

# **GAP Fellowship Presentation**

## **The Development of Paediatric Surgery in Papua New Guinea**

**Mcleo Aite Mathew**

General & Paediatric Surgeon, National Department of Health & UPNG  
Papua New Guinea

Until 1993, complex surgery for children in Papua New Guinea (PNG) was usually conducted by general surgeons or by overseas paediatric surgeons with little coordination and no involvement in teaching programme in the University Department of Surgery. Over the last decade three Australasian surgeons have participated in the Paediatric surgical training, having spent around 40 weeks of teaching and services contributed to the training of the first PNG paediatric surgeon. During the teaching visits, many children have been treated for complex anomalies; strategies have been developed to deal with the more advanced state of the disease for the patients in PNG. Also, protocols have been developed and distributed for anorectal anomalies and Hirschsprung's management with clinical and operative management pathway feasible for PNG and elsewhere in the Pacific Island. Furthermore the choice of colostomy formation, sites, with proactive management of complication is discussed. In general the standard of care for children with surgical condition has significantly improved, new techniques developed, so has the standard of Paediatric Anaesthesia. This paper will explore the lessons learnt through learning curves and personal experiences in PNG with continue support but reduced visits from visiting colleagues from overseas. The success of development of paediatric surgery in PNG is achieved from Royal Australasian College of Surgeons programme funded by AUSAID through Medical Officer, Nursing and Allied Health Programme (MONAHP) and Pacific Island Project (PIP) with support from numerous donors locally assisted with the establishment Dewan Paediatric Surgical Foundation (DPSF) here in Lae, PNG. Other support is ongoing publication of scientific papers, review of Masters thesis and development of a proposal for investigations of the prevalence of renal tract anomalies and overall Incidence of paediatric surgical related congenital anomalies in PNG.

# Oral Presentation Abstracts

## O01

### **Vascular-pedicled costal cartilage graft for the treatment of tracheal stenosis**

**Kohei Hashizume\***, **Yutaka Kanamori**, **Masahiko Sugiyama**

**Tetsuya Tomonaga**, **Hiroko Nakanishi**

The University of Tokyo Hospital, Japan

**Purpose:** To minimize the formation of granulation after tracheoplasty using free costal cartilage graft, a new operative method with vascular-pedicled costal cartilage graft was introduced.

**Method:** Vascular-pedicled costal cartilage graft is prepared using internal thoracic and intercostal vasculatures. After median sternotomy, internal thoracic artery and vein are dissected and taped. They are carefully dissected from the posterior surface of the sternum to the point where 5th intercostal vasculatures are bifurcated. Fifth costal cartilage is dissected with intercostal vasculatures and surrounding muscles. The cartilage graft with vascular-pedicle is ready and is brought to the part of the trachea to be operated.

**Results:** In 3 cases (2 infants, 1 child) with tracheal stenosis, this operation was performed. In all the cases, postoperative bronchoscopy revealed adequate luminal size of the trachea and no granulation tissue at the site of plasty. In one case, however, because of laryngeal dysfunction, tracheostomy became necessary later. At tracheostomy cartilage graft was biopsied, and histological examination revealed normal cartilage structure. In another case, postoperative angiography was performed, which showed well-vascularized cartilage graft.

**Conclusion:** Vascular-pedicled costal cartilage graft is feasible for the treatment of tracheal stenosis in infants and children. With this operation formation of granulation can be minimized.

## The value of lung biopsy in Pulmonary disease in children

Saleh Al Nassar

Pediatric Surgery, King Faisal Specialist Hospital &Research center,  
SaudiArabia

**Purpose:** Open lung biopsy (OLB) claimed to be a sensitive tool for diagnosis of interstitial lung disease. It is reported to be associated with significant morbidity and mortality. AIM: The yield of lung biopsy to make specific diagnosis. , whether it will result in change in therapy. and assessment of morbidity and mortality.

**Method:** This was a retrospective analysis of 91 lung biopsies performed in 83 patients between January 2000 and December 2002. These children were allocated to 3 groups: a. Primary pulmonary pathology (22). b. immunocompromised (49): i. Primary Immunodeficiency (10) ii. Post chemotherapy and BMT (39). c. Pulmonary metastases from solid tumours (20).

**Results:** Overall specific diagnosis reached in 87/91 children (95%) but the change in therapy (excluding lung mets) only in 23/71 (32%). It is lower in those post chemo/BMT 8/39 (20.6%). Postoperative morbidity found to be 11/91 (12%) but procedure related morbidity 3/91 (3.2%). Death within a month of the biopsy was in 6 children (6.5%), only one (1.1%) Procedure related.

**Conclusion:** 1- Open lung biopsy is a safe procedure at our Institution despite reports of high morbidity and mortality. 2- OLB is a sensitive tool to determine the specific cause of pulmonary infiltrate. 3- Change in therapy expected to be only in 32% of patients, and even lower in postchemotherapy and BMT children.

## The Experience of Removal of Palmaz Stent by Rigid Bronchoscope

Chinsu Liu, Taiwai Chin\*, Hsin-Lin Tsai, Wen-Jue Soong  
Choufu Wei\*

Taipei Veterans General Hospital, National Yang-Ming University, Taiwan

**Purpose:** Placement of Palmaz stent for congenital tracheobronchiomalacia and /or trachea stenosis had been reported effective. But the indications of removal are still not determined and the results of removal were only reported in few papers. We report our results of removing Palmaz stent and also discuss the indications of removal.

**Method:** Forty Palmaz stents were placed in 33 patients under flexible bronchoscope during 1997 to 2003 by one pediatrician. Eleven stents in 10 patients were removed by rigid bronchoscope by one pediatric surgeon for various reasons. Most are for expected recovery ( $n=6$ ), some for severe granulations and expected recovery ( $n=2$ ) and some for migration and fracture ( $n=3$ ).

**Results:** The courses of removal are smooth in 8, emergent cardiopulmonary bypass and tracheotomy due to partial collapse of stent during removal in 1, fail due to bleeding in 1, and fail due to strong vagal reflex causing cardiac arrest in 1. At least 6 months follow-up shows satisfactory results in all but the last case that needed multiple operations for complicating tracheoesophageal fistula.

**Conclusion:** Most Palmaz stents can be removed smoothly by rigid bronchoscope. But severe complications still can not be avoided completely. Based on our experience, we suggest the indications to remove the stent are severe granulations, migration, fracture and partial collapse causing clinical airway symptoms rather than expected recovery. The muscle relaxants are avoided during the procedure under general anesthesia. The cardiopulmonary bypass and tracheotomy should be standing by.

## Prenatal Factors Affecting Prognosis in Congenital Diaphragmatic Hernia

**Shinkichi Kamata\***, **Noriaki Usui**, **Masafumi Kamiyama**, **Yuko Tazuke<sup>1</sup>**, **Hirohmi Okuyama<sup>1</sup>**, **Hisayoshi Kawahara\*<sup>1</sup>**  
**Akio Kubota\*<sup>1</sup>**

Department of Pediatric Surgery, Osaka University Graduate  
School of Medicine, Japan

Department of Pediatric Surgery, Osaka Medical Center for Child  
and Maternal Health and Research Ins<sup>1</sup>, Japan

**Purpose:** We have proposed a measurement of lung/thorax transverse area ratio (L/T) as a volumetry of the lung for evaluating the severity of patients with CDH. The aim of this study is to analyze prognostic factors concerning prenatal features including L/T in fetuses with CDH and define the strategy for treatments in fetuses with CDH.

**Method:** Seventy-five fetuses with CDH admitted to Osaka University Hospital and its affiliated hospitals from 1986 to 2002 were retrospectively analyzed. As prognostic factors before birth including sex, gestational weeks at diagnosis, L/T and associated anomalies were analyzed.

**Results:** Thirty-three babies of 80 babies died (41.3% of an overall mortality). Excluding fetuses with associated chromosomal abnormalities, cardiac anomalies, major anomalies and hydrops, 17 of 61 fetuses died (27.9% of mortality). A significant relationship was observed between prognosis and sex, herniated organs in left side hernia, cardiac anomalies, associated major anomalies, fetal hydrops, and L/T, respectively. However, logistic regression analysis revealed that L/T (coefficient 36.3), fetal hydrops (coefficient -17.8), cardiac anomalies (coefficient -3.2), and sex (coefficient 1.5) were significantly and independently associated with the prognosis.

**Conclusion:** While associated anomalies and fetal hydrops are fatal prognostic factors, L/T is useful for evaluating prognosis in CDH patients.

## Paediatric Clean Surgical Wounds: Is Dressing Necessary

Jamal Merei

Jordan University Of Sceince And Technology, Jordan

**Purpose:** The covering of the sutured surgical wound with a sterile dressing is usually considered a routine conclusion to an aseptic operation. The wound is usually left dressed for a minimum of 3-5 days. The main purpose of dressing is protection of the wound against bacterial contamination that remains a significant source of postoperative morbidity. The aim of this study was to compare the infectious local risk when the clean paediatric surgical wounds were dressed or left exposed without dressing after the completion of wound closure

**Method:** Four hundred and fifty one patients with clean surgical wounds were prospectively randomized to receive dressing ( $n=216$ ) or have their wounds left exposed without any dressing ( $n=235$ ) after the completion of wound closure

**Results:** In the group who received wound dressing, 3 patients developed wound infection 1.4% while 4 patients developed wound infection 1.7% in the group who had their wounds exposed without any dressing

**Conclusion:** In children, there was no significant difference in terms of wound infection after applying dressing or leaving the clean surgical wounds exposed without any dressing after completion of wound closure. Dressing clean surgical wounds may be unnecessary

## A usefulness of videofluoromanometry for studying pediatric esophageal motor abnormalities

Hisayoshi Kawahara\*, Natsumi Tanaka, Yuko Tazuke

Takaharu Oue, Hiroomi Okuyama, Akio Kubota\*, Akira Okada\*

Osaka Medical Center and Research Institute for Maternal and Child  
Health, Japan

**Purpose:** Abnormalities in esophageal motor function underlie various gastrointestinal and respiratory symptoms in the pediatric population. Manometry remains an important tool for studying esophageal motility, whereas the conventional methods of its analysis require considerable subjective interpretation. Videofluoromanometry was used in the present study to investigate the characteristics of pediatric esophageal motor abnormalities.

**Method:** Videofluoromanometry was conducted in five patients with primary gastroesophageal reflux disease (GERD)(age,1 - 12 years), four with postoperative esophageal atresia (EA) (3 months - 18 years) and one with diffuse esophageal spasms (DES) (5 years). Videofluoroscopic images were recorded synchronously with manometric digital data in a personal computer. Manometric analysis was conducted by referring concurrent esophageal contour and transportation of contrast medium.

**Results:** Primary GERD patients showed contrast medium moving rapidly into the stomach after swallowing by esophageal peristaltic contractions recorded manometrically, whereas EA patients frequently showed esophageal stasis of contrast medium during defective esophageal peristaltic contractions. GER was seen mostly when the fundus opened during transient lower esophageal sphincter relaxations recorded manometrically. Characteristic corkscrew appearance was seen concurrently with irregular synchronous esophageal contractions in a DES patient.

**Conclusion:** Videofluoromanometry is a useful method to interpret manometric data by analyzing concurrent fluoroscopic images for studying pediatric esophageal motor abnormalities.

## Collis-Nissen fundoplication in patients with long-gap esophageal atresia

Hisayoshi Kawahara\*, Akio Kubota\*, Natsumi Tanaka

Yuko Tazuke, Takaharu Oue, Hiroomi Okuyama, Akira Okada\*

Osaka Medical Center and Research Institute for Maternal and  
Child Health, Japan

**Purpose:** Patients with long-gap esophageal atresia (EA) have an associated short esophagus and often require an antireflux procedure. The Collis-Nissen fundoplication (CNF), which consists of a combination of Collis gastroplasty and Nissen fundoplication, is considered an option in such conditions. The results of CNF were examined in long-gap EA patients.

**Method:** The subjects were four long-gap EA patients (gap > 3 cm) with follow-up from 4 months to 12 years. The CNF was conducted as an antireflux operation in three postoperative EA patients with unreducible sliding hiatal hernia and gastroesophageal reflux (GER) (Gross type A, 2; C, 1), and with primary repair of EA in a Gross type A patient.

**Results:** There were no significant complications except incidental anastomotic leakage in one and anastomotic stenosis that required dilatation in two. GER was adequately controlled in all, but limited esophagitis just above the esophago-gastric junction was observed in a patient with a long mediastinal gastric segment. Swallow-induced pressure reduction in the neo-fundus was detected by manometric examination, whereas swallow-induced peristalsis was not observed in the distal esophagus and gastric segment.

**Conclusion:** The CNF is a safe and effective option for treating EA patients who require esophageal lengthening and control of GER.

## **Circular Myectomy for the treatment of congenital esophageal stenosis due to tracheobronchial remnant**

**Kosaku Maeda\***, Chieko Hisamatsu, Tomomi Hasegawa  
Yutaka Okita

Kobe University Graduate School of Medicine, Japan

**Purpose:** The management of the congenital esophageal stenosis is not well established.

**Method:** We present an infant with critical esophageal stenosis due to tracheobronchial remnant. This lesion was successfully managed by circular myectomy of the esophageal wall without resection.

**Results:** An 1-year-old female was referred for recurrent vomiting and dysphagia. Esophagogram showed an abrupt stenosis and endoscopic ultrasonography showed cartilaginous tracheobronchial remnant within esophageal wall. Initially, she underwent balloon dilatation but had recurrent stenosis after one month. The extirpation of muscular layer, which contained cartilage, was tried. Circular muscular layer was resected in 1cm width leaving mucosal layer intact. Muscular layer was closed horizontally with absorbable sutures. She is free from the symptoms and eats normally for one year after surgery.

**Conclusion:** In case of short segmental stenosis due to tracheobronchial remnant, this may be the preferred approach.

## A simple technique of delayed anastomosis using two magnet-attached tubes in congenital esophageal atresia

Masaki Nio\*, Tomohiro Ishii<sup>1</sup>, Shintaro Amae<sup>1</sup>, Megumi Nakamura<sup>1</sup>, Shigehiko Yoshida<sup>1</sup>, Motoshi Wada<sup>1</sup>  
Yutaka Hayashi<sup>1</sup>

Miyagi Children's Hospital, Japan  
Tohoku University Graduate School of Medicine<sup>1</sup>, Japan

**Purpose:** Primary repair is desirable in cases of esophageal atresia (EA) with tracheoesophageal fistula (TEF), but not always safe enough. We, therefore, developed a simple, new technique.

**Method:** A boy was diagnosed soon after birth as having EA with TEF. His gestational age was 32 weeks and his birth weight 1,750 g. He underwent TEF separation by the extra pleural approach on day 1 of life. He had a long gap and was in a poor general condition. As primary repair was felt risky, the proximal side of the distal esophagus was closed, the two blind ends were roughly approximated with several stitches, and gastrostomy was placed. He developed severe tracheobronchomalacia, and had to be managed on respirator. At 9 months of age, 2 magnet-attached tubes were inserted through a nostril and gastrostomy, respectively. Both ends of the esophagus were continuously compressed together by the magnetic force.

**Results:** Three weeks later, a fluoroscopy demonstrated successful anastomosis. He had no sign of anastomotic stenosis. This boy, now 4 years old, is still being ventilated for his airway problem.

**Conclusion:** This technique, is a safe and useful alternative for high-risk cases of EA, can be utilized to accomplish anastomosis after video-assisted separation of TEF.

## Lap-belt injuries requiring critical care

**Michael Shepherd, James Hamill, Elizabeth Segedin**

Starship Children's Hospital, New Zealand

**Purpose:** To highlight the injuries that result from lap-belt use and make recommendations for prevention, the recent experience at the Paediatric Intensive Care Unit (PICU) of a Regional Paediatric Trauma Centre was reviewed.

**Method:** Retrospective review of admissions to Starship Children's Hospital PICU from 1996 to 2003 following involvement in a motor vehicle crash whilst wearing a lap-belt. Patients were identified from prospectively collected PICU and trauma databases.

**Results:** Ten children were admitted to PICU - 7 of these over the last 3 years. Five were female. Median age was 8.5 years; 4 were  $\leq 5$  years. The median New Injury Severity Score (NISS) was 14, range 10 - 45. Abdominal wall ecchymosis was present in all. Eight underwent laparotomy for hollow viscus injury. Spinal fractures were present in 6 (4 Chance fractures) with 2 children rendered paraplegic. The full lap-belt complex (abdominal wall, hollow viscus and spine) was present in 5. One patient died.

**Conclusion:** Forces transferred to the abdomen via a lap-belt may cause life-threatening injuries or permanent disability. The incidence of serious lap-belt injury does not appear to be decreasing. Three-point harnesses (lap-sash belts) should replace lap-belts in the middle seat of motor vehicles.

## **Adriamycin-induced disruption of Shh pathway associated with abnormal foregut development**

**Dejan Arsic, Bao Quan Qi<sup>1</sup>, Jacqui Keenan, Spencer Beasley\*<sup>1</sup>**

Department of Paediatric Surgery, Christchurch School of Medicine,  
Otago University, New Zealand

Department of Paediatric Surgery, Christchurch Hospital<sup>1</sup>,  
New Zealand

**Purpose:** The hedgehog signalling pathway appears to have a crucial role in embryogenesis. Studies suggest that the Sonic hedgehog (Shh) protein and gene are involved in the differentiation of trachea and oesophagus from the primitive foregut, and that disturbance of the Shh pathway may cause abnormal development of the foregut leading to oesophageal atresia and/or tracheo-oesophageal fistula. This study addresses the exact pattern of Shh gene expression during normal and abnormal foregut development.

**Method:** Expression of Shh gene was investigated by *in situ* hybridization, in control and adriamycin-exposed rat embryos between gestational days 11 and 15, using radiolabelled single-stranded RNA probe.

**Results:** *In situ* hybridization showed that the pattern and the level of Shh gene expression are affected by adriamycin. Adriamycin-treated rats have a deformed notochord and an undivided foregut, and some of the embryos lack the dorso-ventral patterning of the Shh expression seen in control embryos.

**Conclusion:** Shh appears essential to normal foregut development. Mutation in Shh and its signalling components (Gli transcription factors) appear to be responsible for foregut defects. In adriamycin treated rats the abnormal and displaced notochord exhibits high Shh activity, while the activity in the mesenchyme of the foregut was either decreased compared with control embryos, or absent in later developmental stages: these observations support the hypothesis that adriamycin, by interfering with the Shh signalling pathway, disrupts normal foregut development.

**Study on effects of bar placement on the thorax after Nuss procedure for pectus excavatum using bone scintigraphy**

**Koichi Ohno, Yoshiki Morotomi, Masanao Ueda, Hiroto Yamada**

Osaka City University, Japan

**Purpose:** The timing of bar removal has been empirically minimum 2 years after Nuss procedure based on the concept that the operation remodels the thorax gradually. To study effects of a bar on the ribs and sternum, bone scintigraphy was performed.

**Method:** Six boys and five girls (5-21 years of age) were studied with bone scintigraphy, using  $^{99m}\text{Tc}$ -HMDP. Nine patients were studied from 5 to 17 days after Nuss procedure; 3 were done from 20 to 24 months after the operation just before bar removal.

**Results:** In an early phase after the operation, RI accumulation was found at the sternum in 1 of 5 patients under 10 years of age, whereas in all 4 older patients, RI accumulated at the sternum or ribs. On scintigrams before bar removal, hot spots were found at the lateral ribs contacting with a bar and at the ribs where the bar passed through intercostal spaces. Furthermore, chest roentgenograms showed the lateral ribs deformed.

**Conclusion:** Nuss procedure fractures the sternum and ribs, especially in older patients. Furthermore, a bar must restrain growth of the ribs and rub against them. This investigation suggests that the bar should be removed shorter than 20 months after Nuss procedure.

## **: CT Scan and the Pediatric Trauma Patient - Are we overdoing it?**

**Stephen Fenton, Eric Scaife\*, Kris Hansen, Rebecka Meyers\***

**Daniel Vargo, Keith White**

University of Utah, USA

**Purpose:** Recent literature expresses concern regarding the increased risk of cancer in the pediatric population exposed to low-dose radiation during computed tomography (CT) examination. Pediatric hospitals have implemented the ALARA (as low as reasonably achievable) concept, but this is not true at most adult referring institutions. The purpose of this study was to assess the overall use of computed tomography in the evaluation of pediatric trauma patients.

**Method:** A retrospective review of the trauma database at a large, level I, freestanding children's hospital with specific attention to the pattern of CT evaluations was conducted.

**Results:** From 1/1999 to 10/2003, 4659 trauma patients were evaluated with 2588 patients undergoing 3575 CT scans. Overall, 50% of obtained scans were interpreted as normal. Forty-three percent of treated patients were transferred from referring hospitals. Of these children, 40% arrived with previous CT scans with 10% requiring repeat imaging. Repeat CT scanning resulted in increased radiation exposure and it is unclear whether or not it contributed to management. Of the children that underwent repeat imaging, 15% were taken to the operating room, 8% received blood products, and 51% were admitted to the ICU.

**Conclusion:** CT scans are used with regularity in the initial evaluation of the pediatric trauma patient. A substantial number of these scans come from referral institutions that may not comply with ALARA. The purported risk of CT radiation questions whether a more selective approach to CT evaluation of the trauma patient should be considered.

## C-reactive Protein (CRP)-Determined Injury Severity is a Major Outcome Factor in Surgical Infants

Walter Chwals\*, Diya Alaedeen, Donald Liu<sup>1</sup>, Eric Leung<sup>1</sup>  
Aaric Queen<sup>1</sup>

Case Western Reserve University School of Medicine, USA  
University of Chicago School of Medicine<sup>1</sup>, USA

**Purpose:** Serum CRP levels reflect the severity of the metabolic response to injury in critically ill children. During this period, caloric overfeeding can increase complications and delay recovery. We hypothesized that by avoiding excessive caloric delivery, the effect of injury severity would be the major factor determining clinical outcome.

**Method:** After IRB approval, serum CRP levels, measured energy expenditure (MEE), respiratory quotient (RQ), and hospital length of stay (LOS) was obtained in 25 surgical infants (mean 19, range 1-80 days old). Daily energy intake minus MEE (I-E) was recorded. Statistical correlation was established using Pearson coefficient analysis.

**Results:** CRP (4.6+/-6.5 mg/dl) was significantly correlated to LOS (85.6 +/- 85.9 D, p=0.0001). When I-E did not exceed 5 calories/kg/D (9 patients), CRP (6.0+/-5.7 mg/dl) positively correlated with RQ (0.92+/-0.04, p=0.05). When I-E exceeded 5 calories, CRP (4.0+/-6.9 mg/dl) was positively-associated with increased RQ (0.96+/-0.1) but without significant correlation (p=0.33).

**Conclusion:** These data suggest that CRP-measured injury severity is a major determinant of clinical outcome in surgical infants. In addition to CRP, overfeeding may cause additional RQ elevation and further delay recovery in critically ill infants.

## Electron Beam CT is a Valuable Imaging Tool for the Pediatric Surgical Patient

**Mark Holterman, Ai-Xuan Holterman, Vladimir Jelnin**

**Carlos Ruiz**

University of Illinois at Chicago, USA

**Purpose:** Traditional computed tomography has been the mainstay diagnostic tool for complex abdominal and thoracic processes. Increasingly abdominal CT scans are used by emergency room physicians to diagnose common conditions such as appendicitis. In addition to the extra cost of these tests there are increasing concerns about radiation induced malignancies. Electron Beam CT(EBCT) is a relatively new technology with several potential advantages over traditional CT: 1) fast acquisition times, 2) 1/10th the radiation exposure, 3) robust software enabling real-time interactive 3-D visualization, 4) improved surgical planning, and 5) reduced hardware costs.

**Method:** Children with a variety of thoracic and abdominal disease processes were imaged using EBCT and traditional CT. These conditions included: pulmonary hypoplasia, thoracic dystrophy, retroperitoneal masses, and a Wilm's tumor.

**Results:** The new EBCT has comparable spatial resolution to the Multislice CT but much higher temporal resolution, thus providing equivalent diagnostic images with much faster acquisition times, less artifact and much lower radiation. Rendering volumetric 3D images took less than 4 min, and the images could be readily evaluated to guide surgical procedures.

**Conclusion:** EBCT is an effective user- and patient-friendly alternative to traditional CT in the care of pediatric surgical diseases.

**Tracheal obstruction caused by dislocation of an internal metallic stent: A case of successful removal of the stent using ECMO**

**Hiroomi Okuyama, Akio Kubota\*, Hisayoshi Kawahara\***

**Takaharu Oue, Yuko Tazuke, Natsumi Tanaka\***

Osaka Medical Center for Maternal and Child Health, Japan

**Purpose:** Although an internal stent has been applied to tracheomalacia, it can cause serious complications. We report a case of secondary tracheal obstruction caused by a balloon expandable metallic stent (Palmaz stent).

**Method:** A 3-month-old girl with severe tracheomalacia had placement of a Palmaz stent. After repeated balloon dilatation of the stent, she weaned from artificial ventilation at 1 year of age. At 3 years of age, she developed progressive dyspnea due to tracheal stenosis. CT scan showed that her trachea had been almost obstructed because of dislocation of the stent. After initiation of ECMO, the trachea was exposed. The stent was found to penetrate the membranous wall of the trachea. The thick and edematous trachea was divided and the stent was removed along with the 2cm-long membranous wall of the trachea. Slide tracheoplasty was performed to reconstruct the trachea.

**Results:** She weaned from artificial ventilation in a week. The postoperative course was uneventful. She has been free from respiratory symptoms for 1 and half years after the operation.

**Conclusion:** Tracheal obstruction is one of the serious complications caused by a balloon expandable metallic stent. ECMO in combination with slide tracheoplasty is a safe way to solve this problem.

## Case of the Missing Penny: Thoracoscopic Removal of a Mediastinal Coin

**Mehul V. Raval, Brendan T. Campbell, J. Duncan Phillips\***

Division of Pediatric Surgery, University of North Carolina at Chapel Hill, USA

**Purpose:** Coin ingestion by children is common. Although rare, reported complications include esophageal perforation, tracheoesophageal fistula, esophago-aortic fistula, and death. We describe the first reported thoracoscopic removal of a mediastinal coin that migrated extra-luminally from the esophagus in a 23-month-old female.

**Method:** A 23-month-old female swallowed a U.S. penny 12 months prior to being evaluated at our institution for shortness of breath and dysphagia. Chest x-rays showed a coin at the level of the second thoracic vertebra. Rigid esophagoscopy and bronchoscopy revealed that the coin had migrated through the wall of the esophagus and was pushing forward on the membranous trachea causing 50% airway narrowing. Computed tomography (CT) confirmed these findings. Right-sided thoracoscopic exploration using a 3-trocarr technique and 3mm instruments in a modified prone position was used. Coin location was assisted by manipulation of a trans-orally placed Foley catheter and intra-operative fluoroscopy. The coin was in a chronic cavity formed outside of the lumen of the esophagus.

**Results:** The coin was successfully retrieved with neither intraoperative nor postoperative complications. The patient tolerated clear liquids by mouth within 24 hours and was discharged two days after the procedure.

**Conclusion:** Although only the first reported case, thoracoscopy appears to be a valuable approach for mediastinal foreign body removal.

## **Distension enterogenesis: Increasing intestinal size and function**

**Jinyoung Park**

Kyungpook National University Hospital and UCLA Children's Hospital, Korea

**Purpose:** The purpose of this study is to evaluate the feasibility of using mechanical force to lengthen small bowel while preserving intestinal function.

**Method:** Male Sprague-Dawley rats had a 1.5 cm jejunal segment isolated out of continuity. A metallic screw was inserted in the proximal end of the isolated segment and the distal end was oversewn. After 4 days, the screw was advanced into the segments by 4.8 mm every other day for two weeks. Segment length was measured preoperatively and at the time of harvest. Alkaline phosphatase and lactase activities of the harvested segment were determined. Comparisons were made with normal jejunum from control animals. Statistical significance was determined by Student's t-test.

**Results:** Segment length following mechanical distension( $n=10$ ) increased  $222\pm27.2\%$  of the original length( $p<0.001$ ). Compared to the control groups( $n=6$ ), the total alkaline phosphatase activity in the lengthened intestinal segment was not different( $0.70\pm0.12$  versus  $0.51\pm0.27$ ,  $p>0.05$ ). Total lactase activity was significantly lower in the lengthened intestinal segment ( $0.31\pm0.12$  versus  $0.07\pm0.04$ ,  $p<0.001$  ).

**Conclusion:** Mechanical force appears to be a viable method for increasing intestinal length. This phenomenon may provide a new method for the treatment of patients with short-bowel syndrome in the future.

## Primary Closure for the Gastroschisis and Omphalocele

Woo-Sup Ahn

Pohang Hospital, Dongguk University, Korea

**Purpose:** To introduce a new procedure applied in several cases of abdominal wall defect. This procedure was obtained by animal experiment.

**Method:** Skin and three layers of abdominal wall muscles were separated laterally to the posterior axillary line. Externalis was divided vertically along the posterior axillary line. Medial margin of internalis was sutured to the divided line of lateral margin of the externalis longitudinally. Medial free margin of externalis and lateral margin of the rectus sutured longitudinally. By this procedure, three layers of abdominal muscles were arranged to two layers. Then the skin of the abdominal wall was closed.

**Results:** This procedure had been applied to 7 gastroschisis and 1 omphalocele. Six gastroschisis patients are alive. One gastroschisis with extensive hypoganglionosis died 125days after operation. One omphalocele patient died of pneumonia at 10th postoperative day.

**Conclusion:** This procedure enabled primary closure of gastroschisis and large omphalocele, shortened the length of ventilatory support and parenteral nutrition, and prevented the complications of infection and intestinal obstruction.

## Acute gastrointestinal and genito-urinary manifestations in children with Henoch-Schonlein purpura

Nicholas. S.Y. Chao, Beatrice P.Y. Wong<sup>1</sup>, Michael W.Y. Leung, K.W. Chung, W.K. Kwok, Kelvin K.W. Liu\*<sup>1</sup>

Queen Elizabeth Hospital, China

United Christian Hospital<sup>1</sup>, China

**Purpose:** To review the incidence and management of acute gastrointestinal and genito-urinary manifestations in children with Henoch-Schonlein purpura (HSP).

**Method:** The records of children admitted with HSP between 1994 and 2003 were reviewed.

**Results:** 165 children were admitted with HSP. 70 children, aged 2-17 years (mean = 6.6) had acute surgical manifestations. 42 of them presented before onset of purpura or arthralgia. Some children had multiple symptoms. Gastrointestinal symptoms included abdominal pain (61), per-rectal bleeding (9), prolonged ileus with ascites (1) and haematemesis (1). Abdominal ultrasonography (USG) was performed in 14 cases. No intussusception was identified. Surgical intervention was not required in all these patients. Genitourinary symptoms included acute scrotal swelling (9) and frank haematuria (2). Scrotal USG was performed in 2 boys. No scrotal exploration was performed. 27 children required systemic steroid for control of disease.

**Conclusion:** Acute gastrointestinal and genito-urinary manifestations are common in children with HSP. Symptoms frequently precede purpura or arthralgia. Vigilant physical examination and ultrasonography in selective cases enable correct diagnosis. Conservative management almost invariably suffices and systemic steroid is useful in selected cases.

## O21

### The etiologic role of intrauterine intussusception and volvulus in jejunoileal atresia.

**Hiroaki Komuro, Tetsuo Hori, Teruyoshi Amagai, Misako Hirai  
Katsumi Yotsumoto, Yasuhisa Urita, Chikashi Goto, Michio Kaneko\***

Department of Pediatric Surgery, University of Tsukuba, Japan

**Purpose:** Although intussusception has been reported as quite a rare cause of jejunoileal atresia (JIA), pediatric surgeons are impressed with the frequent presence of intussusception as well as volvulus at surgery. The aim of this study is to investigate the contribution of intrauterine intussusception and volvulus to the development of JIA.

**Method:** Forty-seven newborns (24 males and 23 females) treated for JIA at Tsukuba University Hospital from 1978 to 2003 were reviewed. Operative and pathological findings were carefully reviewed.

**Results:** Intussusception was responsible for gap and cord type atresia in 12 cases (25.5%). The cord showed an atrophic intestinal lumen in two cases. Volvulus was observed in 12 cases. Volvulus and intussusception were simultaneously observed in one case. This suggested that intussusception was the cause of the atresia, while volvulus was the secondary event. Neither intussusception nor volvulus was observed in high jejunal, apple peel, or multiple atresia. Constriction by abnormal bands or adhesions was causative in five cases. No significant findings were obtained in the remaining 19 cases.

**Conclusion:** Intrauterine volvulus and intussusception were commonly observed in single low JIA. Intrauterine intussusception might be one of the commonest causes of gap and cord type JIA. Volvulus could be not only a cause of JIA but also a resultant event from anatomical changes after the development of JIA in some cases.

## Effects of N-desulfated non-anticoagulant heparin on ALI induced by abdominal sepsis

**Shan Zheng\***, Junyi Han, Leiwei Zhu, Bo Sun

Children's Hospital, Fudan University, Shanghai, China

**Purpose:** To study the effects of N-desulfated non-anticoagulant heparin on acute lung injury (ALI) induced by abdominal sepsis in piglets.

**Method:** 17 piglets were randomly allocated to three groups, Group A: underwent laparotomy without other interference. Group B: a 2-cm perforation was made in the distal cecum. Group C: all procedures were similar to B except heparin derivatives were infused in early ALI. At checkpoints values for hemodynamics, blood routine, blood gas, pulmonary function were recorded. Blood and peritoneal liquid were cultured. The lungs were lavaged for analysis of neutrophil count.

**Results:** Animals in group B had the appearance of ALI as evidenced by decreases of  $\text{PaO}_2/\text{FiO}_2$  ( $<300\text{mmHg}$ ) and  $\text{Cdyn}$  ( $<30\%$ ) with increases of  $\text{Rrs}$  ( $>40\%$ ). Blood neutrophil count in group B increased initially, however decreasing at 8h after onset of ALI. Compared with Group B, group C had higher  $\text{PaO}_2/\text{FiO}_2$  at 8h ( $p<0.05$ ) and lower  $\text{Rrs}$ . Blood neutrophil count in group C increased continually throughout study and neutrophil count in BALF was lower than group B. At same time, histologic observation were much lighter than group B. Blood and peritoneal liquid culture were all positive after interventive operation .

**Conclusion:** N-desulfated non-anticoagulant heparin could alleviate lung injury in this sepsis model by attenuating neutrophil infiltration and activation in the lung without any severe side effect.

O23

## Total Bowel Atresia: X-Linked disease, Report of three brothers

**Saleh Al Nassar, ZN Al-Hassnan<sup>1</sup>, Maha Faden<sup>2</sup>**

**Abdul Hakiem Kattan<sup>3</sup>**

Surgery, King Faisal Specialist Hospital &Research center, SaudiArabia

Department of medical genetics, KFSH&RC<sup>1</sup>, SaudiArabia

Fellow, Medical Genetics, KFSH&RC<sup>2</sup>, SaudiArabia

Director Neontology Unit, KFSH&RC<sup>3</sup>, SaudiArabia

**Purpose:** Hereditary multiple bowel atresia is extremely rare. Complete bowel atresia involving mid and hind gut is almost unknown.

**Method:** We report three brothers with complete atresia from the second part of duodenum to the anus. The brothers have two normal living sisters confirming it is X-linked inheritance. There was No associated anomaly. Chromasomal studies in two of them, 46,XY. During surgery the atresia was intraluminal with small normal appearing bowel from outside.

**Results:** All three babies died in their first few months of life from sepsis.

**Conclusion:** This is a new disease entity that we could not find similar to this in the English medical literatures.

## A new association of alveolar capillary dysplasia and duodenal atresia with paradoxical dilatation of duodenum

**Noriaki Usui, Masafumi Kamiyama, Shinkichi Kamata\***

**Akihiro Yoneda, Yuko Tazuke, Masahiro Fukuzawa**

Osaka University Graduate School of Medicine, Japan

**Purpose:** We experienced three particular cases of duodenal atresia with paradoxical dilatation of the distal blind end of duodenum associated with persistent pulmonary hypertension of the neonate (PPHN). The aim of this study is to clarify the characteristics of this rare association.

**Method:** Medical charts of the patients were retrospectively reviewed.

**Results:** Case 1 was a 2862g male infant with prenatal diagnosis of duodenal atresia. Dilatation of distal blind end was noticed during duodenoduodenostomy. The infant has developed PPHN after the surgery and expired despite treatments with inhaled nitric oxide (iNO) and ECMO. Lung biopsy revealed alveolar capillary dysplasia (ACD) with misalignment of pulmonary vessels. Case 2 was a 2244g female infant of duodenal atresia. Surgery could not be performed because of PPHN resulted in death. Autopsy revealed ACD with misalignment of pulmonary vessels and duodenal atresia with dilatation of distal blind end. Case 3 was a 2462g female infant with prenatal diagnosis of duodenal atresia and dilatation of distal blind end. She has developed PPHN after the surgery and was extubated successfully by the combined therapy with iNO and intravenous prostacyclin and finally died of refractory PPHN. Retrospective review of prenatal ultrasonography indicated enlargement of high echogenic fetal lungs in all of these three cases.

**Conclusion:** The characteristics of this new association were summarized as a syndrome as follows; fetal lung enlargement, duodenal atresia with paradoxical dilatation of duodenum, refractory PPHN resistant to iNO.

## Prenatal Diagnosis and Management of the Abdominal Diseases in Pediatric Surgery

**Tatsuo Kuroda\***, Yoshihiro Kitano, Toshiro Honna, Haruhiko

Sago, Satoshi Hayashi, Morihiro Saeki\*

National Center for Child Health and Development, Japan

**Purpose:** To study the prenatal courses and management of abdominal surgical diseases.

**Method:** Among 327 patients registered in our fetal treatment board since March 2002, eighty-three fetuses were referred to the surgical department and underwent sequential ultrasonography and magnetic resonance imaging. Their database was reviewed retrospectively.

**Results:** Of 83 patients, abdominal diseases were suggested in 34, chest diseases in 25, genitourinary diseases in 12, and other anomalies in 12. Meconium peritonitis (MP), intestinal obstruction, and abdominal wall defects accounted for casually 65% of the abdominal diseases. Five of 34 developed no gastrointestinal symptoms postnatally. Imperforate anus was seen in 1 of 4 suggested. Five with lung diseases had surgical fetal intervention including open surgery, whereas no intervention was given for abdominal anomalies. MP showed a variety of prenatal courses; low risk patients showed marked decrease of ascites by 35th gestational week, whereas one developed hydrops due to midgut volvulus and massive intestinal necrosis, which indicated the hidden mortality and the potential role of fetal intervention to salvage intestines.

**Conclusion:** Prenatal course of abdominal surgical diseases has a large variety, and close observation may provide the opportunity for fetal intervention as a novel treatment option for the selected diseases.

## Insulin-like Growth Factor-1 Suppresses Ischemia-Reperfusion Injury of Small Intestine in Mice

Wen-Tsung Hung\*, Yun Chen<sup>1</sup>

Department of Surgery, National Taiwan University Hospital and  
Chang-Hua Christian Hospital, Taiwan

Division of Pediatric Surgery, Department of Surgery, National  
Taiwan University Hospital<sup>1</sup>, Taiwan

**Purpose:** Our previous studies demonstrated that enteric luminal perfusion of fetal bovine serum (FBS) protects the small intestine from ischemia/reperfusion injury (IRI) by increasing the intestinal mass and suppression of plasma interleukin-8 level. We also found that FBS treatment can suppress the cellular apoptosis by up-regulated bcl-2 during IRI. Since insulin-like growth factor-1 (IGF-1) is one of the major components of FBS, therefore, in this study, we further investigated the effects of IGF-1 on IRI.

**Method:** The FVB/N mice were treated with IGI-1 (50ug for each) or normal saline intraperitoneally one hour before receiving total I/R of a segment of terminal ileum. The I/R-injured small intestine was obtained at different time points and subjected to terminal deoxynucleotidyl transferase mediated dUTP nick-end labeling (TUNEL) staining, western blot analysis to study the expression of p53, bcl-2, and bax, and caspase activity assay to investigate the activity of caspase 3.

**Results:** In the saline-treated group, the I/R-injured small intestine showed increased cellular apoptosis, suppressed expression of p53 and bcl-2 with decreased bcl-2/bax ratio, and increased activity of caspase 3. In contrast, IGF-1 treatment suppressed the cellular apoptosis, up-regulated p53, bcl-2 and also bax levels with increased bcl-2/bax ratio, and inhibited the activity of caspase 3 ( $P<0.05$ ) at 1hr and 6hr after initiation of reperfusion.

**Conclusion:** The results suggested that IGF-1 can up-regulate bcl-2 and bax level with increased bcl-2/bax ratio and subsequently reduce apoptosis in total IRI of small intestine.

## The Effect of Gastroschisis on the Experimental Diaphragmatic Hernia in Fetal Rabbits

**Yong-Soon Chun, Tae-Kweon Ha, Kwang-Hee Kim**

**Ji-Hoon Yoon, Soo Jin Jung<sup>1</sup>, Woo-Ki Kim<sup>2</sup>**

Department of Pediatric Surgery, Inje University Pusan Paik Hospital, Korea

Department of Pathology, Inje University Pusan Paik Hospital<sup>1</sup>, Korea

Department of Pediatric Surgery, Seoul National University Children's Hospital<sup>2</sup>, Korea

**Purpose:** The purposes of this study is to improve animal model of fetal surgery about congenital diaphragmatic hernia(CDH) using rabbits and to analyze the effect of gastroschisis made by fetal surgery on pulmonary hypoplasia in fetal rabbits with CDH.

**Method:** Twenty-three pregnant New Zealand rabbits underwent hysterotomy and fetal surgery on gestational day 24-27. Two fetuses of each pregnant rabbit were operated. In the fetus of one end of bicornuate uterus, left diaphragmatic hernia was created by excision of fetal diaphragm through open thoracotomy (DH Group). In the fetus of the other end of bicornuate uterus, left diaphragmatic hernia and gastroschisis were created (GS Group). The fetuses were delivered by Cesarean section on gestational day 27-33. Among twenty-three pregnant rabbits, seventeen were born with diaphragmatic hernia and fifteen were born with diaphragmatic hernia and gastroschisis.

**Results:** The most commonly herniated abdominal organ was the left lobe of liver. In DH group, the lungs were hypoplastic with a decrease in lung weight/body weight ratio and an increase of vascular medial wall thickness of pulmonary arteries. The alveolar septae are markedly thickened with increased interstitial tissue and diminished alveolar air spaces. In GS group, the alveolar septae were thickened but narrower than those of DH group and air spaces were increased. In DH group, the pulmonary arterial wall was markedly thickened. In GS group, the pulmonary arterial wall was slightly thickened but was not significantly thickened in comparison with that of DH group.

**Conclusion:** Experimental diaphragmatic hernia results in pulmonary hypoplasia and concurrent gastroschisis formation prevents pulmonary hypoplasia that resulted from diaphragmatic hernia.

## Adrenal Cortical Cell Transplantation

**James Dunn, Yinting Chu, Mandy Lam, Edward McCabe**

**James Atkinson\***

UCLA, USA

**Purpose:** The adrenal cortex is a critical component of the hypothalamic pituitary adrenal/gonadal axis that coordinates the stress response and maintains homeostasis. We hypothesize that adrenal cortical cells can be transplanted in adrenal insufficiency states to regenerate the adrenal cortex.

**Method:** Murine adrenal glands were dissociated into adrenal cortical cells. Cells cultured on a collagen substrate were transplanted under the renal capsule. The implants were procured 1, 4, and 8 weeks later. Total RNA was extracted from the retrieved specimens and was analyzed by polymerase chain reaction.

**Results:** All animals survived the surgical procedure. At implant procurement, a distinct organoid could be visualized under the renal capsule. Histologically angiogenesis was evident in the implant by week 1, followed by glandular formation. The expressions of adrenal specific markers including SF-1, DAX-1, SCC, 21 hydroxylase, 11-beta hydroxylase, and aldolase were detectable up to 8 weeks post transplantation.

**Conclusion:** Primary adrenal cortical cells were capable of function after heterotopic transplantation. Ex vivo gene transfer followed by adrenal cortical cell transplantation could lead to curative therapy for patients with adrenal insufficiency.

## Living-related Intestinal Transplantation For A Patient With Hypoganglionosis

**Tomohiro Ishii, Motoshi Wada, Kotaro Nishi, Takuro Kazama**

**Yoshinori Kawahara, Hideyuki Sasaki, Shintaro Amae**

**Shigehiko Yoshida, Megumi Nakamura, Masaki Nio<sup>1</sup>**

**Tomoaki Kato<sup>2</sup>, Yutaka Hayashi\***

Department of Pediatric Surgery, Tohoku University School of Medicine, Japan

Department of Pediatric Surgery, Miyagi Children's Hospital<sup>1</sup>, Japan  
Division of Liver and GI Transplantation, University of Miami School of Medicine<sup>2</sup>, USA

**Purpose:** Reporting the first case of living-related intestinal transplantation for our patient with hypoganglionosis.

**Method:** A 14-year-old boy with TPN-dependent short-gut syndrome associated with hypoganglionosis underwent living-related intestinal transplantation by using a 150 cm segment of distal ileum taken from a healthy donor. The graft vessels were connected to infrarenal aorta, and inferior vena cava. The immuno-suppressive regimen consisted of daclizumab, tacrolimus, and steroid. The graft surveillance for acute cellular rejection (ACR) was accomplished using zoom endoscopy and mucosal biopsy. The blood trough level of tacrolimus was maintained between 20-25 ng/ml for the first 2 months, followed by 15-20 ng/ml thereafter. The 50mg of daclizumab was administered on day of operation, and same dosage has been repeated once for 2 weeks.

**Results:** The first ACR which was developed POD-9 was progressive, and required a 14-day course of OKT-3 injection. No infectious complication has been occurred. After 2 months of transplantation, graft functions well, and patient seems to be tolerating oral intake, only requiring fluid supplementation of watery stomal output. The donor discharged without any complications.

**Conclusion:** The living-related intestinal transplantation could successfully be performed, and it can be a treatment of choice for patients with short-gut syndrome.

## **Successful Treatment of Gastroschisis with Intestinal Atresia; Report of Two Cases**

**Eriko Yahata, Hideo Yosida, Tadashi Matsunaga, Katsunori**

**Kouchi, Hiroaki Kuroda, Tomoro Hishiki, Naomi Ohnuma**

Chiba University Graduate School Of Medicine, Japan

**Purpose:** Surgical management of gastroschisis associated with intestinal atresia is often difficult. We successfully treated two such cases of gastroschisis, including one with an intestinal rupture.

**Method:** In both cases, primary repair of intestinal atresia was not feasible because of edema of the intestine. The first case had an intestinal atresia at the jejunum. We performed a staged repair of the abdominal wall using a silo and decompressed the gastrointestinal tract through a naso-gastric tube. The second case had a colonic atresia and the caecum was perforated. We constructed a silo pouch and inserted a tube into the intestine through the perforated site.

**Results:** In the first case, the prolapsed intestine was naturally repositioned into the abdominal cavity within six days, and delayed anastomosis was performed on the ninth day of life. In the second case, we made an enterostomy at the perforation site five days after birth. Intestinal anastomosis was performed on day seventy. Postoperative courses were uneventful in both cases.

**Conclusion:** Intestinal atresias in gastroschisis cases can be safely managed by a delayed repair of the atresia. In intestinal atresia cases, a naso-gastric tube is sufficient as a decompression device, whereas a drainage tube is effective in those with intestinal perforation.

O31

## The use of positron emission tomography in detecting hepatoblastoma recurrence : a cautionary tale

**Kenneth Wong, Lawrence Lan, Godfrey Chan, Steve Lin, Paul Tam\***

University of Hong Kong Medical Centre, China

**Purpose:** The use of positron emission tomography (PET) with [18F] fluorodeoxyglucose (FDG) in the detection of recurrences has been well established in many tumour types. Here we present our experience of using this modality in the evaluation of post treatment hepatoblastoma patients.

**Method:** We conducted a retrospective review on patients diagnosed with hepatoblastoma from 1996. FDG-PET imaging was performed together with measurement of alpha-fetal protein (AFP) during post treatment follow-up.

**Results:** 16 patients (8 boys and 8 girls) were identified in our series. The mean age was 23.5 months (range 5 months to 4 years). Three post treatment patients had PET results suggestive of tumour recurrence. One of these patients had normal AFP level and suspected recurrence in the caudate lobe. Radiological guided biopsy was performed three times and there was no evidence of tumour. The other two patients underwent further liver resections because of mildly raised AFP levels. The histology of these showed regenerative liver tissue only with no hepatoblastoma recurrence.

**Conclusion:** Although PET has been gaining popularity as a tool in the detection of tumour recurrences worldwide, we have shown in our series that PET may not be useful in hepatoblastoma patients and caution must be taken in the interpretation of positive results.

## The Prognostic Impact of the Cytoplasmic Survivin Expression in Neuroblastoma

Soo Min Ahn, Jung-Tak Oh<sup>1</sup>, Seok Joo Han<sup>1</sup>, Eui Ho Hwang\*<sup>1</sup>  
Seung Hoon Choi\*<sup>1</sup>

Department of Surgery, Hallym University College of Medicine, Korea

Department of Surgery, Yonsei University College of Medicine<sup>1</sup>, Korea

**Purpose:** The prognostic significance of survivin expression (SE), which has been reported to be an apoptosis inhibitor protein, was examined in order to identify a more accurate prognostic grouping of neuroblastoma.

**Method:** Thirty-seven tumor specimens were obtained between 1992 and 2002. The SE level was examined by immunohistochemical techniques, and was scored positive when more than 5% of the cells reacted with the anti-survivin antibody. The outcome of the stratified potential prognostic groups were evaluated according to age, location, stage, Shimada class and SE. The correlation between the SE level and the prognostic factors were analyzed. The predictive value of SE in the prognosis was determined using a multivariate assessment.

**Results:** The median follow up period was 23 months. The 5-year overall survival rate was significantly reduced with old age, the advanced stage, adrenal primary, unfavorable Shimada class, and positive SE. The positive SE correlated with old age, advanced stage, and an unfavorable Shimada class ( $p=.03$ ,  $.00$ ,  $.03$ ). The relative risk of the SE positive group was 5.20 ( $p=.01$ ). Survivin was expressed in 73% of the recurred cases, but in only 31% of the non-recurred cases ( $p=.04$ ).

**Conclusion:** Survivin expression is a valid independent prognostic factor for neuroblastoma, and it correlates with the disease progression and a poor prognosis.

## O33

### Hepatocyte growth factor enhances angiogenesis and tumor cell migration in human neuroblastoma

Yi Ren, Brian Cao<sup>1</sup>, Wen Li, GCF Chan<sup>2</sup>

George F Vande Woude<sup>1</sup>, Paul KH Tam\*

Department of Surgery, The University of Hong Kong, China

Van Andel Research Institute<sup>1</sup>, China

Department of Paediatrics, The University of Hong Kong<sup>2</sup>, China

**Purpose:** Hepatocyte growth factor (HGF/SF) and its receptor, oncogene c-met play important roles in tumor development. In this study, we explored the significance of HGF and c-Met expression in human neuroblastoma and the effect of HGF on angiogenesis and tumor cell migration.

**Method:** The expression of c-Met, HGF, IL-8 and VEGF in neuroblastoma specimens and cell lines was investigated by Immunohistochemical staining, ELISA and flow cytometry, respectively. Invasion chamber were used to study tumor cell migration.

**Results:** HGF was expressed mainly by stromal cells in neuroblastoma specimens. However, the HGF expression by tumor cells was observed in 4 of 32 cases (11.1%). c-Met positive tumor cells were observed in 27 of 32 cases (84%). The results showed that all neuroblastoma cell lines expressed c-Met and one of four cell lines can secret HGF into supernatant of tumor cell culture (450.8 18.5pg/ml). In addition, HGF induced a significant dose-dependent increase in IL-8 and VEGF production. Both recombinant human HGF and the supernatant of tumor cell line culture (HGF positive cell line) enhanced migration of the neuroblastoma cells. This effect was prevented by the addition of a neutralizing anti-HGF antibody.

**Conclusion:** HGF may act as an autocrine/paracrine-acting factor that stimulates angiogenesis and metastasis, and therefore as a potential therapeutic target for neuroblastoma.

## Effect of Post-operative chemotherapy on the serum alpha-fetoprotein level in hepatoblastoma

**Masayuki Kubota\***, **Minoru Yagi\***, **Satoshi Kanada**, **Naoki Okuyama**

**Satoru Yamazaki**, **Shinji Tanaka**, **Keiko Asami<sup>1</sup>**, **Atushi Ogawa<sup>1</sup>**

**Teruhiro Watanabe<sup>1</sup>**, **Michio Kaneko<sup>2</sup>**, **Yukihisa Saida<sup>2</sup>**

Niigata University Graduate School of Medical and Dental Sciences, Japan

Niigata Cancer Center<sup>1</sup>, Japan

University of Tsukuba<sup>2</sup>, Japan

**Purpose:** It is generally accepted that post-operative chemotherapy does not affect the level of serum alpha-fetoprotein (AFP). We herein report three cases who were supposed to show chemotherapy-related changes in the AFP levels after operation.

**Method:** This study included three patients with hepatoblastoma (one case with PRETEXT III and two cases with PRETEXT IV).

**Results:** One patient with PRETEXT III underwent a complete tumor resection and the post-operative AFP level decreased in the normal range. However, he consistently exhibited a transient, two- to three-fold increase in the AFP after each course of chemotherapy for three courses. Chemotherapy had to be ceased due to drug-induced encephalopathy but he has been followed for five years without any evidence of recurrence and the AFP level was also stable in the normal range. In two cases with PRETEXT IV who underwent the curative tumor resection showed similar chemotherapy-related changes in AFP levels. So, these cases were only observed after administrating the routine post-operative chemotherapy instead of administrating further high-dose chemotherapy. The AFP level remained stable for fourteen months and three months after the cessation of chemotherapy in two cases, respectively.

**Conclusion:** In the post-operative chemotherapy of hepatoblastoma, we have to pay close attention to AFP status during chemotherapy as well as the absolute AFP level.

## A Rare Case of Bilateral Stage IV Adrenal Neuroblastoma with Multiple Skin Metastases in A Neonate: Diagnosis, Management and Outcome

Toshihiro Yanai, Atsuyuki Yamataka\*, Hiroo Fujita, Masahiro Saito, Geoffrey Lane, Yuichiro Yamashiro, Takeshi Miyano\*

Juntendo University School of Medicine, Japan

**Purpose:** To review the diagnosis, management, and outcome of bilateral adrenal neuroblastoma (stage IV) associated with multiple skin metastases.

**Method:** Case: A boy born vaginally at 35 weeks had an abdominal mass, and more than thirty 5-30mm skin tumours all over his body. Prenatal ultrasonography was unremarkable. Radiography showed bilateral adrenal tumours with skin, liver and bone marrow metastases. The right tumor was larger and extended across the midline. Urinary VMA and HVA were elevated, and the provisional diagnosis was stage IV neuroblastoma. A course of systemic chemotherapy was commenced on day 19 of life to alleviate respiratory distress caused by growth of hepatic metastases. The right tumour was still present on day 80 of life, and was excised on day 82. Tumor markers increased again at 5 months but returned to normal, with disappearance of skin metastases at 9 months after three courses of systemic chemotherapy. Histology was unfavourable with no amplification of MYC-N. He is well after a follow-up period of 5 years.

### **Results:**

**Conclusion:** Treatment of stage IV neuroblastoma in neonates remains controversial, but the value of a basic protocol for management is highlighted as a result of the successful outcome of this extremely rare case with multiple skin metastases.

## Less invasive surgery is appropriate for advanced neuroblastoma

**Michio Kaneko\*, Misako Hirai, Mayumi Iwakawa, Kiyoshi Ohara, Haruo Ohkawa**  
University of Tsukuba, Japan

**Purpose:** To elucidate the role of surgery combined with intraoperative radiotherapy for advanced neuroblastoma

**Method:** Twenty-three patients older than one year of age with stage 3 or 4 neuroblastoma with or without MYCN amplification treated in our institution since 1985. They received more than 5 cyclic courses of induction chemotherapy of the Japan Advanced Neuroblastoma study group protocols(JANB 85, JANB91 and JANB91) before the surgery combined with intraoperative radiation therapy(IORT). The original tumor and bulky residual lymph nodes only were resected instead of radical systematic lymph node dissection. The adventitia of the aorta and its main branches were left intact to prevent injury to the perivascular nervous tissue. Dose of IORT was 10-15Gy.

**Results:** Two patients recurred preoperatively during induction chemotherapy. The chemotherapy could start 4 to 7 days after surgery strictly on schedule. Fifteen patients are alive without relapse. Five and ten year relapse-free-survival rates were 72%, and 65%, respectively. Local relapse was observed only in 4 patients with preceding or simultaneous relapse of distant metastases. Bilateral kidneys in all cases could be preserved and no immediate or late surgical complications were encountered.

**Conclusion:** Later and less invasive surgery with IORT followed by immediate chemotherapy for advanced neuroblastoma was appropriate and reasonable.

## Urinary diversion in children with pelvic tumours

**David Ben Meir, David Ashley, John Hutson\***

Royal Children Hospital, Australia

**Purpose:** To investigate children presenting with malignant pelvic tumours obstructing the upper urinary tract.

**Method:** The records of 17 children presenting to 2 pediatric medical centres with an upper tract system obstructed by a malignant tumour were reviewed. The decision to divert the upper tract was made by pediatric oncology and urology consultants, based on ultrasound and CT studies performed at presentation. A nephrostomy tube or Double J (DJ) stent was inserted into each obstructed urinary system. All children were subsequently started on chemotherapy. The stents were removed after tumour shrinkage and/or hydronephrosis regression. Alternatively, the nephrostomies were replaced antegradely by DJ stents when longer periods of time were required.

**Results:** There were 9 boys and 8 girls; the mean age at diagnosis was 5.7 years (range 0-12). The obstructing tumours were rhabdomyosarcoma, lymphoma, teratoma, Ewing sarcoma, neuroblastoma, desmoplastic and endodermal sinus tumours. The most common presentation was an abdominal pain. Seven of 17 (41%) presented with renal failure, all of whom but one had normalised after urinary diversion was performed. Of the children, 12 were initially diverted by nephrostomy tubes, 3 by DJ stents, and 2 patients underwent resection of the tumours with ureteroureterostomy. Complications following the insertion of the stents included febrile urinary tract infections (UTI) in 3, and pyelonephritis in one of the children with DJ stents. In the nephrostomy group, 2 developed febrile UTI, and 1 child had a persistent candiduria. The nephrostomy tube fell out in 1 patient, and had occluded in another child. The median follow-up was 2.5 years (range 0-6). Of the 17 children, 9 (53%) have no evident of disease, 2 (11%) are currently under treatment, and 6 (35%) died of cancer.

**Conclusion:** Different types of malignant tumours can obstruct the upper urinary tracts in children, in whom - unlike most adults - the prognosis after treatment is not necessarily bad. Hence, an urgent and optimal upper tract diversion at diagnosis will enable chemotherapy to be started as soon as possible.

## Effects of Recombinant Human Endostatin on Neuroblastoma Xenograft by Three Different Modes of Administration

**Minoru Kuroiwa\***, **Yoshiaki Tsuchida\***, **Jung Hee Lee<sup>1</sup>**  
**Seung Hoon Choi<sup>\*1</sup>**, **Jyoji Yoshizawa<sup>2</sup>**, **Norio Suzuki<sup>\*</sup>**

Gunma Children's Medical Center, Japan  
Yonsei University College of Medicine<sup>1</sup>, Korea  
Jikeikai Medical College<sup>2</sup>, Japan

**Purpose:** We examined the efficacy of recombinant human endostatin (rhEndostatin) on human neuroblastoma by 3 different ways of administration.

**Method:** When tumors on the nude mice reached a weight of 90-95 mg, rhEndostatin 10 mg/kg/day was administered subcutaneously to the mice daily for 10 consecutive days in the first experiment and administered continuously with infusion pumps for 3 days in the second experiment and for 9 days in the third experiment. In addition to HE sections, factor VIII expressions were studied immunohistochemically.

**Results:** There was a significant difference in relative tumor weight (RTW) between the rhEndostatin-treated mice and controls on day 2 only ( $p<0.05$ ) in the first experiment, on day 2 through 10 in the second experiment ( $p<0.01$  or  $<0.05$ ), and on days 6, 9 and 12 ( $p<0.01$  or  $<0.05$ ) in the third experiment. In the second experiment, a maximum inhibition rate of 60.7% indicating the efficacy of rhEndostatin was seen and the number of the intra-tumoral vessels immunostained with anti-factor VIII antibody was noticeably reduced ( $p<0.05$ ) in tumors in rhEndostatin-treated mice, compared with that in control tumors.

**Conclusion:** These results clarified that continuous administration of rhEndostatin resulted in a more significant anti-tumor effect than intermittent administration. rhEndostatin is effective only when administered continuously.

## A Giant, Highly Vascular Sacrococcygeal Teratoma: Report of an Excision Using the LigaSure Vessel Sealing System with Review of the Literature

Kazuhiro Kaneyama, Atsuyuki Yamataka\*, Hiroyuki Kobayashi  
Geoffrey Lane, Takeshi Miyano\*

Juntendo University School of Medicine, Japan

**Purpose:** Operative management of giant, highly vascular sacrococcygeal teratoma(GHV-SCT) in neonates is often complicated by life-threatening hemorrhage. We first applied the LigaSure(LS) vessel sealing system to GHV-SCT to control intra-operative hemorrhage, and we discuss its efficacy, with review of other GHV-SCT reports.

**Method:** Case: A boy weighing 3716g, with prenatal diagnosis of GHV-SCT at 26 weeks gestation was delivered by an emergency cesarean section at 34 weeks gestation because of the development of high-output cardiac failure on fetal ultrasonography. At birth, GHV-SCT was 20x17x14cm in size. We operated on him 4 hours after birth for fear of development of thrombocytopenia, coagulopathy, and post-natal high-output cardiac failure. Through a Pfannenstiel skin incision, followed by a lower midline fascial incision, the aorta was surrounded with a rubber tape(Vesselloops) just above the aortic bifurcation. Both ends of the tape were brought through a rubber tube to form a snare. The loop was tightened to occlude the aorta, and was secured with a clamp. There was no prominent feeding artery(such as the middle sacral artery) supplying to the GHV-SCT. Excision of the GHV-SCT then commenced using monopolar diathermy, but we encountered difficulty in controlling hemorrhage, especially from veins. Also, the need for the ligation of numerous large vessels was extremely time-consuming. Thus, we started to use the LS. All vessels could be successfully sealed with LS, without the need for vessel ligation, thus greatly accelerating the procedure. Once we began using the LS, the GHV-SCT could be completely excised in an almost dry surgical field. The total operating time was 255 minutes, however, the time for actual GHV-SCT excision was only 16 minutes. The GHV-SCT weighed 1208g, or 325.1g/kg. Total blood loss was 77ml(20.7ml/kg). Hemodynamic stability was maintained throughout the excision, although during operation the platelet count decreased to 59000 from a pre-operative count of 192000/ul. Histology showed an immature teratoma with no malignant elements. Alpha-fetoprotein levels favorably dropped post-operatively. At last follow-up, 7 months after surgery, he remains well.

**Results:** Review of literature: In reported GHV-SCT cases(n=8) in whom intra-operative devascularization was performed by middle sacral artery ligation or aortic tourniquet, the size ranged from 15x10x7 to 25x14x11cm, the tumor weight per body weight ranged from 245.7 to 556.7g/kg, the intra-operative hemorrhage per body weight from 22.5 to 311.0ml/kg, and the operating time from 220 to 330 minutes.

**Conclusion:** Based on this experience, the LS vessel sealing system was extremely useful to control intra-operative hemorrhage in GHV-SCT excision. We strongly recommend its selective use.

## Long-term survival of advanced stage Wilms' tumor correlated with the initial histology

Mikako Ueno, Hiroko Suda, Shintaro Hayashida, Hironori Iwasaki, Satoshi Ida, Takayuki Takeichi, Hideaki Okajima Katsuhiro Asonuma, Yukihiro Inomata

Kumamoto University, Japan

**Purpose:** The aim was to perform a clinicopathologic evaluation of Wilms' tumor patients in our institution.

**Method:** From January of 1990 to December of 2003, eleven patients were diagnosed with Wilms' tumor. We reviewed the patients' profile, clinical stage, pathology, treatment and outcome.

**Results:** Of these 11 patients, 4 were male and 7 were female ; the median age at diagnosis was 29 months (range; 8-143months). The clinical stage of the patients was I in 3, II in 5, III in 1, IV in 1 and V in 1. The tumor showed favorable histology in 9 cases and unfavorable in 2 cases . Five patients were treated following NWTS and six patients following Japan Wilms' Tumor Study (JWiTS) protocols. The recurrence free survival and overall survival rates at 5 years were 81.8% and 90.9% respectively. All patients with stage III, IV, V had favorable histology and have been doing well without recurrence (range; 3.5-7.5years). Both patients with unfavorable histology suffered from tumor recurrence and one of them died.

**Conclusion:** Despite our limited experience, there were long-term survivors even with advanced stages. In contrast, prognosis of the patients with unfavorable histology was very poor.

## Postoperative Corticosteroid use for Bile Drainage in Biliary Atresia-A Nation-Wide Survey

Toshihiro Muraji\*, Masaki Nio<sup>1</sup>, Youkatsu Ohhama<sup>2</sup>

Takashi Hashimoto<sup>3</sup>, Tadashi Iwanaka<sup>4</sup>, Hideo Takamatsu<sup>5</sup>

Naomi Ohnuma<sup>6</sup>, Tetsuo Kato<sup>7</sup>, Ryoji Ohi<sup>8</sup>

Kobe Children's Hospital, Japan

Tohoku University Hospital<sup>1</sup>, Japan

Kanagawa Children's Medical Center<sup>2</sup>, Japan

Nagoya City University Hospital<sup>3</sup>, Japan

Saitama Children's Medical Center<sup>4</sup>, Japan

Kagoshima University Hospital<sup>5</sup>, Japan

Chiba University Hospital<sup>6</sup>, Japan

Akita University Hospital<sup>7</sup>, Japan

Miyagi Children's Hospital for the Japanese Biliary Atresia Society<sup>8</sup>, Japan

**Purpose:** Members of the Japanese Biliary Atresia Society were surveyed to determine their current practice regarding early corticosteroid use after Kasai operation between 1997 and 2000.

**Method:** Questions included background data, dosage, timing, complications, and ultimate outcome. Anicteric survival with the native liver was statistically compared between groups categorized by various dosing using a Kaplan Meier analysis.

**Results:** Of 54 institutions surveyed, a total of 222 cases with uncorrectable type of BA were collected from 52 responders, including 208 cases with steroid use and 14 cases without steroid use. The survival rate in the steroid and non-steroid groups was 58.0% and 35.7%, respectively ( $p=0.052$ ). The initial prednisone dose was between 2.0 and 3.9 mg/kg/day in 38% and between 4.0 and 4.9 in 35% of patients. Prednisone was started in the first postoperative week in 31% and in the second week in 63%. There was no statistical difference in survival rate between any groups. Perforation and peritonitis was noted in one patient given 3mg/Kg/ day of prednisone on postoperative day one.

**Conclusion:** 1. Most surgeons use steroids. 2. While the anicteric survival rate was higher in the steroid groups, the number of patients in the non-steroid group is too few to draw conclusions.

## Preliminary Report of the Home Intravenous Antibiotic Treatment (HIVA) for Intractable Cholangitis in Biliary Atresia

**Seok Joo Han, Myung-Joon Kim<sup>1</sup>, Soo Min Ahn<sup>2</sup>, Jung-Tak Oh  
Seung Hoon Choi, Eui Ho Hwang<sup>3</sup>**

Department of Pediatric Surgery, Yonsei University College of Medicine,  
Korea

Department of Diagnostic Radiology, Yonsei University College of  
Medicine<sup>1</sup>, Korea

Department of Surgery, Hallym University College of Medicine<sup>2</sup>, Korea

Department of Surgery, National Health Insurance Corporation Ilsan  
Hospital<sup>3</sup>, Korea

**Purpose:** : Intractable cholangitis (IC) in postoperative biliary atresia (BA) patients often causes the progressive liver failure and results in liver transplantation. We have reviewed the effects and role of home intravenous antibiotic treatment (HIVA) in 5 BA patients with IC.

**Method:** All five patients had achieved disappearance of jaundice after Kasai's operation but had IC and intrahepatic cysts. The drainage procedures of intrahepatic cyst were performed to control IC in the first two patients but it did not clear IC. We decided no to do further drainage procedure in remaining three patients because it's unsuccessful in the first two patients. We started the HIVA (HIVA) program in all five patients after insertion of central venous catheter.

**Results:** The HIVA program was successful in all five patients with minor complications of central venous catheter. Duration of the HIVA ranged from 69 to 549 days. Three patients are on HIVA (from 69 to 356 days of HIVA) and two patients are off HIVA (from 244 to 549 days of HIVA) without cholangitis. The cholangitis-free interval in HIVA-off group ranged from 171 days to 901 days. Four patients are anicteric and no patient required liver transplantation.

**Conclusion:** HIVA may be an effective primary treatment for IC after Kasai operation in BA.

## Live donor liver transplantation for fulminant hepatic failure in children

Chi-Leung Liu, Htut Saing\*, Sheung-Tat Fan, Chung-Mau Lo  
Kwong-Leung Chan<sup>1</sup>, Paul Kwong-Hang Tam\*, William Ignace Wei  
Boon-Hun Yong, Nai-Shun Tsoi, John Wong

University of Hong Kong Medical Centre, Queen Mary Hospital, China

Department of Surgery, The University of Hong Kong<sup>1</sup>, China

**Purpose:** To evaluate our results of live donor liver transplantation (LDLT) for fulminant hepatic failure (FHF) in children.

**Method:** Patient demographics, clinical and laboratory data, surgical details, complications, and graft and patient survival of children who underwent liver transplantation in our center are reviewed.

**Results:** Between September 1993 and December 2002, primary LDLT was performed for 26 children: 8 of these children had FHF. Four boys and four girls received left-lateral segment (n=7) and full left-lobe (n=1) grafts. Mean age was 2.9 +/- 1.3 years (range, 3 months to 11 years). Causes of FHF were drug induced in 2 patients and idiopathic in 6 patients. One child received a blood group-incompatible graft. Two patients died; 1 patient of cytomegalovirus infection at 8.6 months and 1 patient of recurrent hepatitis of unknown cause at 2.8 months after LDLT. The child who received a mismatched graft had refractory rejection and underwent a second LDLT with a blood group-compatible graft 19 days afterward. He eventually died of lymphoproliferative disease. Another patient developed graft failure related to venous outflow obstruction and survived after retransplantation with a cadaveric graft. With a median follow-up of 13.2 months (range, 2.8 to 60.3 months), actuarial graft and patient survival rates were 50% and 62.5%, respectively. Survival rates appear inferior compared to those of 18 children who underwent LDLT for elective conditions during the same study period (graft survival, 89%; P=0.051; patient survival, 89%; P=0.281).

**Conclusion:** Although survival outcomes are inferior to those in elective situations, LDLT is a timely and lifesaving procedure for children with FHF.

## Surgical Treatment For Anomalous Arrangement Of The Pancreaticobiliary Duct With Nondilatation Of The Common Bile Duct

**Naomi Iwai\*, Shigehisa Fumino, Tomoki Tsuda, Osamu Kimura  
Yukikatsu Gotoh, Eiichi Deguchi, Kazuaki Tokiwa**

Children's Research Hospital, Kyoto Prefectural University of Medicine, Japan

**Purpose:** The selection of operative procedure for anomalous arrangement of the pancreaticobiliary duct (AAPBD) with nondilatation of the common bile duct (CBD) is still controversial. From the viewpoint of pathophysiology of the bile duct, we investigated which procedure was most effective for AAPBD with nondilatation of the CBD.

**Method:** We encountered 60 children with AAPBD (aged 5 months to 17 years) from 1979 to 2002. All 60 underwent excision of the extrahepatic bile duct with Roux-en-Y hepaticojjunostomy. Six of the 60 were classified as the nondilated type (diameter of the CBD; less than 8 mm), and the other 54 were classified as the dilated type (more than 9 mm). Amylase levels in serum, CBD, and gallbladder were examined for the incidence of hyperplasia and Ki-67 labeling index (Ki-67LI).

**Results:** Diameter of the CBD in 6 patients with nondilated type was 4 to 8 mm. Amylase levels of serum, CBD, and gallbladder in the nondilated type were 1,247+/-610, 26,616+/-1,400, and 48,284+/-1,850 IU/l, respectively. Epithelial hyperplasia in the gallbladder mucosa was present in 5 of the 6 patients with the nondilated type, 10 of 20 with the dilated type, and none of the 6 controls. The Ki-67LI of the nondilated type was 6.82+/-2.11, that of the dilated type was 14.46+/-3.17, and that of controls was 4.93+/-1.35.

**Conclusion:** A free reflux of pancreatic juice into the biliary system was found regardless of nondilatation or dilatation, and cellular proliferative activity of the gallbladder mucosa was increased in both nondilated and dilated type. Therefore, excision of the extrahepatic bile duct including cholecystectomy is recommended for AAPBD with nondilatation of the CBD.

## Cholangitis as a late complication in long-term survivors after surgery for biliary atresia

**Masaki Nio\***, **Nobuyuki Sano**, **Tomohiro Ishii<sup>1</sup>**, **Hideyuki Sasaki<sup>1</sup>**

**Yutaka Hayashi<sup>1</sup>**, **Ryoji Ohi\***

Miyagi Children's Hospital, Japan

Tohoku University Graduate School of Medicine<sup>1</sup>, Japan

**Purpose:** To assess the utility of diagnostic imaging (DM), efficacy of treatment and outcome of late cholangitis in long-term survivors after surgery for biliary atresia.

**Method:** Sixty patients surviving without liver transplantation (LTx) for more than 20 years, were divided into 2 groups depending on whether or not cholangitis developed after age 20. Clinical factors including the type of obstruction, the age at the initial operation, and the early complication with cholangitis were compared between the 2 groups. DM such as CT scan and MRI, clinical courses after treatment of cholangitis and current status of the patients were also evaluated.

**Results:** Twelve patients developed cholangitis after the age 20. There was no statistical difference in the clinical factors studied between the 2 groups. Abnormal DM findings including dilatation of intrahepatic bile duct and hepatic fibrosis were demonstrated in 9 patients with late cholangitis. One patient died, and 2 ultimately underwent LTx. The remaining 9 patients including 4 with good liver function tests have survived without LTx.

**Conclusion:** While the majority of the patients had potential predisposing factors to cholangitis such as dilatation of intrahepatic bile duct, a few patients unexpectedly developed cholangitis without such abnormal findings after an excellent, long-term postoperative course.

## **Effective antibiotic regime for post-operative acute cholangitis in biliary atresia: an evolving scene**

**Kenneth Wong, Alex Fan, Lawrence Lan, Steve Lin, Paul Tam\***

University of Hong Kong Medical Centre, China

**Purpose:** The prompt use of empirical antibiotics is vital in managing post-Kasai cholangitis. We published findings of our clinical trial in 1991 and established the use of cefoperazone, with a response rate of 88.9%. Here we reviewed its clinical use since its introduction and assessed the trend in its efficacy.

**Method:** A retrospective review was carried out between 1997 and 2003. All episodes of acute cholangitis in patients who underwent Kasai procedure were recorded. Cholangitis was defined as unexplained fever with derangement of liver enzymes. Cefoperazone was started empirically according to our established protocol and the response to treatment was analysed.

**Results:** There were 19 patients with a total of 49 episodes of cholangitis. Cefoperazone was used as the first-line empirical antibiotic in 40 of these episodes. Only 30 of them showed successful response (75%). For the 10 unresponsive episodes, meropenem was used as second line antibiotic with complete response in all.

**Conclusion:** The efficacy of cefoperazone in the treatment of post-Kasai cholangitis has dropped over the past years. This suggests a need for a more effective first-line empirical antibiotic. From this review, meropenem seems to be a suitable candidate and a future prospective clinical trial is warranted.

## Ligation of a portosystemic shunt resolves hepatopulmonary syndrome in the Abernethy malformation

**Nobuyuki Morikawa, Tatsuo Kuroda\*, Toshiro Honna**

**Kiyoshi Tanaka, Yoshihiro Kitano**

National Center for Child Health and Development, Japan

**Purpose:** We describe a case of congenital extrahepatic portosystemic shunt (Abernethy malformation) associated with hepatopulmonary syndrome, which was completely resolved by shunt ligation.

### **Method:**

**Results:** A 5-year-old boy presented with a 2-year history of cyanosis. The 99mTc-MAA scintigraphy detected a presence of pulmonary right-to-left shunt, and abdominal CT and angiography clearly demonstrated a portosystemic shunt. The levels of serum ammonia, total bile acid, and lactate were elevated while liver function was normal. Based on these findings, he was diagnosed as congenital portosystemic shunt, which might cause pulmonary vasodilatation leading to right-to-left shunt. The shunt ligation resulted in a marked increase of the intrahepatic portal flow and cyanosis was gradually improved.

**Conclusion:** There are two important messages from the clinical course of this case to consider pathogenesis or treatment for hepatopulmonary syndrome. First, hepatopulmonary syndrome can develop in the setting of Abernethy malformation, which is not associated with cirrhosis. This supports the hypothesis that direct exposure of the lungs to substances from the gut that bypassed hepatic filtration should play a major role in pathogenesis of hepatopulmonary syndrome. Second, hepatopulmonary syndrome can be resolved by ligation of portosystemic shunt, indicating that liver transplantation is not the exclusive therapeutic strategy for hepatopulmonary syndrome.

## **Role of Percutaneous Transhepatic Biliary Drainage for Cystic Dilatation of the Intrahepatic Bile Ducts in Postoperative Biliary Atresia**

**Takashi Sasaki, Toshimichi Hasegawa\*, Takuya Kimura  
Yoshiyuki Ihara, Masahiro Fukuzawa\***

Osaka university graduate school of medicine, Japan

**Purpose:** Cystic dilatation of the intrahepatic bile ducts (CDIB) in postoperative biliary atresia (BA) patients causes cholangitis and is often uncontrollable. We have reviewed the effects and role of percutaneous transhepatic biliary drainage (PTBD) in 5 patients.

**Method:** Four of 5 patients had achieved disappearance of jaundice after Kasai's operation. CDIB was detected at the age from 7 months to 15 years. Two patients had solitary cyst and the other 3 developed multiple cysts. Refractory cholangitis was evident in all patients.

**Results:** Duration of the drainage ranged from 3 to 151 days. Drainage of the bile was good in 3, but poor in 2 patients. Only one patient with solitary cyst has been asymptomatic after achievement of reconnection with the intestine. In the other 4 patients, frequency of cholangitis was decreased or unchanged. However, one patient died of respiratory failure, and the other 3 patients eventually required liver transplantation. The extirpated liver in the dead or transplanted patients showed severe cirrhotic change.

**Conclusion:** PTBD may be an effective primary treatment for solitary CDIB. However, in multiple CDIB patient, PTBD may not be effective and extensive treatment such as liver transplantation may be necessary.

## Histological evaluation and clinical significance of liver fibrosis in patients with congenital dilatation of the bile duct

**Toshimichi Hasegawa\***, **Takashi Sasaki**, **Takuya Kimura**

**Yoshiyuki Ihara**, **Masahiro Fukuzawa\***

Osaka University Medical School, Japan

**Purpose:** Liver fibrosis with a various degree is accompanied in patients with congenital dilatation of the bile duct (CDBD) and may occasionally persist after diversion operation, resulting in liver cirrhosis. This study is designed to evaluate the histological classification of liver fibrosis using intraoperative biopsy and its clinical significance.

**Method:** Forty-three patients with CDBD were included. Age at operation ranged from 1 month to 14 years. Histological classification was made as follows: Grade-0; no fibrosis, -1; confined in the portal area, -2; bridge formation with the neighboring portal area, -3; widened bridging fibrosis, -4; pseudolobule formation, i.e. cirrhosis

**Results:** Grade-0:21(48.8%), -1: 17(39.5%), -2:4(9.3%), -3:1(2.3%), -4:0(0%). Among these groups, there was no significance difference in preoperative serum level of total bilirubin, total bile acid, aspartate aminotransferase and gamma-glutamyl transpeptidase, although grade-2 and -3 groups included the patients with lower age at operation, lower serum amylase or lower amylase level in the bile obtained from the biliary system. Postoperatively, clinical symptoms disappeared and laboratory data were normalized in all patients.

**Conclusion:** Mild to moderate liver fibrosis may be associated in about half of the patients with CDBD. However, liver fibrosis, less than moderate grade, may not influence the postoperative clinical course.

## Delayed Vascular and Biliary Complications in Pediatric Liver Transplantation

**Hideo Kawarasaki\***, **Koichi Mizuta, Shuji Hishikawa**

**Satoshi Egami<sup>1</sup>, Yoshikazu Yasuda<sup>2</sup>**

Transplant Surgery Jichi Medical School, Japan

Pediatric Surgery Jichi Medical School<sup>1</sup>, Japan

Gastro Intestinal Surgery Jichi Medical School<sup>2</sup>, Japan

**Purpose:** Purpose of this study is to investigate proper diagnosis and management of delayed vascular and biliary complications(DVBC) which occur 3 months after liver transplantation(LT) in the pediatric patients.

**Method:** 129 pediatric patients under 18 years with live donor LT have been taken care at the outpatient clinic of our institution. DVBC consisted of hepatic vein stenosis in 6(5%), portal vein stenosis in 3(2%) and biliary stenosis in 2(2%). Clinical course, findings of Doppler US, managements and outcomes have been investigated.

**Results:** Hypoalbuminemia, ascitis and hepato-splenomegaly were observed in the patients with hepatic vein stenosis. Melena and splenomegaly were identified in the patients with portal vein stenosis. Frequent cholangitis was associated in the patients with biliary stenosis. Doppler US showed low flow velocity, dilatation of the hepatic vein and scarce positive wave form in the patients with hepatic vein stenosis. Slow portal velocity and splenomegaly were shown in the patients with portal vein stenosis. Dilatation of intrahepatic bile duct was observed in the patients with biliary stenosis. Interventional venoplasty was performed in all of the patients with vascular complications. Two of the patients with vascular complications died of graft failure. Other seven with vascular complications are surviving two months to six years uneventfully. Surgical reconstruction was performed in one and balloon dilatation through intestinal fiber scope whose technique had been newly innovated by our institution was done in one with biliary stenosis. Both of them are doing well for six months to six years.

**Conclusion:** 1) Hypo-albuminemia, ascitis, melena, splenomegaly and cholangitis are early signs for the diagnosis of DVBC. 2) Doppler US is a useful technique for making an early diagnosis of DVBC. 3) Interventional venoplasty is useful management of delayed vascular complications. 4) Newly innovated interventional technique with intestinal fiber scope is useful for delayed biliary complications.

## STAT3 Confers Resistance against Hypoxia/reoxygenation Injury in Hepatocytes via Mn-SOD

**Keita Terui, Hiroaki Kuroda, Katsunori Kouchi, Tadashi Matsunaga  
Hideo Yoshida, Naomi Ohnuma\*, Sanae Haga<sup>1</sup>, Shin Enosawa<sup>1</sup>  
Michitaka Ozaki<sup>1</sup>**

Department of Pediatric Surgery, Chiba University, Graduate School of Medicine, Japan

Department of Innovative Surgery, National Research Institute for Child Health and Development<sup>1</sup>, Japan

**Purpose:** Ischemia/reperfusion (I/R) injury plays a critical role in post-operative complications of liver surgery. Especially in liver transplantation, I/R injury deteriorates initial liver function after surgery and affects the graft survival. Signal transducers and activators of transcription-3 (STAT3) is one of the powerful inducers of liver regeneration, and recently known to protect cells from various pathogens. The present study was designed to examine hepatoprotective effect of STAT3 against hypoxia/reoxygenation (H/R)-induced stresses.

**Method:** Primary cultured hepatocytes were prepared from male rat by collagen-perfusion method. Adenoviral vectors and cytokines (Interleukin-6 and cardiotropin-1) were added 2 days and 1 hr, respectively prior to the H/R insult.

**Results:** Cytokines that locate upstream of STAT3 protected hepatocytes from H/R-induced apoptosis. Adenoviral over-expression of constitutively activated form of STAT3 (S3-C) or addition of antioxidant (N-acetyl-L-cysteine) reduced apoptosis as well as generation of reactive oxygen species. Interestingly, S3-C induced Survivin and Manganese superoxide dismutase (Mn-SOD) both in protein and mRNA levels. Over-expression of Mn-SOD reduced H/R-induced apoptosis by inhibiting redox-sensitive caspase-3 activity.

**Conclusion:** STAT3 confers resistance against H/R-induced oxidative injury in hepatocytes via Mn-SOD upregulation. Considering its strong mitogenic and anti-oxidant/apoptotic properties, STAT3 seems to be a good therapeutic target for preventing injury and promoting regeneration in liver surgery.

## Resolution of Refractory Postoperative Chylous Ascites Treated with Octreotide

Woo-Hyun Park\*, Jin-Bok Hwang<sup>1</sup>, Soon-Ok Choi

Dept. of Pediatric Surgery, Keimyung University Dongsan Medical Center, Korea

Dept. of Pediatrics , Keimyung University Dongsan Medical Center<sup>1</sup>, Korea

**Purpose:** Chylous ascites is a rare disorder with variable causes. This is a case report of refractory chylous ascites caused by intraoperative disruption of lymphatic channels during Kasai procedure, and treated successfully with octreotide which is a synthetic somatostatin analog.

**Method:** A 3-month-old boy underwent Kasai operation for biliary atresia. At surgery, there noted engorgement of lymphatic channel and multiple lymph node enlargements in the hepatoduodenal ligament.. Chylous ascites (200-300 cc/d) occurred after enteral feeding 10 days after operation. Conservative managements with protein hydrolysate, and subsequently, MCT-rich formula were not effective.

**Results:** With each stepwise increase in the dosage of octreotide by 5 to 10 µg/kg/d every 48 to 72 hours, allowing maximum 40 µg/kg/d, It results in the marked decrease of chylous drainage as well as reduction of the ratio of triglyceride in the lymph to that in serum from 9.5 to 3.3.

**Conclusion:** Octreotide appears to be very effective to treat refractory chylous ascites which is not responsive to other conservative measures. The authors believe that this is the first description of chylous ascites treated successfully with octreotide, which developed after Kasai operation in infancy in the English-language literature.

## Biliary Atresia Associated With Meconium Peritonitis: Contribution Of Prenatal Intraperitoneal Inflammation To The Pathogenesis

Yoshiyuki Ihara, Toshimichi Hasegawa\*, Takuya Kimura

Takashi Sasaki, Noriaki Usui, Akihiro Yoneda

Shinkichi Kamata\*, Masahiro Fukuzawa\*

Osaka University Graduate School of Medicine, Japan

**Purpose:** Association of biliary atresia (BA) and meconium peritonitis (MP) has been rarely reported. We have experienced a case of this combination and observed from the prenatal period, to discuss the pathogenesis of BA.

**Method:** Retrospective review of the case was performed from the chart.

**Results:** At 27 weeks gestation, fetal ultrasound revealed massive ascites, which gradually decreased and disappeared at birth, and echogenic brightness of the entire intestinal wall. Although the gall bladder initially showed the patent lumen, it became atrophic at 34 weeks. A 3748g girl was delivered at 37weeks and diagnosed to suffer from fibro-adhesive type of MP. She did not present with bowel obstruction; however, she gradually developed direct hyperbilirubinemia since the age of 20 days. At the exploratory laparotomy at 68 days, the diagnosis of BA was established. Mild to moderate adhesion was observed in the entire abdominal cavity including the porta hepatis. Histology of the liver biopsy showed moderate bridging fibrosis, proliferation of the intrahepatic small bile ducts, marked infiltration of inflammatory cells in the bile ducts and serositis. The extrahepatic bile ducts showed remarkable fibrotic change. She has been free of jaundice after Kasai's operation.

**Conclusion:** Prenatal intraperitoneal inflammation seemed to be the pathogenesis of BA in this case. Our suggestion was supported by the characteristic appearance of the fetal gall bladder.

## Diagnosis of Hirschsprung's disease in the neonatal period

Weihong Guo, Qinming Zhang, Yongwei Chen, Dawei Hou  
Jinzhe Zhang

Beijing Children's Hospital, China

**Purpose:** 80-90% of Hirschsprung's disease (HD) patients presents in newborns. However, the diagnosis of HD in the neonatal period remains some difficulties since the clinical signs and diagnostic examination might be obscure. Barium enema (BE) and rectal manometry (RM) are the two commonly used non-invasive methods in diagnosis of HD that is believed less precisionness in the neonatal period. Our present study aims to evaluate the criteria that might be helpful in the HD diagnosis during neonatal period.

**Method:** 86 HD suspected patients completed our predetermined study protocol in which BE, RM and full-thickness rectal biopsy were performed. They were divided into 2 aged groups: A, 0-3months(n=57); B, 4-12months(n=29). The diagnosis of HD was made according to the pathological result. Clinical signs, anal examination (tight anus), BE, and RM were selected as the 4 independent factors in diagnosis of HD. The correlation of HD diagnosis to these 4 criteria was analyzed.

**Results:** 45 and 22 HD cases were diagnosed in group A and B respectively. Constipation was the major clinical sign in the group B (90%), however the clinical signs were variable in the group A. Delayed meconium was an independent factor correlated with HD diagnosis in the group B. The accuracy, specialty and sensitivity of HD diagnosis between BE and RM were comparable in two groups. BE and RM were the two independent criteria related to the diagnosis of HD. Short segment HD had a higher incidence of false-diagnosis with the non-HD diseases in both groups.

**Conclusion:** In neonatal period, BE and RM are reliable methods in HD diagnosis. When combined criteria are used, HD diagnosis can be made without further invasive study. However, one has to be very careful with short segment HD patients. Rectal suction biopsy or full-thickness biopsy is needed to avoid false diagnosis.

## Serum Basic Fibroblast Growth Factor as a Marker of Reflux Nephropathy

Hiroyuki Kobayashi, Hideshi Miyakita<sup>1</sup>, Atsuyuki Yamataka\*

Hiroyuki Koga, Takeshi Miyano\*

Juntendo University School of Medicine, Japan

Tokai University<sup>1</sup>, Japan

**Purpose:** Interstitial fibroblasts play a critical role in renal fibrogenesis, and autocrine proliferation of these cells may account for continuous matrix synthesis. Basic fibroblast growth factor (b-FGF) is a mitogenic for most cells and exerts intracrine, autocrine, and paracrine effects on epithelial and mesenchymal cells. We established whether the serum b-FGF has a role as noninvasive marker of renal damage in children with vesicoureteral reflux (VUR).

**Method:** The serum levels of b-FGF were measured in 120 patients aged between 4 month and 15 years, with grade III to grade V VUR, and 21 controls, by a standard enzyme-linked immunosorbent assay technique. 65 children had grade III reflux, 39 grade IV and 16 grade V. Renal scarring was demonstrated in 43 of the 120 children on radionuclide scans.

**Results:** There was no significant difference in serum b-FGF levels in different grades of VUR without scarring (grade III: 13.08 17.67 pg/ml, grade IV: 19.05 23.50 pg/ml, grade V: 6.22 8.53 pg/ml) and controls (6.33 9.41 pg/ml). However, serum b-FGF levels in patients with different grades of VUR with renal scarring (grade III: 81.35 45.41 pg/ml, grade IV: 85.11 42.40 pg/ml, grade V: 61.00 37.09 pg/ml) were significantly higher than in patients with VUR without renal scarring ( $p < 0.001$ ).

**Conclusion:** Our report is the first to document serum b-FGF profiles in children with VUR and scarring. Our finding support measuring of the serum levels of the b-FGF as a simple and noninvasive indicators of renal scarring in association with VUR.

## Analysis of the RET and EDNRB genes in patients with Hirschsprung's disease in Taiwan

Trang-Tiau Wu\*, Wei-Tsui Tsai<sup>1</sup>, Tsung-Ta Chu<sup>1</sup>, Chuan Li<sup>1</sup>

Department of Pediatric Surgery, Chung-Shan Medical University Hospital, Taiwan

Department of Life Science, Chung-Shan Medical University<sup>1</sup>, Taiwan

**Purpose:** Hirschsprung's disease (HSCR), or congenital intestinal aganglionosis, is a relatively common disorder of absence of ganglion cells in the nerve plexuses of the lower digestive tract, resulting in intestinal obstruction in neonate. Mutations in genes of the RET receptor tyrosine kinase and endothelin receptor B (EDNRB) have been identified in HSCR patients. In this study we collected genomic DNA samples from 55 HSCR patients in central Taiwan in order to find out whether there are relationships between Taiwanese HSCR and the RET or EDNRB genes.

**Method:** We examined 55 patients (Male 44, Female 11) diagnosed with HSCR (Sporadic 53, Familiar Twin 2). Forty four patients (M:F37:7) with SSA, 9 patients (M:F6:3) with LSA, and 2 patients (M:F1:1) with TCA. DNA samples from these patients have been analyzed by PCR amplification and direct sequencing for mutations and polymorphisms in 20 exons of the RET, and 4 exons of the EDNRB genes. We also used restriction enzyme analysis with 50 normal control DNA samples to confirm the polymorphism.

**Results:** In this study we detected three polymorphisms in exon 2 and 13 of RET, and exon 4 of EDNRB in 55 HSCR patients.

**Conclusion:** This study represents the first comprehensive genetic analysis of sporadic HSCR disease in Taiwan. However, in contrast to most other HSCR investigations, mutations in RET and EDNRB genes have not been detected yet in this study.

## Is Left-Colon Antegrade Continence Enema (LACE) superior to Right-Colon ACE?

**Seung Hoon Choi\*, Soo Min Ahn<sup>1</sup>, Jung-Tak Oh, Seok Joo Han  
Sang Won Han<sup>2</sup>, Eui Ho Hwang\***

Department of Surgery, Yonsei University College of Medicine, Korea

Department of Surgery, Hallym University College of Medicine<sup>1</sup>, Korea

Department of Urology, Yonsei University College of Medicine<sup>2</sup>, Korea

**Purpose:** The success and complication rates of left-colon antegrade continence enema (LACE) are comparable with those of right-colon ACE (RACE). The aim of this study was to determine which procedure was superior in terms of patient convenience.

**Method:** From 1999 to 2003, 17 patients with fecal incontinence or intractable constipation due to meningocele (14), cloacal anomaly (1), anorectal malformation (1), and lipoma of the spine (1) underwent ACE procedure: 12 LACE using retubularized sigmoidostomy and 5 RACE using appendicocaecostomy. The two groups were compared in terms of the duration of enema, interval between enemas, the volume of fluid used, and the severity of abdominal discomfort after fully adjusting to the appropriate enema regimen and achieving continence control.

**Results:** Sixteen patients (88%) achieved fecal continence control and median follow-up was 14±8 months. The mean duration of LACE was shorter (22.0±8.4 minutes to 48±12.5 minutes, p=.01) and the fluid volume was lower (250±136 ml to 1210±287 ml, p=.00) significantly than those of RACE. No patient with LACE showed any abdominal discomfort or pain.

**Conclusion:** LACE provided more convenient continence control than RACE by allowing shorter enema duration, lower fluid volume requirement and less abdominal discomfort

## Effects of early vesicostomy in obstructive uropathy on bladder development

**Yuriko Sato, Hiroaki Kitagawa\*, Kevin.C. Pringle<sup>\*1</sup>, Junki Koike<sup>2</sup>**

**Jane Zuccollo<sup>3</sup>, Richard Robinson<sup>4</sup>, Hideaki Sato, Yasuji Seki  
Koonosuke Nakada\***

Division of Pediatric Surgery, St. Marianna University School of Medicine, Japan

Department of Surgery, Obstetrics & Gynecology, Wellington School of Medicine and  
Health Sciences<sup>1</sup>, Newzealand

Department of Pathology, St. Marianna University School of Medicine<sup>2</sup>, Japan

Department of Obstetrics & Gynecology, Wellington School of Medicine<sup>3</sup>, Newzealand

Department of Urology, Capital & Coast District Health Board<sup>4</sup>, Newzealand

**Purpose:** Creation of a vesico-amniotic shunt for obstructive uropathy removes the normal fetal urination cycle. It is unclear how this affects bladder function at term. We measured the bladder volume and reviewed the bladder histology after fetal vesicostomy.

**Method:** We created an obstructive uropathy in fetal lambs at 60 days gestation by ligating the urethra and urachus. Vesicostomy (female) or urethrostomy (male) were performed at 21 days after obstruction to release the obstruction. The fetuses were sacrificed at term (145 days).

**Results:** Thirteen fetuses were shunted. Seven fetuses miscarried after shunting. Six survived and three had a successful shunt with a very small bladder (5 to 7 ml). Two had incomplete shunts that failed some time after shunting. These both had huge bladders (399ml). In one, the obstruction was unsuccessful. Histological examination demonstrated that the obstruction caused bladder muscle hypertrophy. Shunted lambs had severe fibrosis of the bladder wall and very poor bladder compliance.

**Conclusion:** Shunt operations after obstructive uropathy may salvage the kidney but fail to preserve bladder function. We conclude that the fetus needs the urination cycle for normal bladder development. This requirement exists even when the obstruction is successfully bypassed.

## Early Feeding and Shortened Hospital Stay after Colostomy Takedown in Patients with Imperforate Anus

**Chau-Jing Chen\***

National Cheng-Kung University Hospital, Taiwan

**Purpose:** This study reviewed a 15-year experience of 94 patients to identify an optimal feeding protocol and hospitalization plan for patients with imperforate anus who underwent colostomy takedown (CT).

**Method:** The author treated 145 patients with imperforated anus from June 1988 to August 2003. Among them, 94 underwent a 3-stage operation (colostomy, posterior sagittal anorectoplasty, and CT). Traditionally, patients started feeding on the 2nd or 3rd postoperative day (POD) and went home on the 5th or 6th POD (n=10). In 1991, the author adopted a protocol in which patients received feeding on the 1st POD and were discharged on the 2nd POD (n=21). Then, in 1994, a more aggressive protocol permitted feeding 6 hours after CT. However, one suffered anastomotic leak, which was resolved three days later after abstinence of feeding. Thus the 1st POD feeding protocol was resumed in 1995 and 59 more patients were added to this 1991 program.

**Results:** Of the 80 patients following the 1991 protocol, feeding started at 20.2 hours (range, 17.4-41.5), and the mean postoperative hospital stay was 1.92 days (range, 1.71-3.16). Three cases encountered minor complications but recovered after conservative treatments. Of the 4 patients following the 1994 protocol, one suffered anastomotic leakage.

**Conclusion:** Our 1st POD feeding and 2nd POD discharge plan is safe and efficient for patients with imperforate anus who underwent CT.

## A role of interstitial cells of Cajal for motility disorder in the rat small bowel ischemia and reperfusion injury

**Naoki Shimojima, Yasuhide Morikawa\*, Ken Hoshino  
Toshio Nakaki<sup>1</sup>, Hiroshi Ozaki<sup>2</sup>, Masatoshi Hori<sup>2</sup>, Masaki Kitajima**

Department of Surgery, Keio University School of Medicine, Japan  
Department of Pharmacology, Teikyo University School of Medicine<sup>1</sup>, Japan  
Department of Veterinary Pharmacology, Graduate School of Agriculture and Life Sciences, The University of Tokyo<sup>2</sup>

**Purpose:** The objectives of the present study are to develop the rodent model of ischemia and reperfusion (I/R) injury of small bowels and to assess the role of interstitial cells of Cajal (ICCs) in the motility disorder of the affected intestines.

**Method:** Male Wistar rats (N=5) were anesthetized by isoflurane inhalation. The superior mesenteric artery and collaterals were clamped by microaneurysm clips for 80 min. After 12 h of reperfusion period, animals were sacrificed and the ileum was harvested to assess the mechanical contractile activities (MCAs) in the organ bath. ICCs were stained by the c-Kit immunohistochemical staining with the whole-mount preparations. Sham control rats (N=5) were treated in an identical fashion except for vascular clamping.

**Results:** The frequency of MCAs is diminished in I/R rats (I/R, 17.9 +/- 4.7 cycles per min vs. sham, 26.7 +/- 2.0 cycles per min). The network of c-Kit positive cells were significantly decreased in I/R, whereas sham revealed strong and distinct activities (I/R, 27990 +/- 17884 pixels per area vs. sham, 136397 +/- 31808 cycles per min).

**Conclusion:** The present study suggests that the deficient expression of c-Kit positive cells may contribute to the motility disorder after I/R injury.

## Renal Angiomyolipoma in Japanese Tuberous Sclerosis Patients

**Yoshihiro Kitano, Toshiro Honna, Tatsuo Kuroda\*, Kenji Nihei**

National Center for Child Health and Development, Japan

**Purpose:** Renal angiomyolipoma (AML) is a benign neoplasm but can grow aggressively in tuberous sclerosis (TS) patients. The aim of this study was to document the characteristics of renal AML Japanese TS patients.

**Method:** Medical records of 29 TS patients followed at our center were reviewed for the presence, size, symptom, and treatment of renal AML.

**Results:** Twenty-four patients had undergone more than one imaging study of the kidney. No mass was detectable in eight (Group 0). AMLs less than 1cm in diameter were detected in five (Group 1). AMLs 1-4 cm in diameter were detected in four (Group 2). Massive AMLs larger than 10 cm in diameter were detected in seven (Group 3). When present, AML always affected both kidneys and were multiple. All patients in Group 0-2 (10 male, 7 female) were asymptomatic, and the tumors seemed stable in size. Tumors in Group 3 (2 male, 5 female) tended to grow aggressively causing pain, hematuria, or fever, which was difficult to control. Total or partial nephrectomy, transarterial embolization were performed with limited success.

**Conclusion:** AML in TS patients can be stable or aggressive. Patient care becomes very difficult once the tumors have grown massive. Prospective follow-up program is mandatory for timely intervention.

## Waardenburg-Shah syndrome with SOX10 mutation: Case report and literature review

**Norio Suzuki\***, Fumiaki Toki, Ken Inoue<sup>1</sup>, Makoto Suzuki, Kyoko Hirakata, Kyoko Nagai, Minoru Kuroiwa, James Lupski<sup>1</sup>  
Yoshiaki Tsuchida

Gunma Children's Medical Center, Japan  
Baylor College of Medicine<sup>1</sup>

**Purpose:** Waardenburg-Shah (W-S) syndrome is a term to denote intestinal aganglionosis with a Waardenburg complex

**Method:** We report two cases of W-S syndrome, and one of them with SOX10 mutation.

**Results:** Case 1: A female infant with short segment aganglionosis had a blue iris in the left eye, as her mother did. An auditory-brain stem response (ABR) test showed a pattern of sensory hearing loss, and she underwent radical surgery with success. Case 2 : A male infant with ultra-short segment aganglionosis had circumscribed leukodermia and bilateral iris depigmentation. ABR test showed a pattern of sensory hearing loss, and she received trans-anal radical surgery with success. A heterozygous mutation was found in SOX10 with alteration in the codon 376.

**Conclusion:** A review of 48 cases of W-S in the literature showed that the extent of the aganglionic segment is quite variable, from nearly total to ultra short, and the female to male ratio was 19:23, and these clinical features resemble to those of Hirschsprung disease associated with Ondine curse. Endothelin-3, endothelin-B receptor and SOX10 genes have been identified recently as disease-causative genes, as in Case 2.

## Posterior Sagittal Anorectal Myectomy for Repair of Ultra-short Segment Hirschsprung's Disease

Nahm-gun Oh

Department of Surgery, Pusan National University, College of Medicine, Korea

**Purpose:** In small portion of patients with Hirschsprung's disease, the aganglionic segment extends only up to the mid-rectum. Lynn's myectomy for the ultra-short segment Hirschsprung's disease is well-known procedure, but it has still some technical difficulty in the case of infant or severe anal stricture, due to its narrow operative field. This survey is intended to introduce a new modification as a posterior sagittal anorectal myectomy (PSAM), which offers wider operative field and thus enables the sufficient resection of posterior anorectal muscle easily.

**Method:** A sagittal incision was made between anus and coccyx, so that the rectum could be exposed. A longitudinal strip of muscular layer is removed in 1 cm width of the aganglionic portion of the anorectum from the upper rectum to the dentate line. This approach has been applied to 16 cases of ultra-short segment Hirschsprung's disease at our hospital for past 8 years and an estimate of bowel function was made according to SanFilippo method.

**Results:** 1) Postoperative diagnosis of ultra-short segment Hirschsprung's disease confirmed in 14 cases. 2) Iatrogenic mucous perforation during procedure occurred in 2 cases, but immediate primary sutures were performed without any postoperative leakage. 3) The bowel function was rated excellent in 5 cases(35.7%), good in 7 cases(50%), and fair in 2 case(14.3%). 4) One case of ileal atresia was reported as comorbidity. 5) 1 case was found to be short segment Hirschsprung's disease involving sigmoid colon, and the other 1 case was to be total aganglionic Hirschsprung's disease.

**Conclusion:** This new PSAM is considered to be an easy and effective alternative for ultra-short segment Hirschsprung's disease.

## The early effects of urinary tract obstruction on glomerulogenesis

Hiroaki Kitagawa\*, Kevin. C. Pringle<sup>\*1</sup>, Junki Koike<sup>2</sup>, Jane Zuccollo<sup>3</sup>

Yuriko Sato, Hideaki Sato, Shinichirou Fujiwaki<sup>4</sup>, Mieko Odanaka<sup>2</sup>

Koonosuke Nakada\*

Division of Pediatric Surgery, St. Marianna University School of Medicine, Japan

Department of Surgery, Obstetrics & Gynecology, Wellington School and Health Sciences<sup>1</sup>, Newzealand

Department of Pathology, St. Marianna University School of Medicine<sup>2</sup>, Japan

Department of Obstetrics & Gynecology, Wellington School of Medicine<sup>3</sup>, Newzealand

Department of Obstetrics & Gynecology, Yokohama City Seibu Hospital<sup>4</sup>, Japan

**Purpose:** Our obstructive uropathy model in fetal lambs showed that renal cystic changes appeared 3 weeks after obstruction. In this study, we studied the changes resulting from complete urinary tract obstruction in the first 7 days after obstruction.

**Method:** We created an obstructive uropathy in fetal lambs at 60 days gestation by ligating the urethra and urachus. They were delivered 48 hours, 3 days, 5 days, and 7 days later by caesarian section. The kidneys were removed and processed for histologic examination.

**Results:** Eighteen fetuses were operated upon and 15 (4 at 48 hours, four at 3 days, 2 at 5 days and 5 at 7 days:- 86%) survived. Macroscopically, bladder dilatation and slightly dilated ureters were identified from 48 hours. Microscopically, dilatation of proximal tubules started from 48 hours after obstruction and increased by 7 days. Glomerular cysts in the nephrogenic zone were also identified from 48 hours. Dysplastic changes were not found.

**Conclusion:** The first areas in the developing kidney that suffer damage after obstructive uropathy are the proximal tubule and the nephrogenic zone. This change started 48 hours after obstruction. We conclude that shunting procedures need to be performed considerably earlier than we had previously thought.

## Is there a role for Hedgehog genes in Hirschsprung's disease?

Merce M Garcia-Barcelo, Vincent CH Lui, Mai-Har Sham<sup>1</sup>

Paul KH Tam\*

Department of Surgery, The University of Hong Kong, China

Department of Biochemistry, The University of Hong Kong<sup>1</sup>, China

**Purpose:** Hirschsprung's disease (HSCR) or aganglionic megacolon has a complex genetic aetiology with approximately 50% of the patients unexplained by mutations in the major HSCR genes. Indian Hedgehog (Ihh) mutant mice present with a phenotype reminiscent of HSCR disease (dilation of intestine and absence of ganglion cells) and Sonic Hedgehog (Shh) mutant mice have defective plexus formation. The requirement of IHH and SHH signalling for the proper development of the ENS, together with the evidence presented by the mutant murine models, prompted us to investigate the possible involvement of the human IHH and SHH genes in HSCR disease.

**Method:** By PCR amplification and direct sequencing, we screened for mutations and polymorphisms the coding regions of IHH and SHH of 87 HSCR patients and 96 ethnically matched controls.

**Results:** Neither mutations nor HSCR associated polymorphisms were found in IHH or SHH coding-regions. Only 23.8% of these patients harboured potential disease-causing mutations in other HSCR candidate genes.

**Conclusion:** We propose that faulty cis or trans regulatory mechanisms of these genes may contribute to the defective ENS development. DNA alterations should be investigated at the promoter and DNA regulatory elements of both IHH and SHH and in those genes implicated in the HH signalling pathway.

## Anterior Pubic Osteotomy for Patients With Exstrophy of Bladder

**Hamid Reza Foroutan\***

Nemazee Hospital, Iran

**Purpose:** Urinary incontinence is a common problem in patients with exstrophy of bladder. Different methods have been performed for their treatment. Here we present our experiments with anterior pubic osteotomy for correction of urinary incontinence in patients with exstrophy of bladder.

**Method:** Fourteen patients with exstrophy of bladder underwent primary bladder closure and reconstructive surgery. Anterior pubic was performed in 10 patients as the primary procedure and in four as a corrective surgery in older age.

**Results:** The symphysis pubis was relatively easily approximated after osteotomy. There were 9 male and 5 female. All the male patients needed bladder neck reconstruction and antireflux procedure. However two of the female patients who had anterior osteotomy with their primary bladder closure and the other two who had the osteotomy at 5 years of age, developed urinary continent.

**Conclusion:** Anterior pubic osteotomy is a simple procedure that can approximate intersympheal band and cause urinary continence. The procedure is suitable both at newborn age and older patients. The success rate is higher in females.

## From gunstore to smoking gun: tracking guns that kill children in North Carolina

**Brendan Campbell, Deborah Radisch, J. Duncan Phillips\***

**Daniel von Allmen**

University of North Carolina Hospitals, USA

**Purpose:** The aim of this study was to track firearms from the time of purchase to their involvement in pediatric firearm related deaths.

**Method:** We reviewed autopsy reports for all children that died from firearm-related injuries in North Carolina from January 1999 through December 2002. Data obtained included demographic information, type of firearm, and manner of death. We also reviewed data from the Bureau of Alcohol, Tobacco and Firearms which traced guns involved in crimes and determined the time elapsed from purchase to their involvement in a crime (i.e., time-to-crime).

**Results:** During the study period 40 children died from firearm injuries. Mean age was 7.6 years. Most victims were male (n=26, 65%). Handguns were responsible for the majority of deaths (59%), followed by shotguns (27%), rifles (10%), and unknown (10%). Most deaths were homicides (67%), followed by accidental (18%), suicide (13%), and undetermined (2%). Most crime guns (76%) were purchased from Federally licensed dealers, and many (40%) had a time-to-crime of less than 3 years.

**Conclusion:** Legally purchased firearms pose a significant threat to children in North Carolina. A more restrictive approach to the sale of firearms, particularly handguns, is a logical approach to reducing pediatric firearm related deaths in the United States.

## Protective effects of estrogen on intestinal ischemia/reperfusion injury in pubertal rats

Xianmin Xiao \*, Dehong Liu, Shan Zheng \*

Children's Hospital of Fudan University, Shanghai, China

**Purpose:** This study investigated the role of 17 beta-estradiol (E2) in intestinal ischemia/reperfusion (I/R) injury and possible mechanism.

**Method:** Rats of pubertal age were ovariectomized and injected subcutaneously with vehicle (vehicle group) or E2 (100 or 500ug/kg/every other day, E2 or 5E2 group) for 4 weeks. Other rats underwent sham ovariotomy as control group. The rats in each group (n=15) were subjected to superior mesenteric artery occlusion followed by 60 minutes (n=5), 6 hours (n=5), or 24 hours (n=5) reperfusion. Intestine specimens were then obtained for the determination of histologic score, inducible nitric oxide synthase (iNOS) mRNA, and iNOS activity.

**Results:** Histologic score was found lower in E2 group ( $2.57 \pm 0.12$ ) than in vehicle ( $3.31 \pm 0.12$ ,  $p < 0.01$ ), 5E2 ( $2.98 \pm 0.08$ ,  $p < 0.05$ ) and control group ( $3.00 \pm 0.09$ ,  $p < 0.05$ ). The expression of iNOS mRNA in E2, 5E2, and control group was increased by 14.4-, 1.1-, and 10.9-fold, respectively, compared with vehicle group. This expression was greater ( $p < 0.05$ ) in E2 group than in any other group. The iNOS activity was consistent with the expression of iNOS mRNA.

**Conclusion:** A moderate level of estrogen may exert protective effects on intestinal I/R injury in pubertal rats, probably by enhancing iNOS mRNA expression and its activity.

## Congenital Colonic Atresia

Soon-Ok Choi, Woo-Hyun Park

Department of Pediatric Surgery, Keimyung University Dongsan Medical Center, Daegu, Korea

**Purpose:** Colonic atresia (CA) is a very rare cause of intestinal obstruction usually seen in neonate at term with no other associated pathology. The aim of this study is to delineate the clinical characteristics of CA with special emphasis on high incidence of associated malformations.

**Method:** A retrospective clinical study was performed about six neonates with CA who were treated in our department between 1989 and 2003.

**Results:** 3 boys and 3 girls aged 1 day to 4 days. Two were premature and one boy was one of heterozygotic twins. Three with prenatal diagnosis presented with intestinal obstruction. Plain abdominal X-ray films showed findings of intestinal obstruction in all cases. A barium enema demonstrated a distal atretic segment and microcolon in 4. The types of atresia were I (n=2), II (n=2), and III(n=2). Atresias were located in ascending colon (n=3), in transverse colon(n=2) and in descending colon(n=1). All except one had associated anomalies; multiple small-intestinal atresias (n=1), gastoschisis and multiple ileal atresia (n=1), malrotation with Ladd band (n=1) segmental dilation of the T-colon, ascending colon perforation with volvulus, and accessory spleen (n=1), and meconium peritonitis with midgut volvulus(n=1). The initial management was an enterostomy and segmental resection of small and/or large bowel in 4 patients, and primary anastomosis in the remaining 2. Immediate postoperative death occurred in patients with associated gastoschisis and multiple ileal atresias. Definitive surgery was performed at age of 2-6 months without complication.

**Conclusion:** The type of surgery and associated abnormalities are the major determinants of outcome. A two-stage procedure consisting of an emergency enterostomy for decompression as the first stage and an elective resection with anastomosis a few months later is recommended. The thorough exploration of the abdomen is emphasized because atresias may be multiple or may be associated with additional gastrointestinal anomalies.

## A Preanal Extrasphincteric Inversion Proctoplasty Using An Illuminating Intrarectal Indicator

**Nahm-gun Oh, Jin-yong Sin, Hae-young Kim**

Department of Surgery, Pusan National University  
College of Medicine, Korea

**Purpose:** To introduce a new modification of a preanal extrasphincteric inversion proctoplasty(PEIP) using an illuminating intrarectal indicator, in which the transperineal positioning of the anal canal was performed without division of anal sphincter muscle.

**Method:** Point-A was designated 0.3mm anterior to the anal dimpling center. A semicircular incision was made in the front of the anus. The flap which included from the skin to the upper margin of the external anal sphincter, and then it was everted posteriorly. A quarter of frontal upper rim of the external anal sphincter was exposed and the retrourethra-oriented center of the uppermost anal sphincter was designated as the other point-B. From the point-A toward the point-B, a spinal needle was inserted through the anal sphincter, and its canal was dilated under direct identification of electronic stimulation confirming the sphincter muscle location. Illuminating intrarectal indicator with laparoscopic light source were pushed through the distal stoma of sigmoid colostomy toward its distal lumen so that the blind rectal pouch was protruded to the perineal incision. A cruciated incision(+) was made on the fistula site of the blind pouch, and it was anastomosed to anal point-A(x).

**Results:** In a case, the urethral injury was found intraoperatively, and immediately repaired, and healed without any sequelae. One case of anal stenosis was improved after several Hegar dilatation. The postoperative bowel function by the Kirwan's clinical assessment at 12 months was grade I in 9 cases, grade II in 2 case.

**Conclusion:** This PEIP with illuminating intrarectal indicator shows acceptable clinical results, and is considered to be an effective optional alternative for anorectal malformations.

## Laparoscopy-assisted Surgery for Prenatally Diagnosed Small Bowel Atresia: Simple, Safe and Virtually Scar-free

**Atsuyuki Yamataka\***, **Hiroyuki Koga**, **Akihiro Shimotakahara**  
**Masahiko Urao**, **Hiroyuki Kobayashi**, **Geffrey Lane**  
**Takeshi Miyano**

Juntendo University School of Medicine, Japan

**Purpose:** To describe a new technique for the surgical management of prenatally diagnosed small bowel atresia.

**Method:** The medical records of three neonates with prenatally diagnosed small bowel atresia treated by laparoscopy-assisted surgery between March and July, 2003 were reviewed. Under general anaesthesia, a 5mm trocar was inserted using an open technique through an intraumbilical incision. The proximal atretic bowel end was identified using laparoscopy. An additional 3mm trocar was inserted in the left lower quadrant, and the atretic end was mobilized toward the umbilicus. The umbilical trocar was then removed allowing the atretic end to be seen directly through the umbilical wound. A ring retractor was inserted into the umbilical trocar site and was used to expand the wound easily to deliver both the proximal and distal atretic ends. A midline fascial incision was not required. The excessively dilated portion was then excised, and the entire distal small bowel was examined through the wound. A standard bowel anastomosis was performed, and the repaired intact bowel was returned to the abdomen. The umbilical fascia and skin were closed conventionally.

**Results:** Birth weights were 2910, 3110, and 3512g, respectively. Laparoscopy-assisted surgery for bowel atresia (Las-BA) was performed within 24 hours in all three cases. Preoperative abdominal distension was minimal in two cases. Thus, the atretic bowel ends could be identified easily, and Las-BA completed successfully. Postoperatively, there was minimal abdominal scarring and the umbilicus was normal in appearance. In the third case, abdominal distension was already present at birth, and it was difficult to find the atretic bowel ends. Laparoscopy was converted to laparotomy.

**Conclusion:** Although our experience is limited to three patients, our technique is simple, safe and virtually scar-free, and can be applied for the treatment of neonates with prenatally diagnosed small bowel atresia, especially if there is minimal abdominal distension at birth.

## Laparoscopic Injection of Dermabond Tissue Adhesive for the Repair of Inguinal Hernia: Short- and Long-term Follow-up

Go Miyano, Atsuyuki Yamataka\*, Yoshifumi Kato  
Eri Tei Geoffrey J. Lane, Hiroyuki Kobayashi, Takeshi Miyano\*  
Juntendo University School of Medicine, Japan

**Purpose:** To determine if laparoscopic injection of octylcyanoacrylate tissue adhesive (Dermabond; Db) into the inguinal hernia sac (IHS) is effective for the repair of inguinal hernia.

**Method:** 30 male 4-week-old Lewis rats (mean weight: 85g), were used as subjects because they have large patent IHS. A 5mm laparoscope was inserted in the epigastrium, and patency of both IHS was confirmed. In the right Db (R-Db) group (n=10), an 18-gauge angiocath was inserted in the right lower quadrant, a fine catheter was passed through it into the peritoneal cavity and directed to the right IHS. 0.2 ml Db was injected into the IHS under laparoscopic control to close the orifice of the right IHS. The left side was not treated. In the bilateral Db (B-Db) group (n=10), both IHS were treated. In the no Db (N-Db) group (n=10), only laparoscope insertion was performed, and this group acted as the control group. In all rats, herniography using Gastrograffin was performed just before sacrifice. Rats in the B-Db and N-Db groups were mated 50 days after surgery to check fertility. Half the rats from each group were sacrificed 2 months after surgery to assess short-term outcome, and the remaining half were sacrificed 12 months after surgery to assess long term outcome. At sacrifice, the abdominal cavity was examined, and the entire IHS, the spermatic cord, and the testis were excised and examined histologically.

**Results:** All rats survived until sacrifice. After mating, sperm were identified in the vaginas of all 10 female rats mated with B-Db group rats, and 9/10 delivered normal newborn rats. The fertility rate was the same as for the N-Db control group. Herniography showed that treated IHS were closed (no flow of Gastrograffin) and untreated IHS were patent (hernia sac full of Gastrograffin). At sacrifice, herniography findings were confirmed; at both short- and long-term follow-up, all treated IHS in the R-Db and B-Db groups were successfully closed, and non-treated IHS in the R-Db and N-Db groups were patent. There were no adhesions between the closed IHS orifice and small bowel in 17 of the 20 treated rats, and only minor adhesions in the remaining 3. Histology showed localized mild inflammation within the IHS adjacent to the Db. The spermatic cord and testes were normal in all rats.

**Conclusion:** Our results suggest that our new technique is simple, safe and reliable as an alternative to standard operative repair of inguinal hernia.

## Clinical Outcome of Laparoscopically Assisted Endorectal Pull-through in Hirschsprung's Disease: Comparison of Abdominal and Perineal Approaches

**Akio Kubota\***, **Hisayoshi Kawahara\***, **Hiroomi Okuyama**  
**Takaharu Oue, Yuko Taduke, Natsumi Tanaka, Hisazumi Uenaka**  
Osaka Medical Center for Maternal and Child Health, Japan

**Purpose:** Laparoscopically assisted endorectal pull-through (EPT) via a perineal approach using prolapsing technique (PA) for Hirschsprung's disease (HD) has been reported. However, clinical outcome after this approach is not well documented. The purpose of this study is to compare the clinical outcome of PA and a conventional transabdominal approach (TA).

**Method:** In the period between 1990 and 2001, 21 infants with HD underwent EPT with PA (Group L) and 20 with TA (Group O). There was no difference of age and weight distribution between the two groups. The clinical outcome was evaluated 3 years after surgery.

**Results:** The operation time was compatible in the two groups (5.2 0.8 vs. 4.9 0.8hrs), whereas blood loss (36 30 vs. 98 52ml) and postoperative complications requiring surgical intervention (0% vs. 26%) were significantly lower in Group L. The incidence of postoperative enteritis (28% vs. 27%), and voluntary defecation (>once/2 days; 87% vs. 70%) were compatible in the two groups. Soiling (small amount of involuntary stooling; >1/month) was significantly fewer in Group L (14% vs. 45%).

**Conclusion:** Laparoscopically assisted ETP with PA is less invasive and can provide better clinical outcome compared with TA in terms of postoperative soiling.

## Laparoscopically Assisted Anorectoplasty for High and Intermediate Imperforate Anus - Utility of Perineal Ultrasonographic Guide

**Akio Kubota\***, **Hisayoshi Kawahara\***, **Hiroomi Okuyama**  
**Takaharu Oue, Yuko Tazuke, Natsumi Tanaka, Akira Okada\***  
Osaka Medical Center for Maternal and Child Health, Japan

**Purpose:** As minimal invasive surgery, laparoscopy assisted anorectal pull-through has been reported with new devices. However, it is difficult to create an accurate pull-through canal (PTC), because of the narrow space between the urethra and puborectal sling. The authors describe a new method employing peritoneal ultrasonography.

**Method:** The rectourethral fistula was dissected laparoscopically. Externally, electrostimulation identified the center of the muscle contraction, over which a 1.5 cm of skin incision was made, and the lower part of PTC was created by hemostat forceps amid muscle contraction. An ultrasonographic probe applied to the perineum demonstrated the urethra, and the forceps was advanced behind the urethra into the pelvic cavity using the ultrasonographic guide. Anorectal pull-through was performed following dilatation of the PTC with dilators.

**Results:** The authors applied this procedure in 5 cases of male high and intermediate anomalies. Surgical damages to the levator and vertical muscles were avoided with this procedure. Postoperative fluoroscopic study demonstrated good anterior angulation and intact contraction and relaxation of those muscles.

**Conclusion:** The combination of laparoscopic dissection, pinpointing the center of anal sphincter by electrostimulation and identification of the urethra by ultrasonographic images from the perineum facilitated creation of appropriate PTC in the muscle complex.

## Complications of laparoscopic surgery in neonates and small infants

**Tadashi Iwanaka\***, Hiroo Uchida, Hiroshi Kawashima

Akira Nishi, Sumi Kudou, Ryosuke Satake

Department of Pediatric Surgery, Saitama Children's Medical Center, Japan

**Purpose:** To evaluate the complications of laparoscopic surgery in small infants weighing less than 5 kg, since the difficulty of performing laparoscopic surgery in small infants remains a common complaint of pediatric surgeons.

**Method:** Since 1997, 154 infants weighing less than 5 kg underwent laparoscopic surgery (Group S). During this period, 96 infants weighing less than 10 kg (group M) and 335 weighing more than 10 kg (Group L) also underwent laparoscopic surgery. Intra- and post-operative complications were evaluated among the three groups; especially in 60 cases that underwent Nissen fundoplication.  $P<0.05$  was considered statistically significant.

**Results:** Complications such as gastrointestinal perforation, wound infection, and intra- and post-operative bleeding were observed in 13 (8.4%), 15 (15.6%), and 32 (9.6%) of groups S, M, and L, respectively. There were no significant differences in complication rates among the three groups. However, in Nissen fundoplication, the complication rate in Group S was significantly higher than that in Group L (6/15 vs. 1/23,  $p= 0.006$ ). Gastrointestinal perforation and vagal nerve injury were the major complications in Group S.

**Conclusion:** Laparoscopic surgery is feasible without complications even in small infants. However, Nissen fundoplication requires special care and attention to avoid severe intra-operative complications.

## Laparoscopic transabdominal pericardial window in a neonate: a case report and review of the literature

Kevin Bruen

Primary Children's Medical Center, University of Utah, USA

**Purpose:** Laparoscopic transabdominal pericardial window has been reported with success in adults. We present a case report of a 4 month old, former 31 week premature infant with pulmonary lymphangiectasia. After removal of an extensive mediastinal cystic hygroma, he developed a chronic symptomatic, pericardial effusion. An abdominal approach offered direct access to the pericardial space avoiding previously operated areas.

**Method:** Pneumoperitoneum was established via a left epigastric incision followed by placement of a 5 mm and two 3 mm ports. A congruent button of central tendon of the diaphragm, located to the left of the falciform ligament was removed using the harmonic hook scalpel. An opening measuring 1.5 cm in diameter was made through the pericardium. Interrupted sutures were then placed to discourage closure.

**Results:** The window adequately drained the pericardial space for a four-week period. The child then developed line sepsis with recurrence of the effusion. A pericardial drain was placed and the effusion subsequently resolved.

**Conclusion:** We present the first report of a pediatric laparoscopic transabdominal pericardial window that can adequately drain the pericardial space. The procedure can be performed safely under direct visualization utilizing small incisions. Resolution of the pericardial process may result in closure or occlusion of the window.

## Prospective randomized, single-blinded comparison of laparoscopic versus open repair of pediatric inguinal hernia

**Kwong-leung Chan\***, Theresa WC Hui<sup>1</sup>, Paul KH Tam\*

Department of Surgery, The University of Hong Kong, China

Department of Anesthesia, Queen Mary Hospital<sup>1</sup>, China

**Purpose:** The aim is to perform a single-blinded randomized study to compare open surgery (OS) with laparoscopic repair (LR) for pediatric inguinal hernia (IH).

**Method:** Pediatric patients with IH were prospectively randomized into OS and LR groups. After operation, multiple dressings were placed to blind observers to the operation type. CHIPPS and CHEOPS pain scores were used to assess postoperative pain. Panadol (15mg/Kg/dose, 6 hourly) was given at a fixed pain score. Analgesic doses were compared. Parents also scored their children's recovery and wound appearance.

**Results:** Between February and December 2003, there were 83 IH patients randomized into OS (n=42) and LR (n=41) groups. There were less panadol intake in the LR group than the OS group ( $0.54 \pm 0.84$  doses vs  $1.05 \pm 1.248$ ,  $p=0.032$ ). Laparoscopy was able to detect 11 more bilateral hernias ( $p=0.006$ ). There was no difference in the operative time for bilateral hernia ( $39.08 \pm 13.37$  min vs  $34.0 \pm 11.31$  min,  $p=0.623$ ) though there was difference in unilateral hernias ( $18.38 \pm 5.71$  min vs  $23.25 \pm 6.26$  min,  $p=0.001$ ). There was 5 contralateral hernias detected in the OS group on follow-up but none in the LR group ( $p=0.026$ ). The parents' score for recovery and wound appearance were higher in the LR group ( $p < 0.05$ ).

**Conclusion:** LR for childhood IH is superior to OS with regards to postoperative pain, recovery and cosmesis. LR can also allow detection of contralateral hernias and have them repaired at the same operation. It takes slightly more time for unilateral repair than OS, but not for bilateral hernias.

## Can laparoscopic anti-reflux surgery improve the quality of life in children with neurological and neuromuscular handicap?

**Hisayoshi Kawahara\***, **Hiroomi Okuyama**, **Natsumi Tanaka**  
**Yuko Tazuke**, **Takaharu Oue**, **Akio Kubota\***, **Akira Okada\***

Osaka Medical Center and Research Institute for  
Maternal and Child Health, Japan

**Purpose:** Children with neurological and neuromuscular handicap frequently have various gastrointestinal and respiratory symptoms partly related with gastroesophageal reflux disease (GERD). The long-term efficacy of anti-reflux surgery for the improvement of their quality of life (QOL) is controversial in those handicapped children with GERD. The results of those patients having undergone laparoscopic Nissen fundoplication (LNF) were examined in the present study.

**Method:** During the past 7 years, LNF was conducted in 54 handicapped children (mean, 6 years) and gastrostomy was made concurrently in all except for two. Early two cases required conversion to open procedure. Main symptoms were emesis/hematemesis in 45 and respiratory symptoms, such as stridor, repeated respiratory infection, apnea, in 29. All were documented with excessive esophageal acid exposure.

**Results:** There were neither severe postoperative complications nor operative mortality. Wrap deformity occurred in 10 (19%), five of which required redo fundoplication (9%). Emesis/hematemesis was adequately controlled in patients without wrap deformity. Respiratory symptoms were controlled unsuccessfully in 12 (41%), four of which required laryngotracheal separation subsequently. During the follow-up period, 12 (22%) have died, nine of which was considered due to acute respiratory deterioration.

**Conclusion:** LNF is effective to control emesis/hematemesis without wrap deformity, whereas its efficacy is limited in terms of respiratory symptoms. Early tracheostomy or laryngotracheal separation with LNF may be helpful to improve QOL in the handicapped patients.

## Transinguinal laparoscopy at ultra-low pressure: ideal complement of ambulatory hernia surgery in children

**Peter KF Yip, Paul KH Tam\*, Michael KW Li<sup>1</sup>**

Department of Surgery, The University of Hong Kong, China

Department of Surgery, Pamela Youde Nethersole Eastern Hospital<sup>1</sup>

**Purpose:** Inguinal hernia surgery in children is widely practiced as ambulatory procedure in day surgery centre. On the other hand transinguinal laparoscopy had been introduced to complement conventional paediatric hernia surgery in detecting the co-existing contralateral hernia. However whether this technique can be safely applied in ambulatory surgery, which has the highest demand of safety and fluency, is still under question. We have performed transinguinal laparoscopy as an ambulatory procedure. In order to minimize the respiratory jeopardization result, a very low pressure of pneumoperitoneum was applied and the technique of laparoscopy needed to be modified accordingly.

**Method:** Seventy children of age 3 months to 14 years old with unilateral inguinal hernia was operated in the day surgery centre. After the hernia sac was dissected and opened, a 5mm laparoscopy port was inserted and pointed towards the umbilicus. Pneumoperitoneum was created at a very low pressure of 4mm Hg. Laparoscopic lens was then inserted and advanced along the small peritoneal space created until the contralateral hernia orifice was inspected.

**Results:** All laparoscopies were successful and majority of them could be completed in around 1 minute. In 25 cases contralateral patent processus vaginalis were identified and repaired in the same session. No respiratory distress was resulted and endotracheal intubation was not required. The postoperative recovery was smooth and patients could be discharged safely.

**Conclusion:** Transinguinal laparoscopy in children can be safely performed in the day surgery unit with an ultralow pressure of pneumoperitoneum.

## The experimental study on the utility of a 3-mm ultrasonically activated trocar system

**Masaki Nio\***, Tomohiro Ishii<sup>1</sup>, Shintaro Amae<sup>1</sup>, Motoshi Wada<sup>1</sup>  
Kotaro Nishi<sup>1</sup>, Yutaka Hayashi<sup>1</sup>

Miyagi Children's Hospital, Japan  
Tohoku University Graduate School Of Medicine<sup>1</sup>, Japan

**Purpose:** A prototype of a 3-mm ultrasonically activated trocar (UT) system was evaluated in terms of its utility and safety using piglets.

**Method:** An 8-mmHg pneumoperitoneum was created with Hasson's technique. Trocars were then inserted under laparoscopic control. Eleven punctures were made with a conical disposable 3-mm trocar (CT). Nine punctures were made with a 3-mm radially expanding trocar (ET). Thirteen punctures were made with a 3-mm UT. Time for abdominal penetration, presence of elevated abdominal pressure greater than 5 mmHg at the penetration, and maximal force applied to the trocar to remove from the abdominal wall were recorded.

**Results:** The average time needed for trocar penetration was 11.8 s with CT, 9.4 s with ET and 4.5 s with UT ( $p < 0.05$ , CT vs. UT, ET vs. UT). Elevation of abdominal pressure greater than 5 mmHg was recorded in any punctures of CT and ET, but not in UT. The average maximal force to remove the trocar was 10.52 N with CT, 21.17 N with ET and 21.24 N with UT ( $p < 0.05$ , CT vs. ET, CT vs. UT). No injury related to the UT system was encountered.

**Conclusion:** The 3-mm UT was revealed to be a safe and easy device.

## Roles for Matrix Metalloproteinases and their Endogenous Tissue Inhibitors in Biliary Atresia-Associated Liver Fibrosis

Jiin-Haur Chuang\*, Chao-Cheng Huang<sup>1</sup>, Ming-Huei Chou<sup>2</sup>  
Chia-Lin Wu, Shin-Yi Lee, Chih-Sung Hsieh\*, Chao-Long Chen

Department of Surgery, Chang Gung Memorial Hospital-Kaohsiung, Taiwan

Departments of Pathology, Chang Gung Memorial Hospital-Kaohsiung<sup>1</sup>, Taiwan

The Graduate Institute of Clinical Medicine, Chang Gung University<sup>2</sup>, Taiwan

**Purpose:** Matrix metalloproteinases (MMPs) and their endogenous tissue inhibitors (TIMPs) are major proteases responsible for remodeling the liver tissue, but their roles in biliary atresia (BA)-associated liver fibrosis are not clear.

**Method:** A DNA microarray containing cDNA clones of 10 MMPs and 4 TIMPs was used to compare the expression profiles of the liver cytokines among three patients with BA at the time of Kasai...s procedure (KP) with three at the time of liver transplantation (LT). Liver samples from two children without liver fibrosis were used as normal control. Those genes that were differentially expressed for more than 2-fold between groups were further quantified with real-time quantitative RT-PCR (QRT-PCR) and validated with gel electrophoresis.

**Results:** In normal human liver, mRNAs of TIMP-1, -2 and -3, but not of TIMP-4 and all 10 MMPs studied, were expressed. With progress of liver fibrosis, only mRNA of MMP-7, but not other MMPs, was induced to express at a significantly higher level in the array. There was a 5-fold increase in liver MMP-7 expression in the array, but a more than 90-fold increase in QRT-PCR in LT over control. Despite its low level of expression, MMP-9 mRNA was significantly upregulated in KP but downregulated in LT, while MMP-2, which was not included in the array, was significantly upregulated in LT than in KP and control in QRT-PCR. There was a more than two-fold increase in TIMP-1 and TIMP-2 mRNA expression in LT over control in the array, which was confirmed in subsequent QRT-PCR. The expression of TIMP-3 mRNA was significantly downregulated in KP than in control. These findings were further confirmed with gel electrophoresis.

**Conclusion:** This study verified differential expression of MMPs and TIMPs in different stages of BA, with emphasis on the role of TIMP-1, -2 and -3, as well as MMP-2, -7 and -9 transcripts in remodeling of liver tissue during the progress of BA-associated liver fibrosis.

**Laparoscopic mobilization of testicular vessels  
(LMTV): an innovative step in orchidopexy for  
impalpable and redo undescended testis in children**

**Michael. W.Y. Leung, Nicholas S.Y. Chao, Beatrice P.Y. Wong<sup>1</sup>  
K.W. Chung, W.K. Kwok, Kelvin K.W. Liu<sup>\*1</sup>**

Queen Elizabeth Hospital, China  
United Christian Hospital, Hong Kong<sup>1</sup>, China

**Purpose:** The testicular position after conventional inguinal orchidopexy for canalicular, peeping and redo undescended testes may not be satisfactory despite retroperitoneal dissection. Laparoscopy allows extensive mobilization of testicular vessels to gain additional length. We review our experience of using LMTV in orchidopexy for these difficult undescended testes.

**Method:** From January to December 2003, LMTV were performed in 15 boys. The testicular vessels were mobilized from just proximal to the internal inguinal ring to the level of the right or left colon respectively. 12 patients had clinically impalpable testes. Diagnostic laparoscopy revealed 10 peeping and 2 canalicular testes. In 9 cases, LMTV were performed together with inguinal orchidopexy. Three peeping testes were brought to the scrotum laparoscopically without inguinal dissection. The presence of looping vas deferens at internal inguinal ring determined whether inguinal orchidopexy was necessary. Three cases were redo-orchidopexies. LMTV were performed after inguinal dissection.

**Results:** The median follow-up period is 6 months. All testes are located at a satisfactory position at the base of scrotum. The size is normal in 14 testes, whereas one testis has mild atrophic change.

**Conclusion:** Laparoscopic mobilization of testicular vessels is a safe and efficient adjunctive step in orchidopexies for impalpable and redo undescended testes.

# Poster Presentation Abstracts

## P01

### The impact of computerized prescription on medication incidence rate : experience of a single centre

**Kenneth Wong, Alex Fan, Lawrence Lan, Steve Lin, Paul Tam\***

University of Hong Kong Medical Centre, China

**Purpose:** Medication incidence has potentially serious effects on patients' health. It is therefore important to try to minimize this. It has long been recognized that medication incidence tends to be under-reported. Our centre launched a computerized prescription system in 1996. This system would warn the prescriber of any errors before sending out prescriptions. Any major medication incidence would be logged and presented at monthly audit meetings. Here we reviewed the secular trend of our medication incidence before and after computerization.

**Method:** Between 1994 and 2003, all medication incidence reported in our centre was studied and included in this review.

**Results:** Our centre adopted a voluntary reporting system before computerization. From 1994 to 1996, there were on average 10 medication incidence every year. After the launch of computerized prescriptions, the medication incidence rose sharply to nearly 100 cases per year in the following three years. The incidence rate steadily declined from 96 cases in 2000 to 40 cases in 2003.

**Conclusion:** The increase in medication incidence after computerization is most likely due to a reflection of previous under-reporting. With proper computerized documentation and audit, the overall medication incidence has steadily declined. We therefore conclude that a well-designed computerized prescription system will help reduce the medical incidence rate.

## Characterization of Metabolic responses in Surgical neonate

Hideki Soh, Masafumi Wasa, Yoshiyuki Shimizu, Akio Kubota\*

Shinkichi Kamata<sup>\*1</sup>, Masahiro Fukuzawa\*

Osaka University Graduate School of Medicine, Japan

Osaka Medical Centre for Maternal and Child Health<sup>1</sup>, Japan

**Purpose:** Metabolic response to surgical stress remains unknown in neonates. To clarify this, we measured resting energy expenditure (REE) and compare REE with the other clinical parameters in surgical neonates.

**Method:** In 10 neonates (3.5 +/- 4.9 days) with the diagnosis of intestinal atresia, malrotation, imperforate anus and sacrococcygeal teratoma, REE was measured by indirect calorimetry using Deltatrac II before operation and every 2 hours for 24 hours after operation. Plasma interleukin-6 (IL-6) and hemodynamic parameters such as respiratory rate and heart rate were also measured.

**Results:** Plasma IL-6 significantly increased after surgery ( $p < 0.01$ ). However, there was no significant difference between preoperative REE (38.0 +/-14.4 kcal/kg/day) and postoperative REE at every time point, and no statistical correlation was observed between plasma IL-6 and REE. Respiratory rates and heart rates also did not show significant change after operation.

**Conclusion:** Characteristic metabolic response that REE does not correlate with IL-6 levels was observed in surgical neonates. These results may be beneficial for the determination of perioperative nutritional management in neonates.

## Rapid Effect of Maternal Betamethasone on Hydropic Fetuses with Cystic Adenomatoid Malformation

**Yoshihiro Kitano, Tatsuo Kuroda\*, Toshiro Honna  
Satoshi Hayashi, Haruhiko Sago**

National Center for Child Health and Development, Japan

**Purpose:** Salvage of hydropic fetuses with cystic adenomatoid malformation (CAM) after maternal steroid administration is reported, but its causal relationship remains unknown. The purpose of this study was to document the short-term effect of maternal administration of betamethasone on hydropic fetuses with CAM.

**Method:** Three hydropic (ascites only in two and ascites with skin and scalp edema in the other) fetuses with CAM were followed by ultrasound after maternal intramuscular injections of betamethasone (12mg x 2) at 25-27 weeks.

**Results:** Ascites disappeared three days after initial injection in two fetuses without skin edema (one micro cystic and the other macro cystic). CAM volume ratio (CVR) decreased in micro cystic CAM, but not in macro cystic CAM. They were delivered at term without further intervention. The baby with macro cystic CAM required emergent lobectomy at 2 hours of age. In the third fetus with skin edema, CVR decreased after steroid, but neither ascites nor skin edema responded. Open fetal lobectomy led to a premature delivery and neonatal death at 28 weeks.

**Conclusion:** These observations show that betamethasone has a rapid effect, allowing early judgement on its efficacy. We conclude that maternal steroid administration is a reasonable initial treatment for a hydropic fetus with CAM.

## Ultrasonography guided subclavian vein puncture

**Toshiki Ohya, Arii Shigeki, Iwai Takehisa**

Tokyo Medical and Dental University, Japan

**Purpose:** Subclavian vein puncture is one of the essential procedures for central vein catheter insertion. However, when the patient was in critical condition or with severe coagulopathy, it could be a disaster. In order to avoid the serious complications, we tried to use ultrasonography to see the subclavian vein.

**Method:** Eleven pediatric patients who required central vein catheter insertion underwent ultrasonography to see the subclavian vein before puncture. Age ranged 4 months to 15 years of age. Eight of them underwent ultrasonography guided subclavian vein puncture. Ultrasonography system was NEMIO SSA550A and micro probe PVF-745V, both made by Toshiba co. Japan.

**Results:** Subclavian vein was easily visualized even 4 months old infant. Ultrasonography was useful to hit subclavian vein; however, when we give a little more pressure to the skin, vein easily collapsed and we lost the target. Also puncture the vein while holding probe sufficiently was not easy. Then we are assembling a holding device using thin sticky material between skin and probe. It did not disturb the sight and was helpful to hold probe.

**Conclusion:** Ultrasonography guided subclavian vein puncture is much safer and accurate than usual blind procedure. Our holding device showed high potential for clinical application.

## **Successful EXIT procedure in a fetus with congenital high-airway obstruction syndrome due to laryngeal atresia**

**Hiroomi Okuyama\***, **Akio Kubota\***, **Hisayoshi Kawahara\***

**Takaharu Oue, Yuko Tazuke, Natsumi Tanaka, Noriyuki**

**Suehara, Keiko Kinouchi, Hiroyuki Kitajima**

Osaka Medical Center for Maternal and Child Health, Japan

**Purpose:** We describe a successful management of a fetus with a prenatally diagnosed airway obstruction.

**Method:** Congenital high airway obstruction syndrome (CHAOS) caused by laryngeal atresia was diagnosed by prenatal ultrasound in a fetus at 21 weeks of gestation. Findings included overdistended hyperechogenic lungs, inverted diaphragms, a dilated trachea, polyhydramnios, and ascites. The fetal ascites disappeared at 29 weeks of gestation. At 30 weeks of gestation, a planned ex utero intrapartum treatment (EXIT) procedure was performed because of premature rupture of membrane. Under general anesthesia of the mother, a midline incision of the uterus was performed. The fetal head, left arm and shoulder were delivered. Immediate laryngoscope confirmed the diagnosis of laryngeal atresia and a tracheostomy was performed on the placental circulation.

**Results:** During the EXIT procedure heart rate and arterial oxygen saturation of the fetus were well maintained. After delivery, the baby was found to have duodenal atresia, which was corrected surgically on the 7th day of life. The baby has weaned from the artificial ventilation in a few weeks. There were no maternal complications.

**Conclusion:** The EXIT procedure is a technique for safely managing airway obstruction at birth in fetuses with a prenatal diagnosis of CHAOS.

## Thoracoabdominal duplication associated with a right congenital diaphragmatic hernia; embryology and surgical management

Tomoro Hishiki, Shunsuke Tatebe<sup>1</sup>, Yoshiteru Ohsone<sup>1</sup>  
Naomi Ohnuma<sup>2</sup>, Toshinori Muramatsu

Kimitsu Chuo Hospital, Department of Pediatric Surgery, Japan

Kimitsu Chuo Hospital, Department of Neonatology<sup>1</sup>, Japan

Chiba University School of Medicine, Department of Pediatric  
Surgery<sup>2</sup>, Japan

**Purpose:** Thoracoabdominal duplication is an extremely rare anomaly caused by an abnormal adhesion of the foregut to the dorsal ectodermal tissue. We present a case of thoracoabdominal duplication with a rare form of congenital diaphragmatic hernia (CDH).

**Method:** The case was a newborn female suffering severe respiratory distress immediately after birth. A right sided CDH and vertebral body defects (C4~Th3) were detected on a chest x-ray. She underwent surgery on her third day of life.

**Results:** The entire small intestine and half of the colon was herniated into a sac occupying the right hemithorax. The mesentery was tightly fixed to the cervical vertebra, causing a strong traction of the intestine into the thorax. A thoracoabdominal duplication arose from the jejunum and was also anchored cranially. Since it was unfeasible to reposition the whole hernia content to the abdomen, we removed 40cm of the small intestine including the duplication which remained in the thorax, but left the mesentery intact. The diaphragm was repaired with a Goretex patch.

**Conclusion:** An abnormally anchored mesentery was observed additionally to the thoracoabdominal duplication. The CDH in this case might have been a result of this anchoring rather than the delayed closure of the diaphragm as in the typical CDH cases.

## **Assessment of obstructive apnea by using polysomnography and surgical treatment in patients with Beckwith-Wiedemann syndrome**

**Masafumi Kamiyama, Shinkichi Kamata\*, Toshio Sawai**

**Keisuke Nose, Noriaki Usui, Hisayoshi Kawahara\***

**Masahiro Fukuzawa**

Department of Pediatric Surgery, Osaka University Graduate  
School of Medicine, Japan

**Purpose:** Obstructive apnea is sometimes seen in patients with Beckwith-Wiedemann syndrome. The cause of apnea is not only due to macroglossia, and surgical indication for obstructive apnea has not been established. We have performed polysomnography for the assessment of apnea.

**Method:** Overnight polysomnograms were obtained in 2 patients who developed obstructive apnea after one-stage repair for omphalocele.

**Results:** Case 1: Apnea index (AI), defined as the apneic events per an hour, indicated 17.3, and SpO<sub>2</sub> below 95% occupied 48 to 80% of the total sleep time. CT and MRI indicated obstruction of the airway between macroglossia and the hypopharynx. Central tongue resection and the division of the frenulum linguae were performed 97 days after birth. One month after surgery, SpO<sub>2</sub> below 95% occupied only 1% of the total sleep time. Case 2: AI indicated 10.4. The division of the frenulum linguae and anterior glossopexy were performed 55 days after birth. Postoperative polysomnogram indicated a marked reduction of AI.

**Conclusion:** These results indicated that polysomnography was useful for evaluating obstructive apnea and that an advancement of the tongue by the division of the frenulum linguae may be recommended for the treatment of obstructive apnea in patients with Beckwith-Wiedemann syndrome.

**Dual myotomies for the treatment of fibromuscular thickening type congenital esophageal stenosis (CES) in a two-year-old boy**

**Masahiro Chiba, Yutaka Sanada\*, Shinya Kawano  
Masayuki Murofushi, Yasuo Yoshizawa, Akira Gomi  
Masashi Yatsuzuka Akira Toki\***

Showa University School of Medicine, Japan

**Purpose:** This is a case presentation to show the efficacy of dual myotomies for the treatment of fibromuscular thickening type CES.

**Method:** A-26-month old boy experienced vomiting following ingestion of an Ume (Japanese apricot) seed. The barium swallow showed 1.5-cm-long stenosis at the level of diaphragm. Endoscopic ultrasonography demonstrated the lesion to be fibromuscular hypertrophy. Via abdominal approach, the lower esophagus was exposed. The balloon catheter, introduced to the esophagus through gastrotomy, was connected with the nasogastric tube. The balloon catheter was inflated and moved to confirm the distal and the proximal margins of the stenosis. The 2-cm-long longitudinal myotomies limited to the stenotic area were made at 2 and 10 o'clock. This procedure allowed the inflated balloon catheter to pass the lesion. Thal fundoplication was performed to cover the sites of myotomies and to prevent postoperative GER.

**Results:** The postoperative barium swallow showed sufficient passage through the lesion. He has been doing well 12 months after surgery.

**Conclusion:** Dual myotomies limited to the stenotic lesion is likely to be a safe and effective modality for the treatment of fibromuscular thickening type of CES.

## Surgical Management for Laryngo-tracheo-esophageal Cleft (Type I) Developing Subglottic Airway Obstruction

Yoshiyuki Ihara, Shinkichi Kamata\*, Noriaki Usui  
Masafumi Kamiyama, Hideki Soh, Masahiro Fukuzawa\*

Department of Pediatric Surgery, Osaka University Graduate  
School of Medicine, Japan

**Purpose:** Patients with type I clefts usually present with congenital inspiratory stridor, cyanotic attacks associated with feeding and recurrent respiratory infections due to aspiration. The aim of this report is to describe surgical management for a patient with respiratory distress, a rare complication in type I clefts.

**Method:** A two-years-old boy associated with Opitz syndrome including hypospadias and anorectal anomaly admitted to Osaka University Hospital with oro-tracheal intubation for respiratory distress after respiratory infection. He had diagnosed of laryngeal cleft in the newborn period for symptoms including stridor and aspiration pneumonia. After two times trial for extubation, tracheostomy was made. Fiberscopic examination revealed obstruction of airway from vocal cord to subglottic space by inspired esophageal mucosa and no improvement for 3 months. He underwent anterior repair of cleft, anterior laryngotracheoplasty with a costal cartilage graft and closure of tracheostomy.

**Results:** Endtracheal tube was secured for 3 weeks. Nissen fundoplication was performed for associated gastroesophageal reflux before extubation. Postoperative course was uneventful without feeding and speaking disorders.

**Conclusion:** Subglottic airway obstruction due to inspired esophageal mucosa is a rare complication in type I cleft. Anterior laryngotracheoplasty and Nissen fundoplication should be considered in addition to repair of cleft.

## Three patterns of recurrence from Sistrunk's operation

Minoru Horisawa\*

Anjo Kosei Hospital, Japan

**Purpose:** Sistrunk's operation is acknowledged to have remarkably decreased the recurrence rate of thyroglossal duct cyst operation, but a few recurrences are still can not be avoided. We experienced 4 recurrences after 64 Sistrunk's operations. The purpose of this report is to clarify the cause of recurrence in each case by clinico-pathological study.

**Method:** These 4 cases of recurrence were studied from their clinical courses and pathological examinations. Anatomical reconstructions were made of the thyroglossal duct in each specimen obtained from the respective operation. The clinical and pathological findings were compared to cases with no recurrence.

**Results:** Clinical analysis revealed that infection preceding the operation increases the recurrence rate. By pathological analysis, many branches of thyroglossal ducts in suprathyroid lesion were suspected to be strong recurrence factor. In special case, existence of an ectopic mucous gland communicating to the main thyroglossal duct made cure virtually impossible. Only resection of the ectopic gland could prevent the recurrence.

**Conclusion:** The causes of recurrences after optimal operation Sistrunk's operation were suspected to be three: 1. Preceding infection; 2. Too many thyroglossal duct branches in suprathyroid lesion; and 3. Some ectopic mucosal gland connecting to the main thyroglossal duct.

## **Remission induced by interferon- $\alpha$ in a patient with massive osteolysis and extension of lymph-hemangiomatosis (Gorham-Stout syndrome)**

**Atsushi Takahashi, Chitose Ogawa, Takashi Kanazawa**

**Hideomi Watanabe, Makoto Suzuki<sup>1</sup>, Norio Suzuki<sup>\*1</sup>**

**Yoshiaki Tsuchida<sup>\*1</sup>, Akihiro Morikawa, Hiroyuki Kuwano**

Gunma University, Japan

Gunma Children's Medical Center<sup>1</sup>, Japan

**Purpose:** A treatment of Gorham-Stout syndrome has been controversial.

**Method:** The authors report a patient with massive osteolysis and extension of lymph-hemangiomatosis.

**Results:** A 2 year-old girl had complained of left chylothorax. Thoracoscopy showed an increase in small lymphatic vessel in the chest wall, and the chylothorax was improved by coagulation of the lymphatic vessel. Massive osteolysis appeared in the left 11th and 12th ribs, the TH10-L3 vertebrae and the right femur. There were also a tumor lesion in the left lower chest wall and hemangiomatous change on the skin surface of the left back. OK-432 was admitted to the femur and the chest wall lesions, and the osteolysis of femur disappeared. Then, as right chylothorax appeared, OK-432 was injected into the right pulmonary cavity. The chylothorax disappeared, but pericardial effusion appeared. After steroid pulse therapy, pericardial effusion was disappeared. During these treatments, the progress of osteolysis and the extension of lymph-hemangioma were found. Due to the progress of lesions, interferon- $\alpha$  and steroid therapy was started. Ten months after, hemangiomatous change in the back disappeared, and some disappeared ribs again appeared.

**Conclusion:** Interferon- $\alpha$  therapy should be chosen in this type of patients.

## Effects of probiotic administration in pediatric patients with intestinal failure

Hideo Yoshida, Hiroaki Kuroda, Tadashi Matsunaga

Katsunori Kouchi, Takeshi Saitou, Naomi Ohnuma\*

Chiba University Graduate School Of Medicine, Japan

**Purpose:** The human gut microbiota plays a crucial role in host health. The objective of this study was to determine the effects of probiotics on the nutritional status and the intestinal functions of the intestinal failure patients.

**Method:** In a clinical trial, 6 patients suffering from short bowel syndrome or chronic idiopathic intestinal pseudo-obstruction syndrome were administered living Bifidobacterium breve and Lactobacillus casei orally. Fecal specimens of the patients were examined for their bacterial flora and short chain fatty acid contents.

**Results:** Before the administration of probiotics , the flora in the feces of the patients were facultive anaerobic bacteria dominant. An increase in lactobacilli and bifidobacteria was observed as a consequence of probiotic treatment. At the same time, the amounts of short fatty acids in the feces increased. The administration of probiotics improved the patients' nutritional status.

**Conclusion:** Supplementation with probiotics can improve the clinical picture, and can restore the depletion of lactobacillus and bifidobacterium in stools after therapy.

## Plasma Interleukin-6 and Interleukin-8 in Evaluating Intestinal Gangrene in Children's Adhesion Ileus

Hsun-Chin Chao

Chang Gung Children's Hospital, Taiwan

**Purpose:** To evaluate the predictive value of plasma IL-6 and IL-8 response in intestinal gangrene in children's adhesion ileus.

**Method:** we performed a 5-year (January 1998-December-2003) study of plasma interleukin-6 (IL-6) and interleukin-8 (IL-8) in children's adhesion ileus. The plasma IL concentration was determined by ELISA method, and were routinely measured on admission and per-operative period. We correlated the IL-6 and IL-8 expressions with surgical findings, and evaluated the predictive value of plasma IL-6 and IL-8 in intestinal gangrene.

**Results:** The study comprised of 50 patients (33 males, 17 females; mean age: 8.3 ± 4.7 years) with 53 episodes of adhesion ileus underwent surgical intervention, and 24 episodes were surgically proved to gangrene. Under statistical analysis, no marked differences of initial IL-6 and IL-8 levels were found between gangrenous and nongangrenous patients. Those gangrenous patients had statistical significance of higher plasma IL levels on follow-up period (IL-6:  $54.7 \pm 19.3$  pg/ml; IL-8:  $386.5 \pm 107.6$  pg/ml) and operation day (IL-6:  $133.5 \pm 46.3$  pg/ml; IL-8:  $729.9 \pm 193.4$  pg/ml) than those nongangrenous patients (follow-up period: IL-6:  $37.1 \pm 12.9$  pg/ml, IL-8:  $260.3 \pm 89.5$  pg/ml; operation day: IL-6:  $88.3 \pm 27.2$  pg/ml; IL-8:  $391.2 \pm 102$ . pg/ml).

**Conclusion:** Regular monitoring of plasma IL-6 and IL-8 concentration help to identify the occurrence of gangrene in children's adhesion ileus.

## **Meconium-related Ileus -The Most Common Functional Ileus of Extremely Low-birth-weight Neonates –**

**Akio Kubota\*, Katsuji Yamauchi<sup>1</sup>, Takashi Nogami<sup>1</sup>**

Osaka Medical Center for Maternal and Child Health, Japan  
Kinki University School of Medicine<sup>1</sup>, Japan

**Purpose:** Meconium related ileus (MRI) is functional ileus of neonates characterized by impaired meconium excretion. As the survival rate of extremely low-birth-weights increases, the incidence of MRI is increasing, however, pathogenesis of MRI is not well described. The purpose of this study is to elucidate the pathogenesis of MRI.

**Method:** Sixty-one cases of MRI were reviewed clinically, and additional 10 in which biopsy or autopsy were available were included in the study for histological evaluation of myenteric plexus.

**Results:** Ninety-seven percent of MRI occurred in low-birth-weights. Five required laparotomy, three of them died shortly after the surgery. The other 56 regressed with conservative therapies, and started enteral feeding within six months at most, and presented no more intestinal symptom. Contrast enema demonstrated microcolon, caliber change in the ileum, and meconium impaction not constantly consistent with the caliber change. The nuclear area of ganglion cells was compatible with that of gestational age-matched controls in six, and slightly smaller in four.

**Conclusion:** Meconium impaction may not be the cause of the ileus, but the result of impaired bowel movement of a fetus. Functional immaturity proportional to the gestational age of the ganglion cells may be implicated in the etiology of MRI.

## Ectopic pancreatic tissue: A rare cause of persistent umbilical discharge

Yasuhiro Okada, Masatoshi Miyamoto, Katsuhiko Saito<sup>1</sup>

Department of Pediatric Surgery, Toyama City Hospital, Japan

Department of Pathology, Toyama City Hospital<sup>1</sup>, Japan

**Purpose:** We report a case of ectopic pancreatic tissue in the umbilicus. To our knowledge, there have been only four cases reported previously.

**Method:** Case: A 3-years old girl presented with a persistently discharging umbilicus since birth. She had previously been treated by cauterization with silver nitrate, which did not give long-term improvement. Physical examination showed a small and wet granulation tissue in the umbilicus. Abdominal ultrasound showed no cystic lesion underneath the umbilicus. The patient underwent surgical exploration via an inferior circum-umbilical incision. The undersurface of granulation tissue attached to the obliterated umbilical vessels, however, no further continuity between underlying intra-abdominal viscera. Granulation tissue with umbilical vessels was excised en bloc. The patient made uneventful recovery. Histopathologically, granulation tissue consists of intestinal mucosa and smooth muscle, implying omphalomesenteric duct remnant. In addition, ectopic exocrine pancreatic tissue was also present within the specimen.

**Results:** Persistent umbilical discharge was associated with periumbilical injury, which presumably caused by the local action of pancreatic enzymes secreted by the ectopic tissue.

**Conclusion:** Ectopic pancreatic tissue in omphalomesenteric duct remnant is extremely rare, however, a high index of suspicion is required for patients with persistent umbilical discharge with no apparent cause. Complete surgical resection is necessary.

## The Longest Survival of Megacystis-Microcolon-Intestinal-Hypoperistalsis Syndrome-A case report-

**Shigeru Ueno, Seishichi Yokoyama\*, Hitoshi Hirakawa**

**Tomoya Hinoki**

Department of Pediatric Surgery, Tokai University School of Medicine, Japan

**Purpose:** To describe a history of a female patient with generally fatal Megacystis- microcolon-intestinal-hypoperistalsis syndrome (MMIHS), in whom long-term parenteral nutrition has been successful for 22 years.

### **Method:**

**Results:** She was born at 36 weeks gestation with huge abdomen and the bladder filled with 1200ml of urine. She developed the gastric perforation and her condition was diagnosed as MMIHS. Severe hepatic dysfunction and scurvy like changes were encountered in her early infancy but they resolved after starting oral feeding with cyclic TPN and trace element replacement. Repeated operations failed to achieve a functional intestinal tract, resulting in living with an eventual stoma at the upper jejunum and fluid and nutritional replacement by home parenteral nutrition (HPN). Long-term central venous catheterizations resulted in the thrombosis of both caval veins, which necessitated the percutaneous transhepatic IVC catheterizations for eight years. Her growth curve shows that the HPN has enabled her to achieve adequate physical development of her weight but not of her height. Her intelligence enables her to compete effectively with others and she works as a computer programer at a big company now.

### **Conclusion:**

## Successful Separation of Omphalopagus Conjoined Twins

Hai-Chyi Peng

Buddhist Tzu-chi General Hospital, Hualien, Taiwan

**Purpose:** Conjoined twins provided one of the greatest challenges to modern pediatric surgery. We report successful separation of one-year-old omphalopagus conjoined twins.

**Method:** The babies were born by cesarean section with 39 weeks gestation on July 1, 2002. On physical examination, the twins were conjoined from lower sternum to a common umbilicus. Umbilical hernia was noted, too. Series imaging studies commenced at the age of 9.5 months. Conjoined lower ribs, xyphoid process, small area of pericardia, and an extensive portion of the liver were identified. There was no associated anomaly except protruded chest wall (pectus carinatum) and lordotic spines. Two tissue expanders were placed subcutaneously at the age of 10.5 months and were gradually expanded in 6 weeks. The infants were then separated. The division of the liver parenchyma was accomplished with ultrasonic dissector (CUSA) and bipolar diathermy. The fascia defect was repaired with Vicryl mesh and the skin was closed primarily on each infant. It took about 6 hours to complete this operation and there was almost no blood loss.

**Results:** The babies stood the procedure well and were healthy at follow-up 6 months after separation.

**Conclusion:** If there is no severe cardiovascular malformation, results of omphalopagus conjoined twins separation can be expected to be satisfying concerning current medical management.

## The long term course of chronic intestinal failure in children

**Yuko Tazuke, Akio Kubota, Hisayoshi Kawahara, Takahiro Oue  
Hiroomi Okuyama, Shinobu Ida**

Osaka Medica Center and Research Institute for Maternal and Child Health, Japan

**Purpose:** To identify prognostic factors in children with chronic intestinal failure, which might lead to the intestinal transplantation, especially in the cases of short bowel syndrome (SBS) and chronic idiopathic intestinal pseudo-obstruction syndrome (CIPS).

**Method:** 32 children with chronic intestinal failure were investigated retrospectively (1979-2003). The twenty two of 32 cases were developed into SBS by the extensive bowel resections; small bowel atresia (8), gastoschisis (5), aganglionosis (2), mesenteric ischemia; midgut volvulus (4) and miscellaneous (3). The 13 of 32 cases were diagnosed as CIPS. The follow-up term was 3m-23y (mean 8y) in SBS and 1y-17y (mean 5y) in CIPS.

**Results:** Total operations (without catheterizations) were 3 times in both groups. Intestinal decompression stoma were used in 10 cases of SBS and 7 case of CIPS, and the stoma were closed in 9 cases (90%) of SBS and 2 cases (29%) of CIPS. The PN (>1years) was requested 11 cases of SBS and 5 patients developed into PN dependency. One patient had poor outcome (death) and a SBS case is planning for intestinal transplantation because of her acquired liver dysfunction. In CIPS children, PN (>1years) was requested in 6 cases, while last 6 cases were controlled their abdominal condition by daily enema. Eight CIPS patients had PN dependency during their life. Four poor outcome (death) was seen in CIPS patients within 2years (3) and 11years (1). The inserted numbers of PN catheter a case (mean) were more 3 times of CIPS than 2 times of SBS. During follow term, the mean septic episodes (depend on PN catheter) were seen more 3 times of CIPS than 2 times of SBS. Interestingly, the symbiotic treatment (BF, BBG, etc.), which is noticed to be valuable therapy recently, effected to improve their intestinal malaise (lead to enterocolitis) in 5 cases of SBS and 5 cases of CIPS.

**Conclusion:** Prognostic factors, which lead to transplantation, in children with chronic intestinal failure were undefined because their bowel conditions were well controlled and improved by age dependently in both of SBS and CIPS patients. On the other hand, the bowel transplantation might have to be prompt for 5 dead patients before their having lethal complications.

## High Concentration of Interleukin-6 and its Reduction by Continuous Hemodiafiltration in Infants with Intestinal Perforation<sup>1</sup>

**Katsunori Kouchi, Hideo Yoshida<sup>1</sup>, Tadashi Matsunaga<sup>1</sup>  
Hiroaki Kuroda<sup>1</sup>, Tomoro Hishiki<sup>1</sup>, Naomi Ohnuma<sup>1</sup>**

Chiba University, Japan

Department of Pediatric Surgery, Chiba University<sup>1</sup>, Japan

**Purpose:** Intestinal perforation and its surgical stress induces systemic inflammatory response syndrome (SIRS), which may lead to postoperative multiple organ failure (MOF). Plasma interleukin-6 (IL-6) levels were measured in infants who underwent surgery. Furthermore, we performed continuous hemodiafiltration (CHDF) on infants with severe SIRS to reduce plasma cytokine.

**Method:** IL-6 concentration was measured in eight infants with (n=4, IP group) or without (n=4, NP group) intestinal perforation. A status in which the heart rate (HR) was >200 bpm throughout the eight postoperative hours despite intensive care, with IL-6 >1000pg/ml, was defined as severe SIRS.

**Results:** Plasma IL-6 peaked immediately after surgery and decreased thereafter. Maximum IL-6 was 368 to 394468 (mean 122422) pg/ml in the IP group, whereas in the NP group it was 281 to 6546 (mean 1969) pg/ml. Only a short period of oliguria was experienced in infants with IL-6 <1000pg/ml, which was recovered without CHDF. Two infants in the IP group had severe SIRS and their HR decreased after the induction of CHDF. Both infants recovered from SIRS without any sequelae of CHDF.

**Conclusion:** In infants with IL-6 <1000pg/ml, oliguria was improved without CHDF. In infants with severe SIRS, CHDF should be used to prevent MOF.

## Difference of Adapative Response After Massive Small Intestinal Resection IN The Rat

**Shigeki Hikida, Chiaki Kawaguchi, Yoshiaki Tanaka**

**Hiroyoshi Mizote, Hiromichi Yano\*, Kimio Asagiri**

**Kenjirou Akiyoshi, Tomomitsu Tsuru**

Kurume University Hospital, Japan

**Purpose:** The aim of this study is to evaluate morphological and functional differences between remnant jejunum and remnant ileum after massive bowel resection in the rat.

**Method:** Male Lewis rats(172-224g) underwent 60% resection of the proximal or distal intestine. The studies (weight gain, morphological analysis, transepithelial potential difference evoked by active glucose uptake as a functional marker existing both in the jejunum and in the ileum) were evaluated 1week, 1month, 3months after operation.

**Results:** The weight gain in the jejunum remnat group was significantly worse than that in the ileum remnant group until 2 month after operation. From the results of morphological analysis, the villus height, the crypt depth, and the mucosal thickness in the jejunum remnat group were significantly lower than those in the ileum remnant group until 1month after operation. The transepithelial potential difference evoked by active glucose uptake in the jejunum remnant group was significantly lower than that in the ileum remnant group. All factors in the jejunum remnant group became similar to those in the ileum remnant group

**Conclusion:** These results suggest that the adaptive response in the jejunum reaches the endpoint more than 1month after intestinal resection, while that in the ileum reaches less than 1month. Of the morphological and functional factors evaluated, the delays of adaptations of mucosal growth and the increase of active glucose uptake most likely to explain the inferior nutritional and absorptive outcome associated with resection of the distal small intestine.

## The novel strategy with synbiotics therapy in small bowel transplantation

**Toshihiko Watanabe, Ken Hoshino<sup>1</sup>, Naoki Shimojima<sup>1</sup>**  
**Minoru Tanabe<sup>1</sup>, Yasuhide Morikawa<sup>1</sup>, Masaki Kitajima<sup>1</sup>**  
**Takashi Asahara<sup>2</sup>, Koji Nomoto<sup>2</sup>**

Saitama City Hospital, Japan

Department of Surgery, Keio University School of Medicine<sup>1</sup>, Japan  
Yakult Central Institute for Microbiological Research<sup>2</sup>, Japan

**Purpose:** Recently the therapeutic effect of synbiotics is notable in gastrointestinal diseases. The aim of this study was to elucidate the beneficial effects of synbiotics therapy for acute rejection and bacterial translocation after small bowel transplantation in rats.

**Method:** Orthotopic small bowel transplantations were performed and the animals were divided into two groups of with or without synbiotics therapy, which consists of combined oral administrations of *Bifidobacterium breve*, *Lactobacillus casei* and galactooligosaccharide. Feces were sampled to detect the change of intestinal flora and the concentration of organic acid. At day 10, rats were sacrificed and several organs including liver, spleen, mesenteric lymph nodes, ascites and blood were obtained and cultured to evaluate the degree of bacterial translocation. Histological examination of the intestinal grafts was performed to define the acute rejection.

**Results:** Organ culture revealed that bacterial translocation was completely suppressed in treatment group. The concentrations of total organic acid had significantly decreased with the progress of rejection in non-treatment group. Especially the decreased concentrations of lactic acid and butylic acid are remarkable along with the alteration of intestinal flora.

**Conclusion:** Synbiotics therapy in small bowel transplantation could be a novel therapeutic tactics to get a better graft function by keeping normal enteral flora.

## Long-term quality of life after extirpation of sacrococcygeal tumor

Norihiko Kitagawa, Youkatsu Ohhama\*, Hiroshi Take

Yoshimitsu Fukuzato, Toru Murakami

Kanagawa Children's Medical Center, Japan

**Purpose:** Operation and chemo/radiotherapy for sacrococcygeal tumor occasionally occurs various long-term complications. The aim of this study is to clarify these patients' QOL.

**Method:** We experienced 45 patients with sacrococcygeal tumor from 1970 to 2003. Clinical data of 28 patients who were followed up for at least 5 years were analyzed retrospectively.

**Results:** Histopathological analysis showed that 22 cases were benign teratoma, 4 were York sac carcinoma(YSC), 2 were lipoma. Only one patient with benign teratoma developed recurrence of tumor. Rectoanal problem occurred in 5 patients. Urinary incontinence occurred in 2 patients. These neurological problems were related to the patients who had presacral or spinal cord connecting tumor. Adhesional ileus developed in one patient with YSC. As a sequela of chemo/radiotherapy, growth retardation and subarachnoid hemorrhage developed in different patients with YSC. Scar was cosmetically acceptable for many patients, but 7 patient have problems, 2 patients of these hoped to be collected by plastic surgery.

**Conclusion:** As a result of chemo/radiotherapy, patients with YSC tends to have severe complications. On the other hand, the majority of patients with benign tumor could get satisfying functions. Although, 25% of the patients have cosmetic problems. We should take care the problem more when initial operation is performed.

## The Diagnosis of Pancreaticobiliary maljunction (PBM) using Magnetic Resonance Cholangiopancreatography (MRCP)

Tomomitsu Tsuru, Kimio Asagiri, Yoshiaki Tanaka  
Shigeki Hikida, Hiroyoshi Mizote, Hiromichi Yano\*

Department of Pediatric Surgery, Kurume University, School of Medicine, Japan

**Purpose:** Magnetic resonance cholangiopancreatography (MRCP) is a relatively new technique to visualize the pancreaticobiliary tract. MRCP is a non-invasive procedure and poses little risk of complications, such as pancreatitis, in comparison with ERCP. In the present study, we performed MRCP on mostly pediatric patients with choledochal cyst to evaluate the usefulness of MRCP-based pancreaticobiliary maljunction (PBM) diagnosis using a 0.5 T MRI machine.

**Method:** From 1995 to 2003, 20 patients (9 male, 11 female; mean age, 6.8 years; range, 4 months to 25 years) with PBM were prospectively evaluated using MRCP. MRCP was performed using a 0.5 T MRI machine by the 2D-FASE and 3D-FASE methods, and in 6 patients a breathing synchronization system was employed while MRCP was performed.

**Results:** In 4 patients who could breath hold (10 to 25 years old), MRCP clearly diagnosed PBM and identified the morphology of the pancreaticobiliary duct system. In 11 patients who could not breath hold (4 month to 8 years old), PBM was clearly diagnosed using MRCP in the 5 patients above 6 years of age, but could only be diagnosed by intraoperative cholangiography in the 6 patients younger than 6 years of age. However, in 5 patients , MRCP using the breathing synchronization system clearly identified the PBM.

**Conclusion:** PBM was diagnosed in children as young as 2 years using a 0.5 T MRI machine equipped with a breathing synchronization system. Therefore, we believe that a future version of the breathing synchronization system will facilitate the identification and diagnosis of PBM in younger children.

## **Effect of peroral coenzyme Q10 supplementation on chronic hepatic dysfunction in children with biliary atresia**

**Yoshiaki Tanaka, Horoyoshi Mizote, Kenjiro Akiyoshi, Kimio Asagiri, Tomomitsu Tsuru, Shigeki Hikida, Horomichi Yano\***

Department of Pediatric Surgery, Kurume University School of Medicine, Japan

**Purpose:** We aimed to study whether the peroral administration of coenzyme Q10 (CoQ10) as an antioxidative agent might improve the hepatic dysfunction in biliary atresia (BA) patients.

**Method:** In twelve postoperative BA patients given CoQ10 (1.6mg/kg/day) for 100 days and in twelve healthy volunteers without administration as control, blood levels of superoxide dismutase activity(SOD), manganese-SOD concentration(Mn-SOD), copper concentration(Cu) and hepatic enzymes, and urine levels of 8-iso-prostaglandinF2 (8-Isoprostanate) and 8-hydroxy-2'-deoxyguanosine(8-OHdG) were evaluated before and after administration.

**Results:** Before administration, compared to the normal range, SOD values in all patients and both Mn-SOD and Cu in 8 were high. SOD values, Mn-SOD and Cu were excessively high in the 6 cases with low level of Insulin-like growth factor-1(IGF-1) or high level of TypeIV collagen. In the patients with Mn-SOD values(ng/mL) over 200, 8-Isoprostanate and 8-OHdG were significantly higher than those in the group under 200. Mn-SOD was strictly correlated with ALT, AST and IGF-1 respectively. All oxidative stress markers in BA patients were significantly higher than those in control. After administration, every oxidative stress marker and hepatic enzyme improved.

**Conclusion:** We conclude that the administration of CoQ10 could improve hepatic dysfunction in BA patient, which might be the protection effect of intrahepatic oxidative stress.

## The Effects of Fatty Liver on Hepatocyte Apoptosis And Liver Regeneration after Partial Hepatectomy

**Hong-Shiee Lai\***

National Taiwan University Hospital and National Taiwan University  
College of Medicine, Taiwan

**Purpose:** Nonalcoholic fatty liver is a chronic liver disease that occurs in nondrinkers. The liver-related morbidity and mortality that accompanies with the fatty liver may occur due to an increased apoptosis of hepatocytes and a decreased capacity of the liver regeneration. The purpose of this study is to find out the effects of a fatty liver on the hepatocytes apoptosis and the liver regeneration after partial hepatectomy.

**Method:** Male Wistar rats around 200g were used as a subject. Fatty liver was induced by a high fat diet feeding for 4 weeks. Then the rats were randomly assigned into three groups in which further high fat diet (HF), normal diet (FN) or high fiber diet (Ff) feeding for another 4 weeks were given. Partial hepatectomy (around 70%) were performed in control (C) and all experimental groups and rats were sacrificed at 6, 24, 48 and 72 hours after hepatectomy. The measurements of the following categories were implemented: [1] the ratio of remnant liver weight/ body weight and its associated histological pictures as indicators of fatty liver; [2] *in situ* cell death detection (TUNEL) of DNA fragmentation as indicators of apoptosis; [3] 5-Bromo-2-Deoxyuridine (BrdU) activities; and [4] ornithine decarboxylase (ODC) contents in remnant livers as markers of regeneration.

**Results:** We found that [1] high fat diet feeding for 4 weeks could induce the fatty liver markedly; [2] the apoptosis of hepatocytes was increased in fatty liver when compared with that of normal liver ( $98\pm19$  vs  $36\pm7$ ) at 6h after partial hepatectomy ( $p<0.05$ ); [3] the capacity of the liver regeneration was decreased significantly (BrdU index:  $30\pm5$  vs  $12\pm3$ , and ODC contents:  $604\pm48$  vs  $390\pm42$  mg/dL) in a fatty liver at 24h after partial hepatectomy ( $p<0.05$ ); [4] a normal diet could mildly recover the apoptotic situation and regeneration capacity; [5] a high fiber diet could significantly reduce the increased apoptotic number and enhance the lower liver regenerating capacity in the fatty liver ( $p<0.05$ ).

**Conclusion:** Fatty liver can be induced by high fat diet, and it may increase the apoptosis of hepatocyte and decrease the capacity of liver regeneration after partial hepatectomy. A high fiber diet could reverse fatty liver and recover the negative effects of fatty liver on apoptosis and the regeneration capacity which occurred after partial hepatectomy.

## Laparoscopic diagnosis and treatment for disorders of the umbilicus in children

**Keisuke Nose, Makoto Yagi, Hideki Yoshida, Akio Kubota\***

**Hitoshi Shiozaki, Harumasa Ohyanagi**

Kinki University School of Medicine, Japan

**Purpose:** We have developed a laparoscopic approach to diagnose and treat disorders of the umbilicus. The aim of this study was to evaluate its efficacy.

**Method:** Laparoscopically assisted resection of umbilical structures was performed in three patients with infected umbilicus. The patients age ranged from one month to six years old. An initial 5-mm trocar was inserted through an upper abdominal incision with a pneumoperitoneal pressure of 8-10mmHg. A rigid-type telescope was introduced, and two additional working ports (3 or 5mm) were inserted. Methylene blue was injected through the external fistula orifice. Laparoscopically, the trace of methylene blue was found beneath the fascia and the fistula tract was visualized. The whole tract along with the skin sinus was detached from the surroundings, ligated by using an endoscopic suturing device and excised by using diathermy scissors. The dissected umbilical remnants were removed from through a subumbilical incision.

**Results:** Mean operation time was 105±8 minutes. Excellent laparoscopic view revealed the umbilical structures as urachus remnants in two patients and arteria umbilicalis remnants in one, which were confirmed by histological examination. No major complications occurred during or after surgery. All of the patients were discharged within one week with good cosmesis.

**Conclusion:** The laparoscopic approach is supposed to be effective for accurate diagnosis and appropriate treatment of umbilical disorders in children.

## Expression of RET Protein as an ancillary diagnostic tool for Hirschsprung's Disease

Jung-Tak Oh, Soo Min Ahn, Woo-Ick Yang<sup>1</sup>, Seok Joo Han  
Seung Hoon Choi\*, Eui Ho Hwang\*

Department of Pediatric Surgery, Yonsei University College of Medicine, Korea  
Department of Pathology, Yonsei University College of Medicine<sup>1</sup>, Korea

**Purpose:** The pathophysiology of Hirschsprung's disease (HD) is not fully understood, but recent studies have disclosed that RET protein is involved in differentiation and proliferation of neural crest cells. To evaluate the roles of RET protein in HD, immunohistochemical analysis was performed using formalin-fixed and paraffin-embedded tissue sections.

**Method:** The entire resected specimens of the colon were obtained from 13 patients. The specimen was cut longitudinally into 0.5-1 cm width, and full-thickness strip. All specimens were fixed in formalin and embedded in paraffin wax. Each specimen was stained immunohistochemically for RET protein.

**Results:** With the monoclonal antibody for RET protein, the ganglion cells in both the submucosal and myenteric plexuses were positively stained with varying intensity and the nerve fibers in them also showed some reactivity. However the submucosal and myenteric plexuses in aganglionic segments from all samples were clearly negative for RET protein. The polyclonal antibody to RET protein did not work on paraffin embedded tissue sections convincingly although we tried varying dilutions and several sensitive staining methods.

**Conclusion:** These data suggest that RET protein may play important roles in pathogenesis of HD and immunohistochemical staining for RET protein can be used as an ancillary diagnostic tools for HD.

## A Successful Resection of an Acquired Megarectum with Posterior Sagittal Approach in Redo Pull-through Operation in Hirschsprung's Disease

Seok Joo Han, Sung Kook Sohn, Soo Min Ahn<sup>1</sup>, Jung-Tak Oh<sup>2</sup>  
Seung Hoon Choi<sup>2</sup>, Eui Ho Hwang\*<sup>3</sup>

Department of Surgery, Yongdong Severance Hospital, Yonsei University College of Medicine, Korea

Department of Surgery, Hallym University College of Medicine<sup>1</sup>, Korea

Department of Surgery, Yonsei University College of Medicine<sup>2</sup>, Korea

Department of Surgery, National Health Insurance Corporation Ilsan Hospital<sup>3</sup>, Korea

**Purpose:** To present the posterior sagittal approach during redo pull-through operation in an eight year old boy for severe fecal incontinence due to megarectum after previous surgery for Hirschsprung's disease.

**Method:** The coccyx, the levator muscle, and the striated muscle complex were divided in the midline under the guidance of electrical stimulation. The rectal wall was exposed, and a circumferential incision was made around the rectal wall 1 cm proximal from dentate line. The megarectum was mobilized by dissecting close to the adventitia rectalis, which could be identified by traction on the rectum. The megarectum could be pulled easily though out of the skin incision by moderate traction. The entire dilated hypertrophic segment of rectum was resected, and new anastomosis was reconstructed by relocating the proximal normal sigmoid colon to the anal stump. The levator muscle and the striated muscle complex were closed in layers with help of electrical stimulation. No covering colostomy was done.

**Results:** The patient had increased stool frequency in the early postoperative period but improved with time. The patient has attained normal voluntary bowel actions.

**Conclusion:** The posterior sagittal approach offers excellent exposure, precise dissection, and direct anastomosis in redo pull-through operation in Hirschsprung's disease.

## Can an appropriately-timed vesicostomy prevent the development of dysplasia in an experimental model of complete urinary tract obstruction?

**Hideaki Sato, Hiroaki Kitagawa\*, Kevin. C. Pringle<sup>1</sup>, Junki Koike<sup>2</sup>  
Jane Zuccollo<sup>3</sup>, Yuriko Sato, Mieko Odanaka<sup>2</sup>, Koonosuke Nakada\***

Division of Pediatric Surgery, St.Marianna University School of Medicine, Japan  
Department of Surgery, Obstetrics & Gynecology, Wellington School of Medicine  
and Health Sciences<sup>1</sup>, Newzealand

Department of Pathology, St.Marianna University School of Medicine<sup>2</sup>, Japan  
Department of Obstetrics & Gynecology, Wellington School of Medicine<sup>3</sup>,  
Newzealand

**Purpose:** Many investigators have attempted to bypass in-utero urinary tract obstruction to preserve renal development. Theoretically, bypassing the obstruction will preserve renal function. We created an obstructive uropathy in fetal lambs and created a vesicostomy to salvage renal function.

**Method:** We created an obstructive uropathy in fetal lambs at 60 days gestation by ligating the urethra and urachus. Vesicostomy (female) or urethrostomy (male) were performed 21 days later to release the pressure. Fetuses were sacrificed at term (145 days) and the urinary tract was removed for histology.

**Results:** Thirteen fetuses were shunted. Seven fetuses miscarried after shunting. Six survived and three had a successful shunt with a very small bladder (5 to 7 ml). Two had incomplete shunts that failed some time after shunting. In one, the obstruction was unsuccessful. In fetuses with a successful shunt (n=3) or a temporary shunt (n=2), the histology demonstrated relatively preserved renal architecture with decreasing of nephron mass in two and multicystic dysplastic change (MCDK) in three. None had normal nephrogenesis.

**Conclusion:** Our pilot study of shunt operations to salvage obstructive uropathy partially succeeded. We conclude that earlier shunting may salvage the kidney in complete obstruction but selecting optimal timing for shunting may be difficult to select in human obstructive uropathy.

## **Bladder Auto-augmentation Using Biodegradable Polymer Seeded With Autologous Cells In A Rabbit Model-A Preliminary Result**

**Jin-Yao Lai, Jer-Nan Lin, Pei-Yeh Chang**

Chang-Gung Children's Hospital, Taiwan

**Purpose:** The prolapsed mucosa after bladder auto-augmentation usually collapses and the volume increment is limited. This study is aimed at auto-augmentation covered with polyglycolic acid (PGA) mesh seeded with autologous bladder smooth muscle cells in a rabbit m

**Method:** Auto-augmentation surgery was performed one month after an initial 70% partial cystectomy, including traditional autoaugmentation (TA, n=6), PGA mesh autoaugmentation without cells seeding (PGA-N, n=6), and PGA seeded with autologous bladder smooth muscle

**Results:** PGA-C group showed significant bladder capacity increment as compared to the other groups in all time points ( $p=0.006$ ,  $< 0.001$ , 0.016 at 1st, 2nd and 3rd months respectively). Normal urothelial layer was maintained in all groups. The PGA-C group showed g

**Conclusion:** Cell-seeded PGA polymer could facilitate smooth muscle regeneration and offer good structural support and significant volume increment for the auto-augmentation.

## Esophageal Atresia in Very Low Birth Weight Infant

Ryuichi Shimono, Hideo Takamatsu<sup>1</sup>, Hiroyuki Noguchi<sup>1</sup>

Satoshi Ibara, Yuko Maruyama, Hideki Maruyama

Jiro Kikuchi<sup>2</sup>, Hiromitsu Matsuda<sup>2</sup>

Perinatal Medical Center, Kagoshima City Hospital, Japan

Department of Pediatric Surgery, Faculty of Medicine Kagoshima University<sup>1</sup>, Japan

Pediatric Surgery, Kagoshima City Hospital<sup>2</sup>, Japan

**Purpose:** Primary esophageal repair with gastrostomy had been done routinely in all cases with esophageal atresia with tracheo-esophageal fistula (TEF) including mature infant from 1984 to 1992. But primary repair without gastrostomy has been a standard procedure excepting for the cases with severe associated anomalies, respiratory or cardiac failure. Management of (TEF) in very low birth weight infant (VLBWI) was researched since 1992. In this paper, management of TEF in VLBWI was researched.

**Method:** Nine VLBWIs with TEF were managed in our hospital from 1984 to 2003 (<1000g 2 cases, 1000~1500g 7 cases). Their prognosis, operative procedure and associated anomalies were reviewed.

**Results:** Three of six with 18-trisomy could not be operated because of severe cardiac anomalies. Only gastrostomy was done in one case and lower esophageal banding with gastrostomy was done in two cases. Primary esophageal repair was done in two cases without 18-trisomy including one case with coarctation of the aorta, and their clinical courses have been good. Gastrostomy, lower esophageal banding and duodenoduodenostomy with delayed repair were chosen in one case associated with hypoplastic lung, duodenal atresia and gastric perforation.

**Conclusion:** Primary esophageal repair could be done safety even in VLBIs, but staged operation with multiple procedures is necessary in cases with severe respiratory failure or severe associated anomalies.

## Unusual Causes of Dysphagia and Esophageal Stricture

**Hamid Reza Foroutan\***

Nemazee Hospital, Shiraz University of Medical Sciences, Shiraz, Iran

**Purpose:** The purpose of this study is to present 2 patients with unusual cause of dysphagia and esophageal stricture

**Method:** the first was a three years old boy presented with dysphagia for two months. Upper gastrointestinal series showed dilated esophagus with a short stricture in the distal part of the esophagus. Endoscopy showed normal mucosal of the esophagus with a stricture in the distal part of the esophagus, but the endoscope was passed into the stomach. Chest X-Ray showed pleural effusion. Pleural tap showed non-clotting blood in the pleural cavity and chest tube was inserted and 300 ml blood was drained. Then the patient underwent chest CT Scan, which showed a large space-occupying lesion in the posterior of the mediastinum. The other patient was a 12years old boy with dysphagia for a few months. Chest X-ray showed a calcified mass in right side of the chest. Barium swallow showed severe pressure effect on the esophagus.

**Results:** Thoracotomy was performed for the first patient, which showed a large mass in the posterior mediastinum, invading the diaphragm and pleura. The esophagus was intact with no perforation. Histopathologic study was in favor of Germ cell tumor. Exploration for the second patient showed a bony structure in the chest with invasion to diaphragm and esophagus. The mass was partially excised. Pathology result was inflammatory pseudo-tumor.

**Conclusion:** Although rare, but in mediastinal masses, germ cell tumor and inflammatory pseudo-tumor should be considered in patients with acute onset of dysphagia and proper work up should be done for better diagnosis and treatment.

## Internal Jugular Venous Aneurysm in a Child

Dae-Yeon Kim, Seong-Chul Kim, In-Koo Kim

University of Ulsan College of Medicine and Asan Medical Center, Korea

**Purpose:** Congenital dilatation of jugular veins is uncommon in children. Unlike venous aneurysms in adults, which are usually seen in association with previous trauma or disease involving contiguous veins, in children the etiology is uncertain. Because cervical swelling in children is a common diagnostic problem, venous aneurysm must be included in the differential diagnosis of enlarging mass on Valsalva maneuver. Once the diagnosis of venous aneurysm is suspected clinically, it can be confirmed by ultrasonography or computed tomography.

**Method:** A 16-month-old girl was referred with an intermittent swelling on the right side of the neck, first noted 1 month ago. The patient denied any previous trauma to the region. A 5x4 cm sized mass was painless and non-pulsatile, became more prominent on crying and almost doubled in size on compression of right upper neck. A CT angiography revealed an enlargement of the ipsilateral internal jugular vein with a large aneurismal dilatation. The parents were concerned about the cosmetic nature of the mass.

**Results:** The aneurysm was excised through a vertical incision along the posterior margin of sternocleidomastoid muscle. Postoperative course was uneventful.

**Conclusion:** No known complication, such as rupture or infection, has been reported in these cases. Surgical excision is indicated for cosmetic reasons.

## The Study on the Increasing Effect of the Abdominal Cavity by the Procedure on Abdominal Wall Muscles in Puppies

**Woo-Sup Ahn, Seung Hoon Choi<sup>1</sup>, Sung Kim<sup>2</sup>**

Pohang Hospital, Dongguk University, Korea

Yonsei University College of Medicine<sup>1</sup>, Korea

Samsung Medical Center, Sung Kyun Gwan University<sup>2</sup>, Korea

**Purpose:** The ideal repair of gastroschisis and large omphalocele is complete primary closure without compromising the respiratory state, venous return, or intestinal blood supply. This study is intended to device a procedure for the treatment of these diseases.

**Method:** Five healthy new born puppies were used for the experiment. A silastic balloon was inserted into the abdominal cavity through a small vertical incision about umbilicus in control(pre-op). After extension of the vertical incision, skin and three layers of abdominal wall muscles were seperated laterally up to the posterior axillary line in the experiment(post-op). Three layers of abdominal muscles were arranged to two layers. Pre- and postoperatively, air was blown into the silastic balloon step by step respectively. Pressure variations of inferior vena cava and stomach versus inserted air volume were mesured up to 30 cm H<sub>2</sub>O under the adequate central venous pressure and graphed for comparative analysis.

**Results:** When the input of air volume reached 240 ml, the pre- and postoperative mean inferior vena cava pressure were 32 cm H<sub>2</sub>O and 17 cm H<sub>2</sub>O, respectively.

**Conclusion:** This procedure will make the primary closure easier and minimize the complications in most cases except extremely hage omphalocele

## Ileal Atresia Secondary to Neglected Intussusception in Ventilatory Supported Prematurity

Soo Jong Ko, Jae Wook Seol, Jae Hoon Lee, Young Tag Lee

St. Benedict Hospital, Korea

**Purpose:** Intrauterine intussusception has been known as a cause of small bowel atresia. This case acquired ileal atresia due to disconnection of strangulated intussusception with a fibrotic adhesive band.

**Method:** 1600 gram girl was born at 30 weeks' gestational age. She was supported by a mechanical ventilator and total parenteral nutrition after birth. Meconium was passed normally. Abdominal distension and bloody stool was presented at 51st day of birth. Medical treatment was started under the suspicion of necrotizing enterocolitis including antibiotics. Abdominal distension and respiratory difficulty were wax and wane. At the 80th day of birth, we explored under the suspicion of mechanical intestinal obstruction.

**Results:** The operation was performed through right supraumbilical transverse skin incision. Ileoileal intussusception was identified at 20 cm from ileocecal valve. Proximal bowel was cut clearly without mesenteric disruption. Fibrous band was adherent tightly to the end of blind loop of proximal bowel. Intussusceptum was strangulated and necrotized in the intussuscepiens. Intussuscepted bowel was resected and end-to-end anastomosis was performed. Oral feeding was started at 4th postoperative day. She was discharged well with normal growth.

**Conclusion:** We experienced a case of ileal atresia secondary to neglected intussusception in the prematurity whose respiration was supported mechanically. We report this case with references.

## An Unusual Presentation of an infected Urachal cyst-Case Report and Review of the Literature

Chih-Cheng Luo\*, Chen-Sheng Huang

Department of Pediatric Surgery, Chang Gung Children's Hospital,  
Chang Gung University, Taiwan

**Purpose:** We present an unusual case of an infected urachal cyst in an 1-month-old female infant, who presented with fever, erythema overlying right lower abdominal wall, and a firm, fixed right lower quadrant tender mass.

**Method:** This 1-month-old female infant was admitted to the Chang Gung Children's Hospital due to fever and erythema overlying right lower abdominal wall. Sonography and CT scan revealed a 4X6 cm multilobulated cystic mass located in the right pelvic cavity, suggesting an intra-abdominal mass. She accepted antibiotic treatment underwent exploratory laparotomy under the impression of perforation of appendicitis with abscess formation.

**Results:** During laparotomy, a right lower quadrant inflammatory cystic mass beneath the edematous peritoneum could be found. Dissection was carried out until a cystic mass extending to the dome of the bladder. The cyst was totally excised. Pathologic examination showed acute and chronic inflammation of the urachal cyst with a lining of degenerative transitional cell epithelium.

**Conclusion:** An infected urachal cyst presents with a tender lower midline abdominal mass localized between the umbilicus and the suprapubic area. To our knowledge, this unusual presentation of an infected urachal cyst has rarely been reported.

## Torsion of jejunal duplication cyst: a rare cause of midgut volvulus in neonate

Beatrice P.Y. Wong, Michael. W.Y. Leung<sup>1</sup>, Nicholas S.Y. Chao<sup>1</sup>  
Kelvin K.W. Liu\*

United Chritian Hospital, China  
Queen Elizabeth Hospital<sup>1</sup>, China

**Purpose:** Duplication cyst has various clinical manifestations. We report a case of jejunal duplication cyst presenting with midgut volvulus in a neonate.

**Method:** A female full term neonate presented to us with bile-stained vomiting and abdominal distension on day one. Upper gastrointestinal contrast study showed no malrotation but sluggish of contrast flow at proximal jejunum. Emergent laparotomy was performed. Midgut volvulus leading to multi-segmental small bowel infarct with gut perforation was identified. There was a duplication cyst at the mesenteric border of jejunum 10cm distal to the duodenojejunal junction acting as the axis of volvulus. De-twisting of volvulus was done. Resection of the bowel segment with the duplication cyst and three other gangrenous bowel segments were performed. Primary anastomoses of distal segments with proximal double-barrel stoma were done. Total parenteral nutrition (TPN) was started after the operation. Histopathology confirmed the diagnosis of jejunal duplication cyst with ectopic gastric mucosa. Distal loopogram done post-operatively showed no bowel stricture. Closure of stoma was performed subsequently.

**Results:** The patient enjoyed uneventful recovery. Full oral feeding was resumed with TPN weaned off.

**Conclusion:** Torsion of jejunal duplication cyst is a rare cause of midgut volvulus. High index of clinical suspicion and prompt surgical intervention is the key to successful management.

## **Analysis of cystic duct insertion into extrahepatic bile duct in children with congenital biliary dilatation**

**Naoki Okuyama, Masayuki Kubota\*, Minoru Yagi\***

**Satoshi Kanada, Satoru Yamazaki**

Niigata University Graduate School of Medical and Dental Sciences, Japan

**Purpose:** Literally, the pattern of cystic duct insertion into the extrahepatic bile duct (EHBD) reported to be 10%, 75% or 15% in the upper third, middle third or lower third, respectively. Even though such information on CBD seems to be important regarding its pathology, the presence or absence of an anomalous insertion pattern in CBD has not yet been elucidated.

**Method:** We encountered 51 patients with CBD from 1983 to 2002. Thirty-three of the 51 patients underwent ERCP. The cystic duct and anomalous pancreaticobiliary ductal union was confirmed by ERCP in 23 of 33 patients who were enrolled in this study.

**Results:** The cystic duct was inserted into the upper third, middle third or lower third of the EHBD in 43.5%, 47.8% or 8.7% of the patients, respectively. In 10 patients with cystic type dilatation, the cystic duct was inserted into the upper third or middle third in 60% or 40%, and in 13 patients with fusiform type dilatation, the cystic duct was inserted into the upper third, middle third or lower third of the EHBD in 30.8%, 47.8% or 8.7%.

**Conclusion:** There was a difference in the pattern of cystic duct insertion between cystic and fusiform types, which may suggest a different etiological background of these types.

## Laparoscopic Anterior Gastropexy for Gastric Volvulus in a Child

Seok Joo Han, Choon Sik Yoon<sup>1</sup>, Soo Min Ahn<sup>2</sup>, Jung-Tak Oh  
Seung Hoon Choi, Eui Ho Hwang<sup>\*3</sup>

Department of Pediatric Surgery, Yonsei University College of Medicine, Korea

Department of Diagnostic Radiology, Yonsei University College of Medicine<sup>1</sup>, Korea

Department of Surgery, Hallym University College of Medicine<sup>2</sup>, Korea

Department of Surgery, National Health Insurance Corporation Ilsan Hospital<sup>3</sup>, Korea

**Purpose:** To present the surgical technique of the laparoscopic anterior gastropexy for the gastric volvulus in children

**Method:** A six-year-old female had several attacks of acute abdominal pain and vomiting during the admission for the rehabilitation of the sequelae of hypoxic brain damage. The plain X-ray at the time of symptoms demonstrated the typical findings of gastric volvulus. These findings resolved with insertion of nasogastric tube. The diagnosis of gastric volvulus was made. Laparoscopic fixation was performed at the time of the attack. Three ports were inserted and the anterior stomach wall was fixed to the anterior abdominal wall with three non-absorbable stitches.

**Results:** The patient recovered without complications and has been asymptomatic for 24 months since the surgery.

**Conclusion:** In conclusion, laparoscopic anterior gastropexy is a less invasive and very useful surgical procedure for the correction of the gastric volvulus in children.

## Laparoscopic fixation for cecal volvulus in a child

**Tomio Ogawa\*, Yoshiyuki kamii, Kota Okinaga**

Teikyo University Hospital, Japan

**Purpose:** A laparoscopic approach was used to fix the cecum to the abdominal wall in a ten-year-old girl with a history of cecal volvulus. The patient recovered without complication and has been asymptomatic since the surgery.

**Method:** A ten-year-old female was admitted several times from the age of 5 months for acute attacks of abdominal pain and vomiting. Her symptoms resolved after several days of avoiding oral intake. Barium enema studies at the time of symptoms demonstrated the absence of barium into the ileo-cecal valve and the characteristic bird-beak sign of cecal volvulus. These findings resolved with increased barium pressure, which subsequently resolved symptoms. The diagnosis of cecal volvulus was made. Laparoscopic fixation was performed when the child was 10-years-old and asymptomatic. Three ports were inserted and the cecum was fixed to the lateral abdominal wall with three stitches.

**Results:** The patient recovered without complications and has been symptom free for 18 months since the surgery.

**Conclusion:** Laparoscopic fixation of the cecum is a less invasive and more effective procedure for cecal volvulus. Surgery may be performed when patients are asymptomatic to avoid abdominal distension and ensure a safe procedure.

## Laparoscopic cholecystcholangiography for differential diagnosis of biliary atresia: An alternative procedure to exploratory laparotomy

Satoko Nose, Toshimichi Hasegawa\*, Hideki Soh

Takashi Sasaki, Takuya Kimura, Masahiro Fukuzawa\*

Osaka University Graduate School of Medicine, Japan

**Purpose:** The diagnosis of biliary atresia (BA) is based on ultrasonography, computed tomography (CT), hepatobiliary scintigraphy, analysis of the duodenal fluid, etc. However, these studies are not confirmative because of false negative results with considerable rate. In such cases, exploratory laparotomy with cholecystcholangiography has been performed. We have attempted laparoscopic cholecystcholangiography (LC) as an alternative procedure to laparotomy in three patients with direct hyperbilirubinemia.

**Method:** The age at laparoscopy ranged from 28days to 83days. Analysis of the duodenal fluid and hepatobiliary scintigraphy demonstrated no excretion of bile into the small intestine. However, because ultrasonography and CT showed an atrophic gallbladder with a patent lumen, the diagnosis of BA or other diseases was not confirmative. LC was performed by a direct percutaneous puncture of the gallbladder or insertion of the cholangiocatheter into the gallbladder or cystic duct.

**Results:** In two cases, LC did not show the entire biliary systems, and laparotomy confirmed the diagnosis of BA. However, in the other one case, LC demonstrated normal biliary systems and BA was excluded.

**Conclusion:** LC may be a useful method for differential diagnosis of BA when diagnosis of BA is not confirmative by association of a patent gallbladder, and may avoid unnecessary exploratory laparotomy.

## Shock-Associated Non-occlusive Ischemic Colitis in an Infant

Jin-Bok Hwang, Soon-Ok Choi<sup>1</sup>, Woo-Hyun Park<sup>1</sup>

Dept. of Pediatrics, Keimyung University Dongsan Medical Center, Korea

Dept. of Pediatric Surgery, Keimyung University Dongsan Medical  
Center<sup>1</sup>, Korea

**Purpose:** Ischemic colitis is a common disorder in the elderly and developed predominantly left-sided colon. Colonic ischemia affecting young persons has been documented in rare occasions and has developed predominantly in right-sided colon. We report a 3 month-old-boy with strangulated right inguinal hernia, which causes ischemia of the left-sided colon.

**Method:** At emergent surgery, there noted an ischemic perforation of the cecum and large amount of dirty peritoneal fluid. Right hemicolectomy with ileocolostomy was performed. Postoperatively, septic shock has lasted for about 60 hours. Since 14th postoperative day, abdominal distention with small amount bloody diarrhea has developed. Sigmoidoscopy revealed mucosal ulceration with pseudomembrane and a localized necrosis in the sigmoid colon. Vancomycin was not effective. On 20th postoperative day, 2nd operation was performed. There was generalized dilated entire small bowel loops with tight adhesion to the entire colon, which seemed to develop due to ischemic colon with severe serositis. In addition, a localized necrosis in the sigmoid colon and an ileal perforation with localized abscess were found.

**Results:** Pathologic examination of the sigmoid tissue confirmed full-thickness bowel necrosis, compatible with non-occlusive ischemic colitis.

**Conclusion:** The authors believe that this is the first description of non-occlusive ischemic colitis in infancy in English-language literature.

## Ubiquitous aberrant RASSF1A promoter methylation in childhood neoplasia

Ivy HN Wong, Jacqueline KY Chan<sup>1</sup>, Joyce SW Wong<sup>1</sup>, Paul KH Tam<sup>\*1</sup>

Department of Biochemistry, The University of Hong Kong, China

Department of Surgery, The University of Hong Kong<sup>1</sup>, China

**Purpose:** The role of RASSF1A has recently been elucidated in regulating apoptosis, and cell cycle progression by inhibiting cyclin D1 accumulation. Aberrant RASSF1A promoter methylation has been frequently found in multiple adult cancer types.

**Method:** Using methylation-specific PCR and reverse transcription PCR, we investigated epigenetic deregulation of RASSF1A in primary tumors, adjacent nontumor tissues, secondary metastases, peripheral blood cells and plasma samples from children with 18 different cancer types, in association with their clinicopathologic features.

**Results:** Regardless of the tumor size, ubiquitous RASSF1A promoter methylation was found in 67% (16/24) of pediatric tumors, including neuroblastoma, thyroid carcinoma, hepatocellular carcinoma, pancreatoblastoma, adrenocortical carcinoma, Wilms' tumor, Burkitt lymphoma and T-cell lymphoma. Seventy-five percent of pediatric cancer patients with tumoral RASSF1A methylation were male. Methylated RASSF1A alleles were also detected in 4 of 13 adjacent nontumor tissues, suggesting that this epigenetic change is potentially an early and critical event in childhood neoplasia. RASSF1A promoter methylation found in 92% (11/12) of cell lines largely derived from pediatric cancer patients was significantly associated with transcriptional silencing/repression. After demethylation treatment with 5-aza-2'-deoxycytidine, transcriptional reactivation was shown in KELLY, RD and Namalwa cell lines as analyzed by reverse transcription PCR. For the first time, RASSF1A methylation was detected in 54% (7/13), 40% (4/10), and 9% (1/11) of buffy coat samples collected before, during and after treatment, correspondingly, from pediatric patients with neuroblastoma, thyroid carcinoma, hepatocellular carcinoma, rhabdomyosarcoma, Burkitt lymphoma, T-cell lymphoma or acute lymphoblastic leukemia. Concordantly, RASSF1A methylation was found during treatment in plasma of the same patients, suggesting cell death and good response to chemotherapy.

**Conclusion:** RASSF1A methylation in tumor or buffy coat did not correlate strongly with age, tumor size, recurrence/metastasis or overall survival in this cohort of pediatric cancer patients. Of importance, epigenetic inactivation of RASSF1A may potentially be crucial in pediatric tumor initiation.

## A case of metachronous bilateral virilizing adrenocortical tumors with Beckwith-Wiedemann syndrome

Yoshihiro Hayashida, Hideo Takamatsu\*, Hiroyuki Noguchi  
Hiroyuki Tahara, Takamasa Ikee

Kagoshima Univ., Japan

**Purpose:** It is well known that a child with Beckwith-Wiedemann syndrome (BWS) has increased incidence of tumor development. We report a rare case of metachronous bilateral virilizing adrenocortical tumors.

**Method:**

**Results:** A 3,224-g girl had an omphalocele and dyspnea due to a macroglossia at birth. She was diagnosed as BWS and the repair of omphalocele was performed. At the age of 6 years, the patient developed signs of virilization such as rapid growth and clitoromegaly. MRI revealed a circumscribed mass in the right adrenal region. Hormonal investigation showed elevated levels of dehydroepiandrosterone sulphate (DHEA-S, 6,380ng/ml), and testosterone (547ng/dl). Right adrenalectomy was performed and histological examination showed an adrenocortical adenoma with no malignancy. After removal of the tumor, the hormone levels normalized. Since the age of 9 to 13 years, DHEA-S (6,790ng/ml) and testosterone (120ng/dl) levels gradually elevated. MRI revealed a circumscribed mass in the left adrenal region. Adrenal tumor was enucleated and normal adrenal tissue was preserved. Histological examination showed an adrenocortical adenoma as same as the right adrenal tumor. After removal of the tumor, the hormone levels normalized.

**Conclusion:** Once diagnosis of BWS is confirmed, it is essential to evaluate the child for early detection of tumors periodically. Even after the resection of virilizing tumor, one should be aware of the possibility of the metachronous appearance of contralateral lesion.

## **Tactics in Blunt Pancreatic Trauma in Children**

**Anna N Shapkina\*, Vlavimir V. Shapkin, Alina P. Pilipenko**

Department of Pediatric Surgery, Vladivostok State Medical University, Russian Federation

**Purpose:** Aim was to determine the etiology and outcome of BPT in children.

**Method:** We undertook a retrospective review of patients admitted to Vladivostok University Childrens Hospital from 1979 to 2002. There were 33 cases of BPT (8,6% of blunt abdominal injuries): 23 boys, 10 girls. Main mechanism of BPT were falls in 15, motor vehicle accidents in 5, compressions in 3. The most useful was the analysis of trauma mechanism, presence of the associated injury, increasing of serum amylase level, ultrasonography findings. In 17 children diagnosis of BPT was made intraoperatively

**Results:** Hyperamylasemia was found in 66,7% of children. Isolated trauma was found in 10 children, combined with the other injuries in 23. 21 patients underwent surgery, 12 had medical treatment. The type of surgery was external drainage in 8, suture in 2, explorative laparotomy in 1 and 10 underwent laparotomy for combined injury. 8 complications occurred in the operative group: 2 ?pseudocysts, 1 pancreatic fistula, 1- traumatic pancreatitis, 1- duodenal perforation, 2 ?peritonitis, 1 ?intraabdominal bleeding. After non-surgical treatment cysts occurred in 2 cases. 4 children died. There were no deaths in isolated BPT.

**Conclusion:** So, BPT remain uncommon in children and can be suspected in case of polytrauma. BPT has high morbidity but low mortality.

## An unusual variant of bladder hypoplasia

**Kevin Pringle\*, Richard Robinson<sup>1</sup>, Yasuji Seki<sup>2</sup>, Ross Wilson<sup>3</sup>**

Wellington School of Medicine & Health Sciences, Newzealand  
Urology, Capital & Coast District Health Board<sup>1</sup>, Newzealand  
Seki Hospital<sup>2</sup>, Japan

Paediatrics, Capital & Coast District Health Board<sup>3</sup>, Newzealand

### **Purpose:**

### **Method:**

**Results:** Born at term, this child was noted to have ambiguous genitalia. By 2 days of life, both serum creatinine and potassium were raised. She was transferred. On admission, she was noted to have clitoromegaly, no urethral opening and an anterior ectopic anus. A vaginal contrast study revealed no bladder or urethra. An ultrasound revealed echogenic kidneys. The ureters were not dilated. Vaginoscopy identified 2 cervices and an incomplete vaginal septum. Just below this was an opening in the anterior wall of the vagina. No ureters could be identified. Her creatinine continued rising, reaching a maximum of 432mMol/l. A laparotomy was performed to identify any surgically-correctable lesion. There was a 1.5cm diameter bladder immediately anterior to the vagina, just below the uterus, with 2 normal ureters entering it. When she had recovered from the operation, she was sent home to die. Several months later, she returned with her creatinine having fallen to 72mMol/l. Her chronic renal failure was managed. At 2.75years she had a continent reservoir created using the ileocaecal junction. The appendix was used as a catheterisable stoma, and the right ureter was implanted in a non-refluxing fashion. She has now had a living related renal transplant using this reservoir.

### **Conclusion:**

## Surgical Treatment of Chylothorax Caused by Cardiothoracic Surgery

Hsin-Lin Tsai, Chinsu Liu, Taiwai Chin\*, Choufu Wei\*

Taipei Veterans General Hospital, National Yang-Ming University, Taiwan

**Purpose:** To evaluate the surgical method of chylothorax caused by cardiothoracic surgery.

**Method:** Retrospectively review of 4 patients was done.

**Results:** Chylothorax was induced by operations for congenital heart disease in 3 cases and esophagectomy with colon interposition via intrathoracic route in 1 case. The leak sites located in left chest in 1 sternotomy, right chest in 1 sternotomy and 1 right thoracotomy and left upper neck in 1 left thoracotomy. Time interval between chylothorax and surgical intervention ranged from 1 week to 8 weeks and the reason for surgical intervention was conservative treatment failure. All received right thoracotomy and mass ligation of right thoracic duct without detecting the true leak site. In spite of 1 case died of heart failure one day after operation, the other 3 cases had very quick recovery without any sequale

**Conclusion:** Based on our limited experience, we suggest that if the amount of chyle remains large after aggressive conservative treatment of iatrogenic chylothorax, right thoracotomy with mass ligation of right thoracic duct can successfully cure chylothorax on either side. To identify the leak site is risky for severe adhesion by previous operation. The time to operate should be as soon as possible if conservative treatment fails.