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

PROGRAM
DECEMBER 6-9, 1988
SCOTTSDALE, ARIZONA
PEDIATRIC SECTION
OF THE
AMERICAN ASSOCIATION OF NEUROLOGICAL SURGEONS

17th Annual Meeting
Marriott's Camelback Inn
Scottsdale, Arizona
December 6-9, 1988

PROGRAM COMMITTEE
Harold L. Rekate
Marion L. Walker
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PAOLO RAIMONDI LECTURERS

E. Bruce Hendrick 1978 Derek Harwood-Nash 1983
Paul C. Bucy 1979 Anthony E. Gallo, Jr. 1984
Floyd Gilles 1980 Frank Nulsen 1985
(Panel Discussion) 1981 William F. Meacham 1986
(Panel Discussion) 1982 Dale Johnson 1987

SCHULMAN AWARD

Kim Manwaring 1983
Neonatal Post-hemorrhagic Ventriculomegaly:
Management with Pulsed Lumber Cisternostomy

Arno Fried 1984
A Laboratory Model of Shunt Dependent
Hydrocephalus

Anne Christine Duhaime 1985
The Shaken Baby Syndrome

Robert E. Breeze 1986
CSF Formation in Acute Ventriculitides

Marc R. Del Bigio 1987
Shunt-induced Reversal of Periventricular
Pathology in Experimental Hydrocephalus

PEDiatric Section Chairman

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

PEDiatric Annual Meeting Sites

Cincinnati 1972 New York 1980
Columbus 1973 Dallas 1981
Los Angeles 1974 San Francisco 1982
Philadelphia 1975 Toronto 1983
Toronto 1976 Salt Lake City 1984
Cleveland 1977 Houston 1985
Philadelphia 1978 Pittsburgh 1986
New York 1979 Chicago 1987
Scottsdale 1988
The Pediatric Section of Neurological Surgery of the American Association of Neurological Surgeons gratefully recognizes the support of the following exhibitors for the 1988 Pediatric Annual Meeting.

Acra-Cut, Inc. - Acton, Massachusetts
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Cordis Corporation - Miami, Florida
Lone Star Medical Products, Inc. - Houston, Texas
Pudenz-Schulte Medical - Goleta, California
Wild Leitz USA, Inc. - Rockleigh, New Jersey

All registrants are encouraged to visit the exhibit area frequently during the meeting.
7:50 a.m. - 8:50 a.m. SCIENTIFIC SESSION I

DYSRAPHISM - Moderators: Concezio DiRocco, M.D.
Barry N. French, M.D.

1. "Central Sleep Apnea in the Chiari I Malformation Associated with Hydromyelia; Evaluation and Surgical Management", B. Mullen, T. Berger, K. Crone and L. Goldstick, Cincinnati, OH

2. "Laryngeal Dysfunction in Infants with Myelomeningocele", P. Kane, J. McLaughlin, D. Shurtleff, J. Loeser and M. Berger, Seattle, WA

3. "Current Surgical Treatment of Chiari Malformations - A Survey of the Pediatric Section of the AANS", S. Haines and M. Berger, Minneapolis, MN

4. "Neuroectodermal Appendages: The Human Tail Explained", S. Gaskill and A. Marlin, Durham, NC

5. Progressive Myelopathy Decades Following Surgery for Cervical Meningocele", J. Wu and R.M. Scott, Boston, MA

8:55 a.m. - 10:00 a.m. SCIENTIFIC SESSION II

DYSRAPHISM - Moderators: Thomas Waltz, M.D.
Patricia Aronin, M.D.


7. "153 Cases of Retethering of the Spinal Cord", J. Herman, D. McLone and T. Myer, Chicago, IL

* Indicates Resident Paper

11:45 a.m. - 1:00 p.m. SCIENTIFIC SESSION IV

HYDROCEPHALUS - Moderators: David Klein, M.D., Michael Turner, M.D.


22. "Prospective Evaluation of Outcome of Neonatal Post-Hemorrhagic Ventriculomegaly and Determination of Best Predictors for Good Outcome", A. Canady, S. Shankaran, A. Eisenbrey and T. Koepke, Detroit, MI

THURSDAY, DECEMBER 8, 1988

6:30 a.m. - 7:00 a.m. Registration

6:30 a.m. - 7:00 a.m. Continental Breakfast

7:00 a.m. - 8:24 a.m. SCIENTIFIC SESSION V

BASIC SCIENCE - Moderators: Harold Portnoy, M.D., Andrew Parent, M.D.


27. "Effects of Increased Intracranial Pressure and Hyperventilation on AVO2 and Brain Metabolism--Implications Regarding Management of Head Injured Patients", L. Sutton, A. McGloughlin, S. Dante and M. Kotapka, Philadelphia, PA


8:24 a.m. - 10:00 a.m. SCIENTIFIC SESSION VI

TUMORS I - Moderators: Harold Hoffman, M.D. Maurice Choux, M.D.


32. "Pediatric Oligodendrogliomas", L. Foody, K. Crone, T. Berger, W. Ball and E. Ballard, Cincinnati, OH

33. "Expression of the N-myc, c-myc, c-src, and int-1 protooncogenes in medulloblastoma cell lines: Implications for Neurohistopathology", D. Ross, H. Saya, M. Edwards and V. Levin, San Francisco, CA

34. "Brain Stem Tumors in Children", M. Choux, J.M. Riss, L. Genitori and G. Lena, Marseille, FRANCE


10:00 a.m. - 10:30 a.m. COFFEE BREAK

10:30 a.m. - 11:30 a.m. SCIENTIFIC SESSION VII

TUMORS II - Moderators: John Shillito, M.D. Gail Magid, M.D.

38. "Bone Metastasis of Medulloblastoma in Childhood", T. Tomita, M.A. Radkowski, D. McLoone and K. Bauer, Chicago, IL


40. "Postoperative Posterior Fossa Intracranial Pressure Monitoring", W. Boydston, R. Sanford and M. Muhlbaumer, Memphis, TN


11:30 a.m. - 12:30 p.m. SCIENTIFIC SESSION VIII

TUMORS III - Moderators: Fred Epstein, M.D. Dennis Johnson, M.D.

43. "Primative Neuroectodermal Tumors: Improved Survival with 8 in 1 Chemotherapy", R. Brightman, E. Kosnik and F. Buymann, Columbus, OH

*45. "Brain Tumors Below One Year of Age: A Retrospective Analysis of Twenty-Two Patients (1978-1987)", S. Haddad, A. Menezes and J. Godersky, Iowa City, IA

46. "Intracranial Tumors in 332 Infants Below Age Three", C. Wang, Beijing, P.R. China

47. "High Dose of Chemotherapy After Autologous Bone Marrow Transplantation for the Treatment of Pediatric Malignant Brain Tumors", S. Yu-Chen and W. Dao-Xin, Shanghai, P.R. China

12:30 p.m. - 1:00 p.m. BUSINESS MEETING (Members Only)

7:30 p.m. - 11:00 p.m. ANNUAL BANQUET

FRIDAY, DECEMBER 9, 1988

6:30 a.m. - 7:00 a.m. Registration

6:30 a.m. - 7:00 a.m. Continental Breakfast

7:00 a.m. - 8:00 a.m. SCIENTIFIC SESSION IX

RADIOLOGY AND CYSTS -Moderators: Anthony Gallo, M.D.
Robert Douver, M.D.

Henkelman and H. Hoffman, Toronto, ON, CANADA

Malik and M. Payne, R.N., Charlottesville, VA

50. "Neonatal Cerebral Venous Thrombosis (NCVT)", P.
Tracy, R. Wright, W. Tadros and William Hanigan,
Peoria, IL

8:00 a.m. - 10:00 a.m. SCIENTIFIC SESSION X

SPINE AND TRAUMA -Moderators: Thomas Luerssen, M.D.
Martin Johnson, M.D.

51. "Multiple Arachnoid Cysts of the Cerebral Hemisphere in Infancy", P. Gianaris and T. Tomita, Chicago, IL

52. "Congenital Arachnoid Cysts in Children", B.
Chidambaram, J. Laurent, W. Cheek, N. Feldstein, E.
Rouah and S. Schneider, Houston, TX

Hochhauser, D. Long, Syracuse, NY


56. "Immediate Surgical Exploration for Trans-Tentorial Herniation in Pediatric Head Injury", B. Andrews, L.
Pitts and M. Edwards, San Francisco, CA

57. "Craniocephalic Gunshot Wounds in Children", M.
Muhlbauer, R. Sanford, W.C. Clark and J. Simmons,
Memphis, TN


59. "How are Symptomatic Chronic Subdural Fluid Collections Best Managed in the Pediatric Patient?", S. Litofsky, J.G. McComb and C. Raffel, Los Angeles, CA
10:00 a.m. - 10:30 a.m. COFFEE BREAK

10:30 a.m. - 11:45 a.m. SCIENTIFIC SESSION XI

FUNCTIONAL - Moderators: Bruce Bressler, M.D.
Fred Murtagh, M.D.


64. "Results in Callostomy in Retarded Patients with Drop Attacks", T. Gravitz, G. Meyer, G. Harrington and J. Murphy, Milwaukee, WI

65. "Histologic Changes in Dorsal Rootlets from Children with Cerebral Palsy", R. Rovin, Y. Ersahin and B. Storrs, Chicago, IL


11:45 a.m. - 1:00 p.m. SCIENTIFIC SESSION XII

FUNCTIONAL AND VASCULAR - Moderators: Hal Pittman, M.D.
John Laurent, M.D.
1. CENTRAL SLEEP APNEA IN THE CHIARI I MALFORMATION ASSOCIATED WITH HYDROMYELIA: EVALUATION AND SURGICAL MANAGEMENT

Bradley Mullen, Thomas S. Berger, Kerry R. Crone, Lawrence Goldstick (Cincinnati, OH)

The Chiari I malformation consists of cerebellar tonsillar herniation into the upper cervical canal without brainstem involvement. While accompanying hydrocephalus is infrequently reported, hydromyelia is associated with this anomaly in up to one-third of cases.

Central sleep apnea is defined by a lack of airflow lasting longer than ten seconds without evidence of diaphragmatic excursion. While most often this disorder is not associated with recognizable structural pathology, it may be due to cervicomedullary compression at the foramen magnum level with compromise of the involuntary respiratory centers. Central sleep apnea has frequently been associated with the Chiari II malformation; however, the association of central sleep apnea, hydromyelia, and the Chiari II malformation has only rarely been reported.

We report three children, each with progressive neurologic deficits and severe polysomnogram documented central sleep apnea associated with absent hypoxemic and hypercapnic arousal responses. The apnea/hypopnea indices Chiari I malformation with cervicomedullary hydromyelia in each. Treatment consisted of decompressive suboccipital craniectomy with cervical laminectomy and subarachnoid shunting of the hydromyelic cavities. Intraoperative ultrasound assisted in surgical decision making. Long-term follow-up with MR imaging has demonstrated marked reduction in syrinx size and repeat polysomnography exhibited absolute reduction in apneic hypopneic indices in all.

We suggest that central sleep apnea may become a feature of Chiari I deformity when associated with hydromyelia of sufficient size to cause compression of the medullary and
1. CENTRAL SLEEP APNEA IN THE CHIARI I MALFORMATION ASSOCIATED WITH HYDROMEelia: EVALUATION AND SURGICAL MANAGEMENT
(Con't)

cervical respiratory centers. MR imaging is unparalleled in defining this association. Prompt surgical intervention has proven rewarding in treating these patients.

2. LARYNGEAL DYSFUNCTION IN INFANTS WITH MYELOMENINGOCELE

Paul M. Kaney, Jeff M. McLaughlin, David Shurtleff, John D. Loeser, Mitchell S. Berger (Seattle, WA)

Laryngeal dysfunction (LD) presenting as stridor, apnea, and/or respiratory failure may complicate the clinical course of infants with myelomeningocele (MMC). Thirty-five of 616 MMC patients seen at Children's Hospital in Seattle, Washington, between 1962 and 1982 -- Group I had LD in the absence of infection or shunt malfunction. Since 1982, ten of 225 MMC patients (Group II) developed LD.

In Group I patients, time of onset of LD after birth was: 0-1 month (24 patients), 2-6 months (9 patients), and 7-12 months (2 patients). All children in Group II had symptoms when less than one month of age. LD did not correlate with severity of paralysit, degree of hydrocephalus, sex, or gestational age. 63% of the children in Group I and all of the children in Group II had associated cranial nerve dysfunction (CND), including delayed head control, swallowing dysfunction, and facial or tongue paresis. Severity of LD fluctuated with ICP (determined via shunt manometric) in 14 cases and persisted following shunt revision.

Treatment of children in each group included aggressive management of ICP, airway compromise, and CNS infection. Nineteen children of Group I (54%) and three in Group II (30%) underwent posterior fossa decompression. All children had barium esophagrams and laryngoscopy, and children in Group II underwent polysomnographic monitoring. Mortality in Group I was 69.8%, and LD or CND was present at death in 22 of 24 cases. Nineteen of 24 deaths occurred before the age of 30 months, and LD resolved before 30 months in 10 of 11 survivors. One of the patients whose LD persisted beyond 30 months is alive. Developmental progress is good in seven, intermediate in two, and poor in two survivors. There has
2. LARYNGEAL DYSFUNCTION IN INFANTS WITH MYELOMENINGOCELE
   (Con't)

been one death in Group II, and four children with LD persisting beyond 30 months remain alive. The
developmental progress is good in five, intermediate in three, and poor in one child.

We conclude that LD represents an ominous finding in an infant with MMC. LD contributes to the morbidity and
mortality of infants with MMC regardless of other features, such as motor level, which suggest a good prognosis.
There has been no mortality in the children in Group II who have undergone posterior fossa decompression,
suggesting an improved outcome following surgery.

3. CURRENT SURGICAL TREATMENT OF CHIARI MALFORMATIONS -
   A SURVEY OF THE PEDIATRIC SECTION OF THE AANS

Stephen J. Haines (Minneapolis, MN), Mitchell S. Berger,
(Seattle, WA)

A survey of the membership of the Pediatric Section of the AANS was carried out to assess current treatment practices
for Chiari malformations Type I and II. Of 159 questionnaires initially sent out, two could not be
completed because of retirement or because the member was not a neurosurgeon. Ninety-nine responses were obtained
from the remaining 157 members (63%). Sixty percent of the respondents devote at least 50% of their practice to
pediatric neurosurgery. On the average, the respondents estimate that they evaluate four patients for Chiari Type
I and 15 patients for Chiari Type II problems each year. The total estimated number of procedures done by all
respondents for treatment of Chiari malformations was 1796. They estimate on the average that they have done 4
procedures in children less than 2 years of age, 4 procedures in children 2 to 15 years of age and 2
procedures in adults greater than 15 years of age in the past year.

Only two respondents felt that surgical decompression of a Chiari malformation is never indicated. There was
substantial agreement that surgical decompression was indicated for respiratory, brain stem or cranial nerve
dysfunction. There was also substantial agreement that prophylactic decompression of the Chiari malformation in
asymptomatic patients was not indicated. There was considerable disagreement about the value of decompression
in treating nystagmus and hydro syringomyelia in both Type I and Type II Chiari malformations. The most common
operation performed regardless of symptomatology is a combined cervical laminectomy and suboccipital cranietomy
with dural patch grafting but without significant intradural dissection. Lysis of arachnoid adhesions,
plugging of the obex or placement of a ventriculosub-
arachnoid drainage tube is most commonly done in those
3. CURRENT SURGICAL TREATMENT OF CHIARI MALFORMATIONS - 
A SURVEY OF THE PEDIATRIC SECTION OF THE AANS 
(Con't)

patients with cranial nerve and brain stem symptoms. However, even in these patients a substantial proportion of surgeons report treating only with dural patch grafting and bony decompression.

Forty-six percent of respondents felt that the existing classification of Chiari malformation does not adequately describe the range of abnormalities being seen by MRI. Ninety-five percent feel that surgical treatment of Chiari malformations is not sufficiently consistent that one could claim that there is a "standard" treatment.

There appears to be a substantial variation in the indications for and techniques of surgical treatment of Chiari malformations Types I and II among pediatric neurosurgeons in the United States. Cooperative efforts to better define the natural history and indications for specific types of surgical treatment are needed.

4. NEUROECTODERMAL APPENDAGES: THE HUMAN TAIL EXPLAINED

Sarah J. Gaskill (Durham, NC), Arthur E. Martin (San Antonio, TX)

The human tail has been intermittently described in the literature since the early 1900's. These have typically been isolated cases presented primarily with intrigue and medical curiosity. Presented here is a series of six neuroectodermal appendages with a proposal for their etiological development. The notion of the human tail both as an embryologic remnant and as a consequence of the amniotic band syndrome will be addressed. In addition, the important issue of differential diagnosis of a midline lesion will be considered briefly.

In conclusion the material presented will support a theory of the superficial extension of a dermal sinus tract in the formation of neuroectodermal appendages. These are characterized by: a posterior localization in or near the midline, a tubular or "tail-like" appearance, extension of the appendage into the spinal canal with attachment to neural elements, variable vertebral defects and occasionally an associated appendage which may appear as either a separate entity (probably due to breakage during development) or in connection with the posterior appendage. The appropriate evaluation and treatment of neuroectodermal appendages will also be considered.
5. PROGRESSIVE MYELOPATHY DECADES FOLLOWING SURGERY FOR CERVICAL MENINGOCOELE

Julian K. Wu, R. Michael Scott (Boston, MA)

Two patients who underwent repair of cervical meningocoele early in life presented at ages 37 and 43 with progressing myelopathy. MRI demonstrated dorsal tethering of the cervical cord at the area of the previous repair associated with intramedullary cysts. Surgery confirmed the MRI findings. One patient markedly improved post-operatively. The families of children with cervical meningocoele should be advised of the potential for late neurologic deterioration, and a baseline post-repair MRI should be obtained in most circumstances. Patients with abnormal studies probably warrant life-time neurologic follow-up.

6. SURGICALLY CREATED DYSPHAGISM IN THE FETAL RAT: IMPLICATIONS FOR THE CAUSE OF PARALYSIS IN SPINA BIFIDA

Dan S. Heffez, John Aryianpur, John M. Freeman, Grover M. Hutchins, (Baltimore, MD)

The lower extremity paralysis in children with spina bifida has been attributed principally to the developmental abnormality. Certain pathological findings, however, suggest that the malformed spinal cord may be damaged by exposure to the amniotic fluid. We tested this hypothesis using a model of surgically created dysphagism in the fetal rat.

The Sprague-Dawley rat fetus underwent a 3 level laminectomy on day 18 of the 22 day gestation. The dura was opened and the fetal spinal cord exposed. In the experimental group, the fetal skin incision was left open. In the control group, the skin wound was sutured. The fetus was then replaced in the uterus and the uterotomy was closed. Therefore, in the experimental group the spinal cord remained exposed to the amniotic fluid for the remainder of gestation. In the control group, the spinal cord was never directly exposed to the intrauterine environment. Gestation continued until term.

Each of the 14 liveborn rats from the experimental group had weak and deformed hind limbs. One pup had a congenital kyphosis. In each case, there was extensive necrosis of the exposed spinal cord. In contrast, 6 of 9 control rats had completely healed wounds and were indistinguishable from unoperated littersmates. In the remaining 3 control rats, the wound had dehisced in utero; these pups also had weak or deformed hind limbs.

Our results strongly suggest that the spinal cord can be severely injured by exposure to the amniotic fluid. The clinical and pathological similarities between our model and spina bifida are striking.
7. 153 CASES OF RETETHERING OF THE SPINAL CORD

James M. Herman, David G. McLone, Theresa Meyer (Chicago, IL)

After primary repair of myelomeningoceles and lipomyelo-
meningoceles, deterioration of gait, muscle strength,
urinary control, as well as progressive orthopedic
deforities is well documented. In our experience serial
examination and early correction of tethered cord in these
patients has led to significant improvement in the
patient's overall status.

Of 341 tethered cord releases done from 1981 to 1988, we
are reporting on 153 (45%) patients. Ninety-seven were
performed after primary closure of a myelomeningocele and
56 after repair of a lipomyelomeningocele, the results
reported here are a retrospective review of the
presentation, treatment and outcome. The average age of
the myelomeningocele patient was 6 years old with no
difference in distribution of sex. The most common
presenting symptoms and signs of the myelomeningocele
patients were change in gait and motor strength 51%,
scoliosis 49%, spasticity 41% and pain 29%. Lipomyelo-
meningoceles were an average of 8 years old with female
predominance. These patients most commonly presented with
pain radiating into the legs 60%, subtle atrophy or
orthopedic foot deformities 46% and lumbosacral
38%. Operative dissection was performed with the CO2 laser
in all cases.

Follow-up over a 4 month to 7 year period revealed an
overall improvement in 71%, no change in 21% and
deterioration in 2%. A second untethering was required in
16 patients and a third in one. There were no mortalities.

Close follow-up of myelomeningocele and lipomyelo-
meningoceles patients with serial physical exams and
radiographic evaluations has documented a clearly
correctable deformity. Long term follow-up has shown a
significant overall improvement in both presenting
symptoms and signs.

8. PREVENTION OF SPINAL CORD RETETHERING, DEVELOPMENT OF
AN ANIMAL MODEL

Susanne E. Fix (Louisville, KY), Michael S. Turner
(Indianapolis, IN)

Procedures for the release of a tethered cord have been
done for many years. Several years later many of these
patients are returning with recurrent symptoms of tethered
cord syndrome. Several maneuvers have been discussed to
prevent retethering, but none have been experimentally
tested.

Six juvenile Hampshire pigs weighing approximately 40
pounds were obtained for the study. A two level lumbar
laminectomy was performed on each animal. The dura was
exposed and then mechanically stripped from the most
central portion of the exposed cord. The dura was then
closed. At one month the laminectomy site was evaluated
preoperatively with real time ultrasonography and then
re-opened to grossly document scarring and release any
tethering. At two months, the animals were sacrificed and
the involved cord segment submitted for histo pathologic
evaluation.

Cord tethering was demonstrated in all animals grossly and
historically. Ultrasound proved to be a reliable
noninvasive means of documenting dural scarring,
particularly in the more severe cases.

We have developed an easily reproducible model using young
Hampshire pigs to produce spinal cord retethering. The
retethering is documented using real time ultrasound and
pathological specimens.

Our model will be used in future experiments to test
various arachnoid substitutes, suture materials and graft
materials and their effect on prevention of spinal cord
retethering.
8. PREVENTION OF SPINAL CORD RETETHERING, DEVELOPMENT OF
ANIMAL MODEL
(Con't)

This simple, easily reproducible model of the tethered spinal cord will have numerous clinical applications, especially with regard to cerebral adhesions and the study of scar formation.

9. UROLOGIC OUTCOME FOLLOWING SURGICAL TREATMENT OF
LIPOMYELOMENINGOCELE

Philip H. Cogen, Lionel S. Foster, Barry A. Kogen, Joyce Szymanski, Michael S.B. Edwards (San Francisco, CA)

Surgical treatment for lipomyelomeningocele has been controversial when the patient is asymptomatic. However, urologic testing may reveal disturbances of bladder function unnoticed by the patient, especially in the infant age group. As prior studies have demonstrated that operative intervention for lipomyelomeningocele may not reverse pre-existing neurologic deficits, even when well-performed, we reviewed a series of patients who underwent pre-and post-operative urologic testing to evaluate the functional outcome of therapy.

Thirty-two patients had surgical correction of their lipomyelomeningoceles. This involved excision of as much of the lipomatous mass as was possible without injury to neural structures, untethering of the spinal cord, and correction of hydromyelia when present. All of the patients underwent pre-and post-operative urodynamic and cystometric studies. The results of the analysis were evaluated by age at the time of operative repair. Of 14 infants and children less than 1.5 years of age at the time of operation, 6 (43%) were found to have normal pre- and post-operative urologic studies, and an additional 4 patients (28%) achieved normalization post-operatively. However, in the 18 patients greater than 1.5 years of age at the time of surgery, only 4 (22%) demonstrated normal pre-and post-operative bladder function, and only 1 additional patient achieved normalization following the surgery.

The results of this study demonstrate that: (1) the majority of patients with lipomyelomeningoceles have impairment of bladder function pre-operatively, and (2) normalization of bladder function is possible following repair of the lipomyelomeningocele, but only in the group of patients less than 1.5 years of age. We therefore
9. UROLOGIC OUTCOME FOLLOWING SURGICAL TREATMENT OF LIPOMYELOMENINGOCELE (Con't)

recommend that all patients with lipomyelomeningoceles undergo pre-and post-operative urologic testing, and that definitive corrective surgery be performed at as early an age as is possible.

10. A DESCRIPTIVE ANALYSIS FOR CORRECTION OF LAMBDOMASTOID CRANIOSYNOSTOSIS: "HINGE PROCEDURE"

Jerry O. Penix, Petra Gurtner, Roberta M. Stewart (Norfolk, VA)

From 1984-1988, 59 children from the age of one month to eleven years were evaluated for surgical correction of craniosynostosis. Forty-three patients underwent surgery.

Twenty out of twenty-two infants with premature closure of the lambdomastoid sutures were operated upon. Five patients out of this group underwent modified occipital craniotomies with a "hinged gate" procedure. This procedure consists of the construction of a recontoured occipital plate which is fixed and hinged on the affected side. Here is our description of this technique.
11. MIGRAINE HEADACHES IN HYDROCEPHALIC CHILDREN: A DIAGNOSTIC DILEMMA

Hector E. James, Thomas Nowak (San Diego, CA)

Headaches, vomiting, and altered sensorium can be seen in patients with shunt malfunction as well as in-patients with migraines. We report five patients in which children with hydrocephalus and CSF shunts presented with a variety of recurrent symptoms including headache, vomiting, impairment of consciousness, and in one patient, decerebrate posturing. Various diagnoses were entertained: shunt malfunction, slit ventricle syndrome, and low pressure (over shunting). Repeat procedures were performed in all, including shunt taps, multiple shunt revisions, and a subtotal decompression. When the diagnosis of migraine was entertained, four patients improved on Propranolol therapy; one failed to this therapy but responded to Verapamil.

It is concluded that in those patients with hydrocephalus and repeated bouts of symptoms such as headaches, vomiting, impairment of consciousness, in the face of documented adequate shunt function as well as a family history of migraine headaches, the diagnosis of migraine be entertained before repeated operative interventions are performed and/or changes in valve shunt pressures. This disease entity with hydrocephalus has not been discussed by pediatric neurosurgeons in the past, and should be addressed in the face of repeated publications and discussions on over shunting, slit ventricle syndrome and related.

12. SHUNT MALFUNCTION WITH "NORMAL" INTRACRANIAL PRESSURE

Robert A. Sanford, Michael S. Muhlauer (Memphis, TN)

The elevated intracranial pressure of shunt malfunction in hydrocephalics produce the symptoms of headache, vomiting and lethargy. In the majority of shunt obstruction cases, computed tomography scanning reveals ventricular enlargement. Ten shunted, hydrocephalic patients, nine children and one adult, are described who had symptomatic ventriculomegaly but normal or low intracranial pressure. This group had a variety of underlying etiologies of hydrocephalus including aqueductal stenosis, myelomeningocele, Dandy Walker cyst, intraventricular hemorrhagic of prematurity, post traumatic, post subarachnoid hemorrhage and posterior fossa tumor. Pressure measurements via reservoir puncture in the recumbent position revealed an intracranial pressure measurement of 2 to 16 centimeters of water. In the upright position, the pressure decreased but remained a positive value in contrast to the control population whose pressures reached negative values of -10 to -40 cm of water. Computed tomography scans revealed either severe ventriculomegaly or a pronounced increase in ventricular size when compared to the baseline scans obtained while patients were asymptomatic. Shunt dynamics revealed a free egress of fluid and distal run off. Two children had contrast shuntograms which demonstrated shunt patency. Nine patients required shunt revisions to relieve symptoms and reduce ventriculomegaly. One child with a thoracic myelomeningocele improved after reducing intra-abdominal pressure by changing a valveless ventriculocephalic shunt before improving clinically and anatomically.

Conclusion: A population of shunted patients exist who require a negative pressure to maintain normal ventricular size and remain symptom free. In this group of patients, the etiology of hydrocephalus is as varied as is their age of presentation.
13. WHAT IS THE SIGNIFICANCE OF CSF EOSINOPHILIA IN CHILDREN WITH VENTRICULO-PERITONEAL SHUNTS?

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

To determine if ventricular CSF eosinophilia is meaningful, the charts of 106 patients who presented to Children's Hospital of Los Angeles for shunt related procedures for the calendar year 1985 were reviewed. Seventy patients presented for a shunt revision; their charts were retrospectively reviewed from the time of shunt insertion until January 1988. The remaining 36 patients had a ventriculo-peritoneal shunt inserted during the study period and were subsequently followed to January 1988. The mean follow-up period was 6.9 years (range 2-20 years). Of the 106 patients, 17 had shunt insertion only, 21 had an insertion and a single revision, 24 had three shunt related procedures, and 44 had more than three. The 106 patients underwent a total of 558 shunt related procedures with 21 shunt infections (3.8%).

Ventricular CSF eosinophilia, defined as 3% or more eosinophils in the total white count, occurred in 34 patients. Those patients with eosinophilia experienced a mean shunt revision rate of 8.5, while in the non-eosinophilic group it was 2.5 (p<0.001). Shunt infections were also more frequent in patients with eosinophilia (p<0.01). In no case was there evidence of peripheral eosinophilia or parasitic infection.

In summary, a shunt infection should be considered in any patient with ventricular CSF eosinophilia. This group of patients will experience a significant increase in the number of shunt obstructions over those patients who do not have this finding.

14. FUNCTION OF PARIETAL AND FRONTAL SHUNTS IN CHILDHOOD HYDROCEPHALUS

A. Leland Albright (Pittsburgh, PA), Stephen J. Haines (Minneapolis, MN), Floyd H. Taylor (Pittsburgh, PA)

This study was performed to determine if cerebrospinal fluid (CSF) shunts inserted via the frontal and parietal regions function for similar lengths of time. The medical records of 114 children with CSF shunts and 83 of their computed tomography (CT) scans were reviewed. Ninety percent of the operations were to insert the child's first shunt. The site of insertion, cause of hydrocephalus, age, surgeon, duration of function (time from insertion to malfunction or to latest follow-up), presence of infection, catheter location within the ventricle, and duration of function of the subsequent shunt were recorded. Data were analyzed by chi-square, logistic regression and life-table methods. Shunts had been inserted via the frontal route in 62 children and via the parietal route in 52. Ages, causes of hydrocephalus and infection rates were similar in both groups. Duration of shunt function was predicted by the site of shunt insertion and the catheter position within the ventricles: shunts inserted via the frontal region functioned significantly longer than parietally inserted shunts, both as the initial shunt (Wilcoxon p=0.0008) and after a malfunction, and catheters positioned within the ipsilateral frontal horn functioned significantly longer than those in other ventricular locations (Wilcoxon p=0.03).
This study identifies differences in blood/CSF barrier function during halothane vs. thiopental/sufentanil anesthesia, as measured by CSF/serum ratios of IgG and Albumin. Ten children (both sexes, age range 6 mo. - 15 yrs.) with known hydrocephalus underwent ventriculoperitoneal shunt placement or revision. Those children with known tumor, diabetes, or infection were excluded from the study. Subjects were induced with IV methohexital (0.5 - 2 mg./Kg.) and vecuronium. Sample 1 of serum albumin and IgG, and lumbar/ventricular CSF albumin, IgG, glucose, cell count and differential was obtained under N2O/O2 prior to maintenance anesthesia. Subjects were then assigned either to a sufentanil/thiopental group (n=5) (vecuronium, sufentanil 0.5 mg/kg bolus followed by 1-4 mg/kg/hr infusion) or a halothane group (n=5) (endtidal 0.25-1.25% vecuronium as needed). Mechanical ventilation was maintained at O2 sat >95, endtidal CO2 32-36. Body temperature was maintained at 35 - 37.3° C. Sample 2 of blood and ventricular CSF was obtained at time of insertion of the ventricular end of the shunt. Sample 3 was obtained just prior to surgical closure. Halothane, sufentanil and thiopental levels were monitored at each of these times. A statistically significant difference between the two anesthesia groups was demonstrated at S3 (P < .05). Children undergoing halothane anesthesia showed increased CSF/serum ratios of IgG and albumin, those undergoing thiopental/sufentanil anesthesia did not. It is known that Halothane increases cerebral blood flow and intracranial pressure by decreasing cerebrovascular resistance, whereas narcotics and thiopental decrease cerebral blood flow and increase resistance. According to Reiber, acute elevations in protein quotients (CSF/serum) of the type uncovered by this study are consistent with increased permeability of the blood-CSF barrier. Our study indicates, therefore, that clinical concentrations of halothane but not thiopental/sufentanil disrupt the blood-CSF barrier in humans.
ULTRASTRUCTURAL EXAMINATION OF CSF OUTFLOW PATHWAYS IN ACUTE KAOLIN HYDROCEPHALUS

J. Gordon McComb, Floyd H. Gilles, Shigeyo Hyman (Los Angeles, CA)

To what extent are the lymphatic cerebrospinal (CSF) outflow pathways affected in acute kaolin hydrocephalus? Previous studies have examined the distribution of a kaolin/carbon black mixture injected into the lateral ventricles of rabbits from a gross and microscopic standpoint. The present study extends the observations to an ultrastructural level.

New Zealand white rabbits underwent injection of a kaolin/carbon black mixture into both lateral ventricles followed by a four-hour infusion of artificial CSF. The animals were then infused with Karnovsky's mixture into the heart. The tissues were prepared for electron microscopy using standard techniques.

Both carbon black, amorphous particles .3 x .6μ, and kaolin, crystals .5 x 1.5μ, were found to be present in the lymphatic channels surrounding the olfactory fila, and in the episcleral tissue. The particles did not penetrate beyond the subarachnoid tissue barrier at the exit of other cranial nerves, spinal nerves, major blood vessels at the cranial base, the strum of the choroid plexus, below the pial surface or dural lymphatics.

It is concluded that large particulate matter can enter the extra-cranial lymphatics from the olfactory and optic nerves in good quantity four hours after being injected into the ventricles.

PATHOPHYSIOLOGY OF THE ISOLATED VENTRICLE IN MYELODYSPLASTIC CHILDREN

Mitchel S. Berger, David K. Brewer, David W. Newell, Paul M. Kanev (Seattle, WA)

We have reviewed a group of children with myelomeningocele (MMC) who developed an isolated ventricle following shunt insertion for progressive hydrocephalus. The pathophysiology of this phenomenon has been elucidated with Computed Tomography (CT) and multiplanar (T1/T2-weighted) Magnetic Resonance Imaging (MRI), and will be described in detail.

Eight newborns (4 male, 4 female) with a lumbosacral MMC underwent closure of the defect within 1-3 days after birth. Preoperative cranial ultrasound and/or CT confirmed enlarged, symmetric ventricles. The first shunt was placed within 14 days following MMC closure. In all patients, a low pressure Unishunt (7-posterior, 1-coronal placement) preceded development of an isolated contralateral ventricle. Two of eight patients originally had medium pressure (non-Unishunt) systems placed, however required revision to a low pressure system (Unishunt) due to persistent, symmetric ventriculomegaly. The CT Scan, and MRI in particular, demonstrated a consistent finding, namely, a collapsed ventricle and torqued Foramen of Monro ipsilateral to the shunt, and a dilated (opposite) lateral and third ventricle with a patent Foramen of Monro contralateral to the shunt. MMC patients with medium pressure shunts (all types) and non-MMC hydrocephalic children with low pressure shunts (all types) did not develop this finding. Six of eight children required a second contralateral shunt for symptomatic isolated ventriculomegaly.

In conclusion, MMC children treated with low pressure Unishunts are susceptible to developing isolated ventriculomegaly due to excessive drainage and subsequent torquing of the Foramen of Monro ipsilateral to the shunted ventricle. Distorted anatomy, in MMC patients, adjacent to the Foramen of Monro, i.e., enlarged massa intermedia, eccentric anterior thalamic tubercle, etc., most likely contributes to this phenomenon, thus preventing drainage of the isolated ventricle via the (trapped) contralateral shunt catheter.
18. MORPHOLOGICAL EFFECTS OF HYDROCEPHALUS ON THE CEREBRAL CORTEX IN NEONATAL KITTENS

Lyn Carey Wright, James Patrick McAllister, Stephen Katz, Thomas J. Lovely, David Miller (Philadelphia, PA)

Studies have been undertaken demonstrating the effect of hydrocephalus on the periventricular white matter in adult animal models. This present study evaluates the effects of hydrocephalus on the cortical mantle of our neonatal feline model of hydrocephalus. In order to induce hydrocephalus, the cisterna magna of four to eleven day old kittens was injected percutaneously with a solution of 25% kaolin. Clinically, splitting of the sutures with minimally enlarging anterior fontanelles could be detected at two to four days post-injection. Ultrasonic evidence of hydrocephalus was allowed to progress until day eighteen to twenty-five post-injection at which time the animals were sacrificed. The gross and cytoarchitectural changes of the cortical mantle in affected animals were compared with control age-matched counterparts who had undergone saline injections. Areas 4 (motor), 22 (association) and 17 (sensory) were examined. There was a substantial increase in the severity of changes noted in a rostral to caudal direction with area 4 being least affected and area 17 being most severely involved. A decrease in number as well as a disturbance in orientation was demonstrated in both the neuronal cell population and blood vessels. Neurons in areas least affected by hydrocephalus demonstrated vertically oriented dendritic processes. As hydrocephalus progresses these processes became horizontally directed. In the most severe cases no distinct pattern was observed. Furthermore, the deeper cortical layers were more affected than the more superficial laminae, in that more reactive and pyknotic neurons were present in layers V and VI. As the hydrocephalus became more severe, changes could be observed in the intracortical neurons within layers II and III. There were also notable changes in blood vessels regarding orientation and number. The degree of hydrocephalus correlates well with both the amount of neuronal cell loss and the notable paucity of blood vessels in severely affected areas. We postulate that the motor deficits seen with hydrocephalus as well as subtle cognitive deficiencies can be attributed to these neuronal and vascular changes.
19. VENTRICULOSONIC GUIDANCE FOR PRECISE POSITIONING OF THE VENTRICULOSONIC SHUNT: A NEW TECHNIQUE

Kim H. Manwaring, Harold Rekate, (Phoenix, AZ), Robert Sullivan, Frank Gugliomo (Codman & Shurtleff, Randolph, MA)

The use of an endoscope within the ventricular system has been advocated to facilitate recognition of abnormal anatomy and placement of ventriculoperitoneal shunts. We have developed a new technique of ventriculoscopic guidance to select a position for the proximal catheter and to position it precisely at that location. Further, endoscopic features of the foramen of Monro and the cavity of the III ventricle are appreciated.

Utilizing a #9 French peelaway sheath with its stiffening dilator, the lateral ventricle is entered from either the coronal or occipital approach. Upon removal of the dilator, a Codman prototype ventriculoscope is passed through the peelaway sheath. The peelaway sheath and ventriculoscope are then passed in concert under direct ventriculoscopic guidance using the video monitor into the anterior frontal horn from an occipital approach or into the thalamostriate and septal veins are readily identified.

An abnormal foramen of Monro is often recognized in children with myelomeningocele. Within the III ventricle, features of the optic chiasm, suprachiasmatic recess, and infundibular recess are recognized. The very thinned floor of the third ventricle is often translucent, allowing recognition of mammillary bodies and basilar artery tip occasionally. After selection of the site for catheter positioning, the ventriculoscope is removed and the proximal catheter passed through the peelaway sheath. The sheath is then peeled, leaving the proximal catheter at the selected site precisely.

The ventriculoscope has been employed successfully in over 50 procedures to date without complication. Routinely, such visualization adds five minutes to operating time.

In consideration of extension of visualization beyond the period in which ultrasound is practicable, ventriculoscopic guided shunt placement is a valuable adjunct in all pediatric age groups.

20. TRANSCRANIAL DOPPLER: A NON-INVASIVE METHOD FOR MONITORING HYDROCEPHALUS AND INCREASED INTRACRANIAL PRESSURE

Alexis Norelle, Ann Marie Flannery, Asma Q. Fisher, Paul Frankel (Augusta, GA)

The ability to discriminate between ventriculomegaly and hydrocephalus often relies on clinical impression and parameters such as change in ventricular size and configuration, increasing head circumference, and evidence of developmental delay.

In an effort to confirm clinical impressions, 33 children underwent transcranial Doppler. Twenty-seven had ventriculomegaly confirmed by CT, ultrasound, and/or MRI. Nineteen were infants; 14 were older children with closed fontanels. Three normal children were studied, and 3 children were monitored for increased I.C.P. from head trauma.

Causes of ventriculomegaly or factors which put these children at risk included intraventricular hemorrhage (9), myelomeningocele (4), idiopathic (1), and congenital anomalies including aqueductal stenosis (1), schizencephaly (1), agentis of corpus callosum (1), and Joubert's syndrome (3). Other children seen had postmeningeal ventriculitis (2), thrombosed vein of Galen aneurysm (1), and obstruction secondary to tumor (3).

In all cases the Gosling Pulsatility index (P.I.) was compared with head circumference, clinical and radiographic impressions. In 3 cases, data on ICP was obtained during shunt insertion. In 7 children, simultaneous P.I. and I.C.P were obtained.

In this series, we found that an increase in the P.I. could predict the need for eventual shunting or shunt revision. Simultaneous monitoring of P.I. and I.C.P also showed agreement between these values. Transcranial doppler may prove useful as a non-invasive screening tool to objectively evaluate children for increased intracranial pressure or hydrocephalus.
21. VENTRICULAR GALLBLADDER SHUNTS: AN ALTERNATIVE ROUTE FOR CEREBROSPINAL FLUID SHUNTING

Tariq Javed, John E. Kalsbeck, Joel C. Boaz, Robert L. Campbell (Indianapolis, IN)

Hydrocephalus is a problem frequently encountered by the Pediatric Neurosurgeon and is routinely treated by placement of Ventriculoperitoneal (VP) or Ventriculoatrial (VA) shunts. Other sites for insertion of the distal shunt catheter have occasionally been used including the stomach, intestine, ureter, fallopian tubes and the pleural space. The purpose of this presentation is to present our experience with the ventricular gallbladder shunt (VGB) in 32 patients performed between 1970 to 1987.

Patients age ranged from 6 months to 20 years with 18 girls and 14 boys. Diagnosis included congenital hydrocephalus (9), myelomeningocele (8), postmeningitis/ventriculitis hydrocephalus (7), intracranial tumors (4), intraventricular hemorrhage (3), and Dandy Walker cyst (1).

Indications for surgery included recalcitrant VP shunt infection (20), ascites following VP shunt (5), VA shunt infection following VP shunt infection (4), and distal shunt malfunction due to cysts (secondary to infection; 3). 19 patients (59%) had concurrent multiple medical problems and/or multiple non-shunt related abdominal surgeries.

The number of shunt revisions prior to VGB shunting ranged from 1 to 7 (mean gallbladder atony (2), and ventriculitis (2). Proximal shunt revision was required in three, atony was successfully treated with cholecystokinin in two, and the infected shunt was removed in the remaining two.

Five patients were lost to follow-up and three died from unrelated causes. Of the 24 patients available for long-term follow-up 50% had a functional shunt at 5 years and 29% at 10 years with the longest functional shunt to date of 14 years.

In our experience the VGB remains a viable alternative for hydrocephalus patients in whom the peritoneal or vascular shunt are no longer feasible. Additionally it may provide time for the infected peritoneal surface to re-quire its cerebrospinal fluid absorptive capacity for reverting back to a VP shunt.
22. PROSPECTIVE EVALUATION OF OUTCOME OF NEONATAL POST-HEMORRHAGIC VENTRICULOMEGALY AND DETERMINATION OF BEST PREDICTORS OF GOOD OUTCOME

Alexa Canady, Seejna Shankaran, Arthur Eisenbrey, Thomas Koepke (Detroit, MI)

Prospective evaluation of the neurodevelopmental sequelae of 33 low birth weight neonates with moderate and/or severe hemorrhage and ventriculomegaly (VM group) and 39 neonates with mild hemorrhage only (non-VM group), was performed. Both groups were comparable regarding birth weight, gestational age and socioeconomic status. At a mean age of 50 months, 19 of 33 children in VM group had neurological sequelae as compared to 3 of 39 in non-VM group. Mental developmental scores using Bayley's or McCarthy's were similar. In the VM group 17 of 33 were greater than 2SD below the mean as compared to 8 of 39 in the non-VM group. Among children with shunts higher incidence of sequelae occurred with shunt infection. The most important prediction of mental and motor outcome in the shunted group by multiple regression analysis was lack of ventricular decompression immediately following shunt insertion.

23. EFFECTS OF EXPERIMENTAL INFANTILE HYDROCEPHALUS AND VENTRICULOPERITONEAL SHUNTS ON DENDRITIC AND SYNAPTIC MORPHOLOGY


In our study of hydrocephalic neonatal rats, dendritic deterioration of pyramidal neurons was noted throughout all layers of the cerebral cortex. This type of cytological impairment could provide a cellular basis for the residual neurologic deficits that hydrocephalic children may manifest after surgical decompression. Thus it seemed appropriate to extend this dendritic analysis to our feline model of infantile hydrocephalus, which had proven to be suitable for placement of ventriculoperitoneal (VP) shunts. Hydrocephalus was induced in 4-10 day old kittens by intracisternal injection of kaolin. Hydrocephalic animals (n=7) were sacrificed at 15-25 days post-kaolin, just prior to death, and compared to saline-injected age-matched controls (n=3). Animals confirmed to be hydrocephalic by ultrasound received VP shunts at 10-18 days post-kaolin (n=3), before conspicuous neurologic signs developed. Tissue from the association (area 22) and sensory (area 17) cortex was processed for Golgi-impregnation of dendrites and light microscopic analysis. Additional tissue from these regions, as well as the motor cortex (area 4), was processed for transmission electron microscopy. To date the results are most complete for area 22. Although the typical variation characteristic of developing neurons was observed in all animals, pyramidal neurons from hydrocephalic brains consistently exhibited apical and basal dendrites with fewer surface appendages (spines) and much thinner shafts than normal. In severe cases, dendrites were nearly devoid of spines and frequent constrictions gave the shafts a beaded appearance. In contrast, the spine density and shaft caliber of dendrites from shunted brains were indistinguishable from controls, even in one animal sacrificed only 8 days after shunting.
23. EFFECTS OF EXPERIMENTAL INFANTILE HYDROCEPHALUS AND VENTRICULOPERITONEAL SHUNTS ON DENDRITIC AND SYNAPTIC MORPHOLOGY (Con't)

Preliminary observations suggest that area 17 was more severely affected than area 22. One animal that remained hydrocephalic for 44 days was intriguing in that basal dendrites were extremely long and spine density was equal to or greater than controls, even on drastically stunted apical dendrites. Ultrastructural observations, which focused on the cellular laminae of the cortex, revealed very few synapses in hydrocephalic brains but a qualitatively normal synaptic pattern in shunted animals. Further ultrastructural features will be discussed in correlation with the dendritic alterations. Overall, these results suggest that hydrocephalus causes extensive functional impairment that may be corrected with properly timed VP shunts.

24. MRI ANALYSIS OF THE PROGRESSION OF EXPERIMENTAL HYDROCEPHALUS AND THE EFFECTS OF VENTRICULOPERITONEAL SHUNTS IN NEONATAL KITTENS


In the course of developing a feline model of infantile hydrocephalus suitable for ventriculoperitoneal shunting, we had previously employed ultrasonography to monitor the extent of ventriculomegaly. While this technique assessed ventricular enlargement adequately the resolution of anatomic detail was relatively poor and it could not be used to evaluate the effects of shunting after the skull ossified over the coronal sutures. Therefore MRI analysis was initiated. Hydrocephalus was induced in 7-10 day old kittens by intracisternal injection of kaolin. At 10-18 days post kaolin hydrocephalic animals received VP shunts. Using a Fomar 0.3 tesla unit with a small animal coil, lightly anesthetized kittens were scanned at various pre- and post-shunt intervals and sacrificed for morphological correlation. As early as 20 hours post-kaolin the lateral ventricles (LV) began to enlarge. MRI was consistent with the gross morphology in demonstrating that ventriculomegaly progressed steadily so that by 3 days the temporal and occipital horns were moderately dilated, as was the 4th ventricle (V4) and recess of the inferior colliculus (RIC). At this time the coronal sutures were split and the anterior fontanelle was 1-2 mm in diameter. By 5 days a bilateral communication was established through the septum pellucidum. Expansion of the LV, RIC and V4 became severe over the next 5 days, such that the cerebral cortex was reduced to about 75% of its original thickness, the internal capsule was stretched and edematous, the caudate nucleus was compressed ventrally laterally and the cerebellar hemispheres were compressed dorsally. However, the cortical sulci were still visible as shallow grooves. Non-shunted animals exhibited extreme dilatation of all parts of the LV, reduction of most cortical regions to less than 1mm and Arnold-Chiari

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malformation. In all hydrocephalic animals the cerebral aqueduct was patent, the 3rd ventricle only mildly enlarged and the basal cisterns and V4 outlets blocked by kaolin-induced adhesions. Animals shunted successfully demonstrated a 50-75% reduction in LV size within 1-2 days. Except for the temporal horns, which in some cases remained slightly dilated throughout the entire post-shunt period, the LV became slit-like within a week. One animal whose distal catheter pulled out of the abdomen at about 3 weeks post-shunt exhibited moderate ventriculomegaly on MRI. Following shunt revision, the ventricles returned to their previous size, but over a more protracted period. Discussion will center on the accuracy of MRI and the value of this animal model in evaluating infantile hydrocephalus experimentally.

25. METABOLIC CHANGES IN HYDROCEPHALIC RAT BRAIN

---31p-MRS STUDY

Mitsunori Matsumae, Osamu Sato (Kanagawa, JP), Takayuki Sogabe, Iwao Miura (Tokushima, JP)

Energy metabolism in hydrocephalic rat brain was studied by 31P phosphorous magnetic resonance spectroscopy (31P-MRS). Twelve week Wister male rats weighing from 280 to 320 g were used. Hydrocephalus was produced by kaolin suspension (0.1 ml, 280 mg/ml) injection into the cisterna magna. 31P-MRS studies were made on control rats (n=15), 1 week hydrocephalic rats (n=15), 2 week hydrocephalic rats (n=15), 4 week hydrocephalic rats (n=15) and 6 week hydrocephalic rats (n=14) respectively. 31P-MRS were measured by BEM-140/200 (4.77) with double turn surface coil of 15 mm in diameter at 80.55 MHz, acquired with a pulse repetition time of 2 seconds and pulse width of 30 μ sec; for 25 minutes.

A typical 31P-MRS spectrum of a control rat brain consisted of several peaks as follows; β-ATP, α-ATP, β-ADP, phosphorylcreatine (Pcr), phosphodiesters, inorganic phosphate (Pi) and phosphomonoesters (PME). The data analysis was done as follows; the ratio of Pcr to Pi + PME as well as the ratio of β-ATP and Pi + PME from the area of each peak of 31P-MRS spectrum was computed. The intracellular pH was computed from the chemical shift of the Pi peak in 31P-MRS spectrum.

The effect of hydrocephalic state at 1 week rats were decreases in β-ATP, Pcr and intracellular pH. The decreased β-ATP and Pcr were persistent in following weeks, while intracellular pH was decreased at 2 to 4 weeks but somewhat improved at 6 weeks. The present study indicates that the energy metabolism in hydrocephalic rat is initially dependent on anaerobic glycolysis.
26. SYSTEMATIC STUDIES ON THE EFFECTS OF AN NMDA RECEPTOR ANTAGONIST MK-801 ON CEREBRAL BLOOD FLOW AND RESPONSIVITY, EEG, AND BLOOD-BRAIN BARRIER FOLLOWING COMPLETE REVERSIBLE CEREBRAL ISCHEMIA

Mark K. Stevens, Tony L. Yaksh (Rochester, MN)

The dose-dependent effects of MK-801, a glutamate receptor antagonist, on changes in cerebral blood flow (CBF), CBF-PaCO_2 responsiveness (xenon clearance), EEG recovery, blood-brain barrier (methylene blue), and cisternal cerebrospinal fluid (CSF) T\textsubscript{x}/6-keto ratio were examined before and up to 2 hours after a 15-minute period of reversible complete global ischemia in halothane anesthetized cats. Pretreatment with doses of MK-801 greater than or equal to 0.5 mg/kg had no effect on resting CBF measures and produced a dose-dependent slowing of the dominant EEG frequency. In animals receiving this agent, following ischemia, there was an almost immediate return of baseline EEG patterns upon reinstitution of flow, no hypoperfusion after 2 hours of reflow, preservation of CBF and CBF-PaCO_2 responsiveness and maintenance of blood-brain integrity. In contrast, parallel control animals and animals treated with MK-801 at a dose of 0.1 mg/kg exhibited poor recovery based on the above parameters. With regard to CSF levels of 6-keto (stable metabolite of PGI\textsubscript{2}) and TxB\textsubscript{2} (stable metabolite of TXA\textsubscript{2}) MK-801 diminished in a dose-dependent manner the elevation in the absolute levels of both which are otherwise induced by ischemia. Of particular interest, the CSF T\textsubscript{x}/6-keto ratio in vehicle treated animals was near 2, in animals pretreated with MK-801 at doses of 0.5 mg/kg or above, this ratio was nearly 1. These observations are consistent with a possible triggering role of glutamate release in initiating at least part of the acute sequelae of ischemia. Such release in an electrically silent cell would increase Ca\textsuperscript{2+} influx, and activate free fatty acid metabolism leading to probable changes in vascular function and changes in blood-brain barrier permeability.

27. EFFECTS OF INCREASED INTRACRANIAL PRESSURE AND HYPERVENTILATION ON AVDO\textsubscript{2} AND BRAIN METABOLISM - IMPLICATIONS REGARDING MANAGEMENT OF HEAD INJURED PATIENTS.

Leslie N. Sutton (Philadelphia, PA), Alan McLaughlin (Rockville, MD), Steven Dante, Mark Kotapka (Philadelphia, PA)

Patients suffering from head injury often have accompanying brain swelling and increased ICP. During this acute phase, CMR\textsubscript{O}\textsubscript{2} (cerebral metabolic rate of oxygen) is usually reduced, but CBF may be increased, decreased or normal. Since CMR\textsubscript{O}\textsubscript{2} = CBF x AVDO\textsubscript{2} (where AVDO\textsubscript{2} is the difference in oxygen content in artery and jugular veins),

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AVDO_2 = CMRO_2 / CBF
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and thus the AVDO\textsubscript{2} has been proposed as a measure of matching of blood flow to metabolism: a low AVDO\textsubscript{2} reflects flow in excess of metabolism (hyperemia) and a high AVDO\textsubscript{2} reflects insufficient flow (ischemia). It is also proposed that hyperventilation in the setting of high AVDO\textsubscript{2} may reduce flow and aggrivate ischemia.

To investigate this further, we have studied 5 cats in whom ICP was gradually increased by cisternal infusion of mock CSF, while monitoring AVDO\textsubscript{2} and brain phosphocratine by magnetic resonance spectroscopy. The effects of extreme hyperventilation were then tested.

In all animals increase in ICP was accompanied by an increase in AVDO\textsubscript{2}, reflecting compromised flow. In 2 animals, hyperventilation resulted in further increase in AVDO\textsubscript{2} and significant drop in PCR\textsubscript{a}, which were reversible with normocapnia.

This suggests that under conditions of compromised CBF due to elevated ICP, hyperventilation may result in significant brain ischemia and that this may be detected clinically by measuring AVDO\textsubscript{2}. Venous oxygen less than 2 volumes percent correlated with drop in phosphocratine.
Ann-Christine Duhaime, Douglas T. Ross, J. Parker Mickle
(Gainesville, FL)

The concept that increased intracranial pressure ultimately causes damage through ischemia is well established in the neurosurgical literature. While considerable progress has been made in characterizing the metabolic changes associated with acutely increased intracranial pressure, relatively few experiments have focused on pathology in survivors. In light of recent advances in understanding the delayed pathophysiology of ischemia with respect to toxic accumulations of stroke, we attempted to study similar features of the ischemia associated with increased intracranial pressure using a cisternal infusion model in animals.

20 male Long-Evans rats underwent an acute elevation of intracranial pressure greater than systemic arterial pressure for 5 to 30 minutes and were sacrificed by perfusion fixation at 1 to 7 days. Histology was assessed by light microscopy and by immunohistochemistry (GFAP, AAT, and glutamine synthetase). In four additional animals hippocampal glutamate concentrations were measured before, during, and after 30 minutes of raised intracranial pressure via stereotactic microdialysis.

We found major loss of hippocampal cells, especially in CA1, after intracranial pressure increases lasting from 15 to 30 minutes. In addition loss of cells in the reticular nucleus of the thalamus occurred after shorter durations (5-10 minutes) of increased intracranial pressure. The hippocampal changes were nearly identical to those which have been reported in vessel occlusion models after 5-15 minutes of ischemia, and which appear to be preventable by excitatory amino acid receptor blockers.

It appears likely that excitotoxicity plays a significant role in the pathology associated with increased intracranial pressure. This may be even more important in mechanical head trauma where membrane disruption may add to the extracellular neurotransmitter pool. Whether therapeutic intervention with excitatory amino acid receptor blockers is effective in increased ICP and/or after mechanical trauma remains a promising question for future investigation.
29. HYDROCEPHALUS: ZERO PRESSURE ADJUSTABLE CSF SHUNT

Eldon L. Foltz, Jeff Blanks (Orange, CA)

Evidence has increased that a percentage of patients shunted for hydrocephalus will show disabling symptoms and signs of low intracranial pressure several years after shunt placement. Such patients show a dramatic fall in ICP when in the upright position whereas normal ICP is present in the supine position. The fall to as much as -350 mm CSF when upright correlates with disabling symptoms. To date, shunt valves have been relative pressure valves which operate on a pressure gradient basis for opening pressure. This design favors "overshunting" with the resulting "low ICP syndrome".

This disabling sequence can be avoided by the zero pressure shunt system designed to function on the basis of a closing CSF pressure device which allows ICP to fall only to zero on the upstream side. It is an absolute pressure device when inserted into the shunt system, but must have a one-way valve in the system as well.

The system design is based on the concept that the siphoning process in ventricular shunts is the primary ICP process in need of absolute control. Such hydraulic parameters vary related to body position of supine and upright, and therefore placement of this device in the subgaleal space has two vertical coordinates - 1) the distance from vertex when upright; and 2) the distance from frontal tip when supine. At those specific points, the intraluminal shunt pressure cannot go below zero. At reference points of vertex (when upright) and frontal tip (supine) the ICP can fall only to a negative value equal to the vertical distance the siphon control device (or zero pressure device) is below that point for that body position.

This system has been effectively used in 22 patients suffering from disabling low ICP syndrome. All patients had precise ICP records pre- and post-op. 12 have had brain map EEG's to assess electro-physiologic status. 20 patients were markedly improved clinically, all showed markedly improved ICP status, but 2 patients demonstrated impaired ability to function at "normal" ICP levels, and required a graduated return to such levels.

The basic characteristics of the new shunt will be reviewed. The need of such a system in the initial treatment of hydrocephalus will be obvious in the specific review of the results in 22 cases in which the system is now functioning.
30. CRANIOPHARYNGIOMA IN 37 CHILDREN: A 10 YEAR FOLLOW-UP WITH INFORMATION ON PERFORMANCE AFTER HIGH SCHOOL

Edwin G. Fischer, Keasley Welch, John Shillito, Jr., Ken R. Winston, Nancy Tarbell (Boston, MA)

37 consecutive children with craniopharyngioma were treated between 1972 and 1981. All but 2 patients were contacted in 1988, giving a mean follow-up of 10.5 years. 20 patients are old enough to have finished high school, enabling us to collect data on college or work activity. Of these 20 patients, none of the 4 who had radical excision of their tumor were employed or going to college. Among the 16 treated by conservative operations and radiation therapy, job performance or college attendance varied considerably, suggesting that overall psychosocial performance was impaired with either surgical approach but was better when radical surgery was avoided. School performance and the presence of psychological problems, as perceived by the parents, did not differ between the two types of treatment.

The rate of tumor recurrence or of failure to respond to treatment was 57% (4 of 7 patients) following radical surgery and 7% (2 of 27 patients) after conservative procedures and radiation. The overall mortality was 9%, the 3 deaths being caused by (1) "hypothalamic crisis" one year after radical resection; (2) progressive tumor growth despite 2 attempts at resection and radiation; and (3) a radiation-induced glioma of the brain stem 8 years after treatment.

Further information is needed regarding longterm quality of life following treatment of childhood craniopharyngioma to help define the best therapeutic roles for surgery and radiation.

31. GAMMA KNIFE STEREOTACTIC RADIOSURGERY FOR CHILDHOOD ARTERIOVENOUS MALFORMATIONS

Eric Altschuler, L. Dade Lunsford, David Bissonette
(Pittsburgh, PA)

Stereotactic radiosurgery using the 201-source Cobalt 60 gamma knife permits single treatment, closed-skull obliteration of cerebral arteriovenous malformation (AVM). Of 84 AVM patients treated in the first twelve months of operation of the Pittsburgh Gamma Knife, 12 patients (7 males, 5 females) were 18 years old or younger (mean age = 12.5 years). Five patients had subcortical AVM's, six had diencephalic or brain stem AVM's, and one patient had a cerebellar AVM. Eleven patients had prior intracranial hemorrhages, with good recovery (Karnofsky rating at the time of radiosurgery 90 or greater). Radiosurgery was the initial surgical procedure in 11 patients; one patient underwent four prior intracranial operations, and one patient had intravascular embolization preceeding radiosurgery. In a single treatment session, the radiosurgical dose to the AVM center of the margin varied from 33.3 to 50 Gy. The lesion volume was treated at the 50% or greater isodose. One or more gamma knife shots was used to enclose the malformation. General anesthesia was used in patients less than 14 years of age. No significant immediate postoperative morbidity was encountered. Radiosurgical obliteration was confirmed in one patient at four months after treatment. All other patients are being studied by neuro-diagnostic and imaging protocols which include magnetic resonance imaging (MRI) at six month intervals and cerebral angiography at one year intervals until obliteration is confirmed. Initial MRI examinations in three patients have shown either no change (2 cases) or 50% reduction (one case, six months after treatment). We believe that stereotactic radiosurgical obliteration of AVM's is an important tool to obliterate AVM's when microsurgical techniques are deemed too risky. Development of radiobiological response
31. GAMMA KNIFE STEREOTACTIC RADIOSURGERY FOR CHILDHOOD ARTERIOVENOUS MALFORMATIONS (Con't)

Modifiers eventually may allow reduction of the dose required, decrease the risk of delayed radiation brain injury, and shorten the latency until angiographic obliteration occurs.

32. PEDIATRIC OLIGODENDROGLIOMAS

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

Oligodendrogliomas account for one percent of pediatric brain tumors. Four children, two of each sex with ages ranging from four to 11 years (M 7.5 yrs.), have presented to the Children's Hospital over the past three years. Headache was the most frequent symptom, followed by vomiting, blurred vision, weakness and seizures. Papilledema, hemiparesis and positive Babinski were each found in one-half. Duration of symptoms ranged from 0.5-24 months.

Radiologic investigation included CT and, in one case, MRI. Three tumors were parietal in location and one temporo-parietal. Universal CT findings consisted of large tumors (3.5 x 4.0 - 7.0 x 5.5 cm); broad cortical surface bases; cysts; dense calcium deposits; peritumoral edema; and moderate enhancement. The MRI revealed areas of perinecrotic hemorrhage.

Gross total resection was achieved in all cases without operative morbidity. Histopathology revealed three benign oligodendrogliomas and one anaplastic. All are symptom free seven-46 months postoperatively. The anaplastic tumor was treated with combined therapy and is disease free two years postoperatively.

One patient with a benign oligodendroglioma received two further operations for recurrent local disease. Her histopathology sequentially changed from slight pleomorphism to moderate pleomorphism to characteristics consistent with primitive neuroectodermal tumors. Drop metastases, found by myelogram, were successfully treated with combined therapy. All patients are being currently followed with CT and gadolinium enhanced MRI. Recurrent local disease will be controlled with reoperation.
32. PEDIATRIC OLIGODENDROGLIOMAS
(Con't)

These cases are not consistent with classical oligodendroglomas in regards to location and the mean age is significantly younger than those reported by Dohrmann et al. The two patients with aggressive disease have to date been successfully treated with combined therapy.

33. EXPRESSION OF THE N-MYC, C-MYC, C-SRC, and INT-1 PROTO-ONCOGENES IN MEDULLOBLASTOMA CELL LINES: IMPLICATIONS FOR NEUROHISTOPATHOLOGY

Donald A. Ross, Hideyuki Saya, Michael S.B. Edwards, Victor A. Levin (San Francisco, CA)

The proper classification of primitive neoplasms of the central nervous system, especially medulloblastoma, has been a subject of recent debate. Current histological techniques have not been able to supply the information needed to resolve the debate, but the powerful techniques of molecular biology may assist in classifying the cells of the developing central nervous system and the primitive brain tumors which arise from them. Reports on the expression of the proto-oncogenes of the myc and src families in the developing nervous system have begun to elucidate the molecular mechanisms of differentiation in the brain, and expression of the int-1 gene has been observed in the developing brain and in teratocarcinoma cell lines. We have sought to extend the analysis of the expression of these genes to the primitive neuroepithelial tumors by analyzing the least studied of these, the medulloblastoma.

Using Northern blotting, we found no expression of the N-myc and int-1 genes in two medulloblastoma cell lines, but did find the expression of c-myc gene in both cell lines. We also detected c-src kinase activity in the medulloblastoma cell lines, a feature of cells committed to neuronal differentiation. Based on published data on the time-specific expression of these genes in the developing human brain, we propose that the cell of origin of the medulloblastoma exists late in gestation. Based on published data on the expression of these proto-oncogenes in teratocarcinoma, neuroblastoma, and glioblastoma, we propose a neuropathologic classification system based on the time and differentiation dependent expression of these genes.
34. BRAIN STEM TUMORS IN CHILDREN
CLASSIFICATION, MANAGEMENT, AND PROGNOSIS

Maurice Choux, Jean-Marc Riss, Lorenzo Genitori, Gabriel Lena (Marseille, France)

The authors present a series of 97 brain stem tumors in children under the age of 15. Infants represent 7.5% and 31.5% were under the age of 5. Initial symptoms are: increased ICP (34%), with an initial hydrocephalus in 26%, ocular signs (23%), cerebellar signs (18.5%), motor deficit (10%), torticollis (7%).

Radiological investigations (CT scan: 81, MRI: 30), allow to distinguish 5 types in 81 tumors = Type I (intrinsic, diffuse, hypodense without enhancement): 30 (37%). Type II, with cystic (A) or solid (B) focal component: 21 (26%). Type III (dorsal exophytic component): 12 (15%). Type IV (anterior or lateral exophytic component): 11 (13%). Type V (bulbomedullar): 7 (9%).

Surgery was carried out in 43 cases. The surgical approaches were: midline posterior fossa (26), lateral (4), subtemporal (7), intraventricular (1), craniospinal junction (5). Surgical indications and approaches are related with the anatomical types of the tumor. Total removal was possible in only 12 cases (29%), partial removal in 24 and a biopsy in 7 cases. Operative mortality (first month) is closely related with the quality of surgery: 16% after total removal, 28% after surgical biopsy. 6 patients underwent a stereotaxic biopsy.

Pathological aspects in 42 cases are: benign astrocytoma in 43%, malignant astrocytoma in 33%, intermediate grade in 28%, others histologies in 7 cases.

The interest of hyperfractionning irradiation and chemotherapy are discussed.
35. BRAINSTEM GLIOMAS IN NEUROFIBROMATOSIS

Jerrold M. Milstein, J. Russell Geyer, Mitchel S. Berger, W. Archie Bleyer (Seattle, WA)

Brainstem gliomas in childhood have been well characterized with symptoms and signs of brainstem dysfunction and imaging findings of a brainstem mass. The clinical course has generally been one of a partial, temporary remission after conventional radiotherapy of 50 to 55 Gy, only to have progression reappear leading to death usually within six months. Long term survival of children with brainstem gliomas is only 10-20%.

The association of neurofibromatosis (NF) and brain tumors has been recognized for many years. The majority of tumors are astrocytomas originating within cranial nerves, and in children the majority of these have been optic nerve gliomas with some extending into the optic chiasm and/or hypothalamus. In the past two years we have evaluated and treated five children with NF and brainstem gliomas. These children have had symptoms, signs, and clinical courses which appear to distinguish them from children without NF who are diagnosed to have a brainstem glioma.

<table>
<thead>
<tr>
<th>Age Dx(NF)</th>
<th>Age Dx(BT)</th>
<th>Symptom of BT</th>
<th>Exophytic C.N.</th>
<th>Treatment</th>
<th>Outcome</th>
<th>Duration</th>
</tr>
</thead>
<tbody>
<tr>
<td>Birth</td>
<td>9 yr</td>
<td>Headache</td>
<td>0</td>
<td>+</td>
<td>50 Gy</td>
<td>Alive</td>
</tr>
<tr>
<td>Birth</td>
<td>7 yr</td>
<td>Respiratory arrest</td>
<td>0</td>
<td>+</td>
<td>50 Gy</td>
<td>Alive</td>
</tr>
<tr>
<td>4 yr</td>
<td>15 yr</td>
<td>Hearing loss</td>
<td>+</td>
<td>+</td>
<td>55 Gy</td>
<td>Alive/Dead</td>
</tr>
<tr>
<td>2 yr</td>
<td>7 yr</td>
<td>Speech change</td>
<td>0</td>
<td>+</td>
<td>55 Gy</td>
<td>Dead/Dead</td>
</tr>
<tr>
<td>3 yr</td>
<td>21 yr</td>
<td>Headache</td>
<td>0</td>
<td>+</td>
<td>None</td>
<td>Dead</td>
</tr>
</tbody>
</table>

As a result of the experience reported here, we propose an additional factor associated with a better prognosis: neurofibromatosis. The pattern in our five patients suggests that brainstem tumors in NF are often low grade and slowly progressive.
36. FAMILIAL CHOROID PLEXUS CARCINOMAS

Curtis Dickman (Tucson, AZ), Harold Rekate, Steven Coons, Peter C. Johnson (Phoenix, AZ)

Choroid plexus carcinomas in siblings have not been previously reported. Two cases of this rare tumor are presented in full siblings ages 11-months and 10 years old. Both cases presented with intraventricular masses causing obstructive hydrocephalus.

The tumors were examined by light microscopy and immunohistochemical techniques. These neoplasms were invasive and had variable growth patterns. Mitoses and necrosis was present in one of the two cases. Both displayed identical immunohistochemical reaction patterns. They were immunoreactive for keratin; and were nonreactive for GFAP, S-100, and CEA.

Evaluation of proliferation using Ki-67 monoclonal antibody showed 9% of the cells to be in proliferative phases of the cell cycle; a high value for glial-derived neoplasms. Ki-67 activity appears to be a more sensitive measure of proliferation in malignant choroid plexus tumors than the presence of mitosis and necrosis. This may be helpful in distinguishing between choroid plexus carcinomas and papilloma when the histology is equivocal.

37. DIAGNOSIS AND MANAGEMENT OF CHOROID PLEXUS TUMORS WITHIN THE LATERAL AND THIRD VENTRICLES


Choroid plexus tumors are rare tumors of the central nervous system. They consist from .5 to 1 percent of all intracranial tumors. In pediatric hospitals, they have a higher representation because of referral patterns. At Loma Linda University between 1980 and 1988, seven tumors have been identified having choroid plexus tissue in children less than two years of age. Six of the seven tumors were benign, and one tumor was a choroid plexus carcinoma. Five of the histologically verified choroid plexus papillomas were studied with MRI, as well as pre- and post-contrast CT scans. MRI through high resolution and multi-planer techniques offers an improved form of visualization of these lesions in relationship to the existing anatomy and better planning of surgical resection. There were two females and five males in the series. One tumor was located primarily within the third ventricle, one presented with bilateral-lateral ventricular tumor, while the others remained within the confines of one lateral ventricle. Two patients had marked over-production of cerebral spinal fluid, one patient had marked ascites following the placement of a ventriculo-peritoneal shunt which resolved following total excision of the tumor substantiating the premise of the over production of CSF from the tumor. The common post operative finding was the subdural effusion. Four of the seven patients required subdural to peritoneal shunting, three of the seven patients had seizure disorder; however, one reverted to a normal EEG and was off anti-convulsant medications following the resection of the third ventricular choroid plexus papilloma. MRI has been a very
37. DIAGNOSIS AND MANAGEMENT OF CHOROID PLEXUS TUMORS
WITHIN THE LATERAL AND THIRD VENTRICLES
(Con't)

useful tool in both the surgical planning and postoperative follow up with these patients, and appears to be the method of choice in studying choroid plexus tumors. Angiography, while being helpful in certain cases, is no longer absolutely necessary in the evaluation of all choroid plexus tumors.

38. BONE METASTASIS OF MEDULLOBLASTOMA IN CHILDHOOD

Tadanori Tomita, Mary Ann Radkowski, David G. McLone, Kenneth D. Bauer (Chicago, IL)

This report consists of 6 children with osseous metastasis of medulloblastoma detected from 1980 through 1987. They represent 20% of 30 children treated with a combination of radical resection and radiation therapy. Four presented with localized pain and two with palpable mass. Metastatic medulloblastomas were diagnosed by plain radiographs, radionucleid bone scans and/ or computed tomography and verified by excisional biopsy or bone marrow aspirates. Five patients had no demonstrable CNS recurrence at bone metastasis. Bone metastasis occurred in the variable postoperative period between 5 months and 42 months. All but 1 were treated by chemotherapy with or without local radiotherapy. Metastatic bone disease responded well to the given therapy. However, one patient died of subsequent CNS recurrence and two died of chemotherapy complications. The untreated patient died. Flow cytometric DNA ploidy indicates 5 were diploid and 1 was DNA aneuploid type. Though statistically insignificant (Fisher, p=0.055), diploid tumor tend to spread to the bone (38.5% of diploid tumor vs. 6.7% of aneuploid tumor). Follow-up bone scans in all medulloblastoma patients and chemotherapy for diploid medulloblastoma are recommended.
39. SYMPTOMATIC EPIDURAL MALIGNANCY IN CHILDREN: WHAT IS THE BEST TREATMENT?

Corey Raffel (Los Angeles, CA), Sean LaVine (San Diego, CA), Victoria C. D. Neave (High Point, NC), J. Gordon McComb (Los Angeles, CA)

Epidural malignant tumor deposits are uncommon in children, but have been reported for many different tumor types. Is the outcome from radiation therapy alone improved by decompressive laminectomy? To investigate this question, the records of all children presenting to Children's Hospital of Los Angeles between 1975 and 1988 with symptoms of spinal cord compression by epidural malignant tumor were reviewed. The patients with near complete or complete block on myelography or significant cord compression on CT or MR scans comprise the material for this report.

Four percent of patients had cervical lesions, 64% of patients had thoracic lesions and lumbo-sacral lesions were present in 28%. One patient had myelographic blocks at both T5 and L1. The most common tumor types causing compression were rhabdomyosarcoma, neuroblastoma, and Ewing's sarcoma, although twelve different tumor types occurred in this group of patients. Of the patients treated with laminectomy and post-laminectomy radiation therapy and/or chemotherapy, 62% had improved neurological function post-therapy, 33% were unchanged, and a deterioration occurred in 5%. In contrast, radiation therapy alone resulted in an improvement in 14%, stabilization in 28% and deterioration in 37%. When neurological function is considered as to ambulation and sphincter function, a similar superiority of combined laminectomy and radiation therapy versus radiation therapy alone is evident for both.

Based on this date, we conclude that optimal treatment for children with symptomatic epidural malignancy and evidence of significant tumor in the spinal canal on neurodiagnostic studies are best treated by laminectomy and resection of easily accessible tumor followed by postoperative adjuvant therapy.
40. POSTOPERATIVE POSTERIOR FOSSA INTRACRANIAL PRESSURE MONITORING

William Boydstun, Robert A. Sanford, Michael S. Muhlbauer (Memphis, TN)

Intracranial pressure (ICP) monitoring provides an early warning for postoperative complications. Due to the limited size and poor compliance of the posterior fossa, the morbidity and mortality of any postoperative mass is increased. Between September, 1987 and August, 1988, eleven children who underwent suboccipital craniectomy for tumor had posterior fossa monitoring using a Camino fiberoptic subdural monitor. Two of these children had simultaneous supratentorial ventricular pressure measurement. Monitors were utilized for two to four days postoperatively. There were no complications nor infections directly related to the monitoring.

Our results indicate the mean posterior fossa ICP in the supine position with 30 degrees of head elevation was 25.6 +/- 5.5 mm Hg and in the lateral decubitus position, 16.8 +/- 4.2. Both values increased approximately 7 mm Hg when the child was maintained flat. With simultaneous supratentorial monitoring, the mean infratentorial pressure was 4 mm Hg greater than that supratentorially in the supine position. The pressures were the same in the lateral decubitus and sitting positions. One child who underwent simultaneous monitoring developed obstructive hydrocephalus secondary to blood within the third ventricle. The posterior fossa pressure read 14 mm Hg higher than the supratentorial compartment, regardless of position.

Because of positional fluctuations, changes in ICP are more significant than the absolute values. The authors conclude that posterior fossa pressure monitoring provides a safe method to prevent potential postoperative complications.

41. GADOLINIUM ENHANCED MAGNETIC RESONANCE IMAGING VS. ENHANCED COMPUTED TOMOGRAPHY IN EXAMINING CHILDHOOD POSTERIOR FOSSA TUMORS

Charles L. Y. Cheng, Walker L. Robinson, Fouad E. Gellad (Baltimore, MD)

The use of magnetic resonance imaging (MRI) with and without gadolinium (Gd) enhancement has added another powerful diagnostic modality to the management of childhood posterior fossa tumors. Over the last year, we have followed twelve cases of posterior fossa tumors using dry MRI in conjunction with unenhanced and enhanced computed tomography (CT), after resection. Most recently, we have added gadolinium enhanced MRI to our battery of followup studies. The preliminary results are encouraging. We found that unenhanced MRI is particularly useful in delineating postoperative changes consistent with edema in CO2 laser treated tissue areas. Tumor recurrence demonstrated on enhanced CT, however, has occasionally been invisible in simultaneous unenhanced MRI scans. Gadolinium enhanced MRI, on the other hand, has exhibited several satellite lesions not discernible on enhanced CT in the preoperative workup of a medulloblastoma. Tumor recurrence, not seen on enhanced CT or unenhanced MRI in another case, was readily demonstrated on Gd enhanced MRI. More interestingly, preoperative or recurrent lesions that are clearly discernible on Gd enhanced MRI have been found to be discernible only retrospectively on enhanced CT scans. It appears that while enhanced CT remains the more specific modality in identifying neoplastic lesions, Gd enhanced MRI is more sensitive in delineating abnormal tissue. We conclude that Gd enhanced MRI will become an indispensable adjunct to enhanced CT in the optimal examination and management of childhood posterior fossa tumors both before and after neurosurgical resection.
42. THE MANAGEMENT OF HYDROCEPHALUS COMPLICATING
CHILDHOOD POSTERIOR FOSSA TUMORS

Mark Steven Dias, Dachling Pang, A. Leland Albright,
Donald Krieger (Pittsburgh, PA)

Much of the management mortality and morbidity associated
with childhood posterior fossa tumors is the result of
secondary hydrocephalus. The introduction of
cerebrospinal fluid (CSF) diversion procedures prior to
resection of the tumor has greatly reduced the operative
mortality and improved outcome in survivors. Insertion of
a ventricular shunt (VS) prior to tumor resection is
currently a popular method of treating the hydrocephalus.
Several authors, however, have voiced misgivings about
unnecessarily committing some of these children to a
permanent shunt. Temporary external ventricular drain
(EVD), on the other hand, can be inserted with minimal
risk as an alternate method of preoperative CSF drainage.
To settle the argument concerning the preferred method of
CSF drainage, we undertook a retrospective study on 61
children with posterior fossa tumors and hydrocephalus
treated at the Children's Hospital of Pittsburgh from 1979 -
1988. Twenty-five underwent VS as the initial treatment
of their hydrocephalus, 20 had an EVD, and 16 received no
primary treatment for their hydrocephalus prior to tumor
removal. The average follow-up was 52 months (range 4
months to 9 years). The three groups were comparable with
regards to age, sex, pre-operative and discharge
neurological status, duration of symptoms (both related to
the tumor and to the hydrocephalus), tumor location and
histology, and extent of resection. The hydrocephalus was
less severe in the no treatment group, though there was no
difference between the EVD and VS groups. In both the VS
and no treatment groups, significantly greater numbers of
patients had no dural closure following tumor resection.

Among the patients in the EVD and no treatment groups, 15
(76%) and 12 (75%) respectively, remained free of

hydrocephalus and did not require a shunt during the
follow-up period. Combining the two treatment groups,
only 9 patients subsequently required shunting. These 9
were compared with the 27 patients who did not require
shunting to determine which features might predict the
need for a subsequent shunt. Only two features predicted
the need for a subsequent shunt: (1) those patients who
underwent less extensive tumor removal, and (2) those in
whom the dura was left open at the end of the tumor
resection, were both statistically more likely to require
a subsequent shunt. No differences were found with regard
to age, sex, duration of symptoms, admission or discharge
neurological status, tumor histology, or location,
severity of pre-operative hydrocephalus, or CSF indices
(red or white cells, glucose, protein) during the period
of EVD.

We conclude that three-quarters of patients with
hydrocephalus complicating childhood posterior fossa
tumors do not require permanent CSF diversion. Temporary
EVD is well tolerated; no patient in our series acutely
deteriorated, and complications were few. Those patients
with less extensive tumor resection will more likely
require a subsequent shunt. Finally, we recommend closing
the dura following tumor removal, since a higher incidence
of post-operative hydrocephalus occurs in patients in
which the dura is left open.

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43. PRIMITIVE NEUROECTODERMAL TUMORS: IMPROVED SURVIVAL WITH 5 IN 1 CHEMOTHERAPY

Rebecca Brightman, Edward J. Kosnik, Frederick Buymann (Columbus, OH)

Primitive neuroectodermal tumors (PNET) of childhood are highly malignant and are generally considered to be rapidly fatal. Previously published data from our institution reported a series of 18 children with PNET. The present study updates this experience and emphasizes the impact of our current chemotherapeutic regimen.

Twenty-six children with PNET (22 supratentorial and 4 spinal) have been cared at Columbus Children's Hospital since 1980. Twenty-one of these patients (Group I: 1958 - 1980) underwent surgical excision, radiation and conventional chemotherapy. Eight patients (Group II: 1981 - present) were managed in a similar fashion with the exception that a regimen of eight drugs in one day (5 in 1) preradiation chemotherapy was used.

Mean survival of Group I was 8 months whereas Group II had a mean survival of 29 months with 2 patients still alive. Patient population, surgical excision and radiation dosage were similar in the two groups. The addition of 5 in 1 chemotherapy to Group II appears to have had a significant impact on patient survival. This form of chemotherapy thus may play an important adjuvant role in the treatment of children with these primitive tumors.

44. EPENDYMOMAS IN CHILDREN: ISSUES OF CLASSIFICATION AND TREATMENT

Cynthia A. Delaat, Edgar T. Ballard, Beatrice C. Lampkin, Kerry R. Crone, Thomas S. Berger (Cincinnati, OH)

Seventeen children with ependymomas were treated from 1977 to 1988. The mean age at diagnosis was 3.9 years and the major presenting symptoms were those of increased intracranial pressure. The majority (12/17) of these tumors originated in the posterior fossa, three were supratentorial and two primary spinal cord lesions. The histologic classification was based on degree of cellular anaplasia. All high-grade tumors were infratentorial. All patients underwent surgery with a total of 21 procedures performed: biopsy alone 1 (4%), subtotal removal 10 (48%), and gross total resection 10 (48%). Operative mortality was zero. Thirteen patients received post-operative radiation therapy and nine patients had chemotherapy; five at diagnosis and four following recurrence.

The overall survival for these patients is 76% with a median follow-up time of 2.7 years. When survival rates were considered according to histologic grade no differences were seen; survival was decreased for posterior fossa tumors (67% vs. 100%). Seven patients had recurrence of their tumor, all at the initial site with no spinal column dissemination. Most were treated with repeat surgery and chemotherapy and three remain alive (9/12, 2 and 5-1/2 years). This analysis suggests that: 1) aggressive attempts at complete surgical resectability at diagnosis and recurrence should be made to improve survival; 2) chemotherapy following surgery and radiation appears beneficial; 3) the prognosis of recurrent disease remains poor and the need to optimize frontline therapy should be stressed and 4) a more uniform classification of ependymomas is needed to better direct therapy and improve prognosis.
45. BRAIN TUMORS BELOW ONE YEAR OF AGE: A RETROSPECTIVE ANALYSIS OF TWENTY-TWO PATIENTS

Souhel Haddad, Arnold H. Menezes, John C. Godersky (Iowa City, IA)

Congenital brain tumors have been infrequently reported and their management ill defined. A review of all children with brain tumors with symptom onset before one year of age (1977-1987) was made.

Twenty-two children encountered in the past ten years had the following pathological diagnosis: astrocytoma 7, PNET 6, choroid plexus papilloma/carcinoma 3, malignant teratoma 2, dermoid 2, embryonal rhabdomyosarcoma 1, choroma 1. Fifteen were supratentorial and 7 infratentorial. There were 14 females and 8 males. Presenting symptoms were hydrocephalus (32%), focal neurological deficit (23%), asymptomatic increase in head size (18%), failure to thrive (FTT) (14%) and seizures (5%). The goal of treatment was radical excision where possible, with primary chemotherapy in the last 5 years. Radiation therapy with chemotherapy was the adjunct to surgery in the initial 5 year period.

All patients with choroid plexus and dermoid lesions underwent total resection with no recurrences. All astrocytomas were supratentorial, 6 were diencephalic. Five out of seven patients survived greater than five years.

Primitive neuroectodermal tumors (6) were equally divided above and below the tentorium. Four out of 6 infants received chemotherapy (chemotherapy alone-2, chemotherapy and radiation therapy-2) and are tumor free 7 months to 9 years followup. One child received radiation alone early in the series surviving four months. The other refused further treatment.

Radical resection with chemotherapy appears to be the rational approach, keeping reoperation and radiation as options for later use to avoid the effects of irradiation on the developing brain.
46. INTRACRANIAL TUMORS IN 332 INFANTS BELOW AGE THREE

Chung-cheng Wang (Beijing, CH)

Intracranial tumors in infants are less found than in adults. In Beijing Neurosurgical Institute the admitted cases below age 3 account for 6.5% of patients below the age 15 and 1.1% of the total patients of all age groups with intracranial tumors in the same period. Relatively acute onset, rapid deterioration, short course and high mortality were the features of intracranial tumors in infants. Three kinds of gliomas are more frequently found in this group. They are ependymoma (31.42%), medulloblastoma (25%) and astrocytoma (23.92%). Most tumors are found in the subventricular area (60.24%), 59.93% in middle line. The main clinical manifestations include signs of increased intracranial pressure (87.04%) and signs of hydrocephalus (75.48%), both of which are the important criteria for diagnosing intracranial tumors in infants. 14 patients were diagnosed as having congenital tumors with their symptoms starting within two months after birth, 5 of them combined with other congenital deformities. 323 cases sustained surgical procedures, total removal in 62.66%. The surgical mortality rate was 17.33%.

The period of follow up for 216 patients was from 1 to 23 years. 55.2% died within one year after operation. The survival rate for 1, 3 and 5 years after the operation were 44.8%, 32.5% and 22.7% respectively. Combined therapy is the choice of treatment.

47. HIGH DOSE OF CHEMOTHERAPY AFTER AUTOLOGOUS BONE MARROW TRANSPLANTATION FOR THE TREATMENT OF PEDIATRIC MALIGNANT BRAIN TUMOR

From November 1, 1981 to October 1, 1985, 28 cases of pediatric malignant brain tumors were treated with autologous bone marrow transplantation followed by high dose of chemotherapy. 21 cases were followed up for a period. They were 8 cases of posterior fossa medulloblastoma, 7 cases of astrocytomas, 3 cases of ependymomas, 2 cases of brain stem tumors and 1 case of malignant choroid plexus papilloma.

The idea is to increase the dosage of chemotherapy with the least disadvantage. After operation, patient's bone marrow was withdrawn and preserved. High dose of CCNU (260 mg or 390 mg/m²) was taken by mouth. And the autologous bone marrow was re-infused 3-5 days later.

Different kinds of chemotherapy were used:
1. 5 cases received high dose CCNU for once.
2. 6 cases received high dose followed by 2-5 courses of routine dosage of CCNU.
3. 10 cases received high dose CCNU, followed by 7-10 days of Vincristine, daily Prednisone and several courses of CCNU.

After 5 months to 5 years follow up, the life span of the first and second group were 10 months, and 15 months respectively, whereas, 7 of the 10 patients of the third group were alive after 30 months.

The clinical application of the autologous bone marrow transplantation, the selection of high dose CCNU, and the benefit of their combination were discussed.
48. MEASUREMENT OF SLOW FLOW IN CSF SHUNT TUBING WITH MRI PHASE IMAGING

James M. Drake, Alastair J. Martin, R. Mark Henkelman
(Toronto, ON)

Measurement of the flow in implanted CSF shunts is difficult due to the small diameter of the tubing and the very low flow rates. Current techniques to measure CSF shunt flow are either invasive or relatively inaccurate. MRI phase imaging using a specially designed coil, a small field of view, and flow encoding gradients allows the accurate determination of slow flow in shunt tubing.

Distilled water was infused through CSF shunt tubing at controlled rates between 0-20 cc/hr. Magnitude and phase images were obtained initially on a 2 Tesla small bore unit and subsequently on a 1.5 Tesla clinical unit. Flow was perpendicular to the slice selection gradient. The echo sequence with TR = 500 msec and Te = 80 msec and a field of view of 4 by 4 cm was used. Additional flow encoding gradients were used in the slice selection plane.

At the upper flow rates, flow effects could be visualized on the magnitude images. At the lower flow rates the only changes were on the phase images where a parabolic velocity profile was found at flow rates as low as 2 cc/hr. The feasibility of this technique was further demonstrated with shunt tubing implanted in a cadaver. This method should be useful in measuring CSF shunt flow in patients.

49. MRI ELUCIDATES PATHOGENESIS OF SPASTICITY IN CEREBRAL PALSY

T.S. Park, Lawrence H. Phillips, II, Jacek Malik, Madeleine Ortman Payne (Charlottesville, VA)

Spasticity is produced by diverse lesions at various levels of the neuraxis and a predominant clinical feature of cerebral palsy (CP), afflicting almost 80% of patients with CP. In order to understand the pathogenesis of spasticity in CP, the nature and distribution of underlying brain damage needs to be ascertained.

We performed MR studies on 23 patients with CP who underwent selective dorsal rhizotomy. The range of ages was 2-11 years. All except one patient had preterm births (gestational ages: 25-38 weeks). Nineteen patients had spastic diplegia; 4 patients had spastic quadriplegia. Preoperative ambulatory status was as follows: 6 were independent walkers, 1 walked with crutches, 11 walked with walkers and 5 were non-walkers.

MR disclosed lesions in 21 (91%) of the 23 patients. The lesions were most prevalent in cerebral white matter; 20 patients harbored white matter lesions. The cerebral cortex was affected by lesions in 11 patients. The white matter lesions were consistent with old periventricular leukomalacia with increased signal intensity in the region of trigone of the lateral ventricle, loss of white matter and ventricular dilation. The cortical lesions were suggestive of old infarction and affected parasagittal or watershed zone regions in the frontal and parietal lobes. Two patients showed lesions in the basal ganglia. In all patients, the brainstem and cerebellum appeared normal.

When MR lesions in the cerebral cortex and white matter were grouped into territories of the frontal and parietal lobes, the lesions were found to affect the frontal lobe in 70% and the parietal lobe in 91% of all patients.
49. MRI ELUCIDATES PATHOGENESIS OF SPASTICITY IN CEREBRAL PALSY
(Con't)

These results suggest that in spastic CP secondary to preterm birth, periventricular leukomalacia is a predominant pathological abnormality and spasticity in these patients is due mainly to damage to the pyramidal and extrapyramidal systems at the level of the cerebral cortex and white matter.

50. NEONATAL CEREBRAL VENOUS THROMBOSIS (NCVT)

Patrick T. Tracy, Robert M. Wright, Wadie S. Tadros, William C. Hanigan (Peoria, IL)

An uncommon clinical entity, NCVT has been associated with trauma, dehydration or sepsis, and a uniformly poor prognosis. The increasing use of noninvasive magnetic resonance imaging (MRI) may alter this clinical picture. This report will outline the clinical presentation and follow-up in 5 infants with NCVT diagnosed by MRI.

Scans were performed with a Siemens Magnetom® at 0.5 Tesla with a Multislice, multiecho technique. High-signal intensity with separate echoes verified the thrombotic process.

One infant without previous risk factors presented with seizures at 7 days of age. MRI demonstrated thrombosis of the internal cerebral vein; 40 months later the child is developing normally with resolution of the thrombotic process.

In 3 infants MRI was performed as a diagnostic follow-up to abnormal cranial sonography; one infant presented with an enlarging head circumference. Two infants sustained significant asphyxia at birth. Three children demonstrated respiratory distress, while one child developed a Grade III intraventricular hemorrhage. MRI demonstrated thrombosis of the sagittal sinus in 2 infants; the remaining 2 infants sustained a thrombosis in the transverse sinuses. Follow-up scans in 3 infants demonstrated resolution of the thrombosis. No child required specific treatment for NCVT. Clinical follow-up at a mean age of 10 months demonstrated that one child was developing normally while 3 children showed developmental delays consistent with asphyxia or periventricular hemorrhage.

In summary, the use of MRI in the neonatal period revealed that NCVT may occur as a benign primary entity or a secondary phenomenon associated with respiratory distress.
50. NEONATAL CEREBRAL VENOUS THROMBOSIS (NCVT)

and/or asphyxia. No specific treatment was indicated and prognosis was related to associated conditions rather than a primary thrombotic process.

51. MULTIPLE ARACHNOID CYSTS OF THE CEREBRAL HEMISPHERE IN INFANCY

Peter Gianaris, Tadanori Tomita (Chicago, IL)

We present 3 infants with massive, multiple (2 or more) arachnoid cysts affecting the cerebral hemisphere. All patients presented with progressive macrocephaly. Computed Tomography (CT) scans demonstrated large, multiple cystic lesions occupying the bilateral cerebral hemisphere in 2 and the unilateral hemisphere in 1. A transcortaneous puncture through the anterior fontanel and CT metrizamide cisternograms showed isolated nature of the cysts. Craniotomies were performed and multiple cysts were excised in all patients. Surgical inspection showed that the cysts were composed of thick arachnoidal membrane with traversing blood vessels, and compressing the underlying cerebral cortex. The cystic contents were what appeared to be cerebrospinal fluid. Pathological study showed 2 were arachnoid cyst and the other was epithelial cyst. After resection of the cystic lesions, all patients received a single CSF diversion shunt.

Previous reports regarding arachnoid cysts dealt with a single lesion, and multiple arachnoid cysts are extremely rare. Multiple arachnoid cyst is perhaps a new clinical and pathological entity. Clinical management and pathological features of these lesions will be discussed.
52. CONGENITAL ARACHNOID CYSTS IN CHILDREN

Balasubramaniam Chidambaram, John P. Laurent, William R. Cheek, Neil A. Feldstein, Emile Rouah (Houston, TX), Steven Schneider (New York, NY)

Congenital arachnoid cysts are benign developmental disorders. Thirty children with arachnoid cysts, managed by the Neurosurgery Service at Texas Children's Hospital, were reevaluated. The cysts were marsupialized or excised as the primary procedure, and only those children who showed no improvement had a shunt placed. This study is an analysis of our experience with marsupialization and excision of the cysts. The pathology of the excised cyst was reevaluated to establish a correlation between the nature and the outcome.

53. SPINAL SUBDURAL SUPPURATIVE INFECTION IN CHILDREN

Daniel D. Gaylon, J.A. Winfield, Julia A. McMillan, Leo Hochhauser (Syracuse, NY)

Suppurative infection of the spinal subdural space is a rare clinical entity. Whereas epidural abscesses are well described in the neurosurgical literature, there are only 19 adult and 7 pediatric cases of completely reported spinal subdural empyema. We report a case of infantile spinal subdural empyema which is typical of previously reported pediatric cases in that infection occurred in the setting of a congenital dermal sinus. In contrast, adult cases of subdural empyema occur predominantly by either hematogenous seeding from distant sites of soft tissue infection or by direct subdural contamination from lumbar puncture.

The microbiology of these pediatric infections is also unique with the involvement of enteric organisms compared to adult cases, which characteristically prove to be infections with skin flora. The most useful radiologic study in our case was a contrast enhanced spinal CT showing distinct uptake within the subdural space. MRI was nonspecific with diffusely increased intensity on T2 weighted images.

The management and review of this clinical entity in children and adults is compared. We recommend early surgical intervention with incision and open drainage.
54. SURGICAL MANAGEMENT OF SPINAL NEUROFIBROMATOSIS IN THE PEDIATRIC POPULATION

Andrew Glass, Allen Rothman, Bruce Rosenblum (New York, NY)

Recent developments in the field of genetic analysis have permitted the elucidation of the genotype of patients with both peripheral and central neurofibromatosis. Both types I and II may present with abnormalities of the spinal cord and spinal column.

There is a special subset of patients with neurofibromatosis who demonstrate extensive involvement of the spine. These include hamartoses, benign and malignant tumors, and osseous abnormalities. Because of the disproportionate growth of the spinal cord and vertebral column during childhood, many of these patients manifest symptoms of "spinal neurofibromatosis" during the formative years of life.

These complicated cases may present with mass lesions at multiple sites along the spinal axis, a variety of tumor types, and neoplastic lesions invasive enough to involve adjacent structures, including those in the neck and thoracic and peritoneal cavities. Osseous deformities of the spinal column may be either congenital or develop secondary to pressure effects of underlying masses. Abnormalities of the vertebral axis are particularly prone to occur in these children after extensive removal of their histologically micracytic bones during tumor surgery.

Based upon the treatment of a series of children with "spinal neurofibromatosis", management strategies are outlined. Appropriate timing and selection of lesions requiring surgery is stressed, as are techniques for both internal and external stabilization of the spine. The need for a multidisciplinary team is emphasized and appropriate diagnostic and therapeutic maneuvers are discussed, with particular emphasis on intraoperative monitoring. Screening with MRI and aggressive treatment are crucial.

55. THE MANAGEMENT OF ACQUIRED TORTICOLLIS IN CHILDREN SECONDARY TO ROTATIONAL ATLANTO-AXIAL SUBLUXATION

Mahmoud G. Nagib, Erich S. Wisiol, David E. Tubman (Minneapolis, MN)

Acquired torticollis secondary to atlanto-axial subluxation in children often presents management problems whether spontaneous or as a result of known etiology. Over a period of three years, the authors had the opportunity to attend seven children with such a disorder ranging in age from 7 years to 14 years. Except for one child who presented acutely, a mean interval of three months elapsed from the time of the initial insult to the time the patients were brought to our attention. One child presented with spontaneous torticollis; one child presented with torticollis secondary to an eosinophilic granuloma of the atlanto-occipital joint; one child presented with a large neurofibroma of the C2 root; one child presented with torticollis secondary to severe tonsillitis and cervical adenitis, and three children developed post traumatic torticollis. Emphasis will be placed on this last group. A follow-up period of at least one year was available for all patients. Five patients were eventually treated surgically while two were successfully managed through conservative methods. The two most salient difficulties encountered were: defining the extent of the deformity the child incurred at the time of the insult and selecting the ideal therapy. The reliance on high resolution CT scan and the addition of three-dimensional reconstruction of the atlanto-axial joint have rendered the differentiation between "physiologic" and "pathologic" subluxation more precise. Thus allowing for a better understanding of the pathology involved and a more accurate therapy. Children with a "physiologic" rotation of the C1 and C2 joints are preferably managed through physiotherapeutic methods except when a chronic deformity is at hand. Patients with a "pathologic" rotation are more likely to benefit from an internal immobilization after an initial attempt at
reduction by skeletal traction. In the following discussion, we will present our past experience and the management difficulties peculiar to this age group we have encountered.

56. IMMEDIATE SURGICAL EXPLORATION FOR TRANS-TEMPORAL HERNIATION IN PEDIATRIC HEAD INJURY

Brian T. Andrews, Lawrence H. Pitts (San Francisco, CA)

Immediate surgical exploration with bilateral placement of frontal, temporal and parietal burr-holes was performed in all children admitted to the emergency room with clinical signs of trans-temporal herniation following head injury, between 1981 and 1987. Those in whom an extra-axial hematoma was identified underwent an immediate craniotomy for evacuation of the mass. There were 17 children, 10 boys and 7 girls, between one and 17 years of age (mean 7.3 years). The median Glasgow Coma Scale (GCS) at the time of admission was 3 (range 3-10). Ten children had anisocoria and seven had bilaterally dilated and fixed pupils; five children had hemiparesis and 12 had symmetrical but abnormal motor findings. In 9 children (53%) a significant extra-axial hematoma was identified, in each case confined to the subdural space. In one case a small intracerebral hemorrhage deep within the dominant hemisphere was identified using intraoperative real-time ultrasonography; this lesion was not evacuated.

Postoperatively the twelve children with stable vital signs underwent an immediate computerized tomographic examination, confirming in each case the findings at surgery. The eight children that died were all subjected to autopsy; in no case did CT or autopsy identify an intracranial mass lesion that was not discovered at the time of initial surgical exploration. Overall, nine children survived (53%) and eight died (47%). Among those that survived, seven (78%) had a good outcome and two were left moderately disabled at late follow-up; no children were left with a severe neurological disability or vegetative. This study shows that a significant proportion of children with early signs of trans-temporal herniation following head injury have extra-axial hematomas, which may readily be identified by immediate exploratory burr-hole placement bilaterally; intraoperative ultrasonography also allows identification of
intraparenchymal hematomas. The survival and recovery of these severely head-injured children is much better than in similarly injured adults.

57. CRANIOCEREBRAL GUNSHOT WOUNDS IN CHILDREN

Michael S. Muhlbauer, Robert A. Sanford, W. Craig Clark, James C.H. Simmons (Memphis, TN)

Most series examining cranioencebral gunshot wounds (GSW) are based exclusively on adult injuries. No data has been collected reviewing gunshot wounds in the pediatric age group. This study examined the pediatric intracranial gunshot wounds at our institution, analyzing the existing data and comparing this with the authors' published experience with adult wounds.

Thirty-two children with cranioencebral gunshot wounds were treated by the Department of Neurosurgery at the University of Tennessee, Memphis over a seven year period. Mortality rate was 59%, all by brain death. There were eighteen children with Glasgow Coma Scale (GCS) scores of three or four and all died regardless of therapy. Fourteen patients received surgical debridement and nine had intracranial pressure monitoring. Significant hematomas on computed tomography were present in 28% of patients. The admission GCS score was the best predictor of outcome as categorized by the Glasgow Outcome Scale (Spearman Correlation r = -.90, p < .0005). Mean GCS on admission was 10.1 ± 3.0 in the good outcome group versus 4.1 ± 2.7 for those with poor outcome. Other factors associated with poor outcome included ventricular injury and bihemispheric wounding.

The value of using standardized data scales to prognosticate regarding the injury may be as useful to children as it is with adult injuries. As with the adult injuries, the authors advocate a conservative approach and feel that surgical intervention is probably not indicated in patients with GCS of three. Other prognostic factors were bihemispheric wounding and transventricular wounds. Contrary to the experience in adults, the presence of intracranial hematomas is not a predictor of poor outcome in children.
58. REAR SEAT LAP BELTS -- ARE THEY REALLY "SAFE" FOR CHILDREN?

Scott Paul Falci, Dennis L. Johnson (Washington, DC)

Although 80% of traffic fatalities in children could be reduced by proper use of safety restraint devices, a recent report suggested that rear lap belts may be more harmful than none at all.

Over the past three years during a time when seat belt use has been mandated by state and federal legislation and popularized by private safety campaigns, we have managed 8 lumbar spine fractures in children caused by standard rear lap seat belts. Children with the lap belt syndrome typically complained of abdominal and back pain, and the nature of their injury was underscored by a belt-shaped abrasion across the lower abdomen. Midlumbar spine fracture may be associated with paraplegia and life-threatening visceral injury.

The pelvis is the point of fixation in correct application of the adult lap belt. At impact the force of deceleration is dissipated in torque at the hip and in longitudinal distraction of the entire spinal axis. In a child the belt tends to ride above the pelvis, and the instantaneous axis of rotation moves up from the hip joints. The force of impact is concentrated in the midlumbar spine. If the spine is analyzed as a beam, the full spectrum of the reported injuries is predictable.

Lap belts are better than no restraint, but this study shows that the rear seat belts installed as standard equipment in US-manufactured cars do not meet the special needs of children.

59. HOW ARE SYMPTOMATIC CHRONIC SUBDURAL FLUID COLLECTIONS BEST MANAGED IN THE PEDIATRIC PATIENT?

The records of 103 patients in the pediatric age group diagnosed as having chronic subdural (SD) effusions at Children's Hospital of Los Angeles over the past 11 years were reviewed. Of this group, 75/103 (73%) underwent the placement of a subdural-peritoneal (SD-P) shunt. Immediate shunting was found to be more effective than other treatment modalities (p<0.01).

Of the shunted group, the SD fluid collection was unilateral in 15/75 (20%), and bilateral in 60/75 (80%). Of the 60 patients with bilateral SD effusions, 31/60 (52%) were treated with bilateral SD-P shunts, and 29/60 (48%) with unilateral shunts. Unilateral shunting was found to be just as effective as bilateral shunting when the fluid collection was bilateral (0.5 > p > 0.3).

Of the total group, 9/75 (12%) required shunt revision including 2/75 (3%) who developed infection. Eosinophilia in the SD fluid was associated with shunt obstruction requiring revision (p < 0.05). The shunts have not been removed in 30/75 (51%) of the patients with no untoward effects.

It is concluded that the placement of a unilateral SD-P shunt to treat symptomatic chronic unilateral or bilateral SD fluid collections is the treatment of choice. The shunts need not subsequently be removed.
60. CT FINDINGS OF INFANTILE AND SUBARACHNOID FLUID COLLECTION

Nobuhiro Morota, Shigeaki Kobayashi (Nagano-Pref., Japan), Keizzo Sakamoto (Hyogo-Pref., Japan)

Subdural fluid collection (SdFC) and subarachnoid fluid collection (SaFC) in infants was compared with special reference to the efficiency of contrast enhancement as a diagnostic modality.

Forty-three cases of SdFC and 8 cases of SaFC experienced over the last 5 years at our institutions were studied; each case underwent plain and enhanced CT scanning, and metrizamide CT cisternography.

The following results were obtained: 1.) In the plain CT, most of the cases with SaFC showed an irregular brain surface with deep sulci, while the cases with SdFC failed to demonstrate characteristic findings. 2.) In enhanced CT, vessels in the subarachnoid spaces were opacified as high density spots on the brain surface in SdFC, showing enlarged subdural spaces. In SaFC, such vessels were opacified along the inner table of the cranium. 3.) Co-existence of the two groups were observed on enhanced CT in some cases where vessels in the subarachnoid space were opacified like linear networks between the cranium and surface of the brain. 4.) The first CT sign of improvement in cases with SdFC was expansion of the subarachnoid space, followed by the brain expansion.

In conclusion, enhanced CT is most useful as a diagnostic procedure in differentiating the two diseases. Moreover, it is stressed that contrast enhancement is necessary in a case with SdFC in order to exclude an associated SaFC.

61. NEUROBEHAVIORAL SEQUELAE TO TRAUMATIC BRAIN INSULT IN CHILDREN: DIFFERENTIAL EFFECTS ON THE FAMILY

Seth Warschausky, Patricia M. Meylan, Alexa I. Canady (Detroit, MI)

This presentation involves a discussion of the inter- actions between neurobehavioral sequelae to traumatic brain insult, including commonly noted acquired attention deficit symptomatology and social disinhibition, and the distress experienced by the family at the individual and systemic level. Preliminary results and case examples from a longitudinal study of family adaptation to traumatic brain insult in children ages 6-16, are used to highlight the dynamic interactions between the neurobehavioral profile and family system dimensions over time, in the early phases of recovery. Specifically, data are presented on primary care giver and sibling distress in single versus two parent families, fathers compared with mothers, and self-report measures of family communication, at one month post insult. Preliminary data show an apparent disengagement of fathers' distress from the child's degree of behavioral dysfunction, in contrast to the significant linkage noted in mothers' self-report indices. Siblings in single parent families exhibit a lower degree of dysfunctional behavior than siblings in two parent families. Results are discussed in terms of methodological issues and systemic adaption of the family to a trauma. Data from retrospective child behavior checklists and academic history are discussed as illustrations of the biased sample effects frequently noted in the literature. Repeated measure data from one and four months post insult, including neuropsychological evaluations, and self-report measures of psychological and behavioral dysfunction, are also discussed in terms of implications for psychosocial and educational recommendations for discharge planning.
62. SURGICAL TREATMENT OF UNILATERAL MEGALENCEPHALY

Richard E. George, Harold J. Hoffman, Laurence E. Becker,
Paul Hwang (Toronto, Ontario, Canada)

Unilateral megalencephaly is a rare brain malformation caused by defective cell migration with subsequent overgrowth of one cerebral hemisphere. Six patients with unilateral megalencephaly managed at The Hospital for Sick Children were reviewed.

Clinically, these patients presented with the early onset of intractable seizures, progressive hemiparesis, and severe developmental delay. Cardiac failure was present in two patients and was due to vascular shunting through the enlarged hemisphere. One patient died from heart failure prior to undergoing surgery.

CT and MRI provided clear delineation of the pathology. All patients exhibited enlargement of the involved hemisphere and ipsilateral ventricle, abnormal gyral patterns, and cortical thickening.

Our patients were treated with staged hemispherectomies utilizing intraoperative electrocorticography. Frequently, the involved hemisphere was tough and gliotic and required an ultrasonic aspirator for resection. Younger patients had extremely vascular hemispheres and one died secondary to intraoperative blood loss.

Pathologically, unilateral megalencephaly was characterized by a bulky hemisphere with disorganized gyral pattern. Normal cortical lamination was lost and the cortex was characterized by numerous, disorganized, large neurons. The white matter showed gliosis and occasional ectopic neurons. Focal regions of necrosis and calcium deposits were seen.

Hemispherectomy provided excellent seizure control. Two patients are seizure free and the third actually achieved a marked reduction in seizures. Seizure control was improved in patients treated early. Developmental regression was halted in all patients and was reversed in one treated at 3 months of age. Contralateral hemiparesis and homonymous hemianopsia present pre-operatively persisted following hemispherectomy, however, improved function was frequently achieved.

We advocate treatment of unilateral megalencephaly with staged hemispherectomy performed early in the patient's course. Intraoperative electrocorticography, attention to cardiac status and maintenance of blood volume are essential for successful treatment.
63. HEMISPHERECTOMY FOR INTRACTABLE SEIZURES IN CHILDREN WITH HEMIMEGALENCEPHALY

DiRocco Concezio, Vigevano Federico, Ceddia Antonello, DiTrapani Girolamo (Rome, Italy)

Hemimegalencephaly is a dysplastic disorder characterized by the unilateral enlargement of the brain, which can occur isolately, in association with body hemihypertrophy with skin anomalies. The affected cerebral hemisphere exhibits serious histological alterations, such as abnormalities of the cytoarchitecture of the cortex, with the absence of the cellular stratification, presence of giant neurons, neuronal and glial heteroplas.

The clinical manifestations include hemiparesis, hemianopsia, psychomotor retardation, and epilepsy. Seizures usually appear in the first two weeks of life and become intractable in almost all the cases, eventually leading to precocious death.

In the present report the authors describe their experience on five children with intractable epilepsy, surgically treated through removal of the affected hemisphere. In all cases, seizures disappeared immediately following the operation. A marked improvement in psychomotor development was observed in all but one subject, suggesting that the removal of the abnormal hemisphere may favor the mental and motor development, besides controlling epilepsy. The child, whose improvement was of a relatively minor degree, demonstrated the maximal anatomical abnormalities of the removed cerebral hemisphere. This child was also the only patient who developed a post-operative hydrocephalus, which required a ventriculo-peritoneal CSF shunting procedure.

64. RESULTS OF CALLOSOTOMY IN RETARDED PATIENTS WITH DROP ATTACKS

Thomas G. Oravetz, Glenn A. Meyer, Gregory J. Harrington (Milwaukee, WI), Jerome V. Murphy (Kansas City, MO)

From 1984 through 1988, 10 patients with medically refractory epilepsy underwent partial or complete section of the corpus callosum in an attempt to control their seizures. The study consists of 9 males and 1 female with ages ranging from 9 to 32 years, and with the average age at the time of surgery being 15.2 years. All the patients had some form of bilateral seizure disorder and all had drop attacks which resulted in frequent falls and injury. In addition, all the patients in the study had severe to borderline mental retardation. The period of medical treatment ranged from 5 to 21 years, with the average length of medical therapy being 11.4 years. Preoperative evaluation included CT scanning, angiography, neurological testing, EEG analysis with videomonitoring, and MRI scanning in most of the patients. Intraoperative EEG monitoring was also used in 7 of the 10 patients. There were no operative mortalities and only two cases of operative morbidity. One patient had a postoperative disconnection syndrome which resolved and another had a small left hemisphere medial wall infarct which was not clinically significant. All the patients who underwent surgery showed improvement in either seizure severity or frequency, or both. Contrary to most prior reports, this study did not exclude very retarded patients, and in fact, the most severely retarded patient showed dramatic improvement in functional ability and social interaction.
65. HISTOLOGIC CHANGES IN DORSAL ROOFTLETs FROM CHILDREN WITH CEREBRAL PALSY

Richard A. Rovin (Chicago, IL), Yusuf Ersahin (Bornoval Izmir, Turkey), Bruce B. Storrs (Chicago, IL)

Though cerebral palsy affects both the central and peripheral nervous systems, selective dorsal rhizotomy can improve functional capacity in children with spasticity from CP. The physiologic basis for this decrease in spasticity following rhizotomy is not clear, but may in part be due to the interruption of the facilitatory influences of dorsal root afferents. The anatomic components of the spasticity circuitry are not well defined either. Data from experimental spinal cord injury models reveal that the distribution of the specific axonal populations is altered in dorsal roots and suggests that collateral sprouting from the dorsal root axons is important in the development of spasticity. The authors compared dorsal nerve rootlets from normal controls to those removed from patients with CP at the time of rhizotomy. One-hundred thirty-nine dorsal rootlets from 16 CP patients were examined and compared to 19 rootlets from three normal controls. We found that large myelinated axons are increased and small unmyelinated axons are significantly increased in dorsal rootlets of patients with cerebral palsy (P<0.002 and P<0.001, respectively). We conclude that the pathologic changes occurring in the dorsal rootlets of patients with cerebral palsy are similar to those changes found in animal models of spasticity.

66. SELECTION OF MUSCLES FOR MONITORING DURING SELECTIVE POSTERIOR RHIZOTOMY

Lawrence H. Phillips, II, T.S. Park, Jacek Malik (Charlottesville, VA)

The "selectivity" of Selective Posterior Rhizotomy (SPR) derives from electrophysiological monitoring of responses in leg muscles evoked by stimulation of dorsal rootlets in the cauda equina. One criterion for section of rootlets is spread of reflex activity beyond the spinal segment stimulated. Knowledge of segmental anatomy of the lower extremity muscles is critical for interpretation of the electrophysiology.

We performed a protocol of ventral root (VR) stimulation during SPR in 50 cases. Intramuscular recordings were made in adductor longus (AL), vastus medialis (VM), vastus lateralis (VL), anterior tibial (AT), peroneus longus (PL), lateral gastrocnemius (LG), and medial gastrocnemius (MG), while stimulating VR's from L2 to S2. The compound action potential area was measured, and the percentage of total evoked area for each VR was calculated from the summed areas for each muscle.

The major root supplying each of the muscles was: L2 or L3 for AL, VM, and VL; L4 for AT; L5 for PL; and SI or S2 for LG and MG. Deviations from this pattern in the form of pre- or post-fixed distributions were found in 27% of studies. Individual muscles derived more than 50% innervation from a single root in 40% of muscles. The number of roots innervating a single muscle ranged from 2-5.

We recommend recording from a minimum of 5 muscles in order to adequately cover the segmental innervation from L2-S2. The combination of AL, VL, AT, PL, and MG provides the most comprehensive coverage of these segments.
67. HOW SELECTIVE IS SELECTIVE POSTERIOR RHIZOTOMY?

Alan R. Cohen (Boston, MA)

Although selective posterior rhizotomy has become a popular surgical treatment for spasticity and multiple institutions have documented its therapeutic efficacy, the procedure remains unstandardized with little known about its mechanism of action. The technique is based upon an assumption that intraoperative neurophysiologic monitoring can identify two populations of dorsal rootlets: those which are involved in abnormal circuits contributing to spasticity, and those which are "normal". Abnormal rootlets are divided and normal rootlets are spared, thereby reducing spasticity and eliminating sensory side-effects. However, the definition of normal and abnormal responses varies among centers and it is still not clear what is really being selected in the procedure or even whether selective rhizotomy is superior to non-selective random rootlet selection.

To help clarify these issues we undertook a detailed analysis of intraoperative electromyographic (EMG) patterns in ten children with spastic cerebral palsy undergoing selective posterior rhizotomy. The dorsal rootlets from L2 to S2 bilaterally were individually isolated in the cauda equina and repetitively stimulated. EMG activity was recorded from multiple muscle groups in both lower extremities.

The electrical response to dorsal rootlet stimulation was highly reproducible. However, the response of a given rootlet was significantly altered simply by the sectioning of neighboring rootlets. In some cases this response was facilitated and in others it was inhibited. Thus, the order in which rootlets are sectioned may have a strong influence upon the response of rootlets stimulated subsequently. The "normal response" described by Fasano of temporal inhibition to a repetitive stimulus train was

never seen by us in any case. Therefore, we used other parameters to select rootlets for sectioning. Postoperatively, there was an immediate and significant reduction of muscle tone in the lower extremities in every patient. To a lesser extent upper extremity tone was reduced as well.

We conclude that selective rhizotomy effectively reduces spasticity in children with cerebral palsy. We question the electrophysiologic criteria used to make the procedure selective and suggest ways to use intraoperative monitoring more effectively.
68. GAIT STUDIES IN PATIENTS UNDERGOING SELECTIVE DORSAL RHIZOTOMY

Leslie D. Cahan (Orange, CA), Lauren Beeler (Newport Beach, CA), Jan Adams, Jacquelin Perry (Downey, CA)

A decade ago, Fasano reported selective dorsal rhizotomy to be effective in decreasing spasticity of children with cerebral palsy. While the past two years has witnessed an explosion of interest in this operation in the United States, it is vitally important that documentation of the results of this surgery be based upon observer independent, quantified tools. The availability of sophisticated gait laboratories has allowed us to study in detail 15 patients before and after selective dorsal rhizotomy and to quantify the changes in gait related to the surgery and subsequent physical therapy. Gait analysis includes measurement of: (1) stride characteristics (velocity, stride length, cadence, single and double limb support time); (2) Motion analysis about the hip, knee and ankle; (3) Foot switch analysis of foot placement; (4) Ambulatory EMG monitoring.

Fifteen patients with spastic diplegia ranging from 4 to 20 years have been studied at a single gait laboratory before and 6 to 9 months after surgery. In each case, there has been clear reduction in tone and an improvement in gait as documented by the above studies. Detailed statistical analysis of EMG response to quick stretch, motion analysis and stride characteristics will be presented. In an update to the paper presented to this meeting last year, more patients have now been studied and a more sophisticated analysis of the motion analysis is now available. Studies of this sort will allow comparison of this surgery with currently available orthopedic operations.

69. PAIN CONTROL AFTER SELECTIVE RHIZOTOMY IN CEREBRAL PALSY

J.P. Mickle, Mary Sparkes, Alan Klein, Ann-Christine Duhaime (Gainesville, FL)

Selective posterior dorsal rhizotomy is being utilized more frequently in an attempt to improve the functional status of certain patients with cerebral palsy. The surgical procedure is usually performed through a narrow multi-level lumbaracral laminectomy L2-S1. The dorsal roots of S2-L2 bilaterally are isolated at their foraminal exits intradurally and each dorsal root is gently divided into multiple fascicles for neurophysiological evaluation. Based on various criteria certain fascicles are divided. In our early experience with this procedure post-operative pain control and muscle spasms were a major consideration in recommending this procedure in children. The treatment most effective to date has been the continuous infusion of intravenous narcotics combined with the use of various muscle relaxants. We report here our experience with the use of epidural Duramorph in 15 patients after selective rhizotomy in the treatment of cerebral palsy. All these patients achieved a remarkable level of pain control and none of these patients have suffered serious side effects with the exception of pruritis. Analgesia lasted between 8 and 24 hours per dose of Duramorph (0.5 mg per cc @ 0.05 mg. per kg per dose). The patients that required no other medication for pain control remain in the intensive care environment with the epidural catheter in place an average of 3 days and are sent to the regular hospital ward 12 hours after the last dose of Duramorph. Naloxone has been very effective in reversing the pruritis which may occur with the use of epidural Duramorph.

This anesthetic technique has virtually eliminated one of the major concerns in our recommending this operation for the control of spasticity in certain patients with cerebral palsy.
70. INTERVENTIONAL NEURORADIOLOGIC MANAGEMENT OF THE VEIN OF GALEN MALFORMATION IN THE NEONATE

Samuel F. Ciricillo, Michael S.B. Edwards, Grant Heishma, Randall Higashida (San Francisco, CA)

The authors review their experience treating thirteen infants with vein of Galen malformations who presented with severe congestive heart failure at birth. The first five infants underwent cranitomity and attempted clipping of the feeding vessels. All five of the infants died in the perioperative period. Eight other infants were considered for staged arterial and venous interventional neuroradiologic procedures for treatment of their malformations. Ventilatory support was withdrawn from one child born with severe hydrocephalus and a massive dural and intraparenchymal arteriovenous malformation causing significant neurologic impairment. Two infants died despite treatment—one from congestive heart failure and the other from massive intraparenchymal and intraventricular hemorrhage. One infant sustained a large middle cerebral artery infarction due to the embolization procedure and survives with significant neurologic impairment and developmental delay. Another infant survives with a partial visual field defect from occipital lobe infarction, which was not directly attributable to treatment. Three other children are neurologically and developmentally normal. Follow-up neuroradiologic studies in all five survivors show some degree of flow in the malformations. Sequential magnetic resonance scans reveal progressive thrombosis of the vein of Galen in three patients. Improved understanding of the anatomy and physiology of these malformations using Doppler flow ultrasonography has increased our ability to treat these lesions. Advances in interventional neuroradiologic techniques will continue to improve our management of these difficult malformations. While encouraging, our results using staged arterial and venous embolic procedures attest to the difficulty in managing and treating these patients. Careful assessment of the long-term neurodevelopmental outcome of surviving patients is needed and will help guide our future interventions.

71. INTRACRANIAL ANGIOMAS IN CHILDREN

Marion L. Walker, Mark V. Reichman (Salt Lake City, UT)

During the past 10 years 31 pediatric patients with intracranial angiomata have been evaluated at the Primary Children's Medical Center. These patients have been divided into two groups based on the angiographic appearance of the lesion: low flow and high flow. Twenty-three patients had angiomata with high flow characteristics. Of these 20 cases, 17 were treated with operative resection of the lesion. Three AVM's were deemed unresectable, one of which has been partially embolized. Eight low flow lesions were evaluated. These include 1 capillary hemangiomata, 3 small AVM's, and 4 cavernous angiomata.

Regardless of flow characteristics of the lesions, subarachnoid hemorrhage is the most common presentation of patients with intracranial angiomata. High flow lesions may present as mass lesions or with symptoms of congestive heart failure. Hemorrhage in these lesions is generally more catastrophic and is associated with increased morbidity and mortality than low flow lesions. The second most common presentation of low flow lesions is seizures, however, nearly all patients presenting with seizures had evidence of previous or concurrent hemorrhage on MRI or CT or at the time of surgery.

The natural history of intracranial angiomata is unclear, however, repeated hemorrhage occurred in 4 patients with low flow lesions and in 10 patients with high flow lesions in our series. Our surgical results demonstrate that low flow lesions can be resected with very low morbidity and excellent results. High flow lesions should be evaluated individually, however, most of these can also be successfully obliterated with surgery, embolization, or both with very acceptable results.
INTRACRANIAL HEMORRHAGE DURING EXTRACORPOREAL MEMBRANE OXYGENATION OF THE NEWBORN: THE NEW ORLEANS EXPERIENCE

Susan Reid Hemley, Richard A. Coulon (New Orleans, LA)

Intracranial hemorrhage represents the most serious complication associated with the treatment of respiratory insufficiency in the newborn by extracorporeal membrane oxygenation (ECMO). Ninety-two newborns at Ochsner Foundation Hospital underwent ECMO from 1983 to 1987, after meeting criteria that were formerly associated with close to 100% mortality. Eleven of these infants suffered intracranial hemorrhages. We analyzed these cases and compared them with the infants who did not have this complication, considering them with respect to gestational age, birth weight, age on beginning treatment, and clinical course. We also compared our experience with that of other ECMO centers. The aim of the study was to elucidate the causes and natural history of ECMO-related intracranial hemorrhage, in the interest of reducing its incidence and severity and thus allowing a wider and safer utilization of this very valuable method for treating infants with otherwise intractable pulmonary insufficiency.
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