence of brain death. The use of PET may not be specific for the diagnosis of pediatric brain death.

22. IS THE SELECTIVE POSTERIOR RHIZOTOMY TECHNIQUE VALID?

Rick Abbott, MD, Nobu Morota, MD (New York, NY)

Selective posterior rhizotomy is a procedure which is increasingly gaining acceptance for the treatment of spasticity. The stimulation techniques most widely used are modeled after Fasano's technique first described in 1976. This technique was based upon noninvasive monitoring techniques developed to study spasticity in patients who had sustained injury to their central nervous system and has never been confirmed as being able to identify nerve roots/rootlets responsible for a limb's spasticity. In 1988 we began to include the S2 sensory roots in our testing and lesioning because of a large incidence of postoperative spasticity in the gastrocnemius muscles in patients undergoing testing/lesioning at the L2-S1 levels. Subsequently, we modified our technique by the inclusion of the pudendal neurogram because an increased incidence of bladder dysfunction seen in our patients after the inclusion of the S2 roots. The neurogram identifies nerve roots responsible for the conduction of afferent action potentials of the cutaneous branches of the pudendal nerve, allowing for their preservation. Using this neurogram has caused us to leave abnormally "spastic" roots as determined by Fasano's stimulation techniques. We have contrasted the postoperative tone examinations of patients who had no abnormally responding roots/rootlets at the S2 level vs those who did after their surgeries. Patients who had all abnormal rootlets cut tended to have less residual tone as compared to those who had abnormally responding rootlets left uncut. This would seem to validate Fasano's stimulation technique as labeling rootlets with a high concentration of neurons participating in the generation of spasticity.
23. SELECTIVE DORSAL RHIZOTOMY AFFECTS COGNITIVE PERFORMANCE OF CHILDREN WITH SPASTIC DIPLEGIC CEREBRAL PALSY

Suzanne Craft, PhD, T.S. Park, MD, D.A. White, MA (St. Louis, MO)

Anecdotal reports of improved attention and language in children with cerebral palsy following selective dorsal rhizotomy (SDR) for treatment of spasticity are usually attributed to improved mood or reduced physical discomfort. The present study examined the possibility that SDR may have distal effects on cortical function which enhance cognition. Twenty children with spastic diplegic cerebral palsy received tests of visual attention and other cognitive functions one day prior to and six months following SDR. Visual attention was assessed with an orienting task in which subjects pressed a button when a target appeared to the left or right of a central fixation point on a computer screen. The target was preceded by a valid or invalid cue. Previous investigations indicate that performance on validly and invalidly cued trials is mediated by thalamic and cortical (frontal or parietal) areas, respectively. It was hypothesized that general reduction in response time (RT) across all experimental conditions following SDR would suggest nonspecific effects of improved mood or physical comfort, whereas changes in performance patterns on invalidly and validly cued trials would suggest that SDR affects specific neural systems mediating attention.

Cerebral palsy subjects were compared with 24 normal children tested initially and six months later. Subjects treated with SDR showed a significantly greater decrease in RT for invalidly cued relative to validly cued trials and significantly greater improvement in associative learning than did normal subjects. These results suggest that SDR affects cortically mediated cognitive function, and indicate the need for further study with electrophysiologic or metabolic measures of cortical activity.

24. EXTREME LATERAL APPROACH FOR JOINT RELOCATION IN AARD

Fraser C. Henderson, Myron A. Rogers, H. Alan Crockard, John M. Stevens (Bethesda, MD)

Atlanto-axial rotatory dislocation (AARD) should be regarded as a distinct entity from atlanto-axial rotatory subluxation (AARS).

The nature of fixation in AARD is poorly understood, and relocation of the joints is therefore seldom accomplished in the chronic phase. Furthermore, if reduction is not achieved, then it would seem that posterior fusion serves little purpose. We describe the use of the "extreme lateral approach" in AARD, and discuss the intra-operative findings which shed light on the mechanism of fixation and the reason for failure of closed reduction.

Nine cases of traumatic rotatory injuries at the atlantoaxial level have been treated by the authors over the past 4 years. Two of the cases have been AARD and 7 AARS. Eight of the patients were in the pediatric age group. The 2 patients with dislocations underwent relocation using the "extreme lateral approach."

The "extreme lateral approach" allows direct inspection of the entire involved joint and control of the vertebral artery. The nature and loci of fixation can be visualised, and the joint surfaces mobilised and relocated directly. Intraoperative findings in our 2 operated cases included disrupted joint capsules, profuse soft tissue proliferation between and around the joint surfaces, with part of the transverse ligament interposed between the articular surfaces and bony ankylosis between C1/C2.

These findings contribute to the understanding of why closed reduction may fail in AARD, and suggest that if closed reduction is unsuccessful, then early operative intervention is indicated.

The extreme lateral approach is presented as an effective option in the treatment of selected cases of AARD in children.
25. ATLANTO-OCcipital AND ATLANTO-AXIAL INSTABILITY AFTER POSTERIOR CERVICAL FUSION IN DOWN'S SYNDROME PATIENTS

Donald S. Soloufik, MD, Ana Maria De Villiers, MD, Anvind Ahuja, MD, L.N. Hopkins, MD, N. Razak, BS (Buffalo, NY)

Follow-up of three Down's syndrome patients treated for cervical instability by posterior C1-C2 fusion (two patients) and C2-C5 fusion (one Patient) at Children's Hospital of Buffalo demonstrated instability in the level above the fusion. In one patient the unstable level resulted in progressive neurologic sequela.

Musculoskeletal disorders in Down's syndrome include cervical anomalies with attendant predisposition for instability. The Atlanto-Axial articulations are commonly unstable with a 10-20% radiographic incidence. This instability is not often associated with neurologic findings, but when these findings present posterior cervical fusion (typically C1, to one to three levels down) is indicated. Atlanto-occipital instability is much less frequently described and also not often associated with neurologic findings. Anomalous ligament laxity is implicated in both AA and A-O instability. After fusion the pivot on head flexion may focus on the level(s) above the fusion and potentially exacerbate ligamentous laxity and consequent A-A or A-O instability.

Post-cervical fusion A-A or A-O instability is important to diagnose and follow carefully as further neurosurgical intervention may be required.

26. INTRAMEDULLARY SPINAL CORD LIPOMAS UNRELATED TO SPINA BIFIDA PRESENTING AS QUADRIPARESIS IN INFANCY

Z. Kiss, H.J. Hoffman, S. Chuang, D. Harwood, N. Nash (Toronto, Ontario)

Intraspinal lipomas in the pediatric population are rare in the absence of a dysraphic syndrome. Only three reported cases have presented in infancy, despite the congenital nature of these lesions.

The experience at the Hospital for Sick Children has comprised three cases, all of whom presented in the first eight months of life with quadriplegias. None had spina bifida, but two had posterior cervical subcutaneous fat pads, suggestive of lipomyelomeningoceles. Imaging was performed with CTmyelography in the first case and MRI in the two more recent cases. Intramedullary lipomas extending over a variable segment of cervical cord and rostrally into the posterior fossa were demonstrated. Treatment consisted of a posterior fossa craniectomy and cervical laminotomies with subtotal resection of the subpial lipomas, followed by expansile duroplasty. Our postoperative results are presented as well as the other previously reported cases discussed.

Intramedullary lipoma of the cervical cord must be considered in the infant with either congenital or progressive upper extremity dysfunction. Cutaneous manifestations are not always present to suggest the diagnosis, although with the increasing availability of MRI, earlier diagnosis should be achieved.
27. SINGLE LAYER DORSAL LUMBAR FASCIA: ITS USE AS A DURAL SUBSTITUTE IN UNTETHERING PROCEDURES

Jeffery A. Winfield, MD, PhD, William H. Bell, MD (Syracuse, NY)

Chemical meningitis (CM), evident as elevated temperatures and WBC counts in the CSF several days following primary or redo intradural lumbar procedures for untethering of the spinal cord, remains a persistent problem. CM is most often seen when a dural substitute is used to enlarge the thecal sac, and its occurrence may predispose the child to recurrent tethering from arachnoid adhesions and scarring developed during the inflammatory response. In this report, the authors present their clinical experience and operative techniques for harvesting a single layer of the dorsal lumbar fascia and its utilization as a dural substitute for thecal sac enlargement. When used as a dural substitute in both primary, and in particular secondary untethering procedures, CM occurs infrequently and post operative steroids are not required. We feel that this simple technique of autologous fascial grafting may further reduce the incidence of retethering and its usage important, as increasing concerns arise that a significant number of toddlers born with meningomyelocele will require untethering procedures.

28. THE LACK OF A ROLE FOR p53 IN ASTROCYTOMAS IN PEDIATRIC PATIENTS

Corey Raffel, MD, PhD, N. Scott Litofsky, MD (Worcester, MA)

Approximately one third of astrocytic neoplasms have mutations in the p53 gene, which codes for a cell division regulatory protein. Astrocytomas in pediatric patients have not been previously investigated for p53 mutations.

We evaluated 37 astrocytic tumors (12 pilocytic, 8 diffuse low grade, 15 anaplastic, and 2 glioblastoma) in pediatric patients for p53 mutations. DNA was isolated, and polymerase chain reaction with radiolabeling and labeling was performed to amplify exons 5, 6, 7 and 8 of the p53 gene. These exons constitute 98% of reported p53 mutation sites in other tumors. The tumors were screened by single-stranded confirmation polymorphism (PCR-SSCP) analysis. Tumors with abnormal PCR-SSCP banding patterns were sequenced. The results were analyzed by Chi-square to determine significant differences from previously reported patterns of p53 mutations in astrocytomas. Two (one pilocytic, one anaplastic) of the 37 tumors (5.4%) had PCR-SSCP shifts, indicating p53 mutations. Both mutations occurred in exon 8. The frequency of p53 mutations in pediatric astrocytomas is significantly less than the frequency for all patients reported by others (Chi-square = 7.52, p<.01). Furthermore, the frequency of p53 mutations in higher grade astrocytomas (anaplastic astrocytoma and glioblastoma) is significantly lower in pediatric patients (Chi-square = 3.98, p<.05). Interestingly, the mutation in the pilocytic astrocytoma is the first described in a low grade astrocytoma.

These results suggest that p53 is probably not important in the oncogenesis of pediatric astrocytomas. Oncogenesis in pediatric astrocytomas may occur by different mechanisms than in similar tumors in adults.
29. PERMANENT LOW ACTIVITY IODINE-125 IMPLANTS FOR RECURRENT PEDIATRIC MALIGNANT BRAIN TUMORS

Brian R. Griffin, MD, Mitchel S. Berger, MD, J. Russell Geyer, MD
(Seattle, WA)

Permanent low activity I-125 sources implanted in the brain after aggressive resection of malignant brain tumors can theoretically deliver a very high radiation dose to a highly restricted brain volume. Additionally, because of the low dose rate with permanent implants, brain toxicity may be greatly reduced compared to external beam radiotherapy or high activity brain implants. Five pediatric patients ranging from 3 to 19 years of age with recurrent malignant brain tumors underwent aggressive resection and permanent low activity I-125 implants; all patients had previously received full course external beam radiotherapy.

Tumor histologies included malignant ependymoma, medulloblastoma, primitive neuroectodermal tumor, glioblastoma multiforme and adenoid cystic carcinoma in one patient each. Radiation doses in excess of 8,000 cGy over the active life of the implant were delivered in each case at one cm deep to the resection cavity; brain tissue closer to the implanted sources received significantly higher doses. With a median followup of eight months (range 3-16 months) all patients are alive with no evidence of tumor recurrence at the implanted site as demonstrated by follow up magnetic resonance scans; one patient has developed a new lesion on the ventricular surface remote from the implant site. No toxicity has been evident in any patient, either clinically or on follow up MRI. Permanent low activity implants combined with aggressive resection appears to be a promising new technique for treatment of malignant pediatric brain tumors.

30. PILOCYTIC DORSALLY EXOPHYTIC BRAINSTEM GLIOMAS: A DISTINCT CLINICOPATHOLOGIC ENTITY

Robert A. Sanford, MD, Z. Khatib, MD, R. Heideman, MD, E. Kovnar, MD, E.A. Kirk, RN, MSN (Memphis, TN)

Brainstem gliomas (BSGs) comprise 10-20% of all childhood brain tumors. Although the majority of BSGs can not be approached with intent of surgical resection, patients (pts) with dorsally exophytic brainstem gliomas (DEBSGs) may be candidates for radical surgery. Of 51 pts with BSGs evaluated and treated at our institution between 1983-1991, 12 pts (24%) were found to have dorsally exophytic tumors on neuroimaging. Age at diagnosis of DEBSG was 17-75 mo. (median=38 mo.). Symptom duration prior to diagnosis was 2-24 mo. (median=7 mo.). Vomiting and ataxia were the most frequent symptoms. Four pts presented with torticollis. Cranial nerve VI was affected in 5 pts; CN VII was involved in 2. Seven pts had no CN signs. Long tract signs were notably absent. On MR, these tumors were sharply demarcated from surrounding tissue with decreased signal intensity on T1 sequences and increased signal on T2. All showed bright enhancement after gadolinium-DTPA infusion. All 12 pts underwent suboccipital craniotomy. Five tumors were found to arise from the dorsal medulla, 3 from the pons and 4 from the pontomedullary junction. Based on operative findings and post-op MR, gross total resection (95% removal) was achieved in 6 pts; 6 had subtotal resection (50-95% removal). Histologic examination showed pilocytic astrocytoma in 11 pts; 1 pt had a low grade fibrillary astrocytoma. One pt with sub-total resection received post-operative irradiation (XRT) and 1 pt received chemotherapy (MOPP) and delayed XRT. Ten had no immediate further therapy.

Three of 10 had tumor progression 2-10 mo. after surgery and were treated with local XRT (54.55 Gy); they now remain stable or improved 3-12 months after XRT. Eleven pts are alive today at a median of 22 mo. (5-56 mo.). The only death was due to secondary leukemia in the pt who received chemotherapy; no residual tumor was found on neuroimaging prior to death. Survival for DEBSGs is 100% at 2 years vs. 18% (+7%) for other BSGs, p=0.0003.

In conclusion, children with DEBSGs have characteristic clinical features, often presenting with a prolonged history of increased intracranial pressure and cerebellar signs and a paucity of cranial nerve abnormalities or long tract signs. In contrast to pts with infiltrative BSGs, these children have an excellent outcome even with subtotal surgical resection. The benign biologic behavior associated with pilocytic astrocytoma may underlie this favorable prognosis.
31. PEDIATRIC INTRACRANIAL HEMANGIOENDOTHELIOMAS

Thomas C. Chen, Ignacio Gonzales-Gomez, Floyd H. Gilles, J. Gordon McComb (Los Angeles, CA)

Hemangioendothelioma (HE) is a rare vascular tumor which histologically is intermediate in appearance between a hemangioma and angiosarcoma. Intracranial HE are extremely rare, and have not been previously reported in children. We have encountered intracranial HE in two patients, a seven year old girl with a lesion in the right gasserian ganglion, and a three month old boy with a cervicomедullary junction tumor. Both lesions were iso-intense to grey matter with gadolinium enhancement on T1 weighted images, and hyperintense to grey matter on T2 weighted images. Histologically, endothelial cells forming small vascular lumina, with minimal mitotic activity were seen. Both tumors were Factor VIII positive. Electron microscopy depicted endothelial cells surrounded by basal lamina, and attached to each other by prominent desmosomelike structures. Other primary intracranial tumors were excluded on the basis of morphology and immunohistochemistry. Recently, interferon alpha-2a has been found to be effective in Improving Kaposi's sarcoma and life threatening hemangiomas of infancy unresponsive to corticosteroids. Although the mechanism of action is still unknown, it has been found to inhibit the locomotion of capillary endothelium in-vitro and to inhibit angiogenesis in mice. Our seven year old girl had a gross total removal with no recurrence. The three month old boy is currently receiving daily injections of interferon alpha-2a. Interferon alpha-2a may represent a new chemo-therapeutic agent in the treatment of this rare tumor, and may prove useful in other tumors of vascular origin.

32. ACUTE SUBDURAL HEMATOMA: IS THE BLOOD ITSELF TOXIC?

Ann-Christine Duhaime, MD, Linda Gennarelli, Anthony T. Yachnis, MD, Douglas T. Ross, PhD (Philadelphia, PA)

The finding of a zone of "infarction" under experimental acute subdural hematoma (ASDH) which can be alleviated by glutamate receptor blockade has led to the question of whether blood, with its high intracellular glutamate concentration, is itself toxic to the cortex. Such a process might influence management of thin but extensive acute subdural hematomas such as those commonly seen in infantile head injury.

To test this hypothesis, wide parietal craniotomies were performed in 20 Long-Evans rats. The dura and arachnoid were opened widely in a stellate fashion. Autologous femoral arterial blood was applied to the cortex until an opaque, solid clot covered the entire exposed area. In control hemispheres, the same procedure was performed but no blood was applied, and careful homostasis and irrigation limited contact between cortex and extracranial blood. Animals were sacrificed at 72 hours, serial sections were obtained, and the mean areas of neuronal loss underlying the exposed cortex were compared between the two groups. Cortical lesions were focal and ranged in maximal diameter from one to ten millimeters. Lesions were characterized by neuronal chromatolysis and glial reactivity. There was no significant difference between the mean lesion areas averaged over serial sections between control and experimental hemispheres (1.072 mm² vs. 0.791 mm²). We conclude that lesions result from mechanical factors and that blood on the cortical surface is not in itself toxic in an otherwise healthy brain. "Infarcts" under ASDH may represent pressure, vascular, or mechanical effects, or cumulative excitotoxic insult, but are not due to blood-induced toxicity alone.
33. METABOLIC MANAGEMENT OF NEUROLOGIC TRAUMA: HYPOTHERMIC HYPOKALEMIC COMA

W. Michael Vise, MD (Jackson, MS)

Conventional management of neurologic trauma has no metabolic strategy for the treatment or prophylaxis of injury and has primarily relied upon a mechanical strategy of ICP control. Since the "secondary injury response" is independent upon metabolic support, a regimen was developed to suppress both "basal" and "activation" components of cerebral metabolism and was instituted within a few hours of injury. It was used 43 times in 39 patients, 15 of these were pediatric. Thirty-two were admitted in coma, 31 of which were traumatic. These cases represented the lower end of the coma scale (avg GCS = 4.7). It was also used for neurologic protection during the removal of complex or critically placed lesions within the craniospinal axis. The protocol included: hypothermia 86°F (30°C), hypotension, hypocapnia, pentobarbital, dexamethasone, diphenylhydantoin, cimetidine and fluid restriction. Shivering was blocked with nondepolarizing muscle relaxants and phenothiazines. On average less than 500 ml of 20% Mannitol was given per trauma patient during the hospital stay. Hypothermia was maintained for an average of 4 days in the treatment of coma and limited to the operative period of elective surgical procedures. This regimen produced an altered physiological state characterized by a resetting of acidbase and fluid balance, serum electrolytes, and suppression of the hemodynamic and cellular response to injury. Recognition of these changes and the adoption of new treatment guidelines were essential to successful patient management. Good outcome was improved over conventional management of traumatic coma (GCS>8) and mortality was significantly reduced (P<.0001): 68% good outcome, 3% moderate disability, 6% poor outcome and 23% mortality. No patients were vegetative.

34. COMPLETE CIRCULATORY ARREST, HYPOTHERMIA, AND BARBITURATE CEREBRAL PROTECTION FOR THE TREATMENT OF GIANT VERTEBOBASILAR ANEURYSMS IN PEDIATRIC PATIENTS

Karl A. Greene, MD, PhD, Harold L. Rekate, MD, Robert F. Spetzler, MD (Phoenix, AZ)

Intracranial aneurysms are rare vascular lesions in children, and may occur in a variety of locations. In our series of 17 patients with 21 aneurysms, 4 were located in the posterior circulation; two were complex giant fusiform aneurysms that compressed the verteobasilar junction. Because of their complexity and critical location, hypothermic cardiac arrest was utilized. The patients, a 13-year-old right-handed female and a 9-year-old left-handed male, required far lateral sub-occipital craniotomies, aneurysmorrhaphy, direct clipping, and post-operative external ventricular drainage. Both patients had prolonged post-operative courses requiring tracheostomy, shunting, and extensive multifaceted rehabilitation. The 9-year-old has mild palatal weakness, mild tongue deviation, persistent unilateral vocal cord paralysis, right upper extremity weakness, and a nearly normal gait at 31-month follow-up. Other than competitive athletic endeavors, his activities of daily living are not limited. At 6-month follow-up, the 13-year-old female had multiple mild lower cranial nerve weaknesses, a mild hemiparesis, and gait ataxia. This technique offers several advantages. The surgical field is bloodless. The danger of intra-operative aneurysmal rupture is eliminated. Tension within cerebral vasculature is reduced as is the size of the aneurysm, which permits safer dissection. Aneurysm clips can be precisely applied and/or endaneurysmorrhaphy is possible. This permits preservation of the parent artery and maintains continuity of associated vital branches. Complications involving hemostasis and clotting mechanisms were not encountered in these patients. These techniques can be used to treat complex giant verteobasilar aneurysms in children to prevent the long-term disastrous consequences of aneurysms.
35. MICRO SURGICAL TREATMENT OF ARTERIOVENOUS MALFORMATIONS IN CHILDHOOD

John A. Grant, MD, Abraham Kader, Bennett M. Stein, MD (Chicago, IL)

Between January 1971 and October 1991, 51 patients under 18 yrs. of age underwent operation for ArterioVenous Malformations of the Brain, from a total series of 350 patients treated by the Senior Author. There were 2 poor results. One a child with a massive bilateral AVM who required 2 procedures and in whom a residual AVM was left. The other a child with a medial thalamic lesion who had a Left Hemiparesis post-operatively. All other 49 children did well. One had a short term memory deficit, one a Left arm weakness and one a mild Left spastic hemiparesis. 7 required more than one operation. In 4 patients there was residual after surgery, one of whom was treated with radiosurgery. 9 patients had pre-op embolization of whom 2 had embolization related complications. 3 patients had Aneurysms in association with their AVMs. Follow up averaged 2 years and ranged from months to 13 yrs. These results demonstrate the relative safety of Microsurgical excision of these lesions. In children where, as noted by Matson: “there is expectation of many years in which these lesions may enlarge and cause increasing disability,” and where there is an increased risk of both bleeding and death from bleeding from these lesions an aggressive stance must be taken. Radiosurgery exposes the developing brain to a greatly increased and prolonged risk in children and consequently Microsurgical excision must be considered the best treatment.
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