

PAEDIATRIC SURGERY

**PS01  
RESCUE HYPOTHERMIA REDUCES ILEAL INJURY IN  
EXPERIMENTAL INTESTINAL ISCHAEMIA AND REPERFUSION**

G. STEFANUTTI, A. PIERRO, E. J. PARKINSON, V. V. SMITH AND S. EATON

*Institute Of Child Health, Middlesex, United Kingdom*

**Aim** Moderate hypothermia is beneficial when applied throughout intestinal ischaemia-reperfusion (IR). However, therapeutic intervention is usually possible only after ischaemia has occurred. The aim of this study was to evaluate the efficacy of hypothermia as a rescue therapy for intestinal IR.

**Methods** Adult rats were ventilated via a tracheostomy and underwent either intestinal ischaemia-reperfusion (60 min superior mesenteric artery occlusion, and 120 min reperfusion) or sham operation. Rats in the hypothermia groups were maintained at normothermia (36–38°C) throughout ischaemia, and were cooled at the beginning of reperfusion until reaching target temperature (moderate hypothermia: 30–32°C). Four groups (n = 8 each) were studied: 1) control normothermia; 2) IR normothermia; 3) control hypothermia; 4) IR rescue hypothermia. The degree of histological injury in terminal ileum was assessed on a semi-quantitative scale (1 = low; 5 = high) by three blinded observers. Data (median [IQ range]) were compared by Kruskal-Wallis test with Dunn's post-test.

**Results** Intestinal IR at normothermia caused severe injury to the ileum (4.5 [4–5]) compared to both normothermic (2 [1.5–2],  $p < 0.01$  vs. IR normothermia) and hypothermic controls (2 [1.5–2],  $p < 0.01$  vs. IR normothermia). However, rescue hypothermia offered considerable protection from IR injury, so that intestinal architecture was partly preserved (3 [2.5–4],  $p = n.s.$  vs. control normothermia;  $p = n.s.$  vs. control hypothermia).

**Conclusions** Moderate hypothermia reduces the extent of tissue injury following intestinal ischaemia, even when applied as a rescue therapy. It could be considered as a possible therapy in clinical conditions associated with intestinal IR.

**PS02  
ATRESIA OF THE JEJUNUM AND ILEUM: THE DISTINCTION**

A. T. TONGSIN

*Queen Sirikit National Institute of Child Health, Bangkok, Thailand*

**Purpose** Atresia of the jejunum and ileum is one of the major causes of neonatal intestinal obstruction. Most affected newborn infants present with bilious emesis and abdominal distention. Traditionally, jejunal and ileal atresia have been grouped together as jejunoileal atresia. From our observation and experience, we found many differences regarding clinical features and outcome between these two groups. The purpose of this study is to elucidate the difference between jejunal and ileal atresia.

**Methodology** A retrospective analysis of patients diagnosed with jejunal or ileal atresia during 1991–2005 was carried out.

**Results** There were 68 patients with jejunal atresia and 56 with ileal atresia. The mean birth weight and gestational age of patients with jejunal atresia were significantly lower than those with ileal atresia. Antenatal perforation occurred more frequently in ileal atresia. Postoperative course was more prolonged and mortality was higher in jejunal atresia. Prolonged ileus and anastomotic dysfunction requiring long-term parenteral nutrition were the major causes of complications leading to death.

**Conclusion** There were many differences between patients with jejunal atresia and those with ileal atresia. The more compliant jejunal wall allows massive dilatation upon obstruction with subsequent loss of peristaltic activity, thus poorer outcome in comparison with ileal obstruction. We suggest that atresia of the jejunum and ileum be considered distinctly.

**Reference**

1. Heij HA, Moorman-Voestermans CGM, Voa A: Atresia of Jejunum and Ileum: Is it the Same Disease? *J Pediatr Surg* 25:635–637, 1990

**PS03  
IS THE PREDICTED OUTCOME OF DIAPHRAGMATIC HERNIA  
DIAGNOSED IN UTERO, DETERMINED BY POSTNATAL CARE?**

G. A. B. RUSSELL, N. ANDERSON AND A. KANG

*Canterbury District Health Board, Canterbury, New Zealand*

Outcome of isolated left congenital diaphragmatic hernia (CDH) has been predicted by the position of the fetal liver and lung:head ratio (LHR) on ultrasound scans between 22–28 weeks gestation (1). Variations in postnatal management will determine if the findings of such studies can be generalised.

This retrospective study aimed to (1) determine if outcome, predicted by antenatal findings, was achieved in a small regional centre and (2) identify what postnatal factors could adversely alter a predicted favourable outcome.

**Method** All fetal cases of isolated CDH diagnosed between 1998 and 2006 were included if liver position and LHR could be measured and babies were born alive. Survival to discharge was compared to the predicted chance of survival (1). Significant physiologic disturbances, possibly due to care practices including lung overdistension, were identified.

**Results** Ten cases were identified. The median (range) gestation at earliest estimate of liver position and LHR was 27 (16–35) weeks. The median (range) of LHR was 1.28 (0.63–3.35) and part of the liver was in the thoracic cavity in 2/10. The overall survival was 7/10 (70%). Eight babies had a predicted survival of 70%. Three cases predicted to have a favourable outcome had significant and unexpected perioperative deterioration related to lung overdistension, and 2 died.

**Conclusion** Failure to achieve expected outcome in individual cases may be determined by avoidable postnatal factors.

- (1) Jani J et al Prenatal prediction of survival in isolated left sided diaphragmatic hernia. *Ultrasound Obstet Gynecol* 2006; 27: 18–22

**PS04  
DELAYED DIAGNOSIS OF ANORECTAL MALFORMATION:  
THE NEED FOR CAREFUL NEONATAL EXAMINATION**

C. E. ETHERIDGE, A. J. A. HOLLAND AND S. V. S. SOUNDAPPAN

*The Children's Hospital Westmead, New South Wales, Australia*

**Purpose** To determine the frequency of delayed diagnosis of anorectal malformation in infants admitted to our institution and determine the resulting morbidity and mortality.

**Methodology** A retrospective review was performed of all anorectal malformations treated at the Children's Hospital at Westmead between November 1995 and November 2006. A detailed review was performed of those infants in whom the diagnoses was made greater than 24 hours after birth.

**Results** 175 anorectal malformations were seen at this institution over the 11 year review period. Diagnostic delay occurred in 5 (3%); four of which were male. The four males were diagnosed within the first week of life, presenting with abdominal distension, vomiting and intestinal obstruction. The single female was diagnosed at 6 months of age when at onset of solid foods she became acutely obstructed. Delayed diagnosis was found to complicate the surgical repair and may have contributed to functional and psychological issues for patients and their families. One case of delayed diagnosis resulted in the death of the child.

**Conclusion** Delayed diagnosis of anorectal malformations should be entirely avoidable but occurred in 3% of all cases presenting to our institution. It was associated with morbidity and even mortality. Careful inspection of the neonatal perineum at birth should allow all anorectal malformations to be consistently detected.

**PS05  
LONG-TERM OUTCOMES IN NEONATAL SURGERY**

M. D. STRINGER

*Dept of Anatomy, University of Otago, Dunedin, New Zealand*

Historically, for good reason, the main measures used to assess outcome after neonatal surgery have been mortality and early postoperative morbidity. These remain crucial yardsticks of surgical care. However, as results have steadily improved more refined indicators of outcome are needed. Long-term results

and quality of life outcomes are being demanded increasingly by clinicians, parents, and health economists.

For paediatric surgeons, knowledge of long-term outcomes not only enable us to inform parents and patients better about future health expectations but also help us to anticipate potentially avoidable late complications. Outcomes inform us when to operate and when not to operate. Some neonatal surgical conditions and their treatment have almost no long-term sequelae but most have definite consequences of variable severity.

Common themes run through the analysis of long-term outcomes. These centre around the future impact on growth, risk of malignancy, psychological issues, implications for fertility, sexuality and inheritance, and quality of life. There are many reasons why long-term data are lacking. These include the difficulties of maintaining long-term surveillance within healthcare systems overstretched by lack of manpower and funding. Problems of continuity are exacerbated by geographic mobility in society. Robust long-term studies require careful data collection over many years, the co-operation of ex-patients, and familiarity with tools such as independent quality of life measures not readily accessible to paediatric surgeons.

Only by addressing long-term outcomes will the true impact of neonatal surgical conditions be fully understood.

#### PS06 LONG-TERM OUTCOME OF BILIARY ATRESIA IN NEW ZEALAND

T. YU, P. MORREAU, S. BEASLEY, S. BROWN, U. SAMARAKODY AND  
P. MANDHAN

*Multicentre Study, New Zealand*

**Objective** This is a multicenter study to assess the long-term outcomes of children with biliary atresia (BA) who have undergone Kasai operation in New Zealand.

**Material and Methods** This retrospective analysis includes 97 patients who underwent a Kasai operation in the tertiary paediatric surgical centres between 1989 and 2006 in New Zealand. The demographic data, age at diagnosis and surgery, success of surgical procedure, need for liver transplantation and long-term outcomes were studied.

**Results** BA was diagnosed in 58 Māori and Pacific People, 31 Caucasian and 8 Asian children. The average age and time at presentation and surgery was 50.37(±37.3) and 61.6 (±25.7) days respectively. Eighty-six (89%) children had recoloration of the stool and 71 (68%) became jaundice free (bilirubin <20 µmol/L) 1 year after the surgery. Ascending cholangitis was the commonest complication and occurred in 41% of children. Five and 10-year survival rates with native liver were 47% and 23% respectively. Liver transplantation was performed in 50 patients and 38 (76%) survived. The overall actuarial 5 and 10-year survival rates for BA patients were 45.4% and 27.8% respectively.

**Conclusion** BA was more common in Māori and Pacific People children. There was a trend of less favourable results in children who had surgery late (>90 d). Long-term prognosis was not directly related to age at diagnosis and surgery. The survival of patients with and without liver transplantation was similar to that of other series.

#### PS07 URACHAL ANOMALIES IN CHILDREN

B. R. YAPO, B GERGES AND A. J. A HOLLAND

*The Children's Hospital at Westmead, New South Wales, Australia*

**Purpose** Urachal anomalies present infrequently and may be associated with morbidity. We present the experience of our institution in the diagnosis and management of urachal anomalies.

**Methodology** A retrospective review of children presenting to this institution with symptoms secondary to a presumed urachal anomaly between June 1995 and June 2006.

**Results** There were 25 children over a period of 11 years with a likely diagnosis of anomaly at presentation. Ages ranged from 3 days to 13 years with 14 males and 11 females presenting with a granulomatous polyp (16 cases), umbilical discharge (5), umbilical infections (2), and abdominal pain (2). One case was diagnosed incidentally during a renal ultrasound. The main investigative tool was ultrasound (10), followed by micturating cystourethrogram (MCUG) (2). Fourteen of the 25 children had a patent urachus confirmed

by subsequent further imaging or surgery. There was no adverse outcome from operative intervention.

**Conclusions** This review confirmed that whilst clinical examination remain important in the management of a child presenting with a possible urachal pathology, in 36% of the cases the correct diagnosis was only made with further radiological imaging or at operation. Surgical excision was effective with no morbidity or mortality.

#### PS08 BILIARY ATRESIA: POLITICS AND PROGRESS – A UK PERSPECTIVE

M. D. STRINGER

*Anatomy Dept, University of Otago, Dunedin, New Zealand*

Biliary atresia (BA) is a congenital obliterative cholangiopathy of unknown aetiology. It is a rare condition with an incidence of ~1 in 17,000 live births in the UK. BA must be diagnosed promptly in any infant with conjugated hyperbilirubinaemia since prognosis is improved by early detection and timely surgery (Kasai portoenterostomy).

In the UK, a survey of all infants with BA conducted between 1993 and 1995 demonstrated that outcome was related to centre experience. Consequently, in 1999 the management of BA in England and Wales was centralised to three supra-regional paediatric liver units. The results of this national change in practice were carefully audited. This showed that approximately 60% of all infants undergoing Kasai portoenterostomy for BA in these centres now achieve clearance of jaundice (plasma bilirubin <20 µmol/l). Infants with BA that require a liver transplant remain under the care of the same team and can be listed for transplant at the optimum time.

An additional benefit of centralised management has been to facilitate clinical research as a result of concentrating a relatively rare disease into specialist centres with multidisciplinary teams. Recent advances include improved diagnostic accuracy of BA using ultrasound scanning and a better understanding of the role of adjunctive postoperative corticosteroids in improving outcome.

Despite these advances, BA remains the commonest indication for liver transplantation in children. Five-year survival after liver transplantation for BA is currently 80–90%. Current medical and surgical management has transformed a disease that was almost invariably fatal in the 1960s into one with an overall 5-year survival of about 90%.

#### PS09 CONTEMPORARY SURGICAL MANAGEMENT OF SILALORRHEA

J. HORNIBROOK

*Christchurch Hospital, Canterbury, New Zealand*

Persistent sialorrhea (drooling) is often a problem in cerebral palsy, mental retardation and neurological conditions. Management modalities have included behavioural and postural modifications, pharmacotherapy, radiotherapy and surgery. Early surgical treatments were removal of glands, duct ligations and neurectomies. The newest and most conservative operation is relocation of submandibular ducts to the tongue base and removal of sublingual glands. In a personal series of 16 cases 11 were children.

**Purpose** To review the indications and results of submandibular duct relocations (with sublingual gland removal) in 11 children with uncontrolled sialorrhea.

**Methodology** Review of hospital and outpatient records, and illustration of the operative technique on a video. Anatomical variations can be encountered.

**Results** The median hospital stay was 2 days. There were no significant complications. Caregivers judged the efficacy with a median score of "75%".

**Conclusion** Duct relocation with sublingual gland removal is a safe and reliable technique for sialorrhea, with very good control in most cases. In contrast to botulinum toxin treatment its effects are permanent.

## PS10 PHAECHROMOCYTOMA IN CHILDREN

J. M. GLENGARRY AND J. HAMILL

*Starship Children's Hospital, Auckland, New Zealand*

**Purpose** Phaeochromocytomas are rare tumours of the sympathoadrenal neuroendocrine system. It is known that these tumours are associated with Von Hippel Lindau (VHL) disease, multiple endocrine neoplasia type II and neurofibromatosis type I. Recently an association with succinate dehydrogenase type B and D gene mutations has been described. Discovery of a SDHB mutation in a girl at our hospital prompted us to review our experience with phaeochromocytoma in children.

**Methodology** Retrospective review of case notes of all children treated for phaeochromocytoma over the period 1997–2006 at Starship Children's Hospital, NZ.

**Results** Seven patients with 8 tumours were identified with average age of 10 years and average systolic BP of 161. The tumours were adrenal in location in 37.5% and 62.5% extra-adrenal. Our malignancy rate was 12.5%. Familial disease was noted in two patients with one girl having VHL disease and the other found to carry a mutation in the SDHB gene. All children received preop antihypertensives and were treated operatively with seven open resections and one laparoscopic converted to open resection. One child received adjuvant radiotherapy. There were no recurrences or deaths.

**Conclusions** Phaeochromocytoma in children varies from the adult form. It is more likely to be extra-adrenal and have a greater chance of a familial genetic syndrome. The discovery of SDH mutations has altered the diagnosis of familial disease and carries implications for family screening, prognosis and surveillance. We emphasize the importance of taking a good family history and suggest screening for SDH mutations in all patients with phaeochromocytoma.

## PS11 PANCREATITIS – INDICATIONS FOR SURGERY IN CHILDREN

M. D. STRINGER

*Dept of Anatomy, University of Otago, Dunedin, New Zealand*

**Purpose** With modern imaging techniques, multidisciplinary management, and advances in supportive care, what are the indications for surgery in children with pancreatitis and have outcomes improved?

**Methods** During a 12 year period, 62 children [31 girls, median age 12.0 (2.3–16.9) years] with pancreatitis were managed in a single centre. All were investigated using a combination of ultrasound, ERCP, MRCP, and CT. Management comprised supportive care and treatment of any primary pathology. Severe acute pancreatitis (pancreatic necrosis, pseudocyst/abscess, and/or organ failure) was managed with broad spectrum intravenous antibiotics, H2 antagonists, prophylactic antifungal agents, and early parenteral nutrition followed by naso-jejunal feeding.

**Results** 51 children had acute pancreatitis, 14 (27%) of whom had severe disease. The leading causes were trauma (12), pancreaticobiliary malunion (6), gallstones (5), drug-induced (4) and viral (4). In 10 patients no cause was found. Twenty four (47%) of these 51 children required surgery (20) and/or interventional procedures (4) to treat primary pathology (13) or pseudocysts (11) with resolution of pancreatitis in all. Eleven children had chronic pancreatitis: 6 required surgery (5) or an interventional procedure (1).

All 62 patients survived. Only two experienced surgical complications.

**Conclusions** In this series, using modern investigative techniques, only 20% of children with acute pancreatitis had no identifiable etiology. Half of the total series of children with pancreatitis required surgery either to treat primary pathology or to treat complications of the disease. Selective surgical intervention in children with pancreatitis is both beneficial and safe.

## PS12 SONIC HEDGEHOG PATHWAY – A POSSIBLE THERAPEUTIC TARGET FOR HEPATOBLASTOMA

D. ARSIC, M. J. SULLIVAN AND S. W. BEASLEY

*Christchurch School of Medicine & Health Sciences, Canterbury, New Zealand*

**Background and Objectives** It has been shown that the hedgehog (Hh) signalling pathway that is turned off once organogenesis is complete in the

embryo is re-activated in a number of different cancers. We analyzed re-activation of the Hh pathway in hepatoblastoma to investigate whether this pathway can be used as a therapeutic target.

**Methods** Surgically resected hepatoblastoma specimens and hepatoblastoma cell lines were analyzed by immunoblot and PCR, looking into different components of the pathway.

**Results** We found that pharmacological pathway blockade leads to decreased cell proliferation in hepatoblastoma cells, demonstrating an ongoing requirement of Hh signalling in this tumour.

**Conclusion** The Hh pathway may be a useful therapeutic target for hepatoblastoma and other tumours in which the Hh pathway (or a component of the pathway) are over-expressed or re-activated at an inappropriate time.

## PS13 DEVELOPMENT OF THE URINARY BLADDER IN ETU-EXPOSED FETAL RATS IS AFFECTED BY DOWNREGULATION OF SHH AND ITS TARGET GENES

Q. B. QUAN, M. J. SULLIVAN, S. W. BEASLEY AND P. MANDHAN

*Christchurch School of Medicine, University of Otago, Christchurch, New Zealand*

**Objective** The cloaca develops into the urinary bladder and distal part of the hindgut. Abnormalities of cloacal development result in a variety of anorectal and urogenital malformations. This study evaluated the formation of the urinary bladder in ETU exposed fetal rats in relation to shh and its target genes.

**Methodology** Embryos of pregnant SD females administered 1% ETU (125 mg/kg) on the tenth day of gestation (gD10) were collected from gD12-16 and gD21. Cloacal development was viewed with serial histology followed by qualitative and quantitative gene expression studies of shh and target genes to elucidate the expression pattern of these genes during each time point.

**Results** ETU exposed fetuses have incomplete separation of the cloaca leading to a variety of anorectal and urinary abnormalities eg rectourethral fistula. Shh and downstream genes are expressed during urinary bladder development in fetal rats. Relative quantification showed that expression of shh, BMP4 and Hox genes are reduced in ETU-exposed fetal rats during the process of cloacal separation and development of the urinary bladder.

**Conclusion** This study demonstrates that ETU-exposure in fetal rats produces abnormal cloacal development, including urinary fistulae. Shh cascade is active during the process of cloacal separation and urinary bladder development and down regulation of shh, BMP4 and Hox genes during this process may contribute to the abnormal development of urinary bladder and anorectum.

## PS14 FACTORS INFLUENCING THE TIMING OF THE EXIT PROCEDURE FOR OBSTRUCTING FETAL NECK MASSES

H. M. JEFFERIES, S. W. BEASLEY, R. T. BLAKELOCK, B. V. KYLE AND A. P. DIXON

*Christchurch, New Zealand*

The EXIT procedure (ex-utero intra-partum treatment) has become the standard modality for dealing with a fetus when it is anticipated that there will be major problems establishing an airway at the time of birth. Rapid growth and expansion of cervical masses (most commonly cervical teratoma or lymphangioma) may deviate and compress the trachea and oesophagus, and obstruct fetal swallowing. This may cause severe maternal polyhydramnios and places the fetus at risk of death. Cervical masses first diagnosed between 20–24 weeks gestation may be observed to grow rapidly on serial ultrasonography, and require repeated amnio-reduction and drainage of the cyst. The risks of multiple interventions to prolong the pregnancy, the rate of expansion of the solid component of the tumour, and of uterine death have to be balanced against the risks associated with extreme prematurity and delivery by the EXIT procedure to establish an airway.

**PS15****PAEDIATRIC GUN SHOT INJURIES IN NEW ZEALAND:  
A DEMOGRAPHIC ANALYSIS OF AN AVOIDABLE CAUSE  
OF SIGNIFICANT CHILDHOOD MORBIDITY**

A. M. SKINNER, J. HAMILL, U. SAMARAKKODY, E. KIM, B. BOWKETT AND  
S. BEASLEY

*Starship Children's Hospital, Auckland, New Zealand*

**Purpose** New Zealand has a reputation for an outdoors lifestyle that includes hunting and tramping. New Zealanders own over a million guns and these are widely distributed throughout the rural and urban communities. Although its gun laws are not liberal, children still suffer gun shot injuries.

The aim of this study was to examine the injuries children receive from firearms in New Zealand, their circumstances and long term morbidity.

**Methodology** A retrospective review of all children admitted to the four New Zealand paediatric surgical hospitals (at Auckland, Hamilton, Wellington and Christchurch) with gun shot injuries over 10 yr period 1996–2005 was undertaken. Data were compared with published literature from elsewhere.

**Results** Overall, the incidence of gunshot injuries in New Zealand is low compared with many other developed countries. The majority are caused by air rifles. Most are superficial but some may lead to long term morbidity.

**Conclusions** Although New Zealand gun laws require air rifle owners to be over 18 yrs of age, children often are victims of air rifle misuse, particularly in urban areas. Mortality is rare, but some children suffer long term morbidity. Our data may enable preventative measures to be better focused.