Annual Meeting
Réunion Annuelle

OTTAWA
September 21-24, 1988

Canadian Association of Paediatric Surgeons
l’Association Canadienne de Chirurgie Infantile
programme détaillé

programme schedule

OTTAWA
September 21-24, 1988
The Canadian Association of Paediatric Surgeons was granted its charter in 1967. Its main aim is to improve the surgical care of infants and children in Canada.

There are three main areas in diagnosis, treatment and research which are of special concern to the members,

**Infants Born With Congenital Abnormalities**

Even though the majority of newborn infants who have severe congenital abnormalities can be treated successfully by a surgical operation, often the condition is either not recognized, or if it is diagnosed, the local physician may be unaware of the possibilities for surgical cure. In this situation most of these babies die, or some survive to live a life crippled by their deformity.

**Malignancy in Childhood**

Cancer is the second commonest cause of death in childhood. Now surgical removal of the tumor, combined with x-radiation and chemotherapy provided by an aggressive team utilizing new techniques can achieve a cure in over 50% of these patients.

**Trauma**

Finally, the number one killer of children in North America is accidents. Here again, with modern methods of first aid, transportation, resuscitation, intensive care, and specialized surgical team effort, many of these seriously injured children can be saved.

**EDUCATION PROGRAM**

To accomplish an improvement in surgical care for babies and children, the Canadian Association of Paediatric Surgeons has launched an educational program for doctors, nurses and others working in the paediatric health field. To support this program, an educational fund has been established.
MEETING AND BUSINESS PROGRAM

WEDNESDAY, SEPTEMBER 21, 1988

1030 - 1500 - COUNCIL MEETING
   Westin Hotel, Alberta Room

1600 - PROGRAMME DIRECTORS MEETING
   (Dr. Desjardins, Chairman)

THURSDAY, SEPTEMBER 22, 1988 - WESTIN HOTEL, ONTARIO ROOM

0700 - REGISTRATION

0800 - 1200 - ORIGINAL PAPERS

1200 - 1300 - FRED McLEOD LECTURE
   Dr. John G. Raffensperger

FRIDAY, SEPTEMBER 23, 1988 - WESTIN HOTEL, ONTARIO ROOM

0800 - 1100 - ORIGINAL PAPERS

COMBINED MEETING - CANADIAN ASSOCIATION OF PAEDIATRIC SURGEONS
   & SOCIÉTÉ FRANÇAIS DE CHIRURGIE PÉDIATRIQUE
   -University of Ottawa, Health Sciences Audit.

1230 - BUS FROM WESTIN HOTEL

1300 - 1630 - ORIGINAL PAPERS

1630 - 1730 - RECEPTION

1730 - BUS RETURNS TO WESTIN HOTEL

SATURDAY, SEPTEMBER 24, 1988 - CHILDREN'S HOSPITAL OF
   EASTERN ONTARIO
   - 4 NORTH CONFERENCE ROOM

0800 - 1115 - ORIGINAL PAPERS & CASE REPORTS

1115 - ANNUAL BUSINESS MEETING
   REUNION D'AFFAIRES ANNUELLE

1300 - MEETING OF ROYAL COLLEGE SPECIALTY COMMITTEE ON
   GENERAL PAEDIATRIC SURGERY
   (Dr. Gordon Cameron, Chairman)
SOCIAL PROGRAM

WEDNESDAY, SEPTEMBER 21, 1988:

WELCOMING RECEPTION

Place: Dr. Stanley Mercer's Home
       210 Bueno Vista Road, Ottawa

Time: 19:00 RSVP

FRIDAY, SEPTEMBER 23, 1988:

C.A.P.S. BANQUET

Place: Royal Ottawa Golf Club,
       Aylmer Road, Ottawa

Time: 19:30 RSVP

630 FOR 730
BLACK TIE
FUTURE ANNUAL MEETINGS

21st ANNUAL MEETING
EDMONTON, ALBERTA
September 22-26, 1989

22nd ANNUAL MEETING
TORONTO, ONTARIO
September 14-18, 1990

23rd ANNUAL MEETING
QUEBEC CITY, QUEBEC
September 20-23, 1991
CANADIAN ASSOCIATION OF PAEDIATRIC SURGEONS
L'ASSOCIATION CANADIENNE DE CHIRURGIE INFANTILE

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1st ANNUAL MEETING - January 22, 1969 - VANCOUVER

* DECEASED
0700 Registration - coffee and donuts

0800 - 1000 Co-Chairmen / Les Co-Présidents
S. Mercer, N. Wiseman

1. 0830 SMALL BOWEL TRANSPLANTATION PERMITS SURVIVAL IN RATS WITH LETHAL SHORT GUT SYNDROME
Children's Hospital and Children's Hospital Research Foundation, Columbus, Ohio

2. 0845 BILIARY ATRESIA: THE EFFECT OF SURGICAL TREATMENT ON THE DISEASE
A.H. Khan, H. Blanchard, A.L. Bensoussan, P. Brochu, O. Reinberg
Hôpital Sainte-Justine, Montreal, Quebec, Canada

3. 0830 CHOLECYSTITIS IN CHILDREN
J. Proctor, S. Chou, E. McMullin
Children's Hospital of Eastern Ontario
Ottawa, Ontario

4. 0845 OCCURRENCE OF CONTRALATERAL INGUINAL HERNIAS FOLLOWING UNILATERAL REPAIR IN A PEDIATRIC HOSPITAL
J. Given, S.A. Rubin,
Children's Hospital of Eastern Ontario, Ottawa, Ontario

5. 0900 UNUSUAL PATTERNS OF CONGENITAL NECK MASSES IN CHILDREN
R.E. Sonnino, N. Spigland, F.M. Guttman, J. DesJardins, J.M. Laberge,
C.R. Moir, L.T. Nguyen
The Montreal Children's Hospital/Ste. Justine Hospital, Montreal, Quebec

6. 0915 THE LATE PRESENTING PEDIATRIC MORGANNI HERNIA - A BENIGN CONDITION
L. Berman, D. Stringer, S.H. Ein, B. Shandling
Hospital for Sick Children, Toronto, Ontario, Canada

7. 0930 THE USE OF THE UMBILICAL SITE FOR TEMPORARY OSTOMY: REVIEW OF 47 CASES
R.G. Fitzgerald, G.S. Cameron, G.Y. Lau
Department of Surgery, McMaster University, Hamilton, Ontario

8. 0945 ADNEXAL TORSION IN CHILDREN
N. Spigland, S. Yazbeck, J.C. Ducharme
Hôpital Sainte-Justine, Montreal, Quebec, Canada
15. 0830  Etiology of Bowel Damage in Gastrochisis: Studies in the Fetal Lamb
    J.C. Langer, M.T. Longaker, T.M. Crombleholme, S.J. Bond, C.D. Rudolph,
    W.E. Pinkheiner, E.E. Verrier, M.R. Harrison
    Department of Surgery, University of California
    San Francisco, California, U.S.A.

16. 0845  A Standard of Comparison for Acute Surgical Necrotizing Enterocolitis
    M.N. Ross, E.R. Wayne, J.S. Janik, J.D. Burrelling, J.H.T. Chang
    The Children's Hospital, Denver, Colorado

17. 0850  Rectal Involvement in Necrotizing Enterocolitis (NEC)
    S.Z. Rubin, A. Alawadi
    Children's Hospital of Eastern Ontario, Ottawa, Ontario

18. 0845  Topical and Systemic Antibiotics in the Prevention of Wound Infection
    G. Stringel, M. Savrich, J. Horton, R. Bawdon
    University of Texas, Southwestern Medical School, Dallas, Texas

19. 0900  Surgical Outcome of Necrotizing Enterocolitis
    N. Spigelman, S. Yazbeck, J. G. Desjardins
    Hopital Ste-Justine, Montreal, Quebec, Canada

20. 0915  Idiopathic Intestinal Perforations in the Newborn: An Increasingly Common
    Entity?
    G. Weinberg, S. Kleinhaus, S.J. Boley
    Montefiore Medical Center - Albert Einstein College of Medicine, N.Y.

21. 0930  The Effect of CIS-Platinum on Liver Regeneration in the Rat
    D. May, C. Vindeun, C. Blair, E. Blacklock, P. Rogers, G. Taylor
    British Columbia Children's Hospital, Vancouver, British Columbia, Canada

22. 1015  Experience En Transplantation Hepatique Pediatric (a Montreal)
    H. Blanchard, A.L. Bensoussan, A. Weber et coll., M. Gauthier et coll.,
    J. Charest et coll., G.E. Rivard et coll., G. Delage et coll., H. Patrquin
    et coll., J.M. Laberge, F.M. Gutman, J. Adelson
    Hopital Sainte-Justine et Hopital de Montreal pour Enfants, Montreal, QUE.

23. 1030  Liver Transplantation (LT) in a High Risk Group of Children
    R.A. Superina, R.H. Pearl, E.A. Roberts, N. Graham, P.D. Greig, B. Langer
    Department of Surgery, Hospital for Sick Children, Toronto, Ontario

24. 1045  Orthotopic Liver Transplantation in Children With Polyspleenia Syndrome
    With Absent Inferior Vena Cava
    M.A. Hoffman, S. Celli, P. Ninkov, K. Rolles, R.Y. Caine
    Department of Surgery, University of Cambridge, Cambridge, UK
1300 - 1630

**COMBINED SESSION**

**CANADIAN ASSOCIATION OF PEDIATRIC SURGEONS**

Co-Chairman / Les Co Presidents
S. Yazbek, Serge Juskiewenski

**SOCIETE FRANCAIS DE CHIRURGIE PEDIATRIQUE**

25. 1300
**PEDIATRIC SURGERY - A COMPULSORY UNDERGRADUATE CLINICAL ROTATION. YES OR NOT?**
S.Z. Rubin, S. Mercer, M.M. O'Neil
Children's Hospital of Eastern Ontario, Ottawa, Ontario

26. 1315
**REPAIR OF INFRALEVATOR STRUCTURES IN HIGH AND INTERMEDIATE IMPERFORATE ANUS**
P. Hollard, P. Soucy, D. Louis, P. Mouriquand
Hôpital Debrusse, Lyon, France

27. 1330
**DUODENAL ATRESIA AND ITS ANTENATAL DIAGNOSIS**
B.J. Hancock, N.E. Wiseman
Department of Pediatric General Surgery
Children's Hospital, Winnipeg, Manitoba

28. 1345
**CONGENITAL DIAPHRAGMATIC HERNIA AND PRENATAL DIAGNOSIS (50 CASES)**
J.Y. Kurzene(1), F. Bary(1), E. Sapin(1), P.C. Helardot(1), A. Wajim(1),
F. Levin(2), Ch. Franconal(2), J.M. Baron(2),
Services de chirurgie pédiatrique (1) et gyneco-obstétrique(2) Hôpital
Saint Vincent de Paul, Paris, France

29. 1400
**PRETERM BABIES CAN HAVE HIRSCHSPRUNG'S DISEASE**
O. Reinberg, S. Yazbeck, H. Bart
Service de chirurgie pédiatrique, Hôpital Sainte-Justine, Montreal, Quebec

30. 1415
**MANAGEMENT OF HIRSCHSPRUNG'S DISEASE: CURATIVE SURGERY BELOW THREE MONTHS OF AGE**
M. Carcassonne, J.M. Guyot, G. Morisson-Lacombe, B. Kreitmann
Clinique Chirurgicale Infantile et Orthopedie, C.H.U. Timone-Enfants
Marseille, France

1430 - 1500 Coffee
31. 1500 CAPS LIVER TRAUMA REGISTRY REPORT
From the Trauma Subcommittee, Canadian Association of Pediatric Surgeons
A. Wong
Division of General Surgery, The Hospital for Sick Children, Toronto, ONT

32. 1545 H-TYPE SHUNT WITH AN AUTOLOGOUS VENOUS GRAFT IN THE TREATMENT OF PORTAL HYPERTENSION IN CHILDREN
F. Gauthier, J. Valayer, Ph. Montupet, O. De Dreuzy
Service de chirurgie, Department de Pedriatrie, Hopital de Bicetre et Faculte de Medecine Paris-Sud, France

33. 1550 GASTROSTOMY AND FUNDOPPLICATION IN NEUROLOGICALLY IMPAIRED CHILDREN
G. Stringel, M. Delgado
University of Texas, Southwestern Medical School and Texas Scottish Rite Hospital, Dallas, Texas

34. 1600 ESOPHAGEAL MOTOR EFFICIENCY IN GASTROESOPHAGEAL REFLUX
J. Arana, J.A. Tovar
University del Pais Vasco, Hospital NªSª de Aranzazu, San Sebastian, Spain

35. 1600 ESOPHAGEAL ATRESIA WITH DISTAL TEF: OTHER AROMALIES AND OUTLOOK
S.H. Ein, B. Shandling, D. Wesson, R.M. Filler
Hospital for Sick Children, Toronto, Ontario

C.N. Fekete, F. Bawab, S. Lortat-Jacob, P. Arhan, D. Pellerin
Department of Pediatric Surgery
Hopital des enfants malades, Paris, France

1630 - 1715 Wine Social
0800 - 0930 Co-Chairman / Les Co Presidents
S. Yazbek, R. Filler

37. 0800 THE SPECTRUM OF CHOLELITHIASIS IN THE FIRST YEAR OF LIFE
W.J. Schirmer, E.R. Grisom, M.W.L. Gauderer
Division of Pediatric Surgery, Case Western Reserve University,
Cleveland, Ohio

38. 0815 ISOLATED COMPLETE TRANSECTION OF THE COMMON BILE DUCT DUE TO BLUNT
TRAUMA IN A CHILD
M. Bourque, N. Spigland, H. Blanchard, A.L. Bensoussan, L. Garel
Department de chirurgie, Hôpital Ste-Justine

39. 0820 BILE DUCT STRicture IN AN INFANT WITH GASTROSCHISIS TREATED BY PER-
CUtaneous TRANSPHcatic DRAinAGE, BILIARY STENAGING, AND BALLOon DILATATION
B.J. Hancock, N.E. Wiseman, B.W. Rusnak
Department of Pediatric General Surgery and Radiology
St. Boniface General Hospital, Winnipeg, Manitoba

40. 0830 LONG TERM FOLLOW-UP OF SACROCOCcygeAL TERATOMAS WITH EMPHASIS ON ANO-
RECTAL FUNCTION
J. Bass, S. Yazbek, F. Luks
Hôpital Ste-Justine, Montreal, Quebec

41. 0840 HEREDITARY SACROCOCcygeAL TERATOMA
R.E. Sonnier, S. Chou, F.M. Guttman, C.R. Moir,
The Montreal Children's Hospital, Montreal, Quebec

42. 0850 NEONATAL DIAGNOSIS OF AN ANTERIOR MENINGOCOELE IN THE PRESENCE OF CON-
GENITAL ANAL STENOSIS AND PARTIAL SACRAL AGENESIS
H. Brem, B.L. Beaver, L.R. Scherer, B.E. Carson, J.A. Haller, Jr.,
The Johns Hopkins Hospital, Division of Pediatric Surgery, Baltimore
Maryland

43. 0900 MALIGNANT SMALL CELL TUMOR OF THE THORACO PULMONARY REGION (ASKIN TUMOR)
M. Bourque, M. Di Lorenzo, P.P. Collin, P. Russo, J.N. Laberge, C. Moir
Hôpital Ste-Justine & Montreal Children's Hospital, Montreal, Quebec

44. 0915 ESOPHAGEAL ATRESIA AND RIGHT AORTIC ARCH; RIGHT OR LEFT THORACOTOMY?
G. Stringel
University of Texas, Southwestern Medical School, Dallas, Texas

45. 0920 PULMONARY AGENESIS WITH ESOPHAGEAL ATRESIA AND TRACHEOESOPHAGEAL FISTULA:
A REPORT OF TWO CASES
W.A. Hoffman, R.A. Superina, D.E. Wesson
Department of Surgery, The Hospital for Sick Children, Toronto, Ontario
0930 - 0945 Coffee

0945 - 1115 Co-Chairman / Les Co-Présidents
N. Wiseman, J. Ducharme

46. 0945 THE EFFECT OF INTESTINAL ANASTOMOSIS ON GUT GROWTH AND MATURATION
G. Stringel, R. Uauy
University of Texas, Southwestern Medical School, Dallas, Texas

47. 1000 SEGMENTAL AGENESIS OF INTESTINAL MUSCULARIS - A CASE REPORT
A. Alawadi, S. Chou
Children's Hospital of Eastern Ontario, Ottawa, Ontario

48. 1005 ANATOMICALLY IDENTICAL SHORT SEGMENT HIRSCHSPRUNG'S DISEASE IN THREE MALE SIBLINGS
S. Z. Rubin
Department of Surgery, Children's Hospital of Eastern Ontario, Ottawa

49. 1015 DIAGNOSIS AND INCIDENCE OF CARDIAC INJURY IN CHILDREN WITH THORACIC TRAUMA
Hospital for Sick Children, Toronto, Ontario

50. 1030 TREATMENT OF FOCAL NODULAR HYPERPLASIA OF THE LIVER BY ALCOHOL EMBOLIZATION
P. Soucy, P. Rasuli, S. Chou, B. Carpenter
Children's Hospital of Ottawa, Ontario

51. 1035 GASTRIC HETEROTOPY OF THE BILIARY TRACT
M.J. Martínez Urrutia, F. Rivilla, J. Vazquez, J. Larrauri, J. Diez
Department of Surgery, La Paz Children's Hospital, Madrid, SPAIN

52. 1045 THE VENOCUTANEOUS FISTULA - A NEW APPROACH TO CHRONIC VENOUS ACCESS
C.E. Bagwell
University of Florida College of Medicine, Gainesville, Florida

53. 1050 POSTERIOR SAGITTAL ANORECTOPLASTY FOR PEDIATRIC RECURRENT RECTAL PROLAPSE
R.H. Pearl, S.H. Ein
Hospital for Sick Children, Toronto, Ontario

54. 1100 ESOPHAGEAL LEIOMYOMA IN CHILDREN: TWO CASE REPORTS
M. Bourque, N. Spigland, A.L. Bensoussan, P.F. Collin H. Blanchard O. Reiber
Hospital Ste-Justine, Montreal, Quebec

1115 - 1230 Annual Business Meeting
Reunion D'Affaires Annuelle
abstracts
FRED G. McLEOD LECTURE

"Congenital Anomalies of the Lung"

Dr. John G. Raffensperger, Chicago Illinois

Dr. Raffensperger was born in Joliette, Illinois and did his undergraduate education and medical school at the University of Illinois. He interned at Cook County Hospital followed by his general surgical training at the same institution. A fellowship in cardiac and thoracic surgery followed this training. Prior to beginning his surgical residency he spent two years with the United States Navy.

Dr. Raffensperger has practiced in Chicago since finishing his training. He is currently Surgeon-in-Chief at the Children's Memorial Hospital in Chicago and Professor of Surgery at Northwestern University Medical School. He has published extensively and has been editor of a number of books. In particular, he assumed responsibility for Swenson's Textbook of Pediatric Surgery. He has had a major interest in pulmonary problems in infants and children and has an ongoing interest in postgraduate medical education for interns and residents. He has been an examiner for the American Board of Surgery and American Board of Pediatric Surgery. He has been an active member of the American Pediatric Surgical Association and served on the editorial board of the Journal of Pediatric Surgery. Dr. Raffensperger has been a visiting professor at numerous universities and pediatric centers throughout the world.
SMALL BOWEL TRANSPLANTATION PERMITS SURVIVAL IN RATS WITH LETHAL SHORT GUT SYNDROME. RE Sonino, MD, DH Teitelbaum, MD, DJ Dunaway, BS, RP Harnel Jr, MD Children's Hospital and Children's Hospital Research Foundation, Columbus, Ohio.

The functional integrity of transplanted intestine would most convincingly be demonstrated if transplantation after otherwise lethal intestinal resection permitted survival and growth. We proposed to define the extent of bowel resection necessary for lethality and show that transplantation allows salvage of these animals. Adult Brown-Norway rats (250 g) underwent extensive small bowel resection (SBR) (Treitz to ileocecal valve) (n=5), cecectomy only (n=3), SBR + cecectomy (n=6) or SBR + cecectomy + syngeneic transplantation of 25 cm of jejunum (n=6). All animals with SBR or cecectomy alone survived and grew; all animals with SBR + cecectomy died within 2 weeks (mean 6.4 days); 6/6 animals with SBR + cecectomy + transplant survived and grew. The difference in survival between the lethally resected rats and those with an intestinal transplant was highly significant (p < 0.01). Dietary intake was similar in all groups. We conclude that in the rat, extensive small bowel resection alone is not sufficient to be lethal: most of the small bowel and the cecum must be resected to obtain a lethal model. We have also shown that transplantation of small bowel into rats with an otherwise lethal extent of bowel resection will allow the animals to survive and grow. These results suggest the need for caution in correlating survival after intestinal transplantation in the rat with transplant function, since the preservation of too much native gut might permit survival even if the transplant has no nutritional function.


During 1974-1985, 44 patients with biliary atresia have undergone portoenterostomy or portocholecystostomy at Ste-Justine Hospital, 31 females and 13 males. The mean age at surgery was 8 weeks. Our follow-up period is from 1-12 years. Successful biliary drainage was achieved in thirty patients, their mean bilirubin level was 2 mg/dl. On long term follow-up (3 to 12 years) 11 patients were asymptomatic and have no clinical jaundice with bilirubin level of 0.4 mg to 1.5 mg/dl. One of four patients who stopped secreting and was reexplored is anicteric and asymptomatic. Ten patients secreting at long term and anicteric have evident progression of their disease clinically and biologically.

Portoenterostomy is a valuable procedure to treat biliary atresia; but this evolutive disease progresses in a subdued manner even in the secreting patients. When successful, portoenterostomy or portocholecystostomy permits the patient and the surgeon to gain time. Based on our long term results we believe that most of the postoperative secreting patients of biliary atresia will end up getting a liver transplantation.
Cholecystitis in Children
J. Proctor, S. Chou; E. MacMullin.
Children's Hospital Eastern of Ontario

We have reviewed 35 cases of cholecystitis / cholelithiasis in children, seen at the Children's Hospital of Eastern Ontario, during the period of 1978-1988. There were 30 females and 5 males, their average age was 14.5 year old. A positive family history was present in 46% of the cases. In 51% of the cases, the patient's weight was greater than the 75th percentile for their given age. None of our patients had associated hemolytic disease. Only two patients received TPN. Of the 5 male patients, 4 were atypical in their presentation; such as congenital choledochal cyst, development of necrotizing enterocolitis in the newborn period, necessitating TPN, an acaulous cholecystitis, and the last had a strong family history in male members of the family.

From our study we conclude that our group of patients, although in the pediatric group, is not unlike the adult population, in that there is a female preponderance, with obesity and positive family history as predisposing factors. Blood dyscrasia does not contribute to cholelithiasis in our region. We have only two patients with history of long term TPN, in one of these patients the cholelithiasis was discovered incidentally. We postulate that, even though TPN may produce cholestasis, the incidence of symptomatic cholelithiasis may be low.

4 OCCURRENCE OF CONTRALATERAL INGUINAL HERNIAS FOLLOWING UNILATERAL REPAIR IN A PEDIATRIC HOSPITAL
J. Given, MD and S.Z. Rubin, MB, FRCS(C), FRCS(E), FAAP

Children's Hospital of Eastern Ontario, Ottawa, Ontario

The records of 1,050 children undergoing unilateral inguinal hernia repair at the Children's Hospital of Eastern Ontario during the period from 1985 to 1988 were reviewed. 94.6% had unilateral hernias only. Of these 89.6% were boys and 10.4% were girls. Right-sided hernias were twice as common as left in both males and females.

57 contralateral inguinal hernias were repaired following previous unilateral repair. The age of presentation of the first inguinal hernia ranged from 1 week to 11 years (43% were less than 12 months). 51 were boys and 6 were girls. Left hernias after contralateral repair were seen in 26 boys and 4 girls (4.4% and 4.5% respectively). After left hernia repair, right hernias occurred in 25 boys and 2 girls (8.1% and 5% respectively). Overall incidence of contralateral hernia diagnosis was 5.5% in boys and 4.9% in girls. The mean delay time until presentation of contralateral hernia was 27.5 months.

The low incidence of contralateral hernia does not warrant routine bilateral exploration. An initial left hernia did not increase the risk of a contralateral hernia. Bilateral exploration is not routinely needed in girls. In the first year of life contralateral exploration may be indicated.
UNUSUAL PATTERNS OF CONGENITAL NECK MASSES IN CHILDREN. RE Sonnino, MD, N Spigland, MD, FM Guttman, MD, J DesJardins, MD, J-M Laberge, MD, CR Moir, MD, LT Nguyen, MD. The Montreal Children's Hospital and Ste. Justine Hospital, Montreal, Quebec, CANADA H3H 1P3

Congenital neck masses (Thyroglossal Duct Cysts-TDC, Branchial Cleft Cysts-BCC) are frequently encountered in any pediatric surgical practice. While their diagnosis is usually straightforward, unusual or combined presentations may occur. We report eleven cases of unusual patterns of congenital neck masses in children. In one patient, a lateral neck mass (presumed to be a BCC) was found at surgery to be a TDC. Two patients underwent resection of a BCC. Both subsequently presented with a new mass near the previous scar. These were initially thought to represent recurrent BCC but were found at surgery to be consistent with thyroglossal duct cysts with sinus tracts extending through the hyoid bone. In all cases the diagnoses were confirmed histologically. Eight patients presented with a solitary thyroid nodule. Six of these had intrathyroid branchial cleft remnants and two had intrathyroid TDC. The diagnosis became apparent at operation in six patients, while in two it was made by the pathologist. Ages at presentation ranged from 3 to 14 years. The embryology of these neck structures is closely related and errors in their development may occur. The possibility of an embryologic rest in the neck should therefore be kept in mind with all clinically evident neck masses. TDC & BCC may coexist in the same patient. The histologic differentiation may be difficult in the presence of inflammation, but differences in structure may be characteristic. While it may not be possible to distinguish the intrathyroid lesions from primary thyroid masses pre-operatively, it may be possible to limit the extent of resection once the diagnosis is reached. Such knowledge may assist the surgeon at the time of operation.

THE LATE PRESENTING PEDIATRIC MORGAGNI HERNIA - A BENIGN CONDITION

Lawrence Berman; David Stringer; Sigmund H. Ein, FAAP; Barry Shandling, FAAP.

Hospital for Sick Children, Toronto, Ontario, Canada, MSS 1X8

Fifteen infants and children with late presenting (more than eight weeks) Morgagni hernias over the last 20 years (1966-1986) have been reviewed. Ten were clinically normal on presentation and only one child presented with acute symptoms. Five had previously normal chest x-rays and two others had their initial radiological assessment made incorrectly. Preoperative barium studies were performed in three patients. Twelve had other major congenital abnormalities. Fourteen of the 15 were operated on, usually within days of presentation. At operation, 10 of the 14 hernias contained a hollow viscus, nine had a sac and four had abnormal bowel fixation. Postoperatively, two children had radiological evidence of impaired diaphragmatic motility. There was no mortality in this series.
7 THE USE OF THE UMBILICAL SITE FOR TEMPORARY OSTOMY: REVIEW OF 47 CASES

P.G. Fitzgerald, G.S. Cameron, and G.Y. Lau
Department of Surgery, McMaster University, Hamilton, Ontario

In 1982, we described a technique for umbilical ostomy in neonates. We have continued to use this technique and modified it for use in older children. Our total experience now includes 47 patients (33 neonates, 10 infants and 4 older children) who needed temporary ostomies for Hirschsprung's Disease (21), imperforate anus (9), necrotising enterocolitis (4), meconium ileus (2), intestinal perforation (2), trauma (2), chronic megacolon (2), short gut syndrome (1), and pseudo-obstruction (1). Initially the loss of the normal umbilicus caused some concern, but parents, children and nurses were all assured that the "belly button" was not gone forever! The wide area available around the centrally placed stoma greatly facilitated the application of ostomy appliances. Complications were frequent, but not specifically related to the site of the ostomy. Eighteen patients had 21 local complications consisting of infection (2), prolapse (7), retraction (7), infarction (2), fistula (2), and obstruction at the stoma (1). Three neonatal patients died of sepsis. Surgical revision was required in 4 cases for prolapse and in 3 for retraction. Thirty-three patients have now had their stomas closed for 8 months to 8 years. There have been no late complications related to the use of the umbilical site. In each case the ostomy closure scar resembles a relatively normal umbilicus. We recommend use of the umbilical site for temporary ostomy because it is a convenient location for placement of appliances and ultimately, after closure, the scar is cosmetically superior to that of an ostomy placed elsewhere.

8 ADNEXAL TORSION IN CHILDREN. N. Spigland, S. Yazbeck, J.C. Ducharme. Hôpital Sainte-Justine, Montreal, Quebec.

Adnexal torsion is rare in children and is usually reported as small series or case reports. We reviewed a series of 18 consecutive cases, ages 3-19 years (mean 9.3 years) treated in our institution between 1977 and 1987. Twelve patients presented with torsion of a previously normal adnexa, while 6 presented with torsion of a diseased adnexa. Eighty-three percent of cases involved the right adnexa. De-torsion with recovery of vascularisation of the adnexa was possible in only 4 cases. All patients presented with lower abdominal pain, onset was sudden in 71% of cases with an average of 5.3 days between the first symptom and hospital admission and a mean delay of 31.8 hours between consultation and surgical intervention. A previous history of abdominal pain was present in 8 cases. Nausea or vomiting were present in 88% of cases. An abdominal mass was palpable in 50% of the patients and was associated with a delay in surgical intervention. Ultrasound confirmed the presence of a mass in 93% of cases. The pre-operative diagnosis was accurate in 38% of cases and the most common inaccurate diagnosis was appendicitis or appendiceal abscess. Our series confirms the right sided predominance of these lesions as reported in the literature. It is not clear whether this is an anatomical phenomenon or whether these torsions are more frequently diagnosed because of the suspicion of appendicitis and that indeed many left adnexal torsions are being missed. We therefore advocate pelvic ultrasound in female patients who present with left lower quadrant pain. Finally, early recognition of this clinical entity and accurate pre-operative diagnosis may reduce the number of unnecessary lengthy pre-operative workups and lead to more frequent salvage of torsioned adnexa.
AN EVALUATION OF SURGICAL EXCISION AND INTRALESIONAL OK-432 THERAPY OF LYMPHANGIOMA IN CHILDREN
S.Ogita, E. Doguchi, K. Tokiwa, T. Tsuko
Division of Surgery, Children's Research Hospital, Kyoto Prefectural University of Medicine, Kyoto

The results of treatment for 37 cases of lymphangioma were reviewed. Total or partial excision of the lesions was performed in 11 patients before 1985 (Group I), and after 1986, intralesional OK-432 therapy was performed in 26 patients (Group II). For OK-432 therapy, 0.1 mg of OK-432 [Picibanil (group A Streptococcus pyogenes of human origin); Chugai Pharmaceutical Co., Tokyo] was dissolved in 10 ml of physiological saline, and this OK-432 solution was administered into the lesion at a few sites. The doses of OK-432 depended on the size of the lesion, but did not exceed 0.2 mg of OK-432 at one injection.

In Group I, total excision was possible in 4 of the 11 cases, and resulted in cure. Partial and/or repeated excision was performed in the remaining 7 because of the involvement of the surrounding tissue and wide extension of the lesions, and the results were poor (recurrence in 5 and ugly scars in 2). In Group II, the lesions completely disappeared after 2-3 months in 20 of the 26 patients, and marked regression was noted in the remaining 6. No recurrence of the lesions was noted thereafter (4-25 months). The side effects of OK-432 therapy were fever of 2-3 days' duration and local inflammatory reaction lasting for 3-5 days. However, the local inflammation did not cause any damage to the overlying skin and did not lead to scar formation. The results suggest that intralesional OK-432 therapy is effective for these lesions.


Since 1978, 6 boys with prostatic rhabdomyosarcoma were treated. The tumor was staged by modern techniques including extensive use of CT scanning and ultrasonography. Tissue diagnosis was established mainly by needle biopsy. At presentation, 3 boys (50%) were under 7 years of age and had localized disease with favorable histology. The other 3 (50%) were older than 7, and had metastatic disease with unfavorable histology. The treatment regimen after initial biopsy consisted of 2-4 courses of "pulse" VAC (Vincristine, Actinomycin D and Cyclophosphamide) bladder-sparing surgery with or without irradiation; further chemotherapy for at least 18 months in the 3 patients with localized disease. Two boys had microscopic residual disease undetected by frozen section and unresponsive to early irradiation (3600 and 4140 rads respectively). They required total cystectomy 4 and 7 months later for cure. In one patient, preservation of the bladder was achieved at age 2 months for 8 years. Artificial sphincter inserted to cure his incontinence failed because of ischemia secondary to cuff compression and scar tissue. He is alive with a continent ileocoecccygoplasty. Of the 50% who had metastatic disease, 2 were dead within 12 months despite aggressive chemotherapy and irradiation. The third is currently on treatment. Although chemotherapy has markedly improved the prognosis, surgery is still necessary, in most cases for cure. Bladder salvage is a desirable goal, however, residual microscopic disease, difficulty with frozen section disease detection, submucosal disease infiltration, and poor tissue vascularization for artificial sphincter compression remain significant obstacles. Metastatic disease retains a poor prognosis.
MAJOR PERINEAL TRAUMA IN CHILDREN
O. REINBERG AND S. YABECK
Service de chirurgie pédiatrique, Hôpital Sainte-Justine, Montréal, Quebec

Between 1976 and 1987, 22 children ages 3 to 17 years (mean 9.5) were admitted to our institution for major perineal trauma. There were 14 urethral ruptures and 9 rectal lesions. The lesions were multiple and combined vaginal and urethral or vaginal and rectal injuries were found in 10 females while only one out of 12 males had both urethral and rectal lesions. In most instances perineal physical signs did not reflect the severity of the lesion.

Among the patients with urethral lesions there was no sex or age predominance, and complete rupture occurred in 4 females and 8 males. The insertion of a urethral catheter is of no diagnostic help. Retrograde urethrogram demonstrated a lesion in 7 out of 8 cases. The IVP showed evidence of extravasation in 2 cases out of 8 and in 4 instances there were indirect signs of a lesion. A supra-pubic cystostomy was performed in 12 out of 14 urethral injuries. A primary repair with urethral stenting was carried out twice without bladder drainage. Immediate repair was performed in 8 cases and delayed in 5 others. Within this last group 2 were cured without sequelae by stenting without suture, while the 3 others who underwent only cystostomy necessitated multiple reconstructive operations with poor results. It is of interest that the four treatment failures occurred in the youngest patients (4 to 7 yrs).

The rectal lesions were related to impalement in 7 out of 9 patients. Rectoscopy allowed localization of perforating lesions but did not permit identification of sphincter injuries when the mucosa was intact. Primary repair was possible in all cases. Diverting colostomy was felt necessary in 6 out of 9 cases because of the severity of the lesion even though the pre-operative delay was short. Continuity was reestablished 4 months later. 8 out of 9 patients are cured and continent and one patient still has his colostomy. This series underlines the frequent association of multiple lesions in children with major perineal trauma as well as the necessity of extensive investigations in these cases. Immediate or delayed primary repair should always be attempted in both urethral and rectal injuries if the condition of the child allows it.

FAILURE OF NIHSEN'S FUNDOPPLICATION TO CONTROL GASTROESOPHAGEAL REFUX (GER) IN ESOPHAGEAL ATRESIA PATIENTS
Harry Lindahl
Risto Rintala, Ilmo Louhimo
Helsinki University Central Hospital, Children's Hospital

During 1981 - 1985 Nissen's fundoplication was performed on 13 esophageal atresia patients. One patient with Down's syndrome died because of his cardiac malformation two weeks after the operation and is excluded from the analysis. Of the 12 patients 9 had distal fistula while 3 had isolated atresia. In 8 patients there was a long gap between segments and in 5 Livaditis myotomy was necessary. The median age at the fundoplication was 1.3 years (range 4.5 mo - 10.6 yrs). The main indications were: anastomotic stricture (5), distal oesophagitis (5) and respiratory complications (2). Mean follow-up was 4.1 years (range 2.0 - 6.4 yrs). All patients benefited from the operation. Routine control endoscopy 3 to 8 months after the operation showed a competent fundoplication in all patients. However, in 5 the reflux later recurred and endoscopy revealed a partially disrupted incompetent fundal wrap and oesophagitis. Four patients had Barrett's esophagus at last endoscopic control. There was one late death due to cardiac failure. In conclusion, although the short-term results of Nissen's fundoplication in esophageal atresia patients are good, the risk for late recurrence is high. Regular long-term follow-up is therefore necessary.
GRACILIS MUSCLE TRANSPOSITION FOR ANAL INCONTINENCE IN CHILDREN: LONG TERM CLINICAL AND MANOMETRICAL STUDY.

O. REINBERG, A.L. BENSOUSSAN, ET H. BLANCHARD

Service de chirurgie pediatrique. Hopital Sainte-Justine, Montreal, Quebec

The transposition of the gracilis muscle for the creation of a new anal sphincter in cases of fecal incontinence has been described by Pickrell and many other reports have followed. But very few long term manometrical evaluations in children have been published. 5 patients ages 10 to 15 years old (mean 12.8) have been treated for uncontrollable fecal incontinence since 1976: 3 imperforate anus after multiple surgical interventions, 1 low myelomeningocele with bi-sphincteric incontinence and 1 traumatic destruction of the sphincter apparatus.

Pickrell's procedure was followed as initially described excepted that which concerns the perianal placement. The gracilis was transposed sub-cutaneously without constructing the pubo-anal mechanism through the median raphe. Anchoring to the ischion was performed with special care.

All patients were evaluated by ano-rectal manometries pre-operatively and followed for a period of 1 to 12 1/2 years (mean 5.5) in the post-operative period. All patients are continent with normal defecations, without enema. There is no evidence of fibrosis of the muscle or anal canal. The tension of the transposed muscle is maintained and the voluntary contractions are still efficient.

The age at intervention was an important factor. We believe that the hope of ameliorating their status is a powerful stimulant for the children to participate in their active reeducation pre-operatively and in passive post-operative electric stimulations starting on day 4 or 5. Active reeducation is performed gradually.

Pickrell's operation remains a satisfactory long term alternative for total fecal incontinence when all other therapeutic possibilities related to etiology have been exhausted.

14 BRONCOGENIC CYSTS. M. DiLorenzo, P.-P. Collin, R. Vaillancourt, A. Duranceau, Hopital Ste-Justine, Montreal, Quebec, Canada.

Between 1967 and 1987, 26 patients with bronchogenic cyst were treated in our institution: 15 females and 11 males, ranging in age from 2 mo. to 14 yrs (average 6 yrs, 4 mo.). Only 4 patients (15%) were 1 yr old or less. Seven patients (27%) presented with asymptomatic lesions discovered on chest film. Nineteen patients (73%) presented with respiratory symptoms, 47% of which (9/19) were infectious. Of note are the patients aged 1 yr or less, who presented with dyspnea and/or cyanotic spells. Only 1 of them presented with dysphagia. The interval between the onset of symptoms and diagnosis ranged from no delay to 9 yrs (average 12.2 mo.). All patients underwent routine chest films and tomography and later CT-scan of the thorax once the study became available. Barium esophagagram and bronchoscopy were done as required. Contrary to what has been published concerning the intraparenchymal location of bronchogenic cysts, the majority of them (65%) were located in the mediastinum, usually in close proximity to the carina, main stem bronchi, trachea, esophagus or pericardium. Only 27% (7/26) were intraparenchymal, and 8% (2/26) were situated in the inferior pulmonary ligament. Sixteen of the 26 cysts (62%) were right-sided. A correct preoperative diagnosis was made in 20 patients (77%). Intrapulmonary lesions were removed by segmentectomy or lobectomy. Cysts located elsewhere were simply excised. Among the cysts adhering to the trachea, 50% (2/4) required suture of the membranous trachea after excision. Postoperative complications include 1 pneumonia which responded to antibiotics and 1 transient paresis of the right phrenic nerve. Follow-up averaged 17 mo (range 0-17 yrs). No sequelae were noted and all patients were cured.
ETIOLOGY OF BOWEL DAMAGE IN GASTROCHISIS: STUDIES IN THE FETAL LAMB

Department of Surgery, University of California, San Francisco

Gastrochisis is often complicated by disturbances in small bowel motility and absorption. To study the etiology of this bowel damage, a standardized abdominal wall defect was created. Group 1 was replaced in the uterus, and Group 2 had constricitive tape placed at the base of the extruded bowel. Control fetuses had a small piece of silastic inserted, with no abdominal wall defect.

Results: Three lambs in each group went to term. Groups 1 and 2 had herniated bowel with a fibrous peel. Histologically there was no ischemia and ganglion cells were normal. Differences between groups are summarized:

<table>
<thead>
<tr>
<th>Muscular villi</th>
<th>Group 1</th>
<th>Group 2</th>
<th>Group 3</th>
</tr>
</thead>
<tbody>
<tr>
<td>Micraniel villi</td>
<td>Normal</td>
<td>Focal Blunting</td>
<td>Normal</td>
</tr>
<tr>
<td>Muscle thickness (mm)</td>
<td>0.20±0.07</td>
<td>0.34±0.20*</td>
<td>0.18±0.06</td>
</tr>
<tr>
<td>Circular</td>
<td>0.10±0.06</td>
<td>0.17±0.20*</td>
<td>0.09±0.04</td>
</tr>
<tr>
<td>Longitudinal</td>
<td>18.8±8.2</td>
<td>12.0±3.2</td>
<td>27.0±8.78</td>
</tr>
<tr>
<td>Max. Motility response to acetylcholine (mm)</td>
<td>*p&lt;0.01 vs control</td>
<td>**p&lt;0.05 vs control</td>
<td></td>
</tr>
</tbody>
</table>

Conclusions: (1) The fetal lamb model is reproducible with findings similar to the human condition: (2) The fibrous peel is caused by exposure to amniotic fluid; (3) intestinal damage is not caused by amniotic fluid exposure alone, but appears to require constriction of the bowel.

Comment

A STANDARD OF COMPARISON FOR ACUTE SURGICAL NECROTIZING ENTEROCOLITIS

Mitchell N. Ross, Eli R. Wayne, Joseph S. Janik*, John D. Burrington, and Jack H.T. Chang
The Children's Hospital, Denver, Colorado

Nearly two decades have passed since the first reports on NEC and its treatment. Though reports abound, there still exists some controversy as to the appropriate surgical indications and treatment of NEC. Between 1976 and 1988, 181 neonates underwent treatment for NEC, of which 106 underwent laparotomy for acute surgical complications. A uniform protocol of operative indications was employed. They consisted of: free air, localized mass, single dilated bowel loop on sequential x-rays, abdominal wall erythema, portal air, and clinical deterioration, singly or in any combination. Clinical deterioration was defined as falling platelet count, rising WBC, rising bandemia with falling or rising WBC, persistently low pH or falling pH, and increasing frequency of apnea or bradycardia.

One hundred and six neonates underwent laparotomy following the above indications. All had histologic confirmation of NEC. Ninety had localized disease, sixteen had diffuse. All of the former had resection and diverting enterostomy, all of the latter died. Thirty-eight neonates developed colonic strictures, nineteen of whom did not have acute surgical intervention, all had resection and successful reconstruction of the GI tract.

Employing the above indications and surgical principles resulted in a high degree of diagnostic accuracy and a low degree of surgical morbidity. This is the safe standard of comparison by which all other novel surgical approaches to NEC should be judged.
RECTAL INVOLVEMENT IN NECROTIZING ENTERO-COLITIS (NEC)

S.Z. Rubin, MB, FRCS(C), FRCS(E), FAAP and A. Alawadi, MD
Children's Hospital of Eastern Ontario, Ottawa, Ontario

Although NEC is the commonest cause of neonatal intestinal disease, distal rectal involvement has not previously been reported. We describe 5 children with NEC involving the rectum. In 4 infants, pancolorectal disease occurred; in the 5th patient the ascending colon was spared. A Swenson-type abdomino-perineal resection was performed in 4 children. No definitive cause for the extension of the disease into the distal rectum was noted. There were no immediate post-operative complications. A regular stooling pattern was restored, using a combination of high-fibre diet and loperamide hydrochloride. Integrity of the perianal skin was maintained by regular cleansing with an antacid mixture and application of a barrier paste comprising kerosene, zinc oxide and mineral oil. Long-term follow-up, including ano-rectal manometry, showed normal ano-rectal function and a normal appearing perineum.

Possible rectal involvement should be considered in all neonates with extensive NEC. Contrast studies visualizing the rectum are advised prior to closing colostomies performed in newborns with NEC. Abdomino-perineal resection produces a satisfactory result.

TOPICAL AND SYSTEMIC ANTIBIOTICS IN THE PREVENTION OF WOUND INFECTION.
G. Stirling, M.D., M. Savrich, J. Horton, Ph.D., E. Bawden, Ph.D.
U. of Tx. Southwestern Medical School, Dallas, Texas.

We evaluated topical and systemic antibiotics (Ab.) alone and in combination (T/S) in the prevention of wound infection and measured serum and tissue Ab. levels in the wound and distant tissue. Ab. levels were measured at 2, 4 and 6 hrs. after administration. Sprague-Dawley rats were contaminated on the paravertebral muscles with a standardized inoculum of E.coli, B.fragilis, and Staph.aureus. Systemic administration consisted of a single i.m. dose of a second generation cephalosporin given 30 min. before contamination. Topical administration consisted of a single dose of the same Ab. applied to the wound 30 min. after contamination. Wound infection was confirmed by the recovery of the organism by culture. Statistical analysis was done by a Tukey-type pairwise multiple comparison procedure and repeated measures ANOVA; significance level was 0.05. Infection rate cultures follow:

<table>
<thead>
<tr>
<th>Antibiotic</th>
<th>Staph.aureus</th>
<th>B.fragilis</th>
<th>E.coli</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>20/20</td>
<td>11/11</td>
<td>17/20</td>
</tr>
<tr>
<td>Topical</td>
<td>1/20</td>
<td>1/20</td>
<td>0/20</td>
</tr>
<tr>
<td>Systemic</td>
<td>13/20</td>
<td>1/20</td>
<td>2/20</td>
</tr>
<tr>
<td>Topical/Systemic</td>
<td>2/20</td>
<td>5/20</td>
<td>0/20</td>
</tr>
</tbody>
</table>

Therapeutic levels of antibiotic were achieved in serum and tissue with both Topical and Systemic Ab. Wound tissue concentration of Ab. was significantly higher (p<.05) when topical Ab. was used. Prophylactic Ab. was effective in preventing wound infections in all groups (p<.05). Topical Ab. was significantly better than systemic Ab. in preventing wound infection with E.coli and Staph.aureus (p<.05) and as effective for B.fragilis. Topical antibiotic prevents wound infection.

Forty-six neonates were treated for necrotizing enterocolitis between 1982-87. The mean gestational age was 33.6 wks and the mean birthweight was 1865 grms. Birthweight < 1500 grms and gestational age < 32 wks had an adverse effect on survival. Neonates < 30 wks gestation developed symptoms at a mean age of 10.8 days while those > 30 wks developed symptoms at a mean age of 1.7 days. All infants who never had enteral feedings survived. Twenty patients were cured with medical treatment only. Three patients underwent peritoneal lavage (2 of these survived). Twenty-two patients necessitated surgery. Operative mortality was 9%. Four patients underwent elective surgery for primary strictures. The remaining 18 patients presented with acute perforation (11), obstruction (5), deterioration with medical therapy (1) or peritonitis (1). The most frequent site of perforation was the terminal ileum while the most common site of stricture was the left colon. Seven patients were found to have strictures after prior emergency surgery. In 6 of the patients who underwent primary enterostomy, the strictures were located in the defunctionalized bowel segment. Evidence of progression of the disease to involve most frequently right and transverse colon, necessitated extensive resection. When performing a proximal ileostomy for acute NEC the surgeon must be aware that the chances of conservation of the defunctionalized distal bowel segment are minimal because of progression of the disease. Complications related to resection of the ileocecal valve have been negligible. Our results with peritoneal lavage are encouraging, and our overall mortality rate of 13% in contrast with higher rates in earlier series reflects improvement in management of these neonates.

Idiopathic Intestinal Perforations in the Newborn: An Increasingly Common Entity?
Gerard Weinberg, M.D., Sylvain Kleinhaus, M.D. and Scott J. Boley, M.D.
Montefiore Medical Center-Albert Einstein College of Medicine, N.Y.

Over the past five years we have seen a sudden increase in spontaneous idiopathic perforations of the G.I. tract in newborns. Between 1982 and 1987 seven neonates ranging in age from 24 hrs to 2 weeks were treated at the MMC/AECOM for such perforations. Four of the infants were born premature; three were full-term. Five were being treated in an intensive care nursery when the perforation was diagnosed. All infants whose perforations were due to necrotizing enterocolitis, appendicitis, Hirschsprung's disease, meconium ileus, intestinal atresias, or drug therapy, are excluded from the series. None of the infants had associated anomalies. The sites of the perforations were as follows: jejunum 2, ileum 2, cecum 1 and transverse colon 2. Six of the perforations were on the antimesenteric aspect of the bowel; one was on the mesenteric aspect. The size of the perforations ranged from pinhole to 1 cm in diameter. All the infants did well. Pathologic examination of the resected specimens failed to reveal an etiology for the perforation. There were no cases of duplications or muscular hypoplasia. We believe the etiology of this condition may be ischemic necrosis, secondary to a localized vascular accident in the wall of the affected viscus but do not have a good explanation for the upsurge in cases we are seeing. It would be important to determine if other centers are experiencing a similar phenomenon.
The effect of Cis-platinum on Liver Regeneration in the Rat

D. May, C. Vinden, G. Blair, E. Blacklock, P. Rogers, G. Taylor

British Columbia Children's Hospital, Vancouver, B.C., Canada

Surgical excision remains the mainstay of therapy for childhood hepatoblastoma. Post-operative adjuvant chemotherapy is also employed but some of the key agents such as Adriamycin have a detrimental effect on liver regeneration, and are generally withheld until liver regrowth has occurred. During this period, remaining tumour could potentially grow and metastasize. This experiment was undertaken to study the effect of cis-platinum on liver regeneration. Male Sprague-Dawley rats underwent standard two-thirds hepatectomy and were then randomized either to receive intravenous cis-platinum (3mg/kg) or to act as controls. They were sacrificed at 1, 2 and 5 days after surgery and were compared on the basis of liver weight and mitotic indices. Mean mitotic indices peaked on the second postoperative day in both experimental and control groups. There were no significant differences between experimental and control groups with regard to liver weights and mitotic indices in any of the survival groups. We postulate that that there is no deleterious effect of cis-platinum on liver regeneration in the partially hepatectomized rat.

EXPERIENCE EN TRANSPLANTATION HEPATIQUE PEDIATRIQUE A MONTREAL


Hôpital Sainte-Justine et Hôpital de Montréal pour Enfants, Montreal, Quebec, Canada.

Un programme conjoint de transplantation hépatique pédiatrique a été inauguré à Montréal en 1985. Il s'agit d'un programme multidisciplinaire qui fonctionne selon le modèle de la Nouvelle Angleterre.

Depuis 1985, 9 transplantations hépatiques ont été faites chez 3 garçons et 4 filles, âgés de 13 mois à 5 ans. Les indications de la transplantation ont été en 5 fois échec primaire ou secondaire d'opération de Kasai, 1 fois hépatite fulminante et 1 cas de Crigler Najjar type I. Quatre patients sont vivants 30 à 14 mois post-transplantation. Trois patients sont décédés à 24 heures (mort cérébrale), 2 semaines (non fonctionnement du foie) et à 6 semaines (rupture anévrysmale manchon aortique du donneur). L'indication de retransplantation chez 2 patients encore vivants a été une nécrose hépatique consécutive à une thrombose artérielle. Deux patients sont décédés en attente de transplantation.

Les différents problèmes en rapport avec la collecte des organes, la réanimation opératoire et le postopératoire seront présentés.
Liver Transplantation (LT) in a High Risk Group of Children

PA Superina, RH Pearl, EA Roberts, N Graham, PD Greig, B Langer
Department of Surgery, Hospital for Sick Children, Toronto

Children who weigh less than 10 kg, those with extra-hepatic biliary atresia (EHBA), and those with severe coagulopathy or severe growth retardation have a higher postoperative mortality following LT.

In the 1st year of our LT program, 13 transplants have been done in 11 children. Six children had EHBA, 3 fulminant hepatitis with coagulopathy and encephalopathy, 1 tyrosinemia and 1 Indian childhood cirrhosis. The mean age and weight of the six with EHBA at transplant was 2.1 years and 9.4 kg. Four of the 6 had portal vein atresia and required a portal vein (PV) reconstruction or free interposition vein graft. Those without EHBA were older and larger but usually required transplantation more urgently.

Survival 1 month after LT was 82%. Two children required early retransplantation and both survived. Early deaths included 1 patient in hepatic coma pre-LT who was declared brain dead after successful transplantation, and one with primary non-function after prolonged ischemia during PV reconstruction. Two late deaths occurred in cytomegalovirus (CMV) seronegative recipients who died from CMV pneumonia following LT with livers from CMV+ donors. Another late death occurred from fulminant rejection six weeks following LT.

Our early experience suggests that the incidence of PV atresia in small children with EHBA is high but that the 75% early survival warrants transplantation. We have also adopted a policy of using only CMV-donors for stable children who are at risk for primary CMV infection.

Orthotopic Liver Transplantation in Children with Polysplenia Syndrome with Absent Inferior Vena Cava

M.A. Hoffman, S. Cell, P. Nakou, K. Balles, K.Y. Calne, Dept. of Surgery, Univ. of Cambridge, Cambridge, U.K.

Liver atresia is the most common indication for pediatric orthotopic liver transplantation. Co-existing developmental defects are noted in 10% of children with biliary atresia, and may involve a constellation of anomalies which includes polysplenia, the constant feature of the syndrome, plus several inconsistently associated disorders including intra-abdominal venous abnormalities, mid-gut malrotation, situs inversus, symmetric liver, and bilobed right lung.

The Cambridge/King's College Hospital Liver Transplantation Program has performed 31 liver transplants for biliary atresia in the past 11 years, including 7 patients with failed Kasai portoenterostomies and polysplenia syndrome with absent suprarenal infrahepatic inferior vena cava. The first patient had polysplenia and absent IVC only. The second patient also had a pre-duodenal portal vein, mid-gut malrotation, and anomalous origin of the hepatic artery. The major technical modification was oversewing of the infrahepatic IVC of the liver graft, thus eliminating one vascular anastomosis.

Both patients died in the early postoperative period. The first from infrahepatic thrombosis of the liver with patent vascular anastomoses at postmortem in the third postoperative week. The second died on the sixth postoperative day from hepatic arterial and portal venous thrombosis confirmed at autopsy.

We do not feel, as stated by others, that these anomalies are a contraindication to orthotopic liver transplantation. The anomalies and technical adjustments involved in the operation are illustrated.
This presentation evaluates a compulsory pediatric surgical rotation during the final year of undergraduate medical training. On rotation, two to four students spend an intensive two-week period attached to the Pediatric Surgical service as part of an overall 10-12 week pediatric program at the Children's Hospital of Eastern Ontario (CHEO). The pediatric surgical undergraduate teaching program was evaluated by testing the knowledge of the students at the beginning and end of their stay at CHEO (multiple choice examination MCE). Students were specifically instructed not to identify their own answer sheet. At the end of the rotation, the students anonymously filled in a questionnaire assessing the PST. The MCE results improved from an average of 27% pre-rotation to 50% post-rotation. 80% of students felt the rotation was good, and 2/3 of those replies indicated that the PST was better than expected. Although 3% were disappointed with resident teaching, all students felt the rotation was worthwhile. Pediatric surgery is a clinical specialty relying predominantly on history and physical examination for diagnosis. Treatment is highly specific. These factors may explain the students' satisfaction and indicate the value of a compulsory pediatric surgical rotation.

The repair of high and intermediate forms of rectal stenosis should, logically, preserve and utilize all structures that may contribute to continence. A technique is described which, in addition to definition of the puborectalis sling by the anterior perineal approach, seeks to integrate all infralevator structures as well. Procococutaneous skin is tubulated and inserted within the exposed external sphincter fibers. The full length of the blind rectum is preserved to save any internal sphincter component that may exist. The anastomosis between the skin tube and the bowel is sutured to the puborectalis. In a series of 15 cases with a follow-up of up to 5 years, only one case of mucosal extrusion was seen. Functional results appear to be improved but require more long-term evaluation.
DUODENAL ATRESIA AND ITS ANTENATAL DIAGNOSIS

B. J. Hancock, M.D., N. E. Wiseman, M.D.
Department of Paediatric General Surgery
Children's Hospital, Winnipeg, Manitoba

In a series of 31 infants with congenital duodenal atresia there were 14 in whom the diagnosis was established antenatally with the use of fetal ultrasound. One neonate died of severe associated anomalies and the remaining 13 patients were available for follow-up. Parents were contacted and a questionnaire was filled out. The indication for fetal ultrasound was: polyhydramnios (7), gestational age confirmation (2), premature rupture of membranes (3), and large for dates fetus (1). Diagnoses were established between 21 weeks gestation and term. A physician was present during the ultrasound in 11 cases. In 9 cases parents were informed of the antenatal diagnosis of bowel obstruction at or shortly following the ultrasound and in 8 cases they were advised of the possibility of surgery in the neonatal period. The diagnosis and potential surgical management were discussed by the perinatologist or the obstetrician in each instance. In 9 cases parents felt the information provided to them by the physician regarding the diagnosis was satisfactory. They also felt that the information helped them cope with the post natal care of their infant. In the total group of 31 infants with duodenal atresia 29 had surgical repair with a 93% survival. Four deaths were due to associated congenital anomalies. With antenatal diagnosis surgery was seen to occur at an earlier age in the neonatal period. We conclude that the antenatal diagnosis of duodenal atresia influences parents positively in coping with the anomaly and influences the approach to and the timing of surgical intervention.

CONGENITAL DIAPHRAGMATIC HERNIA AND PRENATAL DIAGNOSIS (50 CASES).

The authors present 50 cases of congenital diaphragmatic hernia (CDH) prenatally diagnosed. All these newborns were treated at Saint Vincent de Paul Hospital (Paris) between 1980 and 1988.

The results suggest that an early prenatal diagnosis (before 30 weeks) is associated with a poor prognosis. Anatomical and clinical data underline the necessity to look for specific factors of poor prognosis.

So it is possible to separate during the gestation the different forms of CDH. Those in which the defect is large enough to permit the intrathoracic development of abdominal viscera, and on the other hand, the CDH with a posterolateral defect and a later compression of the lung.

So, in specific cases, with very precise indications it is possible to discuss "in utero-surgery".

This approach would also permit to better determine the place of extracorporeal membrane oxygenation postoperatively.
PRETERM BABIES CAN HAVE HIRSCHSPRUNG'S DISEASE.

O. REINBERG, S. YAZBECK ET H. BARD
Service de chirurgie pédiatrique. Hôpital Sainte-Justine, Montréal, Québec

"Hirschsprung's disease is usually a solitary anomaly in a full term, otherwise healthy infant" (Wk Sieber, in Pediatric Surgery, YBMP, 1986)

Only 6 cases of Hirschsprung's disease in preterm babies have been reported in the past 20 years.

Between 1980 and 1988, 29 cases of Hirschsprung's disease were diagnosed in neonates in our institution. Among them, 7 were preterm babies, born before 38 weeks gestation (mean 35.4 w). There were 4 males and 3 females weighing between 1285 and 3200 g (mean 2629 g).

Their symptoms were those common to full term babies presenting with Hirschsprung's disease: vomiting was always present and bowel distension was common. There was delayed passage of meconium at a mean age of 44.4 hours. A meconium plug syndrome was suspected twice delaying the correct diagnosis. Flat plate of the abdomen done on day 1 always showed at least dilated bowel loops; barium enema performed at a mean age of 2.3 days demonstrated the typical aspect of Hirschsprung's disease with a transitional zone in 5 out of 7 cases. In 2 cases the diagnosis was that of meconium plug syndrome. Suction biopsy was performed 5 times at a mean age of 7 days and was always diagnostic.

6 patients underwent a colostomy at a mean age of 8.8 days and 1 patient with Down's syndrome was not treated. The definitive procedure (4 Swenson and 2 Somme) was performed at a mean age of 9.6 months and the colostomy was closed either at that time or 2 months later. Pathology confirmed the diagnosis in all cases with a transition zone located at the recto-sigmoid junction in 6 cases and at the splenic flexure in the 7th.

This series demonstrates that Hirschsprung's disease may be observed in preterm babies. Its clinical presentation and management are similar to those of full term babies with Hirschsprung's disease.

MANAGEMENT OF HIRSCHSPRUNG DISEASE : CURATIVE SURGERY BELOW THREE MONTHS OF AGE

H. CARRASONE - J.M. GUIZ - G. HUSSON-LACROIX - B. KEITZMAN
Clinique Chirurgicale Infantile et Orthopédie - Pr H. CARRASONE
C.H.U. Timone-Enfants - 1, boulevard J. Moulin 13005 MARSEILLE F.

Curative surgery of Hirschsprung disease (HD) was performed in 32 infants below three months's old, from 01.01.77 to 12.31.86. There were 25 male and 5 total colonic aganglionosis. Pre-operative relief of obstruction was carried out by enemas in 26 patients (81%), with addition of total parental nutrition from 8 to 27 days in case of severe enterocolitis. Colostomy was never performed as a routine procedure. Pre-operative morbidity is reported in 3 cases : 1 enterocolitis, 2 sigmoid perforation. successfully cured by colostomy. Weight at operation ranged from 3.3 to 6.0 kg (mean 4.9 kg). SIENSON procedure (25), UDENEL (5) and SOMME procedure (2) were performed. There was no mortality. Follow-up varied from 2 to 8 years. Post-operative morbidity is reported in five patients with total colonic HD. All others patients are considered as definitively cured.

Comparison with literature's cases permits to advocate primary corrective treatment of HD mostly without decompression in infants less than three months of age.
CNS Liver Trauma Registry Report

From the Trauma Subcommittee, Canadian Association of Pediatric Surgeons

During the 2 years 1986-1987, 64 patients were enrolled in the CNS liver trauma study (39M, 25F; mean age 7.6 years). Most (94%) had blunt trauma; 25% were pedestrians and 37% were passengers involved in motor vehicle accidents. 47 (73%) had associated injuries including 12 splenic, 3 duodenal and 1 pancreatic injury.

After initial stabilization, 17 (26%) underwent immediate laparotomy, 34 (53%) were transferred to the ICU and 13 (21%) to the ward. None of the patients initially admitted to the ICU or the ward required laparotomy. Operative procedures included simple sutures (9), drainage alone (4), resectional debridement (3), formal lobectomy (2), and hepatic vein repair (2). 41 (64%) required blood transfusion (mean volume 39 ml/kg).

3 patients died; all from head injuries. 8 patients developed sepsis including 4 subphrenic and 2 liver abscesses. The average hospital stay was 29 days. Conservative management remains the mainstay of treatment for blunt hepatic trauma; massive bleeding (> 40 ml/kg) which is usually obvious immediately is the main indication for surgical intervention.

H-TYPE SHUNT WITH AN AUTOLOGOUS VENOUS GRAFT IN THE TREATMENT OF PORTAL HYPERTENSION IN CHILDREN. F. Gauthier, J. Valayer, Ph. Montpetet. O. De Dreuxy. Service de chirurgie, Dept. de PediatrIe, Hôpital de Bicêtre et Faculté de Médecine Paris-Sud, France.

From 1977-87, 127 children aged 1 to 16 yrs were submitted to shunt surgery for correction of portal hypertension. Eighty-seven shunts were performed in 86 children using an autologous venous graft (internal jugular vein in 80 cases); 57 mesocaval, 19 spleno-renal, 4 portacaval and 7 miscellaneous shunts. The indication was a portal obstruction in 61 cases, an intrahepatic obstruction in 21 cases (including 5 cases of congenital hepatic fibrosis) and a Budd Chiari syndrome in 4 cases. One patient of this latter group died from intractable ascites postoperatively, and a second infant died from unrelated disease 2 months postop. The patency of the shunt is routinely evaluated 6 months after surgery by means of ultrasonography and esophageal endoscopy. Four children have a follow-up of less than 6 months. A good result was achieved in 75 of the 81 remaining children, none of whom suffered further gastrointestinal bleeding. In the 2 remaining children thrombosis of the shunt with recurrence of bleeding led to secondary operation with a good result in one case, and another secondary intervention is planned. The H-type shunt brings in most cases a good relief of portal hypertension and should be used as often as possible for those patients without risk of encephalopathy, i.e. cases of portal obstruction with congenital or acquired, congenital hepatic fibrosis and Budd Chiari syndrome without caval hypertension. Iliac vein graft may be used instead of the internal jugular vein if the distance between portal and caval systems is short (spleno-renal and portacaval shunts). The main point to be discussed is the indication of such shunt surgery in children with cirrhosis with the risk of encephalopathy and also additional technical difficulties should the child be candidate for liver transplantation.
GASTROSTOMY AND FUNDOPLICATION IN NEUROLOGICALLY IMPAIRED CHILDREN.
G. Stringel, M.D. and M. Delgado, M.D.
U. of Tx. Southwestern Medical School and Texas Scottish Rite Hospital.
Dallas, Texas.
Children with neurological impairment often do not reach their potential
development because of malnutrition which can be aggravated by inability
to swallow and complications of G.E. reflux. Vomiting and lack of re-
liable route of administration make seizure control difficult. Nasso-
gastric feedings are uncomfortable and can lead to complications when
G.E. reflux is present. Gastrostomy can aggravate G.E. reflux or cause
it in some cases. We report our experience with 79 neurologically im-
paired children treated with gastrostomy and Nissen fundoplication.
There were 50 males and 29 females ranging in age from newborn to 17 y.
Mean age was 4 years. The etiology of static encephalopathy varied.
Malnutrition and developmental delay were present in all patients.
Pharyngoesophageal dyskinesia was seen in 80%, vomiting in 60 %, and
recurrent respiratory infections in 75%. G.E. reflux was demonstrated
radiologically of by pH probe in 62 patients. In 17 children, no reflux
could be demonstrated; in 15 of these, a prophylactic Nissen fundopi-
clation was done with the gastrostomy. In 2 cases, gastrostomy alone
was done; 4 other children had gastrostomy alone done elsewhere. All
6 subsequently required a Nissen fundoplication because of uncontrol-
able G.E. reflux.
Nissen fundoplication was done following a standard technique: preser-
vation of the vagus nerves and their branches, plication of the crura
of the diaphragm, reconstruction of the angle of His, and a 360° wrap.
Complications were few; there were no deaths. Average weight gain
3 months after surgery was 3.2 kg. There was marked improvement in 90% of
the cases. The majority of parents were very satisfied.

ESOPHAGEAL MOTOR EFFICIENCY IN GASTROESOPHAGEAL REFUX
J.A. ARANA, J.A. TOVAR
Univ. del Pais Vasco, Hospital BASQUE de Aranzazu, San Sebastian, Spain

In an attempt to simplify the evaluation of esophageal peristaltic efficiency
we have investigated manometrically 18 normal children and 73 with extended
ph-metering-documented gastroesophageal reflux (GER). The mean pressure and
the frequency of propulsive waves in the body of the esophagus were measured
and both values were multiplied to yield one single figure which reflected in some
way esophageal motor efficiency (EME). We have performed the tests
in basal conditions (EMEB) and after instillation of CH 0.1 N into the esophageal
lumen (EMECIN).

Differences between normal and GER children were statistically significant
for EMEB (13 ± 3.1.8 vs 3.5 ± 4.9, p < 0.01) as well as for EMECIN (15 ± 5.57 vs
10.5 ± 3.83, p < 0.05). These results show that abnormal esophageal motility is not
limited in the presence of non-propulsive waves and they enhance the potential
diagnostic value of manometry.

Furthermore, esophageal motor response after CH instillation can have
some prognostic value because EMECIN was significantly higher in those of the
61 children followed-up for more than one year who responded to medical
treatment (n=23) than in those in whom this was a failure (n=38) (14.5 ± 9.8 vs
7.5 ± 7.3, p < 0.01).

The very limited information currently expected from esophageal
manometry in children (LESP measurement and percentage of non-propulsive
waves) accounts in part for the poor development of this tedious and difficult test
which diagnostic yield has not been fully explored so far.
35 ESOPHAGEAL ATRESIA WITH DISTAL TEF; OTHER ANOMALIES AND OUTLOOK
Sigmund H. Ein; Barry Shandling; David Wesson; Robert M.Filler.
Hospital for Sick Children, Toronto, Ontario, Canada
Ninety-seven newborns with esophageal atresia and distal TEF
were treated between 1979 and 1985 inclusive; there were 54
boys and 43 girls. Their weights ranged from 800 to 4000
grams (average 2.5 gm). There were 26 neonates with cardiac
defects (most common: PDA, ASD-VSD) and 18 survivors (64%);
17 babies with GI anomalies (imperforate anus, duodenal
atresia) and 12 survivors (70%); 12 patients with skeletal
malformations (digits, vertebral), and 11 survivors (91%);
eight newborns with 80 abnormalities (hypospadia,
undescended testis) and six survivors (75%); and 16 infants
with other congenital lesions (Trisomy 18, lung
agenesis-hypoplasia) and three survivors (18%). Forty-six
(2.7 gm) had no other anomalies and all survived. As the
number of systems with defects increased, the weight of the
baby and its survival rate fell. From this entire series of
97 newborns with esophageal atresia and distal TEF, 81 (83%)
survived (2.3 gm). Sixteen babies died (1.6 gm); ten had
defects incompatible with life. Eleven of the 16 were never
operated on (seven Trisomy 18 and one with four systems
involved died within six days, three complex cardiac died
within three weeks). Four of the 16 who were operated on
died between two months and two years from unrelated chest
problems and one newborn died in the operating room.

36 ACHALASIA OF THE CARDIA: SURGICAL TREATMENT OF 35 CASES
AND STUDY OF THE ASSOCIATION WITH FAMILIAL DYSAUTONOMIA
AND SELECTIVE A.C.T.H. INSENSITIVITY.
C.N.-Fékété, F.Bawab, S.Lortat-Jacob, P.Arhan, D.Pellerin
Department of Pediatric Surgery.
Hôpital des enfants malades, Paris, FRANCE.

Although rare in children, achalasia can be the cause of
debilitating symptoms and growth retardation. During a
25-year period, 35 patients (aged 5 months to 14 years)
underwent a modified Heller operation (anterior cardia-
esophageal myectomy with anterior semi-circular
fundoplication). 34 children are cured with no recurrence
of dysphagia, no clinical or radiological evidence of
gastro-esophageal reflux; one has peptic esophagitis in
spite of re-operation for fundoplication. Early
recognition of the disease prevents the onset of mega-
esophagus and is based on esophagoscopy and manometry.
Precise surgical technique obtains immediate and long-
lasting cure.

Familial dysautonomia was observed in 5 cases, one of
which presented with A.C.T.H. insensitivity and
hypoglycemic cerebral sequelae; this severe association
is studied and literature on this subject is reviewed.
THE SPECTRUM OF CHOLELITHIASIS IN THE FIRST YEAR OF LIFE. William J. Schirmer, MD, Enrique R. Grisoni, MD, and Michael W.L. Gauderer, MD. Division of Pediatric Surgery, Case Western Reserve University, Cleveland, Ohio.

Cholelithiasis is being diagnosed with increased frequency in the very young. We encountered twelve patients over a three year span in whom gallstones were identified prior to age one year (birth to 11 months; 7 male, 5 female). The earliest diagnosis was made in utero during the seventh gestational month (of interest - the mother underwent cholecystectomy during the same pregnancy). Known predisposing factors were present in eight: parenteral nutrition-8; prematurity-7; sepsis-4; short gut syndrome-1, yet no recognized predisposing factors could be identified in the other four. Unsuspected gallstones were found in three patients with hypertrophic pyloric stenosis. In three of the twelve, the initial presentation was one of complications requiring prompt surgical intervention (suppurative cholecystitis-2, obstructive choledocholithiasis-1). Two children underwent elective cholecystectomy nearly two years after initial diagnosis. Of the remaining seven, spontaneous stone resolution was documented in four while three remain asymptomatic after over two years of follow-up. The spectrum of presentations in this series suggests that several different etiologic factors may be at play. In addition, it underlines the uncertainty regarding the natural history of gallstones in infants. Therefore, pediatric surgeons must avoid extrapolation from the adult experience but rather individualize their approach to the management and follow-up of this apparently increasingly more common group of children.

ISOLATED COMPLETE TRANSECTION OF THE COMMON BILE DUCT DUE TO BLUNT TRAUMA IN A CHILD.
Michel Bourque, Ntetsana Spigland, Hervé Blanchard, Arié L. Bensoussan, Laurent Garel.

A case of isolated complete transection of the common bile duct due to blunt abdominal trauma in a 3 year old child is presented. The rarity of this injury and its initial presentation as a pancreatic pseudocyst warrant its description. This patient, the 21st child to be reported in the literature, was diagnosed as having a biliary injury following ultrasound guided percutaneous drainage. Choledochal transection was documented at laparotomy and was successfully treated by proximal choleductostomy and choledochojunostomy with Roux-en-Y reconstruction. Delayed diagnosis is common, yet this injury should always be kept in mind and early diagnosis is feasible with the use of the HIDA scan. A review of the literature, modes of diagnosis and surgical repairs are described.
BILE DUCT STRicture IN AN INFANT WITH GASTROESOPHAGEAL reflux TREATED BY PER-
CUTANeous TRANSHEPATIC DRAINAGE, BILIARY STENTING AND BALLOON DILATION

B.J. HANCOCK, N.E. WISEMAN, B.W. RUUSMA
DEPARTMENTS OF PEDIATRIC GENERAL SURGERY AND RADIOLOGY
ST. BONIFACE GENERAL HOSPITAL
409 TACHE AVE, WINNIPEG, MANITOBA, CANADA, R2H 2A6

A newborn twin with an antenatal diagnosis of gastroesophageal reflux underwent staged repair of the abdominal wall defect. She developed multiple small bowel fistulae due to ischemic bowel and then abdominal wall dehiscence requiring additional surgical interventions. Obstructive jaundice, first evident at 3 weeks of age, became progressively severe. A common bile duct stricture was diagnosed by percutaneous transhepatic cholangiography. The stricture was treated by percutaneous biliary drainage, stenting and balloon dilatation of the common bile duct. These procedures, commonly used in adults for biliary decompression, may be useful alternatives to surgical intervention in infants and children with obstructive jaundice due to bile duct stricture.

LONG TERM FOLLOW-UP OF SACRO-COCGYGEAL TERATOMAS WITH EMPHASIS ONANORECTAL FUNCTION. J. Bass, S. Yazbeck, Francois Luks. Hôpital Ste-Justine, Montréal, Québec, Canada.

The outcome of malignant sacrococcygeal teratomas (SCT) is well described in the literature. There are no reports on long term follow-up of patients with benign SCT. The present series is a retrospective study of 49 cases of SCT treated in our institution during the past 27 years. Ten patients died. Six death were related to malignant SCT. One died of medulloblastoma at 2 years of age, one died of intraoperative blood losses, one died of preoperative bleeding and one patient died prematurely. One patient is still under chemotherapy 2.5 years after resection of a yolk sac tumor. Out of the remaining 38 patients, 13 could not be reached and 25 were available for follow-up (66%). The mean follow-up was 10.1 years (1 to 27 years). The extreme disturbances of the pelvic musculature, as regularly noticed intraoperatively, led us to investigate the ultimate outcome of the anorectal function in these patients. All patients available for follow-up were continent for stools and urine. Adequate assessment of rectal evacuation was possible in 21 patients. Ten out of the 21 patients (47.6%) had a significant degree of constipation and five of them presented also documented episodes of urinary tract infections. In this particular group the mean follow-up was 9.5 years (1-24 years). The absence of incontinence in these patients suggests either the return to normal function of the peri-rectal muscular components or a more important role of the external sphincter in the mecanism of continence. The increased frequency of significant constipation in this group of patients have not been previously emphasized, and suggests the need for a longer follow-up.
HEREDITARY SACROCOCCYGEAL TERATOMA
RD Sonino MD, S Chou, MD, FM Guttmann, MD, CR Moir, MD The Montreal
Children's Hospital, Montreal, Quebec CANADA H3H 1P3

A French Canadian family with sacrococcygeal pathology in two siblings and the
father is presented. The first pregnancy resulted in fetal death of unknown
causes two days before term. The second pregnancy resulted in a boy, 33 weeks
gestational age, who came to medical attention in the neonatal period because of
inability to insert a rectal thermometer, failure to pass meconium and
abdominal distention. An attempt at probing the stenotic anus resulted in
perforation of the recto-sigmoid necessitating a colostomy. Re-evaluation at 5
1/2 months of age revealed a pre-sacral mass posteriorly and slightly to the right
side on rectal exam. This was excised and the pathology report revealed a
benign pre-sacral teratoma. The colostomy was closed about one month later. A
girl was born to this couple 2 1/2 years later. She also had anal stenosis and
chronic constipation. Investigations at age 3 months, revealed an anterior
meningocoele and a pre-sacral mass just beneath it. The third sacral segment was
dysplastic. The mass was excised, and the meningocoele closed. The pathology
report was benign pre-sacral teratoma. The father, at age 26, was also
investigated for chronic constipation. A pre-sacral mass was found on rectal
exam, and its presence confirmed on CT scan. The sacrum is dysplastic by X-
ray. To date, he has declined surgical intervention. Continuous monitoring of
this cys over the past 6 years shows no change. All consultants feel that this is
an anterior meningomyelocele. Hereditary pre-sacral teratoma and its
associated anomalies were first described by Ashcraft and Holder. They
reported 17 cases of pre-sacral teratoma in 6 kindred. We report on a similar
family where the father presents with a presumed meningomyelocele and his two
children with sacrococcygeal teratomas.

NEONATAL DIAGNOSIS OF AN ANTERIOR MENINGOCELE IN THE PRESENCE
OF CONGENITAL ANAL STENOSIS AND PARTIAL SACRAL AGENESIS. Harold Brem,
MD; Bonnie L. Beaver, MD; LR Scherer, MD; Benjamin S. Carson, MD;
J. Alex Haller, Jr., MD; FAAP, FACS. The Johns Hopkins Hospital,
Division of Pediatric Surgery, Baltimore, MD.

The simultaneous presentation of clinically symptomatic anal
anomalies and roentgenographically demonstrated sacral dysgenesis
should alert the pediatric surgeon to investigate for the presence
of a presacral malformation. We report on such a case to
illustrate a new radiographic technique which facilitates
diagnosis and management of complex congenital malformations. A
one day white male present with failure to pass meconium after a
full term uncomplicated pregnancy. Significant anal stenosis was
present on physical exam. The coccyx, right half of the sacrum and
the right lamina of L5 were not present. The presence of an
anteriormeningocoele was identified using a computerized program
to interpret transaxial CT scan sequences and reconstruct them
into a three dimensional image. This technique allowed the surgeon
to define the anatomy and borders of the anomaly in detail that
was previously only available under intraoperative vision. A
diverting colostomy was performed. The meningocoele was
successfully corrected utilizing a posterior approach. Two years
post operatively the patient continues to do well. This case
demonstrates that this triad of anomalies (presacral mass, sacral
dysgenesis, and ano-rectal malformation), once considered can be
safely detected with modern radiological techniques and
expeditiously corrected during infancy before further
deterioration occurs.

Malignant small cell tumor of the thoracopulmonary region (MSCT) is an uncommon neoplasm in childhood. We describe 5 cases diagnosed since 1981 which fulfill the criteria put forth by Askin et al. Our group consists of 3 males and 2 females with a mean age of 9 yrs, ranging from 3 to 15 yrs. Three patients presented with a chest wall mass. Pain was present in 2 patients and dyspnea in 2 others. Fever, fatigue, weight loss and coughing were present in one patient. Duration of symptoms averaged one month, ranging from 3 days to 3 mos. Chest X-ray identified the lesion in 4/5 cases and demonstrated periosteal reaction of the adjacent rib. Pleural effusion was present in 2 cases. No calcifications were seen on CT-scan (4/5). Two cases showed bilateral lung involvement. Surgery was performed for diagnosis and/or therapy in all patients. Two underwent open lung biopsy only due to tumor extent. The other 3 had chest wall resections; one synthetic patch reconstruction, one latissimus dorsi muscle flap, and the 3rd patient required no reconstruction. All received radiotherapy and combined chemotherapy. Three patients presenting initially with extensive disease died at intervals of 2.5 to 7 mos after diagnosis. Two patients are alive and disease free at 16 and 24 mos post diagnosis. All five cases were reviewed for standard histology and differential immunohistochemistry. Electron microscopy and tissue cultures were done in 3/5. All 5 patients were neuron specific enolase positive. MSCT is an exclusion diagnosis not always readily made. Clinical course and pathological expertise may point to the correct diagnosis.

ESOPHAGEAL ATRESIA AND RIGHT AORTIC ARCH; RIGHT OR LEFT THORACOTOMY?

G. Stringel, M.D.

J. of Tx. Southwestern Medical School, Dallas, Texas.

Right aortic arch occurs in 5% of patients with esophageal atresia. It is often associated with other anomalies. Repair of the esophageal atresia and associated T.E.F is difficult if attempted through the standard Rt. thoracotomy approach. Traditional teaching has been that when a Rt. aortic arch is found during repair of esophageal atresia through a Rt. thoracotomy, the wound should be closed and repair done through a Lt. thoracotomy. We report our recent experience with 5 infants with esophageal atresia and T.E.F. and associated Rt. aortic arch. There were 4 females and one male, birth weights varying from 900 gm. to 2800 gm. In 3 of the infants, the following anomalies: microcephaly, thoracic atresia, P.D.A., microgastria, microtia, cleft palate and aberrant subclavian artery were all present. In 2, the Rt. aortic arch was the only associated anomaly. Rt. aortic arch was recognized during surgery in 4 infants and successful repair of the esophageal atresia with division of the fistula was accomplished without difficulty through the Rt. thoracotomy. In one other infant, the Rt. aortic arch was diagnosed preoperatively by fluoroscopy and CT scan and repair was done through a Lt. thoracotomy. Three infants recovered uneventfully, requiring no further treatment. Another infant required one post operative esophageal dilatation. One baby died 4 weeks after surgery due to severe associated congenital anomalies. When a Rt. aortic arch is suspected by plain films, it should be confirmed by fluoroscopy of CT scan. Arteriography is not necessary and subjects the patient to unnecessary risk. Cardiac anomalies and vascular rings should be looked for. When confronted with a Rt. aortic arch during repair of esophageal atresia, repair can be done safely and successfully through the Rt. thoracotomy.
Pulmonary Agenesis with Esophageal Atresia and Tracheoesophageal Fistula: A Report of Two Cases

M.A. Hoffman, M.D., R.A. Superina, M.D., D. E. Wesson, M.D.
Department of Surgery, The Hospital for Sick Children, Toronto, Ontario

Unilateral pulmonary agenesis with esophageal atresia is a rare association. There are no reported survivors of definitive repair in the neonatal period. We report two cases of right sided pulmonary agenesis with Type C EA/TEF both definitively repaired shortly after birth. In the second case, there was concomitant duodenal atresia. Both are well at ages 24 months and 12 months.

Case 1: A 2200 gram term, female with mild respiratory distress at birth. Nasogastric intubation was unsuccessful. X-rays revealed a proximal pouch, distal bowel gas, an opacified right chest with mediastinal shift to the right. Definitive repair of EA/TEF was performed at 48 hours age. Chest CT scan confirmed right pulmonary agenesis. The baby was discharged at age 1 month and is well at 24 months follow-up.

Case 2: A 2880 gram full term male with mild respiratory distress at birth. Nasogastric intubation was unsuccessful. X-rays revealed a proximal pouch, an opacified chest, mediastinal shift to the right and large gastric bubbles in an otherwise gasless abdomen. Single stage definitive repair was performed under the same anesthesia shortly after birth. Gastrostomy insertion was followed by duodeno-duodenostomy. The Type C EA/TEF was next repaired. The baby was discharged during the fifth post-operative week. The baby is well at 12 months follow-up.

THE EFFECT OF INTESTINAL ANASTOMOSIS ON GUT GROWTH AND MATURATION

G. Stringel, M.D., R. Dany, M.D., Ph.D.
U. of Tx. S.W. Medical School, Dallas, Texas.

A group of 10 weanling Sprague-Dawley rats at 21 days of age was subjected to intestinal transection and anastomosis in the upper jejunum, 10 cm. distal to the ligament of Treitz(Surg.). A second group of rats of same age and characteristics was used as control(No Surg.). All rats were fed a regular diet and sacrificed 2 weeks later. Body weight, intestinal weight and intestinal length were obtained; the intestine was divided in 2 sections: pre-anastomosis (Section A) and post-anastomosis (Section B) in the surgery group and equivalent sections A and B in the control group. Specimens were taken for morphometric evaluation and mucosal scrapings for biochemical analysis from both sections in both groups. Differences in weight gain, intestinal weight and intestinal length were not statistically significant.

<table>
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<th>Protein</th>
<th>D.N.A.</th>
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<th>Malate</th>
<th>Sucrase</th>
<th>Lactase</th>
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<td>(mg/g of wet weight)</td>
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<td>B</td>
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<td>B</td>
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<td>1.3</td>
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<td>72</td>
<td>7.1</td>
<td>13.2</td>
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<tr>
<td>± S.E.</td>
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<td>0.5</td>
<td>0.3</td>
<td>0.8</td>
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</tbody>
</table>

In the pre-anastomotic segment, muscle thickness, villus height and crypt depth were significantly increased; protein and DNA were higher, but P/DNA was less affected; enzymatic activity was significantly decreased. Intestinal anastomosis has a significant effect on gut growth and maturation in the growing animal and may have important implications in the post-operative management of newborns and infants.
SEGMENTAL AGENESIS OF INTESTINAL MUSCULARIS - A CASE REPORT

A. Alawadi, MD, S. Chou, MD, FRCS(C)
Children's Hospital of Eastern Ontario, Ottawa, Ontario

Congenital absence of intestinal musculature is a rare disease entity. The clinical presentation is variable but most present as intestinal perforation or obstruction.

A case of a female infant with this disease entity is presented. The seven other cases from world literature and the histopathology are reviewed. Moreover, the different theories which have been put forth regarding the etiology of this lesion and its embryogenesis is discussed.

ANATOMICALLY IDENTICAL SHORT SEGMENT HIRSCHSPRUNG'S DISEASE IN THREE MALE SIBLINGS

S.Z. Rubin, MB, FRCS(C), FRCS(E)
Children's Hospital of Eastern Ontario, Ottawa, Ontario

Three male children of a young married couple are being reported with identical short segment Hirschsprung's Disease. There was no positive family history despite an extensive search. There were no associated abnormalities. Although the sex modified multifactorial inheritance, with males having a lower threshold of genes for expression of Hirschsprung's Disease, is accepted, the identical expression of the disorder in the three sibs suggests a dominant, possibly X-linked, gene with variable penetrance explaining the negative family history. Another possibility is that an identical micro-environmental factor has been present pre-natally resulting in all three sibs having Hirschsprung's Disease. This is the first report of three sibs with identical short segment Hirschsprung's Disease.
DIAGNOSIS AND INCIDENCE OF CARDIAC INJURY IN CHILDREN WITH BLUNT THORACIC TRAUMA

Hospital of Sick Children, Toronto, Canada

Cardiac injury has been reported as a frequent occurrence in Blunt Thoracic Trauma (BTT). Over 24 months, we prospectively studied 41 children with BTT, to determine the incidence and morbidity of cardiac injury in this population. Ages were 3 months to 15 years (mean 8.2 years). Four patients died, and in 10 the data were incomplete, leaving 28 for analysis. Serial EKG was abnormal in 8/22 (36%), serial CK-MB ratio was elevated in 8/22 (36%), echocardiogram showed septal dysfunction in 3/21 (14%), and pyrophosphate scan showed grade 1 uptake in 3/21 (14%). There was poor correlation among tests, however, as only 4 patients had more than one abnormal test. There was no significant difference in ISS or thoracic AIS between patients with and without abnormal investigations.

No patients in the study developed arrhythmias or cardiac failure. In 13 trauma-related deaths going to autopsy over the same period, (including the 4 with thoracic trauma), none had any evidence of cardiac injury.

These results indicate a striking lack of consistency in the diagnosis of posttraumatic cardiac injury in children using standard investigations. The absence of adverse cardiac events in surviving patients, and the lack of autopsy evidence of cardiac injury in the trauma deaths, suggest that the clinical significance of this lesion in children is lower than generally reported.

Treatment of Focal Nodular Hyperplasia of the Liver

By Alcohol Embolization

Soucy, P., Rasuli, P., Chou, S.Y., Carpenter, B.
Children’s Hospital of Ottawa, Ontario.

Focal nodular hyperplasia (FNH) is a benign, asymptomatic liver tumour, with no predisposition to spontaneous hemorrhage. In most cases, expectant treatment is recommended. However, in a small number of patients, due to marked increase in size of the tumour, and compression of normal liver or porta hepatitis, surgical resection may become necessary. Alcohol embolotherapy offers a safe and effective alternative to surgery. FNH lends itself to embolotherapy because it is usually fed by a single end artery, with no intratumoral arterio-venous shunting, nor parasitic blood supply. This feature reduces the risk of non-target embolization. Alcohol is superior to particulate emboli because of its deep vascular penetration and ability to create effective sterile tumour necrosis.

An illustrative case is presented. Diagnosis was established by ultrasound, CT scan, radionuclide scan, arteriography, and confirmed by biopsy. Alcohol embolization was carried out using a balloon occlusion technique, and was well tolerated, with only a mild post-embolization syndrome. Over an 18 months follow up period, the 14cm tumour in this patient has shrunk to a small calcified nodule.
GASTRIC HETEROTOPY OF THE BILIARY TRACT

M.J. Martinez Urrutia, F. Rivilla, J. Vazquez, J. Larrauri, J. Diez
Department of Surgery, La Paz Children’s Hospital, Madrid, Spain.

We present the case of a 12 year-old girl with cholestatic syndrome. Transparietal hepatic cholangiography showed an image of obstruction at the confluence of the common hepatic and cystic ducts. At the surgery, cholecystectomy was performed and the common hepatic duct was removed. Both hepatic ducts were anastomosed to the duodenum. The histologic exam reported heterotopic gastric mucosa containing chief, parietal and mucoocytes at level of the gallbladder neck. The biliary fragment studied showed hyperplasia of the heterotopic pyloric glands and an important fibrous and inflammatory component. The postoperative course of the patient was favorable. Three weeks after surgery, administration of HIDA Tc 99m showed a homogeneous distribution and uptake of radioisotope in the liver and normal excretion to the duodenum. Up to date the patient is doing well.

THE VENOCUTANEOUS FISTULA-A NEW APPROACH TO CHRONIC VENOUS ACCESS

Charles E. Bagwell, MD
University of Florida College of Medicine, Gainesville, FL

The use of central venous catheters in children has increased dramatically due to difficulties in maintaining chronic peripheral access as well as for administration of drugs, chemotherapy or withdrawal of blood samples. However, many complications have been associated with the catheter itself, including catheter thrombosis, sepsis or thrombus formation in great vessels or the atrium. A surgical technique has been devised for creating access to the central venous circulation, yet avoiding a chronic, indwelling central venous catheter. Using this approach, a venocutaneous fistula has been successfully created in a series of 6 pygmy scrub goats. In each experimental animal, the external jugular vein was ligated, divided and the proximal end anastomosed to the cervical skin, creating a stoma-like orifice. This open fistula allowed easy access of a soft, blunt-tipped catheter into the right heart for repeated central venous access. No significant bleeding occurred from the fistula following removal of the catheter with infusion of intravenous agents thrice weekly, and no infectious complications ensued. Reliable patency of the fistula could be maintained for a period of 4-6 weeks without dressings or any external appliance. While far removed from actual clinical application, the possibility of catheter-free access to the central venous circulation offers great potential for ease of care and lessened infection rate from catheter-related sepsis.
POSTERIOR SAGITTAL ANORECTOPLASTY FOR PEDIATRIC RECURRENT RECTAL PROLAPSE
Richard H. Pearl; Sigmund H. Ein
Hospital for Sick Children, Toronto, Ontario, Canada

The recent use of the posterior sagittal anorectoplasty for repair of high imperforate anus has demonstrated several advantages: elimination of laparotomy, more direct approach, easier division of recto-urethral fistula, more exact identification of the muscles of fecal continence, proper relocation of ano rectum within these muscles and sphincters and virtual elimination of postoperative anal prolapse. It is this latter advantage that attracted us to use this procedure for the repair of a recurrent rectal prolapse in a one year old girl who also had a recurrent bladder extrophy. The latter probably contributed to her constantly pushing out her rectum which easily admitted two fingers. Two attempts were made to repair the rectal prolapse using the subcutaneous Thielsch perianal technique, however, each was successful for only six weeks. When her recurrent bladder extrophy was repaired, we also repaired her recurrent rectal prolapse using the posterior sagittal anorectoplasty. The midline incision was carried down to but not through the external sphincter, and the patulous rectum was plicated back to a normal size. Reapproximation of the levator sling and lower muscle complex also incorporated the plicated rectum. Both repairs remain intact after one year.


Leiomyoma of the esophagus is prevalent among the adult population and yet is rare within the pediatric population with only 19 cases reported so far within the pediatric surgical literature. We would like to report two cases of esophageal leiomyoma in female patients ages 6 and 13 years respectively. Both cases represented preoperative diagnostic dilemmas. The first patient, age 13, was explored via right thoracotomy with a false diagnosis of bronchogenic cyst. She was subsequently found to have a large distal invasive esophageal mass and underwent resection of this mass. Follow-up at 6 months reveals gastro-esophageal reflux which is currently being controlled by conservative means. The second even rarer presentation was that of a six year old girl who was diagnosed as having achalasia preoperatively. An abdominal exploration revealed the presence of a large esophageal mass which was subsequently biopsied. She was explored via left thoracotomy and at that time she was unexpectedly found to have a diffuse esophageal leiomyoma extending from the cervical esophagus to the cardia. The extent of the tumor required a total esophagogastrectomy with reverse gastric tube. She has done well at 3 months follow-up. Reporting this rare entity and reviewing the literature brings to mind the difficulty encountered with the preoperative diagnosis and intra-operative management of these lesions. Esophageal leiomyoma should be kept in mind as part of the differential diagnosis of mediastinal masses in the pediatric population in order to facilitate preoperative diagnosis and surgical approach. Interestingly, our data supports the evidence that the sex ratio is reversed in the pediatric population.
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Le fond d'éducation permet d'inviter chaque année d'experts chirurgiens pédiatiques étrangers pour enseigner dans différents centres médicaux du Canada. Il permet également à notre Association de déléguer un conférencier en chirurgie pédiatrique lors de la réunion de la Société Canadienne de Pédiatrie. Il rend possible une participation élaborée de notre Association au programme scientifique du Congrès Annual du College Royal des Médecins et Chirurgiens du Canada. Il nous aide enfin à défrayer le coût de la réunion annuelle de l'Association Canadienne de Chirurgie Infantile.

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Montreal, Quebec
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Hospital for Children
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Colborne One
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44122
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Montreal Children's Hospital  
2300 Tupper Street  
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H3H 1P3  
(514) 934-4498

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750 E. Adams Street  
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Angus and Marion  
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A1C 5J9  
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Department of Surgery, DIII
Kingston General Hospital
Kingston, Ontario
K7L 2V7
(613) 544-6133
(613) 544-1426

John and Beryl
41 Cooperfield Drive
Kingston, Ontario
K7M 1M3
(613) 544-0341

Dr. Sylvain Kleinhaus *
Montefiore Hospital and
Medical Centre
111 East 210th Street
Bronx, New York 10467
(212) 920-4758
(212) 920-4214

Sylvain and Anna
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302 Greensboro Place
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9711-141 Street
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T5N 2M5
(403) 452-6520

Dr. Irwin H. Krasna *
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Academic Health Sciences Centre, CN 19,
New Brunswick, New Jersey 08903

Dr. Jean-Martin Laberge
Montreal Children's Hospital
C1137-2300 Tupper Street
Montreal, Quebec
H3H 1P3
(514) 934-4497
Dr. Dennis J. Lafer *
Pediatric Associates, P.A.
820 Prudential Drive,
Jacksonville, Florida 32207
(904) 396-2048

Dr. George Y.P. Lau
25 Charlton Ave. East, #704
Hamilton, Ontario.
L8N 1Y2
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Dr. Henry Lau
The Izaak Walton Killam
Hospital for Children
5850 University Avenue
Halifax, Nova Scotia
B3H 3G8
(902) 428-8194

Dr. Gordon M. Lees
8215-112th Street, Ste. 602
Edmonton, Alberta
T6G 2C8
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G1V 4G2
(418) 656-8168

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Gordon
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G1T 2H4
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N6C 5J3  
(519) 685-2476

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Fairmont Medical Bldg., #1314  
750 West Broadway  
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V5Z 1J3  
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Children's Hospital of  
Eastern Ontario  
401 Smyth Road  
Ottawa, Ontario  
K1H 8L1  
(613) 737-7600  
Stan and Sylvia  
210 Buena Vista Road  
Ottawa, Ontario  
K1M 0V7  
(613) 741-3049

Dr. David R. Murphy **  
David and Beatrice  
219A Main street  
Barriefield Village  
Kingston, Ontario  
K7K 5S5

Dr. Harold O. Nason  
Harold and Norma  
R.R. # 3  
Middleton, Nova Scotia  
BOS 1P0  
(902) 825-6279
Dr. Luong T. Nguyen  
Montreal Children's Hospital  
C-1132-2300 Tupper Street  
Montreal, Quebec  
H3H 1P3  
(514) 934-4438  

Luong  
326 Leacross  
Montreal, Quebec  
H3P 1M1  
(514) 737-5065  

Dr. Herbert F. Owen **  
Montreal Children's Hospital  
C-1134-2300 Tupper Street  
Montreal, Quebec  
H3H 1P3  
(514) 934-4468  

Herb and Christine  
28 Picardy Road  
Baie D'Urfe, Quebec  
H9X 3G4  
(514) 457-5201  

Dr. Gian B.G. Paloschi  
Department of Surgery  
Hotel-Dieu Hospital  
Kingston, Ontario  
K7L 5G2  
(613) 548-8600  

Gian and Susan  
30 Seaforth Road  
Kingston, Ontario  
K7M 1E2  
(613) 546-5093  

Dr. John B. Pietsch *  
Department of Pediatric Surgery  
Vanderbilt University Hospital  
Nashville, Tennessee  37232  

Dr. Raymond Postuma  
Children's Hospital  
840 Sherbrook Street  
Winnipeg, Manitoba  
R3A 1S1  
(204) 787-4203  

Ray and Jane  
232 Glenwood Crescent  
Winnipeg, Manitoba  
R2L 1J9  
(204) 668-7498  

Dr. R. Hampton Rich *  
2545 Chicago Ave. So., Suite 104  
Minneapolis, Minnesota  55404  
(612) 871-4551  

Hampton  
1201 Yale Place #1808  
Minneapolis, Mn.  55403  
(612) 333-7777  

Dr. Bradley M. Rodgers *  
Department of Surgery, Box 181  
University of Virginia School of Medicine  
Charlottesville, Virginia  22908
Dr. Steven Z. Rubin  
Children's Hospital of Eastern Ontario  
401 Smyth Road  
Ottawa, Ontario  
K1H 8L1

Dr. C. Geoffrey F. Seagram  
1820 Richmond Road S.W.  
Calgary, Alberta  
T2T 5C7

(403) 245-7211

Geoff and Shirley  
1110 Sifton Blvd. S.W.  
Calgary, Alberta  
T2T 2L1

(403) 243-0031

Dr. Barry Shandling  
The Hospital For Sick Children  
526-555 University Avenue  
Toronto, Ontario  
M5G 1X8

(416) 597-0705

Barry and Mary  
10 Sunnydene Crescent  
Toronto, Ontario  
M4N 3J6

(416) 484-1811

Dr. Pierre Soucy  
Children's Hospital of Eastern Ontario  
401 Smyth Road  
Ottawa, Ontario  
K1H 8L1

(613) 737-2396

Pierre  
7 Valecrest  
Ottawa, Ontario  
K1B 4G2

(613) 824-7766

Dr. Clinton A. Stephens **  

Clint and Jean  
68 Hillhome Road  
Toronto, Ontario  
M5P 1M5

(416) 488-8642

Dr. Gustavo Stringel *  
3912 Colgate  
Dallas, Texas  
75225

(214) 363-9249

Gustavo and Lina  
10460 Remington  
Dallas, Texas  
75229
Dr. Peter Sudermann
The Winnipeg Clinic
425 St. Mary Avenue
Winnipeg, Manitoba
R3C 0N2
(204) 957-1900

Dr. Riccardo A. Superina
The Hospital For Sick Children
555 University Avenue
Toronto, Ontario
M5G 1X8

Dr. William H. Taylor

Bill and Jane
26 Daleberry Place
Don Mills, Ontario
M3B 2A7
(416) 445-4436

Dr. S. Venugopal *
Department of Surgery
University of the West Indies
Mona, Kingston 7
Jamaica, W.I.

Dr. Carl Eric Walburgh *
400 W. Brambleton Ave., Suite 200
Norfolk, Virginia 23510

Dr. John F. Waldron *
2545 Chicago Ave. So., Suite 104
Minneapolis, Minnesota 55404
(612) 871-4551

Dr. David E. Wesson
The Hospital For Sick Children
555 University Avenue
Toronto, Ontario
M5G 1X8
(416) 597-1500
Dr. Nathan Wiseman  
Children's Hospital  
685 Bannatyne Avenue  
Winnipeg, Manitoba  
R3E 0W3  
(204) 787-2682

Nathan and Eva  
43 Folkstone Boulevard  
Winnipeg, Manitoba  
R3P 0B4  
(204) 832-2805

Dr. Philip J. Wolfson  
1025 Walnut Street  
Room 607-A  
Philadelphia, Pennsylvania 19107  
(215) 928-7635

Dr. Salam Yazbeck  
Hôpital Ste. Justine  
3175 Chemin Cote Ste. Catherine  
Service de Chirurgie  
Montreal, Quebec  
H3T 1C5  
(514) 737-5448

Salam and Diane  
1672 Beaudet  
St. Laurent, Quebec  
H4L 2K6  
(514) 744-5229

Dr. Sami A. Youssef  
444-5757 Decelles Avenue  
Montreal, Quebec  
H3S 2C3  
(514) 737-5448

Sami and Donna  
21 Bellevue  
Westmount, Quebec  
H3Y 1G4  
(514) 933-7221

* - Associate Member  
** - Life Member