Flannery

Section of Pediatric Neurological Surgeons

of the

American Association of Neurological Surgeons

13th ANNUAL MEETING

Hotel Utah

Salt Lake City, Utah

December 11-13, 1984

PROGRAM SUMMARY

This program has been approved by the Joint Committee on Education of the American Association of Neurological Surgeons and Congress of Neurological Surgeons for a maximum of 13 hours of Category I credit toward the Continuing Education Award in Neurosurgery.

PAOLO RAIMONDI LECTURERS

E. Bruce Hendrick	1978
Paul C. Bucy	1979
Floyd Gilles	1980
(Panel Discussion)	1981
(Panel Discussion)	1982
Derek Harwood-Nash	1983
Anthony E. Gallo, Jr.	1984
David G. McLone	
(Panel Discussion)	

PEDIATRIC SECTION CHAIRMEN

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

PEDIATRIC ANNUAL MEETING SITES

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

The Pediatric Section of Neurological Surgery of the American Association of Neurological Surgeons gratefully recognizes the support of the following exhibitors for the 1984 Pediatric Annual Meeting:

American V. Mueller (American Heyer-Schulte)—Chicago, Illinois
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All registrants are encouraged to visit the exhibit area frequently during the meeting.

PROGRAM

PEDIATRIC SECTION

AMERICAN ASSOCIATION OF NEUROLOGICAL SURGEONS

Hotel Utah
Salt Lake City, Utah

December 11-December 13, 1984

TUESDAY, DECEMBER 11, 1984

6:00-9:00 p.m. Registration—Mezzanine
7:00-9:00 p.m. Reception—Empire Room

WEDNESDAY, DECEMBER 12, 1984

7:00 a.m. Registration—Mezzanine
7:00 a.m. Breakfast—Jade Room
7:45 a.m. Meeting—Lafayette Ballroom
Opening Remarks
Harold J. Hoffman
7:50 a.m. Welcoming Remarks
M. Peter Heilbrun
Marion L. Walker
8:00 a.m. Annual Paolo Raimondi Lecture

BRAIN TUMORS—Moderators: David G. McLone, Donald Reigel

8:30 a.m. 1. "Long Prodromal Presentations of Pediatric Brain Tumors"
Thomas G. Luerssen, Kathy R. Siegel, Roger J. Packer, Leslie N. Sutton, Derek A. Bruce, Luis Schut
Philadelphia, Pennsylvania

8:45 a.m. 2. "Cerebral Primitive Neuroectodermal Tumor in Childhood"
Tadanori Tomita, David G. McLone
Chicago, Illinois

9:00 a.m. 3. "Intraventricular Tumors—Treatment with the CO_2 Laser"
Walker L. Robinson, Michael Salcman
Baltimore, Maryland

9:10 a.m. 4. "The Feasibility and Advisability of Resecting Midline Intra-axial Gliomas in Children"
A. Leland Albright, Robert J. Sclabassi
Pittsburgh, Pennsylvania

9:20 a.m. 5. "Brain Stem Tumors: A Reappraisal"
Fred Epstein, Lawrence L. McCleary
New York, New York

9:30 a.m. 6. "The Microcystic Medullary Juvenile Astrocytoma: A Pathologic Study",
Stuart Goodman, Marion L. Walker, J. J. Townsend, Bruce B. Storrs, W. M. Gooch
Salt Lake City, Utah

9:40 a.m. Panel Discussion for Papers 3-6

10:00 a.m. **COFFEE BREAK—Mezzanine**
View Exhibits—Jr. Ballroom

BRAIN TUMORS—Moderators: Harold J. Hoffman, Kenneth Shapiro

10:30 a.m. 7. "A Review of 26 Children Treated for Medulloblastoma"
Mark O'Brien, Daniel L. Barrow, Yoshio Takei
Atlanta, Georgia

10:45 a.m. 8. "Management of Subependymal Giant Cell Astrocytoma"
Robert A. Sanford
Jackson, Mississippi

11:00 a.m. 9. "Gliomas Associated with Radiotherapy: Review of 4 Cases"
Thomas S. Berger, Bolesiaw H. Liwnicz
Cincinnati, Ohio

11:15 a.m. 10. "Preliminary Experience with 125-Iodine Brachytherapy in Children"
Michael S. B. Edwards, Phillip H. Gutin, Steven Leibel, Sharon Lamb
San Francisco, California

11:30 a.m. 11. "Dumb-bell Neuroblastoma in Children: Review and Treatment"
Edward J. Kosnik, Martin P. Sayers
Columbus, Ohio

11:45 a.m. 12. "Pituitary Hyperplasia Due to Hypothyroidism"
Leslie N. Sutton, Thomas Moshang, Larissa Bilanink, Lewis Samuel
Philadelphia, Pennsylvania

12:00 noon **LUNCH—Empire Room**

CT/MRI—Moderators: Michael Edwards, Gordon McComb

1:15 p.m. 13. "Morphological Study of Frontonasal Encephalocele Using 3-Dimensional Reconstruction of Computed Tomography"
David C. Hemmy, David J. David, Donald A. Simpson
Milwaukee, Wisconsin/Adelaide, South Australia

1:30 p.m. 14. "Obliteration of Basal Cisterns in CT, The Cardinal Sign of Life-Threatening Shunt Failure"
Dennis L. Johnson, Charles Fitz, David C. McCullough
Washington, D.C.

1:45 p.m. 15. "Ultrasonic Anatomy of the Posterior Fossa: Anatomic—Sonographic Correlations"
Thomas P. Naidich, David Yousefzodeh
Chicago, Illinois

2:00 p.m. 16. "Imaging the Craniovertebral and Cervico-medullary Junction in Children: A Prospective Evaluation of Multimodal Investigative Techniques"
Arnold H. Menezes, Wendy R. Smoker, Lindell Gentry
Iowa City, Iowa

2:10 p.m. 17. "Magnetic Resonance Imaging in Spinal Dysraphism,"
Joan L. Venes, Alex M. Aisen, Michael A. DiPietro, James Knake
Ann Arbor, Michigan

2:20 p.m. 18. "Surgical Correlates of Nuclear Magnetic Resonance (NMR)"
Nazih Moufarrij, Joseph F. Hahn, Michael T. Modic, Meredith Weinstein
Cleveland, Ohio

2:30 p.m. Panel Discussion, Papers 16-18

3:00 p.m. **COFFEE BREAK—Mezzanine**
View Exhibits—Jr. Ballroom

DEVELOPMENTAL ANOMALIES—Moderators: Derek Bruce, Joan Venes

3:30 p.m. 19. "The Type I Chiari Malformation in Childhood and Adolescence"
W. Jerry Oakes
Durham, North Carolina

3:45 p.m. 20. "Real Time Ultra Sound Evaluation of Chiari II Malformation in Infants"
Bruce B. Storrs, Marion L. Walker
Salt Lake City, Utah

4:00 p.m. 21. "Dermoid Tumors at the Site of Previous Myelomeningocele Repair"
R. Michael Scott, Louis Bartoshesky, Samuel M. Wolpert, Seymour Zimbler, George Klauber
Boston, Massachusetts

4:15 p.m. 22. "Progressive Cauda Equina Syndrome in Children with the Caudal Endodermal Syndrome"
Arden F. Reynolds, Michael Pollay, Patrick Barnes, A. Sullivan, E. Smith, W. Tunell, W. Barnes, P. Lester
Oklahoma City, Oklahoma

4:30 p.m. 23. "Vein of Galen Aneurysm: Combined Treatment by Embolization and Surgery"
Jeffrey H. Wisoff, Alejandro Berenstein, Fred J. Epstein
New York, New York

4:40 p.m. 24. "The Transtorcular Approach to Vein of Galen Aneurysms—A Preliminary Report"
J. Parker Mickle
Gainesville, Florida

4:50 p.m. 25. "Cerebrovascular Malformations in Children: A Retrospective Analysis of 28 Cases"
Francisco A. Gutierrez, David G. McLone, Thomas P. Naidich
Chicago, Illinois

5:00 p.m. Discussion of Papers 23-25

5:15 p.m. Annual Business Meeting—Members Only

7:00 p.m. Reception—Salt Lake Art Center

8:00 p.m. Annual Banquet—Salt Lake Art Center

THURSDAY, DECEMBER 13, 1984

7:00 a.m. Breakfast—Jade Room

HYDROCEPHALUS—Moderators: William Cheek, Fred Epstein

8:00 a.m. 26. "A Laboratory Model of Shunt Dependent Hydrocephalus"
Arno Fried, Kenneth Shapiro
Bronx, New York

8:10 a.m. 27. "An Experimental Model Showing Why the Subtemporal Decompression Should Work in the Slit Ventricle Syndrome With Some Suggestions As To Why It Doesn't Work Often Enough"
Kenneth Shapiro, Futoshi Takei
Bronx, New York

8:20 a.m. Discussion of Papers 26-27

8:30 a.m. 28. "Management of Expanding Multiple Loculated CSF Filled Cavities Following Enteric Organism Ventriculitis"
Robert Breeze, J. Gordon McComb
Los Angeles, California

8:40 a.m. 29. "Loculated Ventricles—Aren't"
David G. McLone, Thomas P. Naidich
Chicago, Illinois

8:50 a.m. Discussion of Papers 28-29

9:00 a.m. 30. "Ventriculo-peritoneal Shunts in the High-Risk Newborn Under 2,000 Grams of Weight—Clinical Report"
Hector E. James, R. Bejar, L. Gluck, R. Coen, A. Merritt, F. Mannio, P. Bomberger, B. Saunders, H. Schneider
San Diego, California

9:15 a.m. 31. "Outcome in Children with Prenatally Diagnosed Congenital Hydrocephalus"
Charles C. Duncan, Laura R. Ment, David T. Scott, Richard A. Ehrenkranz
New Haven, Connecticut

9:30 a.m. 32. "SER & BAER in Experimental Hydrocephalus. Effects of Ventricular Dilation and Pressure Waves"
Leslie N. Sutton, Byung-Ku Cho, Gretchen Avery, Jurg Jaggi
Philadelphia, Pennsylvania

9:40 a.m. 33. "Use of BAER in the Management of Neonatal Hydrocephalus"
Meredith Olds, Bruce B. Storrs, Marion L. Walker
Michael J. Cevette, Nanette Newberg
Salt Lake City, Utah

9:50 a.m. Discussion of Papers 32-33

10:00 a.m. **COFFEE BREAK—Mezzanine**
View Exhibits—Jr. Ballroom

HYDROCEPHALUS—Moderators: Robin Humphreys, Mark O'Brien

10:30 a.m. 34. "One Year Follow-up of the Prospective Randomized Double Blind Trial of Antibiotic Prophylaxis in Cerebrospinal Shunt Infection"
Mel H. Epstein, K. Kumar, P. Lietman, W. Hughes
Baltimore, Maryland

10:40 a.m. 35. "Decreased Risk of Infection in Cerebrospinal Fluid Shunt Surgery Using Prophylactic Antibiotics: A Case-Control Study"
Beverly C. Walters, Harold J. Hoffman, E. Bruce Hendrick, Robin P. Humphreys
Toronto, Ontario, Canada

10:50 a.m. Discussion of Papers 34-35

11:00 a.m. 36. "Contribution of the Spinal Compartment to Cerebrospinal Fluid Drainage"
J. Gordon McComb, Shigeyo Hyman, Martin H. Weiss
Los Angeles, California

11:15 a.m. 37. "The Pathophysiology of IVH in the Beagle Puppy"
Dennis L. Johnson, Catherine Shaer, David C. McCullough
Washington, D.C.

11:30 a.m. 38. "In Vitro Evaluation of CSF Shunt Function by Radionuclide Clearance Studies"
Bruce D. Pendleton, P. Alex Roberts, Arden F. Reynolds, Michael Pollay
Oklahoma City, Oklahoma

11:45 a.m. 39. "Abdominal CSF Cyst Syndrome: Diagnosis and Management"
Yoon S. Hahn, Herbert H. Englehard
Chicago, Illinois

12:00 noon **LUNCH**—Empire Room
Brain Tumor Study Group Report
Moderators: Harold Rekate, Larry Page

1:00 p.m. 40. "Moya Moya: Experience at the Hospital for Sick Children"
Meredith V. Olds, Harold J. Hoffman
Toronto, Ontario, Canada

1:15 p.m. 41. "Extreme Hyperventilation and Cerebral Bioenergetics in the Cat"
Patricia Gibbons, David Swedlow, Frank Welsh, Derek A. Bruce
Philadelphia, Pennsylvania

1:30 p.m. 42. "Operative Planning For Congenital Cerebral Malformations and Hydrocephalus: Benefit of Stable Xenon Computed Tomography"
Kim H. Manwaring, T. J. Tarby, John Hodak, Hal W. Pittman
Phoenix, Arizona

1:45 p.m. 43. "Temporal Lobectomy in Children with Epilepsy"
W. Richard Marsh, Fredric B. Meyer, Edward R. Laws, Frank W. Sharbrough
Rochester, Minnesota

2:00 p.m. 44. "Spontaneous Epidural Cervico-Thoracic Hematoma Extending From C-3 to T-3 in a Child"
Joan H. Humphreys, John P. Laurent, William R. Cheek, Hatem Megahed
Houston, Texas

2:15 p.m.	45. "Selective Posterior Rhizotomy for the Relief of Spasticity" W. J. Peacock Cape Town, South Africa
2:30 p.m.	46. "Intracranial Arachnoid Cysts in Children" G. R. Harsh, Michael S. B. Edwards, Charles B. Wilson San Francisco, California
2:45 p.m.	47. "The Manifestations and Management of Hydrosyringomyelia in Childhood" John Neill, Harold J. Hoffman, E. Bruce Hendrick, Robin P. Humphreys Toronto, Ontario, Canada
3:00 p.m.	**COFFEE BREAK — Mezzanine** Moderators: Hector James, Mike Scott
3:30 p.m.	48. "Occipital Plagiocephaly Due to Unilateral Lambdoid Synostosis — A Reappraisal and Some New Thoughts" Clinton F. Miller, John W. Skinner, Joseph M. Nadell New Orleans, Louisiana
3:45 p.m.	49. "Terminal Myelocystocele" Thomas P. Naidich, David G. McLone Chicago, Illinois
4:00 p.m.	50. "Increase in Ventricular Size with High Pressure Infusion of Artificial CSF into the Lateral Ventricle of Dogs" Harold L. Rekate, Scott Erwood, Howard Chizeck Cleveland, Ohio
4:15 p.m.	51. "Hydrocephalus: Symptomatic Low ICP and Asymptomatic Slit Ventricle Syndromes Treated by Flo-Control Valve Shunting" Eldon L. Foltz, Jeffery Blanks Orange, California
4:30 p.m.	52. "Craniofacial Techniques in Pediatric Neurosurgery" Larry V. Carson Augusta, Georgia
4:45 p.m.	53. "Modified Prone Position and Cross-Bar Craniotomy For Total Cranial Vault Reshaping" T. S. Park, Charles S. Haworth, M. Sean Grady, John A. Jane Charlottesville, Virginia
7:00 p.m.	Mormon Tabernacle Choir Rehearsal

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SCIENTIFIC ABSTRACTS

1. LONG PRODROMAL PRESENTATIONS OF PEDIATRIC BRAIN TUMORS

Thomas G. Luerssen, M.D., Kathy R. Siegel, P.A.C., Roger J. Packer, M.D., Leslie N. Sutton, M.D., Derek A. Bruce, M.D., Ch.B., Luis Schut, M.D. (San Diego, CA)

Fourteen children are presented who fulfilled the following criteria: 1) The onset of neurological disturbances early in childhood. 2) Normal or non-diagnostic neurodiagraphic studies at the time of initial presentation. 3) The subsequent demonstration of CNS neoplasms after an interval of greater than 2 years. Three general initial presentations were found: 1) Obstructive hydrocephalus (4 children), with the onset at an average age of 5 years. All of these children were subsequently shown to harbor posterior fossa tumors after an average interval of 6 years. 2) Epilepsy (6 children) with the average age of onset being 5.2 years. All of these children were subsequently shown to harbor hemispheral tumors after an average interval of 6.2 years. 3) Neuro-psychiatric presentations (4 children), such as "Learning Disabilities", "Cerebral Palsy", or "Psychogenic Vomiting". The average age of initial presentation was 1.5 years; the average interval until tumor diagnosis was 5.5 years. These findings suggest that a small percentage of children with CNS neoplasms can present initially as a more common pediatric neurological syndrome and carry that diagnosis for prolonged periods. Thus, a high index of suspicion is required in order to correctly diagnose the underlying problem, so that definitive treatment can be undertaken at the earliest opportunity.

2. CEREBRAL PRIMITIVE NEUROECTODERMAL TUMOR IN CHILDHOOD

Tadanori Tomita, M.D., David C. McLone, M.D., Ph.D. (Chicago, IL)

Ten children with cerebral primitive neuroectodermal tumors (PNET) were treated from 1979 through 1983. There were 5 males and 5 females and age ranged from 22 months to 13 years. All were diagnosed by computed tomography (CT) and angiography which indicated a large partially cystic, calcified mass with variety of vascularity. Nine patients underwent craniotomy with gross total resection in 4, partial resection in 4 and biopsy alone in 1. One patient received only CSF diversion shunt and whole radiation therapy and autopsy confirmed cerebral PNET. All except one patient who died 4 days after partial resection of the tumor received radiation (5018-5500 rads over whole brain: plus focal boost with 1080 rads in one, and plus additional spinal radiation 2518 rads in 1). Beside one post-operative death, 6 patients died of tumor 6 to 25 months after diagnosis. Three patients are alive with tumor control over 18 to 36 months. Two of these received intracavitary injection of methotrexate and AZQ with disappearance of tumor on CT.

Current concept of PNET and management of the patients with PNET will be discussed.

3. INTRAVENTRICULAR TUMORS-TREATMENT WITH THE CO_2 LASER

Walker L. Robinson, M.D., Michael Salcman, M.D. (Lutherville, MD)

The CO_2 laser has proven to be a valuable surgical adjunct in the treatment of intfacranial and intraspinal tumors. In intracranial lesions, the laser's unique qualities have proven especially useful in lesions deep to eloquent cortex, basal meningiomas, and intraventricular tumors.

Over the past 36 months, 472 tumors were resected in our clinic with 144 of the cases being carried out utilizing the laser (30.5%). Intraventricular lesions were encountered in 21 (14.6%) of the laser cases. The 21 cases were distributed as follows:

Sex:	15 males/6 females	
Pathology:	Medulloblastoma	-10
	Astrocytomas (Gr. I/II)	-5
	Choroid Plexus Papilloma	-4
	Hemangioblastoma	-2
Location:	4th Ventricle	-15
	Lateral Ventricle	-5
	3rd Ventricle	-1

Our operative results show no mortality, a 4.8% (1/21) morbidity rate (an epidural hematoma after ventricular decompression), gross total resections in 86% (18/21), near total resections in 14% (3/21), and no recurrences to date (3 year maximum follow-up).

The surgical techniques necessary to allow successful resection of intraventricular lesions include:

- Variations in spot size and wattage.
- Concomitant use of the two-point suction cautery (Scarf/Greenberg) to clear blood and vapor, remove char and to coagulate larger blood vessels.
- Clackened and roughened retractor blades to present reflection of the beam and inadvertent damage to adjacent tissues.
- Extensive use of constantly moistened cottonoids to protect adjacent brain and to cover reflective retractor blades.

The CO_2 laser provides the surgeon with a delicacy of dissection, a major reduction in tissue manipulation, and an ability to resect distant lesions not afforded by any other surgical adjunct. The benefits, unfortunately, are not without limitations. Operating time is increased, surgery is tedious when dealing with large lesions and hemostasis is poor when large blood vessels are encountered.

4. THE FEASIBILITY AND ADVISABILITY OF RESECTING MIDLINE INTRA-AXIAL GLIOMAS IN CHILDREN

A. Leland Albright, M.D., Robert J. Sclabassi, M.D., Ph.D. (Pittsburgh, PA)

Children with midline intra-axial gliomas--in the optic chiasm, basal ganglia, midbrain, and brain stem--are often treated by radiotherapy without a tissue diagnosis. If tissue is obtained, it is usually obtained by a limited biopsy, whether open or stereotactic. Two recent technological advances--the CUSA and evoked potentials--have enabled neurosurgeons to remove difficult tumors with less morbidity than previously. During the past two years we have used the CUSA to extensively resect midline intra-axial gliomas in ten children. The feasibility results and the indications for such operations are the basis for this report.

Clinical information of the ten children is summarized in the table

Age, Yrs	Tumor Site	Diagnosis	% Resection	Evoked Potentials	Outcome
0.9	Chiasm	Astrocytoma	60	VEP	No deficit
5	Chiasm	Astrocytoma	90	VEP	No deficit
1.8	Thalamus	Astrocytoma	95	-	No deficit
8	Thalamus	Glioblastoma	50	BSEP	No deficit
11	Thalamus	Astrocytoma	75	-	Obtunded
16	Thalamus	Glioblastoma	75	-	Died
5	Midbrain	Astrocytoma	95	BSEP	III paresis
8	Pons	Glioblastoma	75	BSEP	VII paresis
9	Pons	Glioblastoma	90	BSEP	No deficit
13	Pons	Astrocytoma	80	BSEP	No deficit

The extent of tumor resection was documented by CT scans which will be shown. No patient had a serious postoperative neurologic deficit if evoked potentials were monitored intra-operatively and were unchanged by the resection. These cases demonstrate that extensive resections can be done on enhancing midline intra-axial tumors with mass effect, with serious, but probably acceptable, morbidity if evoked potentials are monitored.

The advisability of such operations certainly cannot be determined from ten patients. The strongest evidence supporting such resections comes from a recent Children's Cancer Study Group study of children with astrocytomas of grades II, III, IV (CCSG-943), which concluded that the extent of resection is significantly associated with the length of survivial: children with total resection survive longer than those with partial resections, and those with partial resections survive longer than those who are only biopsied.

5. BRAIN STEM TUMORS: A REAPPRAISAL

Fred Epstein, M.D., Edward Larry McCleary, M.D. (New York, N.Y.)

We present our preliminary experience utilizing a new approach to children with brain stem tumors. This was motivated by the fact that the dismal outlook for these children has remained unchanged for 50 years. Because the therapeutic options available for children with brain stem and intramedullary tumors were similar, we felt justified in expanding our operative experience with spinal cord tumors to include brain stem neoplasms. We have utilized technological advances including the operating microscope, cavitron, ultrasonic aspirator, laser, and brain state analyzer to make this feasible.

Prior to this we had operated on approximately 100 children with benign spinal cord astrocytomas, many of whom have returned to normal, active lifestyles. Only long term follow-up will allow us to document prolonged efficacy.

It has been our experience that operation changed neither the biology nor the clinical course of malignant brain stem tumors.

We observed that 80% of the tumors at or above the pontine level were malignant and roughly this same percentage in the lower brain stem were benign. In the latter group we included tumors that fell into Hoffman's category of exophytic fourth ventricular tumors arising most likely from the subependymal zone without diffuse brain stem infiltration. The pontomedullary tumors we encountered were frequently calcified and of low grade. Tumors at the cervicomedullary junction appeared transitional in nature, having characteristics of many of the rostral cervical astrocytomas by being grade I-II and having rostral cysts.

We are currently advocating operative resection for localized lower brain stem neoplasms. Utilizing this approach we have noted frequently quite satisfying reversal of long tract and certain crainial nerve findings. We have also experienced amelioration of several chronic pain problems and improvement in nausea, vertigo and diplopia.

Statistics in approximately 25 patients will be reviewed and results discussed.

6. THE MICROCYSTIC MEDULLARY JUVENILE ASTROCYTOMA: A PATHOLOGIC STUDY

Stuart G. Goodman, M.D., Marion L. Walker, M.D., J. J. Townsend, M.D., Bruce B. Storrs, M.D., W. M. Gooch, M.D. (Salt Lake City, UT)

Several series now report improved resectability and survival in children with medullary brainstem tumors as opposed to a more severe outcome in children with pontine tumors. Brainstem tumors with more benign morphological characteristics and behavior have also been reported.

Microscope slide preparations of nine operative specimens, from a recent series of children with brainstem tumors, were examined by two university affiliated pathologists, without knowledge of anatomic location or patient outcome. The tumors were described morphologically in addition to their pathological grading. Four of the nine cases were medullary tumors. Three of these four cases, all coming from the posterior medulla, just inferior to the obex, were described as microcystic juvenile astrocytomas with an appearance similar to the cerebellar astrocytoma of childhood. The other child, with documented Von Recklinghausen's neurofibromatosis, and also an acoustic schwannoma, had a glial tumor with characteristics of both an astrocytoma and an ependymoma. Four of the five cases of pontine tumors had a hypercellular appearance with pleomorphism. One child demonstrated a hypocellular appearance with radiation changes.

Three of the four children with medullary tumors are alive today, 12-40 months postoperatively. The other succombed to a viral pneumonia seven months post-operatively. Four of the five children with pontine tumors died within seven months of their surgery. The one child with radiation changes remains alive now six months postoperatively. All of the children have had radiation therapy. This study supports the findings of other suggesting a better outcome in children with medullary tumors. These tumors appear more amenable to surgical resection. The entity of a microcystic juvenile astrocytoma of the medulla is suggested. A videotape demonstrating the surgical resection of a medullary tumor is presented.

7. A REVIEW OF 26 CHILDREN TREATED FOR MEDULLOBLASTOMA

Mark S. O'Brien,M.D., Daniel L. Barrow,M.D., Yoshio Takei,M.D. (Atlanta, Ga.)

The authors present a series of 26 children with medulloblastoma treated by the senior author and review the results of treatment. All children with hydrocephalus underwent ventricular shunting prior to definitive therapy. The latter consisted of either biopsy, subtotal or gross total removal of the neoplasm, and post-operative radiation therapy. In 14 patients a regimen of chemotherapy was also utilized for treatment, in 6 as part of the initial therapy and in 8 as treatment of tumor recurrence and/or metastases.

Nine of the 26 patients had a recurrence of the tumor, 9 developed metastatic disease and 5 had both tumor recurrence and metastases, leaving 12 free of either of these complications.

At the present time 13 of the patients are living and 13 are deceased. Survival of the living patients ranges from 5 months to 13 years 3 months with an average of 5.38 years. In the deceased patients survival ranged from 2 weeks to 6½ years with an average of 2 years.

Discussion will include the clinical presentation, ventricular shunting, and the results of treatment. A biopsy or subtotal removal of the tumor followed by craniospinal radiation appears to be as beneficial as gross total removal. We postulate that individual differences in tumor biology and host response are more important factors influencing outcome than the aggressiveness of surgical removal or use of chemotherapy. A correlation of the histopathology and patient survival will be analyzed.

8. MANAGEMENT OF SUBEPENDYMAL GIANT CELL ASTROCYTOMA

Robert A. Sanford, M.D. (Jackson, Ms.)

Seven cases of subependymal giant cell astrocytomas in patients with tuberous sclerosis have been managed at the University of Mississippi Children's Hospital since the advent of CAT scanning.

These tumors are characteristically located at the foramen of Monroe, producing obstructive hydrocephalus as they progressively enlarge. Four of the children required bilateral CSF diversionary procedures to treat their hydrocephalus. Three children underwent surgical removal of a subependymal giant cell astrocytoma to control the increased intracranial pressure produced by the tumor mass.

This progressive growth of the subependymal giant cell tumor is contrasted with the multiple subependymal nodules of the ventricular wall which calcify but do not enlarge. The charactertistic location of the tumor at the head of the caudate nucleus and progression differentiates the astrocytoma from the more benign tubers.

Our seven cases demonstrated a deliberate rate of growth. Progression of seizures, mental retardation, systemic involvement and neurologic impairment is unpredictable in the child with tuberous sclerosis. The predictably slow rate of tumor growth allows the surgeon to delay surgery until absolutely necessary.

The presentation outlines our surgical approach and compares the results with the 26 cases reported in the English literature. [1]

1. Nagib, Mahmoud G., Haines, Stephen J., Erickson, Donald L., Mastri, Angeline R., Tuberous Sclerosis: A review for the neurosurgeon. Neurosurgery, 14:93-98.

9. GLIOMAS ASSOCIATED WITH RADIOTHERAPY: REVIEW OF FOUR CASES

Thomas S. Berger, M.D., Boleslaw H. Liwnicz, M.D., Ph.D. (Cincinnati, Oh.)

Irradiation associated intracranial tumors are uncommon. Among these tumors, sarcomas are most often encountered and, until 1983, only 18 cases of gliomas were reported. During the past seven years, we have encountered five cases, of which four were irradiated in childhood. All of our pediatric cases were males, and there was a 5 to 25 year latency period between radiotherapy and the diagnosis of glioma. CT scan findings were all abnormal with a differential diagnosis of tumor, radionecrosis, or abscess. The diagnosis was based on tissue biopsy. They were all high grade astrocytomas, which is unusual for this age group. The cases are summarized in the table:

First Tumor		Radiation Dose (rads)	Second Tumor	
Diagnosis	Age		Diagnosis	Latency
Retinoblastoma	2 wks	5500	Glioblastoma	13 yrs
Ependymoblastoma	2 yrs	3500	Glioblastoma	13 yrs
Burkitt's lymphoma	5 yrs	1800	Anaplastic astrocytoma	5 yrs
Craniopharyngioma	12 yrs	5900	Glioblastoma	25 yrs

10. PRELIMINARY EXPERIENCE WITH 125-IODINE BRACHYTHERAPY IN CHILDREN

Michael S. B. Edwards, M.D., Philip H. Gutin, M.D., Steven Leibel, M.D., Sharon Lamb, R.N. (San Francisco, Ca.)

The initial treatment of malignant supratentorial tumors in children is usually surgical resection followed by external beam radiation therapy (RT). Doses of more than 4000 R improve time to tumor progression and survival in children with supratentorial tumors, and use of chemotherapy adjuvant to RT significantly extends median time to tumor progression and survival for supratentorial malignant astrocytomas in children. However, RT remains the most effective treatment available.

A clear-cut dose relationship has been found for adult patients with malignant gliomas treated with RT. This therapeutic approach is limited because escalation of the dose of radiation delivered by external beam teletherapy to levels that control or even possibly cure local disease produces severe normal brain toxicity. Interstitial implantation of radiactive sources (brachytherapy) can be used to increase the radiation dose delivered to the tumor, which increases the therapeutic ratio, but also to obviate the deleterious effects of whole brain RT.

We have treated twelve children/adolescents with 125-Iodine brachytherapy. Eight patients were treated at the time of tumor recurrence and four were treated as a 'boost' adjuvant to external beam RT. Ten patients harbored supratentorial malignant tumors and two had posterior fossa tumors. High activity 125-I seeds were placed sterotactically in nine patients and low activity 125-I seeds were placed in three patients under direct vision at the time of tumor resection. Radiation doses ranged from 3120 to 7600 R for high activity seed and 10,000 to 15,000 R for low activity seeds.

Eight patients are fully active without evidence of tumor progression or recurrence for up to 3 years post-implantation. Two patients have died of tumor recurrence (medulloblastoma; choroid plexus carcinoma) and two are too recent to evaluate. Five patients have required reoperation for focal radiation necrosis and all are stable and doing very well.

A number of problems must be fully solved before brachytherapy can be used to its fullest advantage. The first, accurate dosimetry is being solved by a computer adapted program that is mated to the CT scanner. Second, techniques to evaluate tumor progression and response need to be better clarified. Possibly magnetic resonance imaging and/or spectroscopy will be of assistance in making these determinations. The appropriate interstitial dose that controls tumor growth must be determined, and whether this dose can be used more properly as a single treament or as a boost to conventional external beam irridation must be studied. The use of brachytherapy in combination with chemotherapy or localized hyperthermia, and the regions of brain that can be safety implanted must still be determined. Based on our experience in this small series and a much larger series in adults, we feel that brachytherapy, used either alone or in combination with other treatment modalities (chemotherapy, radiosensitization, hyperthermia) will become the treatment of choice for selected brain tumors in children and adolescents.

11. DUMB-BELL NEUROBLASTOMA IN CHILDREN: REVIEW AND TREATMENT

Edward J. Kosnik,M.D., Martin P. Sayers,M.D. (Columbus, OH.)

Neuroblastoma and ganglio-neuroblastoma are among the more common malignant solid tumors seen in childhood. A variant of this disease process involves not only the retroperitoneal or mediastinal space but, also, by extension, the spinal canal.

In 1974 we reviewed our experience with 19 patients with this dumb-bell neuroblastoma. We have seen an additional 15 patients with this disease process over the past 10 years.

We plan to review the entire series of patients with particular reference to management, not only directed specifically at eradication of tumor tissue, but also orthopaedic deformity seen very commonly in these children.

12. PITUITARY HYPERPLASIA DUE TO PRIMARY HYPOTHYROIDISM

Leslie N. Sutton, M.D., Thomas Moshang, M.D., Larissa Bilaniuk, M.D., Lewis Samuel, M.D., (Philadelphia, Pa.)

Primary hypothyroidism can result in reactive enlargement of the pituitary gland, which on CT scan is indistinguishable from primary pituitary lesions. Two teenage girls and 1 adult are presented in whom enhancing masses in the pituitary region were found on CT scan, and in whom surgery was averted by endocrinologic testing and follow-up CT scans after the administration of thyroid replacement. In retrospect, the CT findings are characteristic showing a brilliantly enhancing cone-shaped intra and suprasellar mass on the coronal view. The importance of the pre-operative endocrinologic evaluation in these patients is stressed.

13. MORPHOLOGICAL STUDY OF FRONTONASAL ENCEPHALOCELES USING 3-DIMENSIONAL RECONSTRUCTION OF COMPUTED TOMOGRAPHY

David C. Hemmy, M.D., David J. David,F.R.A.C.S., Donald A. Simpson,F.R.A.C.S. (Milwaukee, Wi.)

12 children ranging from 1.5 to 14.2 years of age were included in this study. 10 patients were Malay and 2 were Asiatic Indians. Only cases with true frontonasal encephaloceles were included. Cases with craniofacial clefts and resultant brain herniation were excluded. One half of the patients had neurosurgery in early life in Malaysia. Procedures performed included simple transsection of the neck of the encephalocele and repair of the dura in 4 cases and shunting for hydrocephalus in 2. The radiologic investigation provided the most pertinent diagnostic information particularly through the use of 3-dimensional reconstruction of computed tomography.[1] CT scanning revealed normal cerebral anatomy in 8 cases (with the exception of the encephalocele) and hydrocephalus in 3 cases. 3-dimensional reconstruction revealed that, in all cases, the cribriform plate was significantly depressed with the exit of the encephalocele through an enlarged and often asymmetric foramen cecum. The bony deformities of the facial region fitted into the 3 categories described by Suwanwela.[2] In the nasofrontal types the skull defect was noted to be at the root of the nose displacing the nasal bones inferiorly. In the nasoethmoidal types the nasal bones remained contiguous with the frontal bone and the encephalocele exited between the normal nasal bones and the nasal cartilage and septum. Lastly, in the nasoorbital type, the frontal and nasal bones were contiguous with some depression of the nasal cartilage noted. The encephalocele bifurcated posterior to the nasal bones to enter the orbit and erode the medial orbital walls. In summary, the use of 3-dimensional reconstruction of computed tomography has permitted precise classification of frontonasal encephaloceles preoperatively, facilitating surgical planning, the avoidance of surgical pitfalls,and the reduction of operative morbidity.

[1]Hemmy, D.C., D. J. David, G. T. Herman. 3-dimensional reconstruction of craniofacial deformity using computed tomography. Neurosurgery,13:534-541, 1983.

[2]Suwanwela, C., Suwanwela, N. A morphological classification of sinicipital encephalomeningocele. J. Neurosurg, 36:201-211, 1972

14. OBLITERATION OF BASAL CISTERNS ON CT, THE CARDINAL SIGN OF LIFE-THREATENING SHUNT FAILURE — also in in Post trauma

Dennis L. Johnson, M.D., Charles Fitz, M.D., David C. McCullough, M.D. (Washington, D.C.)

Although it is rare that a child dies from shunt malfunction, every experienced pediatric neurosurgeon is aware of patients who precipitously deteriorated on the ward while being observed for diagnostic confirmation or while awaiting scheduled surgery. We are presenting 8 cases of shunt malfunction who were arousable on admission and scheduled for shunt revision the following day. A precipitous decline in neurologic status prompted unscheduled, emergency shunt revision in each case. In fact 6 of the 8 patients demonstrated decerebrate posturing just prior to surgery; 3 patients had a cardiopulmonary arrest; and 2 of the patients died. Clinical history did not suggest extreme shunt dependence. Most children had been revised several times before but never under urgent circumstances. Most of the older children had headaches for 5 days or more, and a majority had been vomiting for more than 24 hours. Furthermore, there were no physical signs which suggested marginal compliance. No patient had papilledema and only 2 children had new eye signs; no pupillary changes were manifest. Review of the CT scans revealed ventriculomegaly with previously documented smaller ventricles. More importantly, the basal cisterns were not seen on any preoperative scan but were visible postoperatively in the cases which survived. To weigh the importance of this observation we reviewed the histories and CT scans of 50 cases of hydrocephalus randomly taken from the neuroradiology files. Obliteration of the basal cisterns proved to be a reliable radiographic sign of shunt malfunction which required emergent attention.

15. ULTRASONOGRAPHIC ANATOMY OF THE POSTERIOR FOSSA ANATOMIC-SONOGRAPHIC CORRELATIONS + thru squamous bone to age 19 m

Thomas P. Naidich, M.D., David Yousefzodeh, M.D. (Chicago, Il.)

Correlation of advanced, computer-assisted sonograms (Acuson^R) with fresh and myelin-stained brain sections documents that sonography routinely displays the cerebral peduncles, substantia nigra, aqueduct and colliculi of midbrain in axial section, and may occasionally image the red nuclei. Saggittal sonograms routinely display the bony and dural walls of posterior fossa, the echogenic pars basilaris of pons, the distinctly different modestly echoic tegmentum of pons, the pontomedullary sulcus, the medulla including olive, the individual lobules of vermis, the central white lamellae, and all the basal cisterns. The authors illustrate the sonograms, correlate them with brain sections, and demonstrate their use for the diagnosis of neuropathology.

16. IMAGING THE CRANIOVERTEBRAL AND CERVICO-MEDULLARY JUNCTION IN CHILDREN: A PROSPECTIVE EVALUATION OF MULTIMODAL INVESTIGATIVE TECHNIQUES.

Arnold H. Menezes, M.D., Wendy R. Smoker, M.D., Lindell Gentry, M.D. (Iowa City, Ia.) MRI sometimes failed to demonstrate cavity Syrinx

The craniovertebral junction (CVJ) with the encompassed neurovascular structures of the cervico-medullary junction (CMJ) make this the most complex region of the axial skeleton. A surgical physiological approach to pathology at the CVJ was described by us in 1980 based on imaging by dynamic pleuridirectional myelotomography, computed tomography and CT metrizamide myelography. Magnetic Resonance Imaging (MRI) has demonstrated excellent neuroanatomic morphology of the posterior fossa due to lack of image degradation by bone and superior tissue discrimination. A prospective study is undertaken to investigate the role which MRI can play here.

Twenty children with symptoms referable to the CVJ-CMJ were studied by current investigational methods and MRI over a four month period. MRI images were obtained on Picker 0.5 Telsa super conductive unit using sagittal, coronal and axial planes. Saturation, recovery and spin echo techniques were utilized. Lesions included basilar invagination, abnormal clivus-odontoid articulations, Chiari malformations (ACM), hydromyelia, brain stem and cervical cord tumors, demyelination and arachnoidal bands.

MRI provided superior visualization of brain stem, cerebellum and cervical cord lathology, and altered treatment course in 5 children. It failed to demonstrate hydromyelia in two ACM (lumbar puncture done previous day); in one patient with high cervical cord tumor misdiagnosis of hydromyelia with ACM was made. Osseous relationships were best demonstrated on pleuridirectional tomography and CT.

Proton Muclear MRI is non-invasive, versatile and particularly suited to study the posterior fossa. It's short comings must be appreciated as it rightfully becomes the primary procedure of choice in accessing lesions of CVJ and CMJ.

17. MAGNETIC RESONANCE IMAGING IN SPINAL DYSRAPHISM

Joan L. Venes, M.D., Alex M. Aisen, M.D., Michael A. DiPietro, M.D., James Knake, M.D. (Ann Arbor, Mi.)

The advantage of a non-invasive study which does not require exposure of the infant to radiation energies hardly needs comment. However, in addition to these obvious advantages, Magnetic Resonance Imaging (MRI) appears to be uniquely suited to the resolution of two of the major problem areas confronting the neurosurgeon in evaluation of the child with spinal dysraphism:

1) The presence of multiple midline anomolies in the child with repaired meningocele and symptoms of late onset cord ischemia.
2) The exact relationship of the cord to the intraspinal lipoma in cases of lipomyelomeningocele.

We would like to briefly present five representative cases in which the remarkable clarity of detail of the MRI contributed to the diagnostic evaluation. Four/five are children and one is an adult with repaired meningomyelocele. The cases include examples of a dermal sinus with an intraspinal lipoma, a lipomyelomeningocele, hydromyelia, Arnold-Chiari malformation and postoperative syrinx formation. Comparison with High Resolution Computed Tomography (HRCT) demonstrates the advantages of MRI in patients with these disorders. Increasing familiarity with this modality and the use of presently available modifications, e.g., the spine coil, will likely result in MRI becoming the diagnostic procedure of choice in those areas of the world in which it is available.

18. SURGICAL CORRELATES OF NUCLEAR MAGNETIC RESONANCE (NMR)

Nazih Moufarrij, M.D., Joseph F. Hahn, M.D., Michael T. Modic, M.D., Meredith Weinstein, M.D. (Cleveland, Oh.)

The purpose of this paper is to analyze our experience with NMR imaging in children and young adults (≤ 33 years) with special emphasis on the NMR of the spine in congenital anomalies and extramedullary tumors. Of note is that our head NMR data in tumors of childhood are similar to what was reported last year at this meeting by Luerssen et al (Paper 17, 12th Annual Meeting of the Pediatric Section of the AANS). We should, however, add that NMR is extremely helpful not only in tumors of the posterior fossa and cervicomedullary junction but also in demonstrating small, less common lesions causing hydrocephalus (colloid cysts, subependymonas) and in isodense subdural hematomas. Other imaging modalities had failed to reveal these lesions.

A variety of techniques were used to obtain the NMR images. The so called T_2 weighed images are especially helpful in spine NMR because the CSF appears white hence acting like a contrast medium. A total of 24 cases are presented. These are subdivided for convenience into 5 groups:

1. The Chiari malformation-syrinx group, 15 cases, the anomalies were clearly depicted by NMR, and this is in agreement with the experience of others.
2. The neurofibroma-hemangioblastoma group, 4 cases, three patients had more than one lesion and the sector CT with metrizamide showed the small lesions to a better advantage.
3. The lipoma-dermoid group, 3 cases, well demonstrated by NMR.
4. One case of C_1-C_2 sublaxation in a child with achondroplasia. The NMR flexion extension views were particularly helpful.
5. One case of cord AVM, poor visualization by NMR.

The NMR appears to be a safe, radiation-free and noninvasive imaging technique of the spine. Metrizamide sector CT and aniography are still superior in certain instances.

19. THE TYPE I CHIARI MALFORMATION IN CHILDHOOD AND ADOLESCENCE

W. Jerry Oakes, M.D. (Durham, N.C.)

Twelve patients aged six to sixteen years of age with radiographically proven Type I Chiari malformations have been evaluated at Duke University Medical Center between April 1, 1979 and June 30, 1984. Eleven of the twelve have undergone suboccipital and high cervical exploration for progressive symptomatology. The symptoms have ranged from lower cranial nerve disturbances to classical syrinx symptoms. Isolated cerebellar disturbances were not appreciated. The presentation of many of the patients appeared unique and not stereotyped. Following operation 4/11 patients developed protracted vomiting which resolved within four weeks. No other major postoperative complications were noted. Objective evidence of neurological improvement was noted in 7/11 patients and 4/11 have remained unchanged in the follow up period.

The changing attitude of the goal of surgery will be discussed in light of Williams' theory of altered equilibration of CSF pressure across the foramen magnum.

20. REAL TIME ULTRASOUND EVALUATION OF CHIARI II MALFORMATION IN INFANTS

Bruce B. Storrs, M.D., Marion L. Walker, M.D. (Salt Lake City, UT)

Chiari malformation occurs in a high percentage of patients with myelomeningocele and presents as a clinical problem in a few percent. We were interested in evaluating non-invasive radiologic techniques for evaluating the Chiari malformation in newborns with myelomeningocele in an attempt to predict clinical progression of symptoms.

Twenty patients had their cervico-medullary junctions evaluated using real time ultrasound. This non-invasive and cost effective method proved helpful in a high percentage of patients allowing us to diagnose medullary kinking and fourth ventricular herniation. These changes were observed to respond to shunting of those patients without symptomatic hydrocephalus, and to cervical decompression for those patients without symptomatic hydrocephalus. A videotape demonstrating the technicque and the real time images will be presented.

21. DERMOID TUMORS AT THE SITE OF PREVIOUS MYELOMENINGOCELE REPAIR

R. Michael Scott, M.D., Louis Bartoshesky, M.D., Samuel M. Wolpert, M.D., Seymour Zimbler, M.D., George Klauber, M.D. (Boston, MA)

The author has recently treated three children with dermoid tumors occurring at the site of myelomeningocele repair. Two ambulatory children presented at ages 9 and 13 with progressive back and leg pain, increased pain and sensitivity in the area of the myelomeningocele repair, progressive leg weakness and loss of continence while on intermittent catheterization regimens. One paraplegic child with a complicated neurologic history presented at age eight years with progressing scoliosis. Myelography in two patients demonstrated irregular lobulated filling defects in the area of the initial surgery. Metrizamide CT scanning demonstrated hypodense irregular masses attached to the cord or filling in the thecal sac. In one patient an unenhanced CT scan suggested either dermoid or hydromyelia. The same surgeon operated on two of these children at birth, but review of his operative notes disclosed nothing unusual regarding initial findings or technique of repair. The tumor in two patients proved impossible to remove because of tight adherence to functioning nerve roots, and only partial removal of the capsule and gutting of tumor contents could be achieved. In the paraplegic patient, the lesion along with neural tissue was excised. All three patients improved considerably postoperatively, with disappearance of pain and improved leg strength and sensation, but urinary disfunction remained unchanged at six month follow-up in the nine year old youngster. Operative findings suggest that these lesions relate both to congenital dermal inclusions in the area of the placode and to incomplete excision of dermal elements at the time of myelomeningocele repair.

22. PROGRESSIVE CAUDA EQUINA SYNDROME IN CHILDREN WITH THE CAUDAL ENDODERMAL SYNDROME

Arden F. Reynolds, M.D., Michael Pollay, M.D., Patrick Barnes, M.D., A. Sullivan, M.D., E. I. Smith, M.D., W. Tunnell, M.D., J. Bodensteiner, M.D., W. Barnes, M.D., P. Lester, M.D. (Oklahoma City, OK)

It has been known for many years that children with bony sacral anomalies have neurological dysfunction. It has also been known for many years that bony sacral anomalies are associated with the caudal endodermal syndrome which is manifested by imperforate anus, anorectal atresia or stenosis, ileoatresia, or choanal extrophy. This syndrome has been considered static, and these children have been cared for in myelomeningocele clinics. Recently we have had three children and one adult develop delayed neurological progression after repair of imperforate anus in three and ileoatresia in the other. All four of these patients had sacral anomalies on plain films and myelography revealed surgically amenable dysraphic myelodysplasia in three and a dural sac stenosis.

As a result of these patients and because of a few case reports from other institutions we have begun systematic reevaluation of all children with caudal gastrointestinal and genitourinary syndromes. Imaging techniques have included plain films in 96, spinal ultrasound in 4, magnetic resonance imaging in 18, metrizamide myelotomography in 11, digital subtraction myelography in 6, and computerized tomography in 9. Of the 96 patients with caudal endodermal syndromes between July, 1976 and April, 1984, 36 had findings of sacral and/or other spinal anomalies including agenesis, hypoplasia, fusion-segmentation anomalies and dysraphic defects. As of August, 1984, 24 children have undergone spinal cord imaging with confirmation of dysraphic myelodysplasias in 14. The findings in these children have included lipomeningocele in 3, lipoma plus thick filum in 2, thick filum in 2, dural sac stenosis in 2, dysplastic conus in 1, lipomyelocystocele in 1, low conus with dural ectasia in 1, and presacral meningocele in 1. To date surgery has been accomplished in 13 of the 14 children with our standard technique of microneurosurgical approach and intraoperative stimulation of nerve roots while recording anal EMG. None of the children have been made worse by surgery, 5 of the children have had improved leg function and 2 have had improved bladder function. Progression has been halted in the remainder.

As with lipomyelomeningoceles prevention of neurological deterioration is of paramount importance because once deterioration occurs it is infrequent that recovery is possible after surgical release. Our adult patient deserves special mention because he had a stable neurological condition for 30 years and then had deterioration of his leg function with spasticity secondary to an intradural lipoma with tethered cord syndrome. Our recommendation is to image these children in early infancy and to repair them between 3 and 6 months of life to prevent delayed neurologic deterioration.

23. VEIN OF GALEN ANEURYSMS: COMBINED TREATMENT BY EMBOLIZATION AND SURGERY

Jeffrey H. Wisoff, M.D., Alejandro Berenstein, M.D., Fred J. Epstein, M.D. (New York, N.Y.)

Between June 1977 and July 1984 seventeen patients with vein of Galen aneurysms were seen at the New York University Medical Center. Five of the seventeen patients were neonates, eight were infants (2-12 mos), three were older children, and one was an adult. All of the neonates presented with intractible congestive heart failure. Neurological and developmental deterioration was the predominant presentation among the infants and older children. CT scans demonstrated progressive cerebral atrophy and dystrophic calcification consistent with a progressive ishemic encephalopathy.

The treatment strategy that has evolved is based on whether the patient presents with heart failure or ischemic deficits. In the neonate with intractible failure embolization of a significant portion of the fistula to allow resolution of the heart failure has been successful in 4/5 patients. In the older patient with ischemic encephalopathy, sequential staged embolizations are utilized to obliterate the majority of the malformation. Surgery is utilized to obliterate anterior cerebral feeders by direct clipping and intraoperative embolization and to control hydrocephalus.

The mortality in our series was 18% (3/17) including one neonate. The dramatic improvement in case mortality and morbidity using embolization technicques over conventional surgical approaches and our methods of treating these lesions will be discussed.

24. THE TRANSTORCULAR APPROACH TO VEIN OF GALEN ANEURYSMS - A PRELIMINARY REPORT

J. Parker Mickle, M.D. (Gainesville, Fl.) can be done in Neonate

The surgical treatment of Vein of Galen aneurysms continues to be difficult and at times controversial. Interventional efforts for these deep Galenic malformations range from direct surgical approach to feeding arteries to direct instillation of various thrombogenic agents into the malformations.

Two patients (three months and four years) with large Vein of Galen aneurysms were treated at the University of Florida Medical Center by the transtorcular deposition of occluding spring emboli (Gianturco springs). A total of 22 Gianturco coils were placed in these two patients in a total of four procedures. The surgical procedure consisted of localizing the torcula and straight sinus and Vein of Galen aneurysm with real time ultrasound after a small craniectomy over the torcula. Through a standard 8 cm. angiography catheter placed into the Vein of Galen, the Gianturco coils were deposited within the Vein of Galen aneurysm with a special plunger delivery set developed at the University of Florida. This system makes the delivery of these coils safe and specific. Pre and post embolization the patients have been studied with digital angiography and ultrasonic doppler. used by gen surg

Both patients have had dramatic changes in their pre-embolization seizure patterns. The flow changes induced by the coils were dramatic. The potential for continued thrombosis exists around these thrombogenic wires. The feasibility of this approach for Vein of Galen aneurysm will be discussed.

done on children — age 4 — survivors already seletty

25. CEREBROVASCULAR MALFORMATIONS IN CHILDREN: A RETROSPECTIVE ANALYSIS OF 28 CASES

Francisco A. Gutierrez, M.D., David G. McLone, M. D., Thomas P. Naidich,M.D. (Chicago, Il.)

Twenty-eight pediatric patients with documented arteriovenous malformations of the brain which were reviewed between December 1974 and December 1983. Patients older than 16 years, patients with arteriovenous malformations (AVM's) of the Vein of Galen or the spinal cord were excluded from this study.

The clinical signs and symptoms were analyzed at presentation and after "definitive therapy". The malformations were classified as wholly pial-parenchymal AVM's (23) and as dural AVM's (5). Spontaneous subarachnoid hemorrhage was the most frequent finding at presentation (18 patients, 64%). In six patients (21%), the initial hemorrhage was accompanied by acute seizures. Chronic seizures (epilepsy) were present in 5 patients (18%). An audible bruit was detected in 4 patients (14%). Recurrent episodes of headache were reported by the majority of patients. The angiographically occult vascular malformation was present in 4 patients (14.7%). Twenty-four of the 28 patients underwent surgical treatment for AVM; 18 patients had total excision of the AVM, 6 had partial excision of the AVM, and 4 patients were treated conservatively. Five patients required more than one surgical procedure to achieve partial or complete obliteration of the malformation. Analysis of outcome after treatment for the entire group of 28 patients, regardless of lesion location or type of treatment, indicates that 19 children (68%) were normal, 5 (17%) had mild neurologic deficit, 2 (7%) were disabled, and 2 (7%) died. The two deaths in our series were associated with partial resection of the AVM.

We consider seizures to be sufficient indication for "prophylactic" resection of the lesion, provided such resection is anatomically feasible.

26. A LABORATORY MODEL OF SHUNT DEPENDENT HYDROCEPHALUS

Arno Fried, M.D., Kenneth Shapiro, M.D. (Bronx, NY)

In order to understand the clinical phenomena characteristic of shunt dependent hydrocephalus, a laboratory model of the shunt dependent child was required. Once developed, the biomehcnical profile of the shunted model could be compared to a previously described group of shunt dependent children.

Kaolin hydrocephalus was induced in twelve cats using the craniectomy-durectomy model. After four weeks, CSF dynamics were studied using the Pressure Volume Index (PVI) technique of bolus manipulation of CSF to define the neural axis volume buffering capacity (PVI) and CSF outflow resistance (R_O) in these untreated hydrocephalic animals. A valveless ventriculo-pleural shunt was then inserted. After four weeks, shunts were occluded and CSF studies were repeated.

Steady state ICP in the untreated hydrocephalic cat was 11.8±2.2mmHg. After shunt occlusion, ICP was 14.0±0.5.1mmHg. PVI in the intact normal cat was 0.76±.04 (SEM)ml which increased 3.5±0.4 (SEM)ml. ($p < .001$), in the untreated hydrocephalic cat indicating enhanced volume buffering capacity. With reconstitution of the cortical mantle, PVI in the shunted animals approached normal at 1.1±.07ml. In addition, as ICP was raised by successive bolus injections or slow infusion, ICP became unstable in a range of 28-38mmHg which corresponded to the beginning of the steep portion of the P-V curve.

CSF outflow resistance (R_O) in the intact cat was 91.3±7.5 (SEM)mmHg/ml/min which decreased to 28.8±4.5 (SEM) in the untreated hydrocephalic cat, ($p < .001$). After shunting, R_O was increased and varied as a function of ICP, in contrast to the normal or hydrocephalic cats. As ICP was elevated, R_O increased linearly ($r = .87$, $p < .001$).

At sacrifice, the temporalis muscle was densely fibrotic and adherent to the brain. The stabilization of container properties of the brain by the muscle and scalp had a similar effect on volume buffering capacity as suture closure in the shunt dependent child. The ventricles were normal to slightly enlarged in all animals despite the occluded shunt.

The biomechanical profile, and ventricular response to shunt occlusion in this model resembles that previously described in twenty shunt dependent children. In both, shunting results in normalization of PVI which persists with shunt malfunction. Based on the changes in PVI and CSF dynamics with shunting, the sequence of events leading to shunt dependency are discussed as well as ongoing studies using this model.

Shunted - constant infusion fails to plateau

Shunt - unable to augment ICT ~~or~~ absorption

27. AN EXPERIMENTAL MODEL SHOWING WHY THE SUBTEMPORAL DECOMPRESSION SHOULD WORK IN THE SLIT VENTRICLE SYNDROME WITH SOME SUGGESTIONS AS TO WHY IT DOESN'T WORK OFTEN ENOUGH

Slit ventricles

Kenneth Shapiro, M.D., Futoshi Takei, M.D. (Bronx, NY)

subtle deteriorations - no plateau waves

A variety of techniques have been described for treating the shunt dependent child with symptomatic slit ventricles. "Subtemporal craniectomy" was originally proposed as a surgical adjunct which would result in dilatation of the ipsilateral ventricle and enhance shunt patency. Often the ventricle does not dilate after this procedure and over time, the subtemporal decompression does not confer immunity from the slit ventricle syndrome. While several investigators have shown that children with slit ventricles may have reduced neural axis compliance, our own studies indicate that neural axis compliance in these children is normal. We used a laboratory model to help understand the effects of craniectomy on neural axis compliance in the hope of extending these observations to the shunt dependent child.

(1) acute deterioration 2° to Press/Vol instability

In earlier laboratory investigations of feline hydrocephalus we found that opening the skull and dura acutely enhances volume buffering capacity or PVI. In chronic experiments performed on non-hydrocephalic animals this enhanced volume buffering capacity decreased which we explained by the formation of a pseudo-dura and scarring over the craniectomy site. In order to develop a model in which the enhanced volume buffering capacity persisted in the chronic state, we sutured silastic over the cut edges of the dura. One to two months later enhanced volume ~~buffering capacity was preserved~~ and was three times that of normal ~~baseline controls and similar to that found in the acute phase immediately after altering the~~ container of the brain. Despite this enhancement of volume ~~buffering capacity~~, ventricular size remained normal. Interestingly, the absorption of CSF was enhanced as found in the acute stage.

(2) Improve c ↑ compliance

From the present studies we infer that by preventing dural adhesions from occurring after cranial decompression, the enhanced volume buffering capacity conferred by this maneuver will remain and may be a useful adjunct to dealing with the shunt dependent child. By implanting a dural substitute, the immediate effect of subtemporal craniectomy and dural opening will be preserved over time and may be a useful way of preventing the acute deterioration of these children.

(3) craniectomy → bony regrowth, (4) silastic duroplasty ↑ duration

28. MANAGEMENT OF EXPANDING MULTIPLE LOCULATED CSF FILLED CAVITIES FOLLOWING ENTERIC ORGANISM VENTRICULITIS.

Robert Breeze, M.D., J. Gordon McComb, M.D. (Los Angeles, Ca.)

Severe ventriculitis in an infant or child can cause extensive and widespread ependymal and parenchymal damage. The subsequent glial and connective tissue reaction sometimes produces multiple isolated expanding CSF filled cavities within or adjacent to the ventricular system. Four patients, with ages ranging from two months to seven years, sustained severe ventriculitis caused by one or more gram negative enteric organisms. In two patients the ventriculitis followed bowel perforation by a coiled spring type of peritoneal catheter, in a third the ventriculitis developed before an open thoracolumbar myelomeningocele was closed, and in the fourth no known predisposing factor was present. After the ventriculitis was successfully treated each patient developed multiple isolated expanding CSF filled cysts as documented by metrizamide ventriculography.

If the patient is to be treated, the alternatives are multiple shunts or fenestration of the cavities into one another, ideally requiring only one shunt to control the hydrocephalus. Previous difficulty with the placement and maintaining function of multiple shunts suggested that a direct surgical approach might be advantageous. A unilateral posterior parietal craniotomy with a 2-3 cm cortical incision allowed for adequate visualization and fenestration of the various loculated cavities with use of the operating microscope. Multiple procedures were necessary in some cases. Repeated CT scanning, with metrizamide ventriculography as necessary, was used to document the success of the fenestrations. In all four patients the hydrocephalus has been controlled with a single shunt without increase in neurologic deficit or associated surgical complication.

under microscope
suction/cautery, 2-3 cm fenestration
ultrasound not used - but could be useful

29. LOCULATED VENTRICLES - AREN'T. glial lined cavities

David G. McLone, M.D.,Ph.D., Thomas P. Naidich, M.D. (Chicago, Il.)

In addition to the developmental deficits inflicted on children by ventriculitis, loculated noncommunicating cerebrospinal fluid chambers make the management of the child's hydrocephalus nearly impossible. Multiple shunts, frequent revisions, recurrent infections, and, often, craniotomy with laser fenestration are needed to control the intracranial dynamics.

In an attempt to illustrate the mechnisms involved, normal and hydrocephalic mice had silastic catheters deliberately contaminated with gram positive and gram negative bacteria placed within their lateral ventricles. Both types of organisms produced abscesses at the catheter tip. The infection spread preferentially along the extracellular space of the white matter tracts deep to the ependyma and subependymal glial layers. A sequence of edema, small cysts, and coalescence of cysts to form large periventricular cavities invariably occurred. Immaturity and hydrocephalus facilitated this process.

Lazar fenestration

Review of CT scans of children progressing to compartmentalization suggest a similar if not identical process in humans. Periventricular lucencies progressed to cysts. The ventricular catheter was often displaced toward the midline as the multiple cysts coalesced and compressed the ventricular system. Electronmicroscopic examination of the septa between cavities revealed myelinated axons, indicating that the septa are indeed brain and not glial scars or ependymal adhesions. Just as in the animal model, the cysts often assumed a size equal to or greater than the shunted ventricular system. Preliminary evidence would indicate that aggressive treatment of ventriculitis, including ventricular irrigation, might abort this process.

30. VENTRICULOPERITONEAL SHUNTS IN HIGH-RISK NEWBORN UNDER 2,000 GRAMS OF WEIGHT - CLINICAL REPORT

Hector E. James, M.D., R. Bejar, M.D., L. Gluck, M.D., R. Coen, M.D., A. Merritt, M.D., F. Mannino, M.D., P. Bromberger, M.D., B. Saunders, M.D., H. Schneider, M.D. (San Diego, Ca.)

700-2000 failed on LP

53 low birth weight high risk newborns who developed progressive hydrocephalus despite a trial period of intermittent lumbar punctures, underwent cribside ventriculoperitoneal shunt replacement. They were all under 2,000 g. at the time of shunt (mean: ~~1,308.6g ± 398.2 S.D.~~). The operative procedures were performed at a mean age of 31.5 days ± 16.1 (S.D.). There were no deaths in this series. During the nursery stay, 14 patients required operative revisions for obstruction. The most common problems were infection which occurred in 13 (24.5%) after the primary intervention, and another 5 of the 14 (35.7%) of the patients that required revision. The overall infection rate per patient was 33.9%. Shunt removal and intensive antibiotic therapy cured the infection in all but one patient. Premature, low birth rate, high-risk newborns may undergo ventriculoperitoneal shunts understanding that certain requirements need to be met: a stable systemic course, close follow-up of complications such as obstruction and infection, and if these are met the procedures can be done with a low incidence of morbidity and mortality.

CSF comes as a one way valve
Mechanism - not understood

30 wound dehis - 1 pt tenuous skin
frequent obstruction
best mortality 5% → 30%
morbid 10 → 30%

31. OUTCOME IN CHILDREN WITH PRENATALLY DIAGNOSED CONGENITAL HYDROCEPHALUS

Charles C. Duncan, M.D., Laura R. Ment, M.D., David T. Scott, Ph.D., Richard A. Ehrenkranz, M.D. (New Haven, Ct.)

Maternal realtime ultrasound carried out on a widespread basis has permitted the prenatal diagnosis of congenital hydrocephalus with an increasing frequency. At our institution in a 5 year interval between 1978 and 1983, all 14 children with prenatally diagnosed congenital hydrocephalus, defined as excessively increasing ventricular size and biparietal diameter on at least two sequential examinations, were available for outcome evaluation. Others in whom this diagnosis was made either did not survive gestation or the initial neonatal period due to interruption of pregnancy or intrapartum demise. This report details the diagnoses, treatment, complications and follow up neurologic and developmental evaluation in this surviving group.

Gestation age at diagnosis varied from 26 to 38 weeks with a mean of 31 weeks. Age at delivery varied from 32 to 40 weeks with a mean of 36 weeks. Thirteen were delivered by Cesarean section. Four of these children had myelomeningocoele, 6 had hydrocephalus without other anomalies, three had intracranial cysts, and one had an encephalocoele. All of the children underwent ventriculo-peritoneal shunt placement. Five had shunt related infections. Six had Bayley mental or Stanford-Binet scores > 80 (43%), 2 had scores between 65 and 80 (14%), and 6 had scores <65 (43%). The poorer outcome of this group in comparison to other series of congenital hydrocephalus may relate both to the early stage at which the diagnosis was made such that children with more severe conditions have survived and the high rate of infections.

32. SER AND BAER IN EXPERIMENTAL FELINE HYDROCEPHALUS. EFFECTS OF VENTRICULAR DILATATION AND PRESSURE WAVES.

Leslie N. Sutton, M.D., Byung-Ku Cho, M.D., Gretchen Avery, Jurg Jaggi, M.S., (Philadelphia, PA)

Recent studies in human and animal subjects have suggested a relationship between ICP and ventricular dilatation and multimodality evoked responses which, if substantiated, would be of value in clinical practice as a non-invasive way of assessing the need for shunting in selected patients in whom the CT scan is not definitive.

In an attempt to better define these changes, BAER and SER were performed on 30 cats as a baseline following which they were made hydrocephalic by the cisternal injection of Kaolin. CT scans were performed 2-6 weeks later, and in those animals in whom ventricular dilatation was noted, repeat evoked responses were recorded. In an additional group of 5 hydrocephalic cats, the ventricle was punctured to measure ICP which in all cases was less than 5 mmHg. The lumbar spinal dural sac was then ligated, which resulted in periodic plateau waves up to 75-100 mmHg after 4-6 hours, lasting up to 1/2 hour.

In neither group of cats was any change in either BAER or SER observed until preterminally when ICP was in the 75-100 range and cerebral perfusion pressure was compromised. This suggests that the BAER and SER are not sensitive to either ventricular dilatation or intracranial hypertension and that prior studies using infusion techniques to induce intracranial hypertension perhaps were measuring a temperature-induced artifact.

Latency?

33. THE USE OF THE BRAINSTEM AUDITORY EVOKED RESPONSE IN THE MANAGEMENT OF NEONATAL HYDROCEPHALUS

Meredith Olds, M.D., Bruce B. Storrs, M.D., Marion L. Walker, M.D., Michael J. Cevette, M.S. (Toronto, ON)

Brainstem auditory evoked responses were measured in thirty patients before and after cerebrospinal fluid shunting procedures for hydrocephalus. The results show that an increasing interpeak latency is a sensitive indicator of clinically significant hydrocephalus in neonates. The test has been helpful in determining which patients with ventricular dilatation require shunting procedures, and useful in monitoring shunt function longitudinally.

true hydro 10-30%
Ventriculomeg 50-80%

Latency ↑↓ ō age
to monitor shunt function
recent baseline studies are needed

34. ONE YEAR FOLLOW-UP OF THE PROSPECTIVE RANDOMIZED DOUBLE BLIND TRIAL OF ANTIBIOTIC PROPHYLAXIS IN CEREBROSPINAL SHUNT INFECTIONS

M.H. Epstein, M.D., K. Kumar, M.D., P. Lietman, M.D., W. Hughes, M.D. (Baltimore, MD)

After the conclusion of the prospective randomized double blind trial showing that shunt prophylaxis with interoperative and two days of post-operative treatment resulted in a dramatic decrease of shunt infections, an additional 215 shunt operations were performed with all patients being placed on the treatment protocol. The infection rate of this group was 0.93%. The recovered organisms in the two patients having infections will be discussed in detail; however, none of them showed resistance to Cephalothin and both were typical infections. There were no late surpressed infections in the study group. A discussion of the doses of antibiotics as they have been adjusted will be addressed together with the family of new penetrating antibiotics which may avoid the need for post-operative shunt taps. Drug allergies, seizures, and other complications of antibiotic therapy will be discussed in detail.

Our overall conclusion is that the use of prophylactic antibiotics in cerebrospinal fluid spine shunting operations significantly reduces the major morbidity and causes of mortality that has plagued these procedures in the past. The use of newer drugs promises to eventually eliminate the need for shunt taps and have the patient on therapeutic systemic treatment for the perioperative period. total infection 0.45% 3 others - break in protocol

35. DECREASED RISK OF INFECTION IN CEREBROSPINAL FLUID SHUNT SURGERY USING PROPHYLACTIC ANTIBIOTICS: A CASE-CONTROL STUDY

Beverly C. Walters, M.D., Harold J. Hoffman, M.D., F.R.C.S.(C), E. Bruce Hendrick, M.D., F.R.C.S.(C), Robin P. Humphreys, MD., F.R.C.S.(C) (Toronto, Canada)

Four randomised controlled trials have focussed on the use of prophylactic antibiotics in the prevention of cerebrospinal fluid (CSF) shunt infection. None of these studies has been able to demonstrate a statistically significant difference in rate of infection. This may be due to the inadequate power of the studies to detect a clinically significant difference, as a result of small sample size. This retrospective case-control study was designed using the same basic principles of avoidance of bias which make randomised controlled trials the model studies for generation of scientific evidence. It has found a highly statistically significant difference in the use of antibiotics among infected compared to non-infected patients, suggesting a three times greater risk of infection among patients who did not receive antibiotics. This study provides the strongest evidence to date regarding the usefulness of perioperative antibiotics in CSF shunt surgery. Excellent Stat's

36. CONTRIBUTION OF THE SPINAL COMPARTMENT TO CEREBROSPINAL FLUID DRAINAGE

J. Gordon McComb, M.D., Shigeyo Hyman, M.S., Martin H. Weiss, M.D. Los Angeles, CA.

In order to evaluate the contribution of the spinal compartment to cerebrospinal fluid (CSF) drainage, three groups of white New Zealand rabbits underwent a series of infusion with artificial CSF containing indigo carmine and radioiodinated serum albumin (RISA). In the first group the spinal sub-arachnoid space (SAS) was blocked at the C-2 level followed by infusion of artificial CSF into the spinal SAS distal to the block. In the second group the ventricles were infused after blocking the spinal SAS at the same level. As a control, a third group underwent ventricular infusion without blockage of the spinal SAS. The intracranial pressure (ICP) was monitored in each animal. The distribution of the dye and RISA was determined following four hours of infusion.

The ICP did not rise in the group being infused distal to the block in the spinal SAS. The ICP rose to the same level during ventricular infusion whether or not the spinal compartment was excluded. CSF drainage from the ventricles, as noted by dye and RISA distribution, was similar irrespective of isolating the spinal compartment. The spinal nerve roots appeared to be the site of CSF drainage during infusion of the isolated spinal SAS. The spinal compartment does not seem to be an important route for CSF drainage in acute infusion experiments.

37. THE PATHOPHYSIOLOGY OF IVH IN THE BEAGLE PUPPY

Dennis L. Johnson, M.D., Catherine Shaer, M.D., David C. McCullough, M.D. (Washington, DC)

The term beagle puppy has been used extensively as an experimental model of IVH in the preterm infant. We have reproduced this model in our own laboratory and are reporting our analysis of 16 preparations. In order to substantiate variables contributing to the model, the pathophysiologic sequence of events was divided into 3 distinct phases: 1) hemorrhagic hypotension, 2) hyperperfusion, 3) postinfusion. We selected variables hypothesized to play a role in IVH and submitted subsets selected in the context of these pathophysiologic phases to 3 separate discriminant function analyses. In the first analysis, only the measurements for variables representing pre-reinfusion events were utilized. Variables included weight, volume of bleed, mean temperature, the difference between the pre- and posthemorrhage hematocrit, arterial pH, arterial p02, arterial pCO2, baseline blood pressure prior to induced hypotension, the level of hypotension, and the degree to which the BP recovered prior to reinfusion. Three variables -- temperature, baseline BP, and pCO2 -- provided for perfect (100%) hemorrhage group classification. In the second discriminant analysis variables representing the events associated most closely with hyperperfusion, the BP after infusion and the duration of hypertension were added to the previously considered variables for determining their potential classification ability. Again perfect group classifications were obtained using only the four variables of weight, change in hematocrit, pH, and the BP after reinfusion. In the third phase, the aftermath of reinfusion/hypertension, additional variables included the BP after 10, 20, 30, 40, 50 and 60 minutes. The animal blood volume, pH, pCO2, and BP after 50 minutes perfectly classified all animals into their groupings. This data supports our hypothesis that membrane injury is the fundamental pathophysiologic basis of IVH and that hemodynamic changes are secondary.

38. IN VITRO EVALUATION OF CSF SHUNT FUNCTION BY RADIONUCLIDES CLEARANCE STUDIES.

Bruce D. Pendleton, M.D., P. Alex Roberts, Ph.D., Arden F. Reynolds, Jr., M.D.. Michael Pollay, M.D. (Oklahoma City, OK)

In order to evaluate shunt function in the hydrocephalic patient, a study was undertaken to relate the clearance of a radio-labeled marker from the reservoir or chamber of a ventricular shunting system and flow through the valve system. An in vitro stidy was conducted on a number of commonly used shunting devices which included: adult and pediatric Cordis (Hakim) valve, the standard and experimental Pudenz-Schulte Valve, Holter-Hausner Cruciform slit valve, Holter mini LP valve, a Heyer-Schulte mini LPV, a Heyer-Schulte Standard LPV, and a Heyer-Schulte Foltz reservoir.

The method used was based on the injection of 0.5 ml of ^{113m}In-DTPA into the valve reservoir or chamber as a bolus. Following this, using a constant flow pump, a nonisotopic fluid was infused at flow rates varying between 0.46 and 0.034 ml/min. The radioactivity was counted with a NaI crystal and counts accummulated at five second intervals on a multi-channel analyzer. The studies were done in triplicate at each flow rate and a working graph of the log values of counts versus time in seconds was constructed. From these graphs a mean $t_{\frac{1}{2}}$ was computed for each flow rate and this value used to construct a second graph in which flow rate was presented versus $t_{\frac{1}{2}}$ (seconds). Each of the valve systems studies demonstrated a different and complex mathematical relationship between flow rate and half-time of clearance. A best fit curve of flow rate versus $t_{\frac{1}{2}}$ was constructed for each system so that an unknown flow rate could be determined for each valve based on the measured half-time ($t_{\frac{1}{2}}$ of isotope clearance). The graphical solution from these experimentally derived curves is proposed as a quick method for evaluating shunt function in the clinical situation.

39. ABDOMINAL CSF CYST SYNDROME: DIAGNOSIS AND MANAGEMENT.

Herbert H. Engelhard, M.D., Yoon S. Hahn, M.D. (Chicago, IL)

Twenty cases of abdominal cerebrospinal fluid cyst syndrome presenting to Chicago Children's Memorial Hospital over the past two years were reviewed. The syndrome occurring in patients with ventriculoperitoneal (VP) shunts, consists of the findings of an acute abdomen and symptoms, to a greater or lesser extent, of a shunt malfunction. Physical examination reveals fever, lethargy, and diffuse abdominal tenderness, often with drawing up of the knees. Vomiting, diarrhea, or abdominal distension were sometimes present. The clinical picture was most often confused with the diagnosis of peritonitis resulting from bowel perforation.

Diagnostic studies included abdominal x-rays, head CT scan, abdominal ultrasound and CDF cultures. Head CT scans were either unchanged from prior studies, or showed mild dilatation of the ventricles. Abdominal ultrasound studies revealed the presence of a cystic fluid collection in all cases. CSF cultures were usually negative. When positive, the most common organism found was S. epidermidis. Treatment consisted of replacing the VP shunt with an external ventricular drain (EVD). When cultures were consistently negative, the shunt was replaced. In all cases, the VP shunt was found to be functional, and the abdominal cyst to be under pressure. Prophylactic antibiotics were used while the patient had the EVD. In rare instances, conversion to the ventriculoatrial shunt was required. In all cases, the symptoms quickly resolved after EVD placement. The pathophysiology of the cyst formation is poorly understood. Most patients had had prior shunt revisions. The cyst is believed to result from loculation and inadequate absorption of the CSF due to adhesions within the abdominal cavity. It is hoped that a better appreciation of the nature of the abdominal CSF cyst syndrome will aid in its rapid diagnosis and treatment, with the avoidance of needless procedures such as exploratory laparotomy.

40. MOYA MOYA: EXPERIENCE AT THE HOSPITAL FOR SICK CHILDREN, TORONTO.

Meredith V. Olds, M.D., Harold J. Hoffman, M.D. (Toronto, Canada)

Between 1970 and 1983 ten verified cases of Moya Moya were treated at the Hospital for Sick Children in Toronto. The children presented between one and twelve years of age. There were four males and six females. One patient was oriental and the rest caucasian.

Seven patients underwent revascularization procedures, with STA-MCA anastomosis or superficial temporal dural synangiosis. Follow-up has ranged from one to six years. All patients with STA-MCA anastomosis are neurologically intact. The patients with synangiosis are stable and functional with mild deficits. Two patients without revascularization procedures died. Another untreated patient is alive but retarded and hemiplegic.

41. EXTREME HYPERVENTILATION AND CEREBRAL BIOENERGETICS IN THE CAT

Patricia Gibbons, M.D., David Swedlow, M.D., Frank Welsh, Ph.D.,
Derek A. Bruce, M.B. Ch.B. (Philadelphia, PA)

Nine cats were anesthetised with intraperitoneal ketamine (10 mg/Kg), paralyzed, intubated and artifically ventilated with 30% PO_2 and room air. Anesthesia was maintained with continuous infusion of I.V. ketamine and galamine. End-tidal CO_2 and temperature were continuously recorded. A femoral artery and vein were catheterized and used for continuous recording of blood pressure and for intermittent sampling of arterial blood for measurements of $Pa0_2$, $PaCO_2$, pH and nitrous oxide during cerebral blood flow. The cat was then placed in a steriotatic frame, the scalp reflected and two screw electrodes placed, one in either frontal area and a third placed over the frontal sinus with reference electrodes to both ears. Both median nerves were stimulated percutaneously using needle electrodes and the median nerve stimulation and frontal recordings were used to measure somatosensory evoked responses (SSER). The sagittal sinus was then cannulated with a PE20 tubing for sampling of venous blood for oxygen and cerebral blood flow (CBF). CBF measurements were performed by the addition of 10% nitrous oxide (N20) to the inspired gas. Arterial and cerebral venous blood samples were obtained over a 20-minute period and measurements of nitrous oxide concentration made. Blood flow was calculated using the modified Kety-Schmidt technique.

Cerebral blood flow and arteriovenous oxygen differences across the brain were measured at an arterial CO_2 baseline of 38. The animal was the hyperventilated for 45 minutes to a $PaCO_2$ of around 20. A second blood flow and AV difference of oxygen measured, and finally ventilation was increased to drop the $PaCO_2$ to approximately 10 torr for 45 minutes at which time a third CBF measurement and AV difference was drawn. The brain was then superfused with liquid nitrogen and frozen in situ. The brain was removed and measurements made at ATP phosphocreatine and lactate in the grey and white matter.

CBF decreased from 40 ± 8 ml/100 gm/min at $PaCO_2$ of 38 to 26 ml/100 gm/min at $PaCO_2$ 18. There was little further decrease with a drop in $PaCO_2$ to 10, CBF equalled 24 ml/100 gm/min. The initial AV oxygen difference was 44 at $PaCO_2$ of 18. It fell to 24 at $PaCO_2$ of 10, slightly further to 20. There was no change in the somatosensory evoked responses.
Table 1 shows the results of cortical metabolites:

	----Cortical----		----White Matter----	
	Experimental	Control	Experimental	Control
ATP mmol/kg	2.08 (.09)	2.29 (.02)	1.65 (.10)	2.06 (.04)
PCR mmol/kg	3.80 (.67)	5.40 (.27)	2.26 (.47)	3.11 (.07)
LAC mmol/kg	4.80 (1.4)	0.63 (.14)	4.20 (1.7)	0.70 (.17)

CONCLUSION

No evidence of brain ischemia was found.

42. OPERATIVE PLANNING FOR CONGENITAL CEREBRAL MALFORMATIONS AND HYDROCEPHALUS: BENEFIT OF STABLE XENON COMPUTED TOMOGRAPHY.

K. H. Manwaring, M.D., T. J. Tarby, M.D., J. A. Hodak, M.D., Hal W. Pittman,M.D.
Phoenix, AZ)

Congenital cerebral malformations and hydrocephalus may be accompanied by aberrations in cerebral blood flow. These aberrations may result from parenchymal maldevelopment, malformation of the cerebral vasculature, or result as a secondary process. We have utilized stable xenon computed tomography to evaluate regional cerebral blood flow (rCBF) in children with four different cerebral pathologies in anticipation of surgical intervention: a thrombosed Vein of Galen aneurysm, hemimegalencephaly, Sturge-Weber syndrome, and hydrocephalus of posthemorrhagic origin.

In the patient with a Vein of Galen aneurysm, no aneurysmal flow was demonstrable; impression of thrombosis was confirmed by angiography. The only surgical intervention was ventriculoperitoneal shunting.

In hemimegalencephaly, the abnormal hemisphere showed markedly increased rCBF when compared to the putatively normal hemisphere. This increased flow correlated with increased vessel caliber noted at angiography and subsequent hemicorticectomy.

In the case of trigeminoencephaloangiomatosis, CT scan showed only a mild asymmetry in hemispheric size; cerebral angiography was unremarkable. Stable xenon CT revealed a marked deficit in parenchymal blood flow in the hemisphere underlying the leptomeningeal vascular malformation, suggesting a clinically significant deficit in rCBF.

Six cases of neonatal posthemorrhagic hydrocephalus showed asymmetry in rCBF, most pronounced in the parietal-occipital regions of the cortex on the side of previous hemorrhage. While two of three post-drainage scans showed improvement in flow, significant asymmetries persisted suggesting a primary ischemic process.

We believe these examples represent the first demonstration of the utility of stable xenon computed tomography in assessment of rCBF in neurologic conditions of childhood.

6/1000 ½ temporal

43. TEMPORAL LOBECTOMY IN CHILDREN WITH EPILEPSY 1970-1983

W. Richard Marsh, M.D., Fredric B. Meyer, M.D., Edward R. Laws, Jr., M.D.,
Frank W. Sharbrough, M.D. (Rochester, MN)

Neurologist

The surgical results of temporal lobectomy for medically refractory seizures in 50 patients under the age of twenty are analyzed. There were 29 males and 21 females with an average age of 15.8 years. The average age at onset of seizures was 7½ years. The median time between the onset of seizures and surgery was 8.3 years. There was a significant history of antecedent factors in 35 patients. Preoperatively, five patients had 1-4 sz/month, ten had 5-10 sz/month, fifteen had 11-20 sz/month, and twenty had over 20 sz/month. Postoperatively, 27 patients (54%) were seizure free, 12 patients (24%) had only auras, 5 patients (10%) were improved, and six patients (12%) were unchanged. Therefore, 78% were essentially seizure free, and 88% benefited significantly from the operation. Thirty-eight out of forty patients who responded to a questionnaire felt that their social interaction and intellectual development had improved related to the operation. Overall, there was no significant change in Weschler Intelligence Tests. However, there was a greater likelihood of the patient improving (p .01) in both verbal IQ and perceptual IQ if the operation was done earlier. Six patients had tumors, three of which were unrecognized by preoperative CT scan and angiogram. Complications included 2 transient upper extremity paresis, 4 with transient anomic aphasia, and 1 with a transient expressive aphasia. Since the surgical results are excellent in terms of seizure control and improved social function, it is recommended that temporal lobectomy be considered early in the treatment plan for medical refractory temporal lobe seizures in children.

10-25% may be candidates

Febrile 19 cryptogenic 3 | 6 tumors | 2 infections
Trauma 7 Idio 9 | grad II astro 2 | mono paresis 2
Tumor 6 | oligo 2 | dysphasia 5
Menin 5 | ganglí 2

44. SPONTANEOUS EPIDURAL CERVICOTHORACIC HEMATOMA EXTENDING FROM C3 TO T3 IN A CHILD

Joan H. Humphreys, M.D., John P. Laurent, M.D., William R. Cheek, M.D., Hatem Megahed, M.D. (Houston TX)

Spontaneous epidural spinal hematoma is extremely rare, and only 3 cases have previously been reported in children under 10 years of age. In adult patients 37% have died when the hematoma extended above C5. Although minor trauma has been entertained as the etiology, the cause remains unknown. A 6 year old presented with a 3 week history of severe interscapular and neck pain, progressive arm and leg weakness, and low grade fever. There was no history of trauma. Physical examination confirmed quadriparesis with atrophy of intrinsic muscles of the hand. Magnetic resonance (MR) and metrizamide computered tomographic scans showed an epidural mass extending from C3 to T3. Laminectomies were performed and revealed an encapsulated mass underlying the ligamentum flavum. It contained fluid characteristic of a chronic hematoma under pressure. Examination of the tissue showed no evidence of vascular malformation, neoplasm or infection. Cerebrospinal fluid, coagulation and collagen vascular studies were normal. In defining the diagnosis of an epidural spinal mass in children, non-invasive diagnostic tests are preferable. This is the first reported case of a chronic spinal epidural hematoma diagnosed by magnetic resonance and demonstrates its superiority in directing surgical intervention.

45. SELECTIVE POSTERIOR RHIZOTOMY FOR THE RELIEF OF SPASTICITY

W. J. Peacock, B.Sc., M.D., Ch.B., F.R.C.S.(Edin) (CAPE TOWN)

In the 1890's Sherrington showed that experimentally produced spasticity could be relieved by sectioning posterior spinal roots. However, in clinical practice the resultant sensory loss made the procedure unacceptable until selective posterior rootlet section could be performed whereby rootlets not involved in abnormal circuits might be preserved and only those associated with abnormal responses were divided.

10 mamp single/ train

At the University of Cape Town, between 1981 and 1984, 67 patients suffering from the effects of spasticity were subjected to selective posterior rhizotomy. There were 58 cases of cerebral palsy, 31 being spastic diplegics, 24 spastic quadruplegics and 3 dystonics. Seven patients were spastic due to spinal cord disease, 2 due to a stroke and 1 was hemiplegic following cerebral trauma.

The procedure is performed via lumbar laminectomy exposing the cauda equina. Posterior lumbar and sacral rootlets are individually stimulated with a nerve stimulator using specific intensity and pulse duration settings and those rootlets with an abnormal motor response are divided. Postoperatively spasticity has been reduced in all cases and gross movements such as those involved with sitting, standing, and walking were improved as were fine movements. In almost all cases an improvement in mood was noted.

Indications for and contra-indications to selective posterior rhizotomy and details of technique will be discussed. A film illustrating patients' pre-operative and post-operative abilities will be shown.

6y old neck pain
inter scapular pain
neck posture kyphos
left hand weakness muscle wasting
gait disturb bone
fever

clear sensory bicep trigs atroph
clawhand
C5-T1 paresis (L) spotty sensation

46. INTRACRANIAL ARACHNOID CYSTS IN CHILDREN

G. R. Harsh, IV, M.D. M.S.B.Edwards, M.D., C. B. Wilson, M.D. (San Francisco, CA)

Controversy in the literature concerning appropriate therapy for intracranial arachnoid cysts prompted this review of 16 pediatric cases. Presentation reflected anatomic distribution: middle cranial fossa cysts (4) caused headache, seizure, or focal neurologic deficit; suprasellar cysts (5) presented with intracranial hypertension, visual symptoms, precocious puberty, or bobble-head doll syndrome; diffuse supratentorial (2) and posterior fossa (5) cysts led to intracranial hypertension or developmental delay. Ventriculomegaly occurred in 7 of 11 supratentorial and 4 of 5 infratentorial cysts. CT scanning or magnetic resonance imaging (MRI) alone was diagnostic in most cases.

Three of seven supratentorial cysts treated by craniotomy for fenestration recurred; each responded to subsequent cyst-peritoneal shunting. Two patients with posterior fossa cysts treated by fenestration failed to benefit; one who subsequently received combined ventriculo-peritoneal and cyst-peritoneal shunting improved. Cyst fenestration was frequently followed by lethargy and fever suggestive of aseptic meningitis. The four patients with supratentorial and the three with infratentorial cysts treated initially by shunting did well. Following shunting there was decrease in cyst and/or ventricular size and no morbidity. Individual cases showed the value of prenatal diagnosis, MRI in excluding tumor and delineating cyst boundaries, ultrasonographic monitoring of asymptomatic and postoperative infants, and ultrasound guidance of shunt catheter placement.

Cyst-peritoneal shunting should be performed on all symptomatic patients. If ventriculomegaly is present, combined cyst and ventricle shunts are indicated. Craniotomy for fenestration and studies assessing cyst communication with ventricular and subarachnoid spaces should be reserved for cysts refractory to shunting and, in older patients, for cysts adjacent to larger basilar cisterns.

47. THE MANIFESTATIONS AND MANAGEMENT OF HYDROSYRINGOMYELIA IN CHILDHOOD

John Neill, M.D., Harold J. Hoffman, M.D., E. Bruce Hendrick, M.D.
Robin P. Humphreys, M.D. (Jackson, MS)

Fifty patients were treated surgically for hydrosyringomyelia from February 1973 to April 1984, at the Hospital for Sick Children in Toronto. This report is based on review of their radiographic and medical records. Forty-three patients had radiographic or surgically confirmed malformations of the hindbrain. These included 25 patients with the Chiari type II malformation, 12 patients with the Chiari type I malformation and 6 patients with "acquired" Chiari type I malformation. Those patients with the Chiari type II malformation presented with classical syringomyelic syndromes, atypical syringomyelic syndromes (i.e. progressive lower extremity dysfunction with normal upper extremities), or scoliosis. Five of the "acquired" Chiari malformations had undergone lumbo-peritoneal shunting as infants. Forty-one of the patients were studied initially with metrizamide myelography and computered spinal tomography. Six of the other 7 patients had other abnormalities of the craniospinal axis, but no demonstrable hindbrain abnormality. We present the clinical and radiographic findings in each group followed by our surgical methods and results. We conclude that virtually all cases of hydrosyringomyelia are associated with hindbrain anomalies and that posterior fossa decompression with occlusion of the central canal at the obex is a safe and effective form of therapy.

CT mass effect
cervical
thoracic
MRI better
signal

CSF ok
CV ok
coags ok
culture Ø

48. OCCIPITAL PLAGIOCEPHALY DUE TO UNILATERAL LAMBDOID SYNOSTOSIS - A REAPPRAISAL AND SOME NEW THOUGHTS

Clinton Frederick Miller, M.D., John W. Skinner, M.D., Joseph M. Nadell, M.D. (New Orleans, LA)

There has been a trend toward earlier correction of craniofacial syndromes in order to take advantage of extensive "passive" remodeling of the craniofacial skeleton during the rapid phase of infantile brain growth. Of the craniosynostoses, one syndrome - occipital plagiocephaly due to unilateral lambdoid synostosis - has received less attention than others and is more likely to receive delayed or no treatment. This lack of attention correlates with diagnostic insecurity attributable to many factors, some of which are: perennial pathogenetic confusion with so-called "positional flattening"; falsely reassuring radiographic findings; dissatisfaction with cosmetic results of (often delayed) surgery; cavalier assumption of innocuous cosmetic impact if left untreated; and, historical prejudice engendered by its reported rarity (2% of Boston Children's Hospital series vs 9% of Toronto Sick Children's Hospital series). Seven pathologically verified cases of typical unilateral lambdoid synostosis in less than one year's time (infants aged 3 to 8 months) caught our attention and prompted this reappraisal. We would like to present some old and new observations, clinical and radiological, helpful in making an earlier and more secure diagnosis; and to propose a new mechanical explanation of how the observed pathology (parasutural enostotic ridging) results in occipital flattening - plagiocephaly.

49. TERMINAL MYELOCYSTOCELE

Thomas P. Naidich, M.D., David G. McLone, M.D., Ph.D. (Chicago, IL)

Terminal myelocystoceles constitute approximately 5% of skin-covered lumbosacral masses and are especially common in patients with cloacal exstrophy. Pathologically, terminal myelocystocele consists of (a) skin-covered lumbosacral spina bifida; (b) an arachnoid-lined meningocele which is directly continuous with the spinal subarachnoid space; and (c) a low-lying, hydromyelic spinal cord which tranverses the meningocele and then expands into a large terminal cyst. The terminal cyst bulges into the extraarachnoid compartment caudal to the meningocele and forms a distal sac which does not communicate with the subarachnoid space. The terminal cyst is lined by ependyma and dysplastic glia, is directly continuous with the dilated central canal of the cord, and likely represents a ballooned terminal ventricle. Simple neuroradiologic procedures now permit accurate identification of this specific form of dysraphism. Since patients with terminal myelocystocele have normal intellectual potential and are usually born without neurological deficit, they must be identified and repaired early, prior to onset or progression of lower extremity pareses.

50. INCREASE IN VENTRICULAR SIZE WITH HIGH PRESSURE INFUSION OF ARTIFICIAL CSF INTO THE LATERAL VENTRICLE OF DOGS

Harold L. Rekate, M.D., Scott Erwood, M.D., Howard Chizeck, D.Sc. (Cleveland, OH)

The question of the mechanism of hydrocephalus produced by choroid plexus papillomas has been thoroughly debated in the literature and it is clear that some choroid plexus papillomas do produce large volumes of CSF. Whether this overproduction of CSF without obstruction to outflow or pulse pressure augmentation is enough to cause hydrocephalus has yet to be delineated and is the central issue in these experiments.

Five adult mongrel dogs were anesthetized and underwent cannulation of each lateral ventricle. One cc of Conray^{R} was infused into each lateral ventricle for initial volumetric determination. Pressure was allowed to return to normal and an infusion of artificial CSF (Eliot's B solution) was begun using a pump with a feedback control mechanism to maintain ICP at 50 torr. After 36 hours a second Conray^{R} ventriculogram was performed in each dog and then the ventricles casted with Stycast^{R}. Ventricular volumes increased in all dogs by an average of nine percent.

There are three important results of this study.

1. High pressure infusions of artificial CSF will produce ventriculomegaly
2. The volumetric measurements used can measure these differences
3. The viscoelastic model of ventricular volume can predict these results.

51. HYDROCEPHALUS: SYMPTOMATIC LOW ICP AND ASYMPTOMATIC SLIT-VENTRICLE SYNDROMES TREATED BY FLO-CONTROL VALVE SHUNTING

Eldon L. Foltz, M.D., Jeffrey Blanks, B.A. (Orange, CA)

I. Clinical symptoms and signs of low ICP in shunted hydrocephalic patients are not widely recognized. Eight shunted hydrocephalic patients suffering from symptoms usually attributed to shunt obstruction with increased ICP were studied. Four of these had undergone fruitless shunt revisions for these symptoms elsewhere. All suffered from headaches, nausea and occasional emesis when upright, and were largely relieved of these symptoms when lying down or in head down positions. Four patients developed blurring vision and mild diplopia after being upright, and three patients showed paresis of upward gaze. All shunts were VP shunts. Accurate CSF pressure studies showed low pressure, which fell to significant negative pressure in the upright position from minus 150 to minus 200. Pressure measurements in prone, upright and head dependent positions were critical data. Treatment with Flo-Control valves, medium (70-80 mm) and high pressure (80-120 mm) opening pressure gave prompt relief in all instances. The clinical aspects of such low ICP problems in shunted hydrocephalic patients needs emphasis.

II. Twenty-five patients with "slit ventricles" by CT scan, but without significant clinical symptoms, were studied as probable ICP problems. Twelve of these patients showed low ICP by measurement. All were treated with the same Flo-Control valve inserted into their shunt system. This increased intracranial pressure significantly but none showed any symptoms of such. Twelve patients showed enlarged ventricles to normal within three months by CT scan, but thirteen did not show ventricular enlargement subsequent to the revised shunt, presumably due to "stiff-slit" ventricles. This entity is as yet not clearly predictable.

The Flo-Control valve is a reliable pressure valve with flow control or flow rate characteristics which seem to handle the siphon problem in VP shunts.

The syndrome of symptomatic low ICP in hydrocephalic patients needs continued emphasis.

52. CRANIOFACIAL TECHNIQUES IN PEDIATRIC NEUROSURGERY

Larry V. Carson, M.D., (Augusta, GA)

Tumors involving the base of the cranium and face are sometimes difficult to expose and treat. The residual deformity after the treatment of such lesions may be significant. Craniofacial techniques provide improved exposure, facilitate extirpation of the lesion, and allow for reconstruction of the defect.

To provide improved exposure, the standard bicoronal flap may be split down the midforehead. The incision may then be carried directly down the midnose or laterally around the nose and down the philtral ridge to the lip. Circumferential elevation of the periorbita allows reduced retraction of the globe and surrounding structures. This exposure combined with a frontal craniotomy lends itself to en bloc resection of anterior cranial base tumors with orbital or facial extension. After verification of margin status, reconstruction may be performed using autogenous materials. Reconstruction minimizes the deformity and re-establishes the normal anatomy which may prevent comorbity and complications.

A case of a child with osteogenic sarcoma involving the midface, orbit and the anterior fossa is presented to illustrate the advantages of craniofacial techniques in pediatric neurosurgery.

53. MODIFIED PRONE POSITION AND CROSS-BAR CRANIOTOMY FOR TOTAL CRANIAL VAULT RESHAPING

T.S. Park, M.D., Charles S. Haworth, M.D., M. Sean Grady, M.D., John A. Jane, M.D. (Charlottesville, VA)

Total cranial vault reshaping is indicated for successful correction of multiple suture synostosis associated with craniofacial dysmorphism and macrocephaly due to advanced hydrocephalus. The major technical problems encountered during the operation are related to 1) positioning of the patient's head and consequent limited access to the calvarium, 2) proper replacement of the fragmented calvarium following craniotomy, 3) excessive blood loss.

The authors have overcome the problem in positioning the patient's head by utilizing a newly developed face frame that secures the patient in the prone position with hyperextended neck. This position allowed easy access to the calvarium from the supraorbital ridge to the foramen magnum and the total cranial reshaping to be performed at one stage.

Reconstitution of the calvarial contour following the extensive craniotomy was significantly facilitated by employing a single craniotomy flap in a curved cross-bar fashion. The cross-bar craniotomy flap consists of four bony bars that are continuous with each other at its central portion. It serves as a basic bony frame that holds the remaining bone flaps in the corrected position.

Blood loss throughout this extensive operative procedure was minimized by separation of the scalp from the pericranium followed by removal of the calvarium with pericranium in large segments and use of scalp clips and infiltration of the scalp with epinephrine.

The authors' experience with the operative techniques and postoperative results will be discussed.

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