The Sky's the Limit

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PRIMARY IMMUNODEFICIENCY IN SOUTH AFRICA – LESSONS FROM A REGISTRY

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Primary immunodeficiencies (PIDs) are rare diseases caused by inherited defects. Established prevalence figures for European, American and Australasian countries are available on electronic databases. No such figures exist for South Africa or for the majority of African countries. With the clinical burden of secondary immunodeficiencies due to HIV and malnutrition these potentially treatable deficiencies are easily missed. General awareness is poor with suboptimal outcome for many. Routine molecular diagnosis is not available in SA, but previous studies have highlighted novel mutations of BTK deletions in agammaglobulinaemia and clustering of complement C6 deficiencies.

Data of over sixty live consenting patients were recorded at the PID Registry, Tygerberg Hospital since November 2006. They reflect a wide spectrum of immunodeficiencies from all over South Africa. Age at diagnosis varies from 3 to 727 months, with delayed diagnosis for common variable immunodeficiency especially. Male patients predominate as expected with X-linked inheritance pattern of the more common antibody deficiency states. Only one black patient is recorded and the majority of patients reside in the Western Cape with sample bias to an area with PID clinic services. Interesting subgroups of hereditary fevers, disseminated warts and ectodermal dysplasias were included. A separate group of 45 complement C6 deficiency patients is known to the Allergy Institute of the University of Cape Town where a cohort with hereditary angioedema is also followed up.

Over sixty percent of patients are on immunoglobulin replacement therapy, which is frequently haphazard and insuffiently monitored. Resultant infectious morbidity such as bronchiectasis, deafness, growth failure and developmental delay are frequently observed.

Parental advocacy and level of informed involvement in childhood PID are crucial predictors to outcome in countries with low level of awareness. With data collection the registry aims at networking between doctors, healthcare workers and patients for earlier diagnosis and improved outcome.

PENICILLIN ALLERGY IN CHILDREN – OFTEN MISDIAGNOSED?

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Background: Penicillin antibiotics are commonly implicated in allergic reactions in children without tests being performed to confirm or refute allergy. Many children are inappropriately labelled penicillin allergic and are treated with alternative antibiotics that may be less effective and more expensive.

Objective: The aim of this retrospective study was to determine the prevalence of true penicillin allergy in children with self-reported allergy.

Methods: Clinical and laboratory data of children referred to the Allergy Clinic at Red Cross Hospital for evaluation of suspected penicillin allergy between July 2002 and June 2007 were analyzed. *Clinical data* included sex, age at first reaction, co-existing atopy and nature of adverse reaction. *Tests* included CAP-RAST® for penicillin V, penicillin G, ampicillin, and amoxil, skin prick tests (SPT) as well as penicillin challenge test. The time interval between adverse drug reaction and evaluation was recorded.

Results: Data of twenty subjects were analyzed. Penicillin allergy was confirmed in 5/20 (25%) subjects. Four were SPT +ve and 2 CAP-RAST +ve. The median age at reaction was 2 years; all were atopic, all presented with urticaria ± angioedema. The median time interval from reaction to evaluation was 2 months.

Penicillin allergy was excluded in 15/20 (75%) subjects. CAP-RAST, SPT and challenge tests were negative in all patients. Median age at reaction was 2,5 years. 3/15 were atopic; 6/15 presented with a maculopular rash, 6/15 with urticaria ± angioedema and 3/15 with an unidentified rash. The median time interval from reaction to testing was 20 months. All non-allergic patients subsequently received penicillin without adverse events.

Conclusions: Penicillin hypersensitivity is relatively uncommon in children. SPT and challenge testing is required to confirm or refute the diagnosis. Accurate diagnosis avoids the morbidity, mortality and economic cost associated with unnecessary withholding of penicillin therapy.

EFFECT OF DESLORATADINE ON EXERCISE-INDUCED BRONCHOCONSTRICTION

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Background: Exercise-induced bronchoconstriction (EIB) is a significant problem in asthmatic patients. The link between allergic rhinitis and asthma is now well established. Patients with allergic rhinitis may have EIB.

Objective: This study compared the effects of desloratadine and placebo on EIB in a group of patients with allergic rhinitis and EIB.

Methods: This was a double blind placebo controlled, randomized, crossover study. Exercise challenge tests were performed before and after 7 days of treatment with either 5 mg desloratadine or placebo. Patients then underwent a washout period for 7 days and were crossed over to receive either 5 mg desloratadine or placebo. The exercise challenge tests were repeated.

Results: Desloratadine had no effect on the reduction in percentage fall in FEV_1, the AUC (0-60min) and the time to recovery.

Conclusions: Desloratadine had a no effect in attenuating the bronchoconstriction caused by exercise in patients with allergic rhinitis and exercise induced bronchoconstriction.

ATOPY IN HIV-INFECTED AND NON-INFECTED CHILDREN IN PRETORIA, SOUTH AFRICA

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Introduction: The relationship between the development or aggravation of a pre-existing atopic state and HIV has not been thoroughly investigated in the South African context. HIV-positive patients have been shown to have a higher prevalence of atopy in some international studies in the early stage of their disease but this has not been documented in children.

Methodology: A prospective convenience sample study of children aged 3 months to 12 years attending the HIV clinic were recruited into the study. Information regarding the child's personal and family history of atopy was recorded. The WHO HIV clinical staging, CD4 counts were recorded. An age and sex-matched control group of healthy children attending routine follow up at the cardiology and the neurology clinic were included. Spin prick tests (SPT) for common aeroallergens were conducted in all patients.

Results: A total of 100 patients were included in the study with 50 in each arm. 10% of the HIV-infected patients in comparison to 16% of controls had positive SPT for aeroallergens. Of the HIV-infected patients a high number of patients had allergic rhinitis and eczema (60% and 68% respectively). There is a lack of correlation between CD4 count and any SPT positivity (r=0.011), CD4 count and presence of reported asthma (r=-0.020), and CD4 count and reported presence of dermatitis (r=-0.06). CD4 count was not statistically different between children with and without family history of atopy p=0.68.

Only the oral presentations at 'The Sky's the Limit' appear in SAJCH, *owing to space constraints.*

FOUR CASES OF GENITO-URINARY SCHISTOSOMIASIS IN CHILDREN

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Schistosomiasis (STS) is a parasitic infestation found in north, sub-Saharan and southern Africa, the Middle East and western Asia. *Schistosoma haematobium* is one of the three common species of the parasite. It has a life span of 20-30 years, resides in the pelvic venous plexus and affects the genito-urinary tracts. Painless terminal haematuria is the characteristic manifestation of urinary STS. The basic lesion is an inflammatory granuloma resulting from the host's reaction to *Schistosoma* eggs. Long standing infestations may result in fibrosis and calcification.

Patients present to surgeons when there is intense tissue reaction and/or when complications set in. We present four cases of genito-urinary STS in children. In two cases there was painless terminal haematuria. One patient had an orchidectomy for what was thought to be a testicular tumour. The fourth patient had an incidental finding of calcified nodules on the pelvic peritoneum on laparoscopy that were found to be due to STS.

We discuss the presentation, investigations, endoscopic/ surgical findings, histology of these four patients with genito-urinary STS and review the relevant literature.

Conclusions: Genito-urinary STS is a common problem in children in this part of the world. It should be considered in the differential diagnosis of genito-urinary pathologies in children. Urine examination, ultrasound, endoscopy and biopsy are the mainstay for the diagnosis. Mutilating surgery could be avoided if the diagnosis of STS is considered preoperatively. Treatment with praziquantel is curative.

SURGICAL MANAGEMENT OF NECROTIZING ENTEROCOLITIS: OUTCOME OF PERITONEAL DRAINAGE IN BLOEMFONTEIN ACADEMIC HOSPITALS

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Peritoneal drainage for NEC has been used since 1975. It was initially described as a temporary procedure, but is increasingly used as the sole surgical intervention. Being very fragile patients, the insult of drainage is much smaller than a laparotomy. Management of NEC with drains alone versus drains followed by surgery once the patient is stable, is controversial.

We assessed patients treated with drains to determine the outcome and to see how many needed definitive surgery afterwards. Case records of 29 patients in need of surgical treatment over the past five years were retrospectively reviewed. Three were taken to theatre directly. In 26, a 6 mm pencil drain was placed in each lower quadrant. Four patients died – two before any further intervention and two after laparotomy. Of the remaining 22, eleven needed no further surgery. In the other eleven the indication for surgery was as follows: worsening of general condition: 8; resection of stenotic areas established on contrast enemas before discharge: 2; persistent entero-cutaneous fistula: 1.

In conclusion: 50% of our NEC-patients requiring surgical intervention and treated with peritoneal drainage did not need any further surgery. This is in keeping with other authors (27 – 46%). In 14% an emergency was turned into an elective operation on a healthy baby with good feeding status. Although big randomized, controlled studies are still pending, the outcome of this retrospective review emphasizes the value of using peritoneal drainage in the surgical management of NEC.

PRE-OPERATIVE FACTORS ASSOCIATED WITH MORTALITY AMONG NEONATES WITH GASTROSCHISIS IN ZIMBABWE'S PAEDIATRIC SURGICAL UNIT

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Background/purpose: Previous studies of gastroschisis in Zimbabwe showed survival rates below 42%, which contrasts with high survival rates in developed countries. The difference shows there is potential for improvement, and the aim was to identify those factors associated with mortality pre-operatively.

Methods: An 8-year retrospective analytical study was carried out on neonates with gastroschisis from January 1996 to December 2003. Twenty-five potentially predictive variables were analysed against mortality. Statistical analysis included a univariate and multivariate analysis. Relative risks were estimated by computing the odds ratio.

Results: 140 cases were analysed. 1) *Overall survival:* 35%. 2) *Demographics:* Delivery at the central hospital was associated with a better outcome (p=0.049, OR=0.35), than at a peripheral hospital, clinic or home. A longer time from birth to admission was associated with an adverse outcome (p=0.0006, OR=1.085). 3) *Obstetric and neonatal:* There was no association with the age of mother, parity, delivery mode, Apgar, sex, gestation and birth weights. 4) *Presenting condition:* Dehydration (p=0.041, OR=2.26), respiratory distress (p=0.015, OR=3.19), gangrenous bowel (p=0.011, OR=5.79) and abnormal fibrin grade (p=0.031, OR=3.34) were associated with an adverse outcome. Hypothermia, perforated bowel, bowel atresia, eviscerated organs, and abnormal blood parameters were non-significant. 5) *Multivariate analysis:* Only presenting with respiratory distress (p=0.044, OR=2.81), and a longer time between birth and admission (p=0.002, OR=1.083) remained significantly associated with mortality.

Conclusion: Survival remains poor. Presenting with respiratory distress, and a longer time from birth to admission were independent predictors of mortality. For every hour delay, there is an 8% estimated increase in risk of mortality, therefore doubles every 12 hours. It is recommended that cases diagnosed prenatally be delivered at the central hospital, or transfers be immediate and direct to the Paediatric Surgical Unit.

OPERATIVE FACTORS ASSOCIATED WITH MORTALITY AMONG NEONATES WITH GASTROSCHISIS IN ZIMBABWE'S PAEDIATRIC SURGICAL UNIT

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Background/purpose: With previous operative survivals of gastroschisis in Zimbabwe below 42%, the aim was to identify those operative factors associated with mortality.

Methods: An 8 year retrospective analytical study was carried out from January 1996 to December 2003, and twenty potentially predictive variables were analysed against mortality.

Results: 120 cases were analysed. 1) *Operative survival:* 41%. 2) *Times for surgery:* The time from birth to surgery was marginally non-significant (p=0.058, OR=1.04). There was no difference in the times between admission and operation for the two groups, and no association as to the time of day surgery, or NICU operations. 3) *Surgical procedures:* Primary closure predicted for a better outcome (52% versus 29% survival, p=0.003, OR=3.7), and abnormal fibrin grades were associated with silo closure (p=0.011), and an adverse outcome (p=0.021, OR=4.3). Procedures to the bowel predicted for mortality (p=0,019, OR=7.6). 4) *Ventilatory support:* There was no association between the type of closure and need for ventilation, however the need for ventilation predicted for mortality (CPAP p=0.006, OR=4.0) (IPPV p=0.001, OR=6.1). 5) *Multivariate analysis:* Silo closure (p=0.008, OR=3.38), procedure on bowel (p=0.034, OR=6.6), CPAP (p=0.006, OR=4.4), and IPPV (p=0.001, OR=8.1) remained significantly associated with mortality.

Conclusion: There has been no improvement in the operative survival (41%). Silo repair predicts for a higher mortality, and its use was associated with abnormal fibrin. There was no association between the type of abdominal closure and need for ventilatory support, however the need for ventilation predicted for mortality, as does as a surgical procedure to the bowel. It is recommended in our setting to attempt a primary closure, but not forced if there is respiratory compromise, in order to reduce the reliance of ventilatory support.

POSTOPERATIVE FACTORS ASSOCIATED WITH MORTALITY AMONG NEONATES WITH GASTROSCHISIS IN ZIMBABWE'S PAEDIATRIC SURGICAL UNIT

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Background/purpose: With previous operative survivals of gastroschisis in Zimbabwe below 42%, the aim was to identify those factors associated with mortality postoperatively.

Methods: An 8-year retrospective analytical study was carried out from January 1996 to December 2003, and twenty-five potentially predictive variables were analysed against mortality.

Results: 120 cases were analysed. 1) *Post-operative survival:* 41%. 2) *Post-operative complications and management:* Respiratory distress >48 hrs post-op, receiving blood transfusion and parenteral nutrition were not associated with an adverse outcome. The development of a surgical complication was marginally non-significant (p=0.054, OR=3.44), however, those who required re-laparotomy (p=0.037), developed post-op haemorrhage

(p=0.044, OR=4.1), and sepsis (p=0.018, OR=2.9) were associated with mortality. There was no association between sepsis and the type of closure, fibrin grade, gangrenous bowel, or procedure to the bowel. 3) *Times:* The time to developing a surgical complication was sooner for silo closure but marginally non-significant (p=0.058, OR=1.15), and the time to removal of silo was not associated with outcome (p=0.137, OR=1.32). Primary closure was associated with starting oral feeds earlier (p=0.0001), and a shorter hospital stay (p=0.018, OR=0.93). 4) *Multivariate analysis:* Only post-operative sepsis (p=0.004, OR=4.1) remained significantly associated with mortality.

Conclusion: There has been no improvement in survival (41%). Post-operatively, sepsis was found to be an independent predictor of mortality, but was not associated with the type of abdominal closure, the fibrin grade, procedure to the bowel, or gangrenous bowel. Primary closure was associated with a shorter time to oral feeds and shorter hospital stay. It is recommended to take special care in aseptic techniques, as well as close monitoring for sepsis, and its prompt treatment.

PRIMARY ANTERIOR SAGITTAL ANORECTOPLASTY IN THE TREATMENT OF LOW ANORECTAL MALFORMATIONS IN FEMALES – THE DURBAN EXPERIENCE

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Surgical correction of anorectal malformations is traditionally a staged procedure which involves a colostomy. Recently, this view has been challenged, with single stage procedures being employed for anorectal malformations. We reviewed our experience with primary anterior sagittal anorectoplasties (ASARP) for anterior ectopic and vestibular anus in females.

Method: A retrospective review of all female patients who underwent a primary ASARP for vestibular or anterior ectopic anus between January 2003 and December 2007 was undertaken.

Results: 20 patients were included in the study. 14 patients (70%) had vestibular anus and 6 patients (30%) had anterior ectopic anus. Median age of presentation was 14 days (birth-9 months). In 14 patients (70%) the diagnosis had been missed at birth. 3 patients (15%) had associated abnormalities. Primary anterior sagittal anorectoplasty was performed at a median age of 4 months (6 weeks-15 months). 19 patients (95%) had preoperative bowel preparation. 1 patient had a low bowel washout. Prophylactic antibiotics were administered in 19 patients (95%). Patients were starved for a period of 4 to 5 days post-operatively. Total parenteral nutrition was administered in 9 patients (45%) post-operatively. 1 patient developed TPN line sepsis. 2 patients (10%) developed perineal sepsis that required a colostomy with 1 patient requiring a redo ASARP. These were subsequently closed after adequate wound healing. There were no long term sequelae. 2 patients (10%) developed superficial wound sepsis which resolved with local wound care. Their outcome was not compromised. Post-operative constipation occurred in 7 patients (35%) and was managed with laxatives. Median follow-up was 9 months (0-42 months). 13 patients (65%) had normal stool consistency. 3 patients (15%) were toilet trained.

Conclusion: Primary ASARPs are a viable option for females with vestibular or anterior ectopic anus. Total parenteral nutrition did not affect outcome. Local sepsis is a problem.

INTUSSUSCEPTION AND THE EFFECT OF RESECTION OF THE TERMINAL ILEUM ON VITAMIN B12 LEVELS, AT THE PRETORIA ACADEMIC HOSPITAL – A PRELIMINARY REPORT

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In the 5 year period 1999 - 2003 data were collected on all patients with the diagnosis of intussusception, as a retrospective study. In some patients vitamin B12 levels were done postoperatively but at the end of the study there were not enough data to come to any conclusion. Since 2004 vitamin B12 levels were done routinely on patients post ileal resection. This was done at regular intervals, namely 1, 4, 12 and 24 months post operatively.

Many patients who did well were lost to follow up and some data were lost in the hospital record system. In the first 5 years 68 patients were evaluated and vitamin B12 levels done on 26 of these patients. Hb and MCV were also done in most cases. The normal range for vitamin B12 in the local laboratory is 193 - 982 pg/ml. Values detected ranged from 87 - 1184 and in patients with a lower than normal value, MCV was still normal and patients not anaemic. In some cases vitamin B12 levels dropped but then raised again to normal. Follow up of 8 patients was longer than 2 years and in 2 patients longer than 3 years.

We still do not have enough data to come to a conclusion but it seems that the majority of children will adapt after ileo-caecal resection and will not develop a vitamin B12 deficiency.

INITIAL EXPERIENCE WITH A VIRTUAL REALITY TRAINER FOR MINIMALLY INVASIVE SURGERY

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Minimally invasive surgery requires a set of skills which are significantly different to those used in open surgery, and which ought to be developed prior to undertaking work on patients.

The East London Health Resource Centre has recently acquired a LAPSIM (Surgical Science) Virtual Reality Trainer (VRT) with a basic skills module for this purpose. Exercises which can be undertaken include a range of simple tasks such as camera and instrument navigation, grasping and co-ordination to more advanced skills such as dissection, clip applying, suturing and intra-corporeal knot tying.

The VRT is able to digitally record procedures and to evaluate each user's performance thus permitting an objective method of establishing when a user has reached a standard acceptable for certification.

The average time taken to reach the standard to permit certification has been 11 hours.

At the time of the preparation of this abstract, three surgeons have been certified as having achieved the required standard; ten are currently undergoing training while a further seven are waiting to be enrolled on the program. Prior surgical experience does not appear to provide an advantage in the speed of acquisition of skills on the VRT. Skills developed on the VRT have been shown to be transferable to the operating theatre.

LAPAROSCOPIC HAND-ASSISTED RLD RENAL HARVEST IN THE PORCINE MODEL

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Background: Laparoscopic retrieval has found widespread international application for the procurement of related living donor kidneys. While clinical outcomes are equivalent, donor satisfaction is purportedly better. Established laparoscopic units train junior staff using the human model, however in units where this experience does not exist, an alternative training method is required. Our model was designed to use human size pigs on which to mirror the human setup, with a view to:

1. Surgically achieve competence in a new procedure, in the absence of appropriately trained supervisors.
2. Familiarize ourselves with equipment, develop an operative sequence, and acquaint ourselves with nuances of the operation.
3. Train scrub nurses, emphasizing equipment, theatre and patient set-up, as well as operative sequence.

Methods: After study approval from the Research Review Board for Animal Ethics at the University of the Witwatersrand, twelve pigs were selected and appropriately investigated for anaesthetic contra-indications. Each pig was anaethetised and bilateral nephrectomies performed, using an identical operative approach to the human. Dissection time, warm ischaemic time, and all operative complications and injuries were documented. At the completion of each procedure the pig was euthanized.

Results: Animal weight varied between 31.3 and 55 kilograms. 22 nephrectomies were completed in 12 pigs. 2 were abandoned due to iatrogenic vascular injuries that necessitated conversion. Dissection time was a mean of 78.66 minutes (Range 32-108). Mean warm ischaemic time was 115.71 seconds (Range 62-221) and blood loss averaged 69.76 millilitres.

Conclusions: This was an excellent pilot and our end points were achieved, namely gaining competence performing a new procedure and attaining confidence using instrumentation and equipment specific to the operation. Our theatre staff were appropriately trained and as a unit we are comfortable moving forward to safely perform the operation on humans.

MANAGEMENT OF CANDIDIAL ESOPHAGEAL STRICTURES IN CHILDREN WITH AIDS

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Background: Causes of acquired esophageal strictures in children include reflux esophagitis, caustic ingestion, eosinophilic esophagitis, and chronic infections, causing mucosal injury and subsequent stenosis. Immunocompromised children are at particular risk for the infective

aetiologies often developing difficult-to-treat strictures. Among these children, repeated Candida species infection is the commonest agent. Despite the fact that a declining incidence of esophageal candidiasis in paediatric patients with HIV has been suggested, persistence in a subgroup of patients who do not respond immunologically and virologically to HAART has recently been highlighted. In addition, the poor role-out of anti-retroviral therapy in South Africa enhances the incidence of these opportunistic infections. For these reasons we still have to deal with candidiasis-induced esophageal strictures in pediatric AIDS at our institution. Diagnosis remains a clinical one. We report on a group of patients with AIDS, diagnosed with esophageal candidiasis. Each had been treated appropriately but subsequently developed oesophaeal strictures that required either dilation or replacement. This report, focuses on the role of the paediatric surgeon as part of a multidisciplinary team in the treatment of this under-reported clinical association, and highlights the fact that major thoracic and intra-abdominal surgery can be successfully performed with minimal morbidity despite the purported risk of major complications.

Methods: Retrospective chart review.

Results: Four patients were identified, all of whom were HIV positive, had been treated for oesophageal candidiasis, and had subsequently developed strictures. After appropriate counselling, initiation of anti-retroviral therapy, and systemic fungal therapy, the strictures were treated. 3 patients responded to oesophageal dilatation with or without gastrostomy, while the fourth ultimately required oesophageal replacement.

Conclusions: Children with HIV/AIDS are at particular risk of developing candida oesophageal strictures. Whilst the majority of these can be managed by conservative measures, a small subset may require segmental resection or oesophageal replacement, with the potential for good outcome.

HOW FAR ALONG THE ROAD IS GENETIC COUNSELLING FOR HIRSCHSPRUNG'S DISEASE?

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Hirschsprung's Disease (HSCR) represents a complex disorder of signalling molecules, with at least eight known susceptibility genes. Families carry 180 times higher risk, but genetic counselling via pedigree analysis may be affected by a small family size, poor history, adoption and no identifiable genetic mutation. Mutations of major susceptibility genes (RET /EDNRB) account for 50% (familial) and 30% (sporadic) cases. The significance of other genetic variations is unclear and most genetic counseling is based on incomplete information.

This study set out to evaluate a set of HSCR patients to identify factors of greatest value in genetic counselling.

Patients and methods: 114 HSCR patients(including 16 kindreds) were screened for genetic variations of the 2 major susceptibility genes (RET and EDNRB) and compared with. 60 control samples (20/ethnic groups). PCR and HEX: SSCP analysis was performed on microdissected tissue samples. SSCP variants were validated with automated sequencing techniques on PCR products

Results: Families with recurrence had an increased incidence, with longer aganglionic segments [especially total colonic aganglionosis (TCA)] Increasing gene penetrance was noted in 3 (female-male). Identified genetic variations included deletions, frameshifts and missense mutations as well as a number of significant SNP variations. Transmitted RET mutations occurred in 5 of 16 kindreds (30%). Splice RET mutation (IVS1) plus in variants of exon 17 (973L) both affected children with identical TCA. In a 3 generation family, variations in RET exon 6, 13, &18 (928) affected 3 male children with increasing penetration to recur as total intestinal aganglionosis.

Conclusions: Mendelian transmission appears mediated by the RET proto-oncogene. EDNRB mutations suggest haplotypic gene-gene interaction Genetic counselling remains a challenge in Hirschsprung's disease due to its multifactorial etiology.

BILIARY ATRESIA AT RED CROSS CHILDREN'S HOSPITAL: A 5-YEAR REVIEW

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Introduction: Biliary atresia represents the most common treatable cause of cholestasis in the neonatal period of life.

Aim: To review the age at presentation to hospital, course and outcome of children presenting or referred to Red Cross Hospital with biliary atresia.

Setting: A retrospective folder review of children coded for biliary atresia at the records department from January 2002 to December 2006 was done at Red Cross Hospital.

Results: Of the 57 patients identified with biliary atresia from the folders available at records department, 24 [42%] of these were seen primarily at Red Cross hospital, with 33 [58%] having been referred from other centers with biliary atresia for liver transplant assessment.There were a total of 20 males and 37 females [m:f ratio 1:1.85]

Age at presentation to Red Cross hospital ranged from 7weeks-7months [mean age 11.6 weeks].Despite the patients jaundice being noted quite early in life [mean age 17days,range of birth-3 months]they still presented much later for intervention to be instituted.

Portoenterostomy was done on 19 [79%] of the 24 at age of between 8-17 weeks [mean age 10 weeks] at Red Cross hsopital,with the other centres having had operated on 24 [72%] of the 33 children referred at 6 weeks-8 months age [mean age 12 weeks].

Of the 5 children at Red Cross Hospital who had no surgery, 3 [60%] died between 9-13 months of age due to liver failure while 2 [40%] are alive and on transplant list.

Conclusion: Early recognition of and referral to specialized centres is of utmost importance in the successful management of biliary atresia

PAEDIATRIC LIVER TRANSPLANTATION IN JOHANNESBURG: A REPORT OF THE FIRST FOUR YEARS OF A NEW SERVICE

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We present the results of paediatric liver transplants performed at the Donald Gordon Medical Centre in Johannesburg from August 2004 until end February 2008. During that time 63 liver transplants were performed in recipients of all ages. All organs were from deceased donors. Of these, 14 were in recipients under the age of 19 years and 7 of these were under 12 years. These patients form the basis of this review. The youngest recipient was an 11 month old 5,6 kg infant. In two recipients, transplants were performed for fulminant hepatic failure. 8 grafts were performed with size matched whole organs and 6 were technical variant grafts including 3 recipients of split livers and 3 of reduced size grafts. One patient received a simultaneous combined liver kidney transplant for congenital hepatic fibrosis related to polycystic kidney disease. Other indications for transplantation included: 5 biliary atresia; 1 hepatocellular carcinoma; 2 metabolic liver conditions; 1 autoimmune hepatitis; 1 cirrhosis with cystic fibrosis; 1 Allagilles syndrome; 1 Budd Chiari syndrome and one patient with ductopaenic rejection following a successful transplant at another unit. There were 9 females and 5 males. 13 (93%) recipients are alive at a mean follow up of 292 days post transplant. One patient (retransplanted for rejection) died due to fulminant sepsis and haemodynamic instability 6 days after a technically successful graft. Immunosuppression used was a standard Tacrolimus and steroid based dual therapy protocol. Three patients were given daclizumab as pre transplant induction therapy. Surgical complications include: 2 anastomotic biliary strictures, 2 bile leaks (1 from raw surface), 1 early postoperative acute rejection, 1 recurrence of hepatoma in the incision. These results confirm the successful evolution of the infrastructure and skills required to provide a comprehensive paediatric service in the new unit.

SOCIAL AND INSTITUTIONAL FACTORS IMPACTING ON THE MANAGEMENT OF PAEDIATRIC SURGERY PATIENTS IN AN ACADEMIC PUBLIC HOSPITAL IN SOUTH AFRICA

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Paediatric surgeons do not treat patients in a vacuum. Gauteng's state hospitals are increasingly under resourced and overburdened. The impact of socio-economic factors and insufficient institutional resources on the quality of our patients' care has never been systematically assessed. Unscheduled admissions to the paediatric surgical unit at Chris Hani Baragwanath hospital were analysed prospectively from 1 November 2006 - 28 February 2007. 96 patients with a disease severity deemed to be of equal or greater severity than an uncomplicated acute appendicitis were included. There were 46 males and 50 females; 31 neonates; 19 infants. The mean age was 2.79 years. The group was analysed for housing status, income, level of education of caregivers, congruency of belief systems amongst other parameters. Reasons for delays in effective surgical treatment were investigated. The mean duration of delay in making a diagnosis and transfer was 4,6 days for neonates. 52% of neonates

experienced a delay in surgical treatment due to unavailability of an ICU bed (average waiting period was 4,9days). 12 neonates were born to HIV positive mothers. None had been given treatment to prevent mother to child transmission. Adverse growth indices and physiological parameters such as acid base status were also prevalent in this group. 10 neonates (32%) died, all from nosocomial sepsis. Mortality for older children was 7% (5/65). Conclusion: In the largest hospital in the world, serving an extremely indigent community, surgical neonates are bearing the greatest burden of morbidity and mortality. Delays in diagnosis, referral and accommodation in neonatal ICU beds as well as the poor physiological condition of patients and high rates of nosocomial sepsis were the most significant factors. Innovative solutions to the unavailability of neonatal beds have been necessary. The capacity of the neonatal intensive care unit needs to be improved as a matter of urgency.

LAPAROSCOPIC ASSISTED ANO-RECTAL PULLTHROUGH (LAARP): LESSONS LEARNT FROM THE FIRST 10 CASES AT TYGERBERG CHILDREN'S HOSPITAL

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The introduction of LAARP has since its first description in 2000 raised new surgical options in the management of high ano-rectal malformations (ARM).

We present the first 10 cases of LAARP treated at the Tygerberg Children's Hospital and reflect on lessons learned. The male to female ration was 9:1 and 8 males had a high ARM with a recto-bulbar urethral fistula and one a recto-prostatic fistula. The female patient had a high blind ending rectum without a fistula. Six of the 10 patients had associated abnormalities. One patient presented with symptomatic midgut malrotation and required a Ladd's procedure, performed laparascopically at the same time.

Setting up the procedure was time consuming, but the average surgical duration was shorter than a PSARP, less than two hours.

Six early complications occurred in 4 patients. These included ongoing oozing of blood from releasing adhesions, an ischaemic sigmoid colon (requiring a redo laparascopic pull-through), one early adhesive bowel obstruction requiring laparatomy and inadvertant intra-operative injury of the vas deferens. The cause of the necrotic sigmoid was attributed to a division of the vascular arcade at the time of colostomy.

Other lesser problems included a small residual uretheric diverticulum (that hasn't caused any problems), a mild mucosal prolapse and an early tension-retraction requiring redo ano-plasty.

All patients recovered quickly, feed early and required very little pain management. Most suffer from constipation and some leakage, but toilet training has not yet been completed in the majority.

LAARP is a feasible procedure in an African setting. It is of paramount importance that the original sigmoid colostomy is sited at the right level without compromising length and vascularity. Improvement of long-term follow up is essential in order to improve functional outcome.

NEONATAL CIRCUMCISION AND HIV PREVENTION: IS THERE A CASE TO REDUCE HIV INCIDENCE?

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Non-therapeutic circumcision is the most commonly published surgical procedure that is still lacking a real significant indication in many cases. Neonatal circumcision has recently been advocated as the main stem of an UNAIDS/WHO policy for the prevention of HIV/AIDS and is being advocated for widespread use.

Although it is agreed that the HIV/AIDS crisis demands extraordinary means to curtail it, it is nevertheless questionable how circumcision and particularly how neonatal circumcision, could achieve such goal. There is no rational theory explaining why circumcision ought to reduce HIV acquisition. Further research is needed. Current evidence is that male circumcision confers no benefit to female partners of HIV-positive men and even increases their risk. It furthermore ignores and discourages the ABC method and correct condom use, which is the most effective means to date of reducing HIV transmission.

Practicing widespread genital surgery on non-consenting infants and children raises legal and human rights issues. It is also an inappropriate public health policy and arguably unethical to expend scarce resources on futile surgery in countries with deficient public health infrastructures and lack of basics like adequate food and clean water.

NUTRITIONAL IMPROVEMENT FOLLOWING THE LAPAROSCOPIC INSERTION OF A BUTTON COLOSTOMY AS A VENTING STOMA AND ACCESS PORT FOR THE ADMINISTRATION OF ANTEGRADE ENEMAS IN AFRICAN DEGENERATIVE LEIOMYOPATHY

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Background: African Degenerative Leiomyopathy, an uncommon condition primarily affecting the colon and rectum of children in Southern, Eastern and Central Africa, manifests as gross gaseous abdominal distension, an inability to evacuate flatus and faeces, anorexia and malnutrition with death in adolescence or early adult life. Surgical intervention has previously been recommended only in the management of complications such as volvulus. We have placed a MIC-KEY skin level device (button) laparoscopically as a colostomy in four children with this disease. A size 8 feeding tube passed through the button is used each day to deflate the colon and to instil antegrade enemas to evacuate stool.

Patients and methods: Qualitative nutritional scores (appetite, food intake, pain, nausea, vomiting and diarrhoea) and quantitative measurements (triceps and sub-scapular skin fold thickness and mid-arm circumference) determined at the time of surgery have been compared to the status nine months after the insertion of the button.

Results: All patients have demonstrated an improvement in both qualitative and quantitative scores. Qualitative scores improved from a mean of 11/25 to a mean 18/25. Increases in mid-arm circumference and triceps sub-scapular skin fold thickness were demonstrated.

Conclusion: Nutritional improvement has been demonstrated by both qualitative and quantitative means following the laparoscopic insertion of a button colostomy as a venting stoma and access port for the administration of antegrade enemas in patients with African Degenerative Leiomyopathy. The quality of life has improved remarkably with this intervention.

OBSERVATIONS OF THE ANATOMICAL PATHOLOGY OF CHILDHOOD HYDROCOELES AT LAPARASCOPY AND THEIR IMPLICATION FOR MANAGEMENT

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With the experience gained in the laparoscopic correction of childhood inguinal hernias, we have begun to manage the patent processus vaginalis (PPV) of childhood hydrocoeles in the same manner. In this procedure, the processus is separated from the peritoneum by hook diathermy and the peritoneal defect thus created is closed with a single intracorporeal absorbable suture. When present, a non-communicating hydrocoele (NCH) is then simply aspirated.

In doing these procedures, we have become aware of differences between our findings at laparoscopy and the descriptions of the anatomical pathology of childhood hydrocoeles as found in the standard paediatric surgical texts.

Our observations of their pathological anatomy include the following:

1. All children with hydrocoeles whether communicating hydrocoeles (CH) or non-communicating hydrocoeles (NCH) whom we have examined have a patent processus vaginalis.
2. The PPV in CH extends down to the testis while in NCH the PPV is of variable length and extends some or all of the way down the inguinal canal to the outer wall of the tunica vaginalis of the closed hydrocoele.
3. The diameter of the PPV varies but in at least one quarter of patients is greater than 10 mm.
4. In some patients thought clinically to have a NCH a pinhole opening between the hydrocoele and the PPV was found.
5. There appears to be a higher than expected occurrence of early abdominoscrotal hydrocoeles.

These observations raise various questions:

1. Is aspiration of laparoscopically confirmed NCH an acceptable form of treatment?
2. Are these PPVs of a variable calibre seen in all patients with congenital hydrocoeles forerunners of indirect inguinal hernias in older children, adolescents and adults?
3. If so, is conservative treatment of infant hydrocoeles appropriate?

THORACOSCOPIC REPAIR OF A PLEURO-PERITONEAL COMMUNICATION IN A CHILD ON PERITONEAL DIALYSIS

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Acute massive hydrothorax is a well-recognized complication of peritoneal dialysis. Reported incidence is 1 - 2% of patients on peritoneal dialysis. Congenital communications between the pleura and peritoneum have been hypothesized to be the source of this hydrothorax. Surgical correction should be considered for an identified defect on long-term dialysis patients.

We describe a six-year-old child on peritoneal dialysis that presented with a massive right hydrothorax. A Tenckhoff catheter was placed laparoscopically two weeks prior to the development of this hydrothorax. The defect was successfully corrected with video-assisted thoracoscopic surgery. Creating a pneumoperitoneum through the Tenckhoff catheter enabled us to identify the defect during surgery.

Peritoneal dialysis was resumed two weeks after this laparoscopic repair. The thoracoscopic method is a save and effective method to close this defect. It avoids a thoracotomy and vision is superior to that of the open procedure. To our knowledge, this is the first report of a video-assisted thoracoscopic repair of this defect in a child.

LESSONS LEARNED FROM THE FIRST 500 LAPAROSCOPIC NISSEN FUNDOPLICATIONS IN INFANTS AND CHILDREN

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Gastro-esophageal reflux disease is a common problem in infants and children. It is a complex problem affecting the health and development of these patients.

Esophageal gastric fundoplication is one of the most common procedures performed by Paediatric Surgeons today. This is a report of the technical lessons we learned from the first 500 laparoscopic Nissen Fundoplications we performed.

The major steps in performing these operations are:

- Mobilization of the intra-abdominal esophagus
- Short gastric mobilization where necessary
- The creation of a 360° short floppy wrap
- Crural repair at all times.

We believe that the technical advantages offered by the laparoscopic approach have contributed to a low recurrence and high success rate. The exposure of the hiatus and gastric anatomy is superior with the laparoscopic method and allows for the correct anatomic formation of the wrap.

VESICO-EJACULATORY DUCT REFLUX (VER): A REVIEW OF THE LITERATURE

Rinus Wiersma

Vesico-ejaculatory duct reflux (VER) is the reflux of urine into the ejaculatory ducts of the prostate, seminal vesicles and the vas deference. Although the relationship between VER and epididymitis is well established in sexually active men, this is not so in young children [1]. VER is not common in children, but is found among the group of boys who present with uro-genital anomalies, and infections of the urinary tract or ejaculatory duct system. The long term effects of VER and the resulting sepsis of the ejaculatory duct system, are post-micturition dribbling, abscess formation, testicular infarction, chronic pain and infertility [2]. The aim of this paper is to present our experience of VER, and review the literature on this condition and its management.

Seven patients with VER have locally been identified in the last 5 years (2003-2007). Their clinical features are presented.

Urine is normally prevented from refluxing into the ejaculatory system by the small duct size and the angle of entry into the urethra. Dilatation of the ejaculatory ducts, congenital anomalies or urethral obstruction of the urethra are thought to give rise to VER [3]. Although the literature on VER is dated, it is still relevant today, and divides the underlying conditions into 4 main groups [3]

1. Anorectal malformation
2. Hypospadias
3. Prune belly syndrome
4. No anomalies.

A management protocol is presented for children who present with either an acute scrotum or UTI. Once VER is diagnosed the options for further management are included.

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South African Children's Cancer Study Group (SACCSG)

BURKITT LYMPHOMA TREATED AT A SINGLE CENTRE WITH LMB-BASED CHEMOTHERAPY

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Objective: To audit the results of an LMB-based protocol used to treat Burkitt Lymphoma at the Red Cross Children's Hospital.

Methods: The study was a retrospective analysis of all HIV-negative patients with biopsy-proven Burkitt Lymphoma treated with LMB-based chemotherapy between July 1988 and June 2007.

Results: Seventy-four patients were diagnosed during the study period, ranging in age from 1.16 to 13.95 years [median 5.83]. 80% had abdominal or pelvic masses, 11% had jaw masses, 17% had CNS disease, and 31% had leukaemia. Three patients were Group A, 42 were Group B (57%) and 29 were Group C (39%). Overall estimated 5-year survival for the whole group was 80.4%; 100% for Group A, 85.5 % for Group B and 70.4% for Group C. Ten patients failed to achieve remission. Six (four of them Group C) had refractory disease and four patients suffered toxic deaths during induction; two from infection (Klebsiella septicaemia and mucormycosis) and two due to haemorrhage. Among the 64 patients who achieved remission, four relapsed and one of these, a Group B patient with a late ovarian relapse, was retreated and survived. The others were Group C patients with extensive early relapses who were palliated. There was more morbidity among Group C patients compared to Group B patients. There were more neutropaenic fevers per patient (3.1 vs 2.2), more positive blood cultures (1.3 vs 0.7) and more episodes of Grade III or IV mucositis (2.2 vs 1.7). Group C patients also required more red cell (5.9 vs 3) and platelet transfusions (6.4 vs 2.3) per patient.

Conclusions: LMB-based chemotherapy is effective therapy for Burkitt lymphoma, but is associated with considerable toxicity that necessitates access to modern supportive care. Analysis of our Group C results suggests that attention to treatment intensity is as important as avoidance of toxicity in improving the results for these patients.

PAEDIATRIC NON-HODGKIN LYMPHOMA PRESENTING WITH BONE INVOLVEMENT

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Introduction: Bone involvement in non-Hodgkin lymphoma is a rare, though well known clinico-pathological entity. Bone involvement may occur primarily or by direct extension from soft tissue masses. Clinicians may experience difficulty distinguishing between primary NHL of bone, and bone involvement by direct extension of a soft tissue mass.

Methods: A retrospective review was performed of records of patients at the Johannesburg Hospital diagnosed with non-Hodgkin lymphoma to determine incidence of bone involvement, modes of presentation, diagnosis and management of these cases.

Results: In total, 111 patients were diagnosed with NHL from 1 January 1996 to 31 December 2007. Of these, 14 (12.6 %) had bone involvement, diagnosed on the basis of clinical, radiological and/or histological grounds. Of this group, 3 (2.7 % of the total) had primary bone involvement, and 11 (9.9 %) had bone involvement by direct extension. The three patients with primary bone involvement were HIV negative and the subtypes were pre B NHL, B cell NHL and Burkitt lymphoma. 2 of the group with bone involvement by direct extension were HIV positive, 9 were HIV

negative, and the HIV status of the remaining 2 patients was unknown. The subtypes in this group included B cell NHL (2) Burkitt lymphoma (3), large cell anaplastic lymphoma (6) and plasmablastic lymphoma (1). The most common modes of presentation were with bony masses, and/or neurological deficits as a result of vertebral invasion. All patients were treated according to standard chemotherapy protocols.

Conclusion: 2.7% of the patients diagnosed with non-Hodgkin lymphoma presented with primary bone involvement, while 9.9% had bony lesions by direct invasion at first presentation. This correlates with incidences published in other series.

G-CSF DOES NOT CONFER A SURVIVAL ADVANTAGE IN PATIENTS WITH BURKITT LYMPHOMA (GROUP B) TREATED ON A LMB-BASED PROTOCOL AT RED CROSS CHILDREN'S HOSPITAL

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Objective: To assess the efficacy of G-CSF in an LMB-based protocol used to treat Burkitt Lymphoma at Red Cross Children's Hospital.

Methods: The study was a retrospective audit of G-CSF usage in a cohort of Group B Burkitt lymphoma patients between 1998 and 2007. The LMB protocol was adopted in 1998 and G-CSF began to be used at a dose of 5mcg/kg daily for 14 days after COPADM 1 and 2 in 1995.

We compared two cohorts of Group B Burkitt lymphoma patients; those prior to 1995 who did not receive G-CSF and those after 1995 who were given G-CSF.

Results: Seventy six patients were diagnosed with Burkitt lymphoma during the study period, 3 with Groups A disease, 44 with Group B disease and 29 with Group C disease. Two of the Group B patients died during induction and one was changed to COMP due to toxicity. These were excluded. Forty one patients were eligible for analysis; 13 did not receive G-CSF and 28 did.

Comparing the two groups, there was no significant difference in the mean number of days of neutropaenia and the mean number of days of delay before starting subsequent chemotherapy. Similarly, there were no significant differences between the groups with respect to percentage of infections and the mean number of blood and platelet transfusions. Mucositis was almost twice as prevalent in the group of patients who received G-CSF compared to the group who did not (58.62% vs. 31.25%).

The overall survival (OS) of the group not receiving G-CSF was 84.6% compared to 96.3% in the group who did. This was not statistically significant ($p = 0.2$). OS for the entire group was 85.5%.

Conclusions: In a non-randomised retrospective analysis, patients who received G-CSF following COPADM 1 and 2 did no better in terms of survival, than those who did not receive G-CSF.

ADOLESCENTS TREATED IN A SINGLE SOUTH AFRICAN PAEDIATRIC ONCOLOGY UNIT BETWEEN 1997 AND 2007

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Introduction: Before 1997 only children up to the age of 12 years were treated in the Kalafong POU. There is a need to establish who and where adolescents should be treated.

Aim: The aim of this audit was to determine the number of adolescents who received cancer treatment in the Kalafong paediatric oncology unit, their cancer diagnoses and their outcome.

Methods: Data was collected retrospectively from the South African Childhood Cancer registry forms, annual reports and individual patient files for the period 1997-2007. The data analysis included demographic data (age, sex, home province) diagnosis, stage of disease and outcome.

Results: There were 45 adolescents treated in the paediatric oncology unit, which was 5% of the total newly diagnosed patients, but 14% in the last year of newly diagnosed patients per year. The median age was 14.25 years (range: 13 years to 17 years); and the male:female ratio was 3.5:1. The diagnoses were as: Acute lymphoblastic leukaemia n=13; Hodgkin lymphoma n=5; acute myeloid leukaemia n=5; osteosarcoma n=5, T cell lymphomas n=3; B cell lymphomas n=3; chronic myeloid leukaemia n=4; various other diagnoses n=7. Forty four percent had advanced stage or high risk disease with a poor outcome.

Conclusion: Five percent of newly diagnosed children with cancer were adolescents, who were treated in a paediatric oncology unit, which has steadily increased in the last 5 years. There is currently no dedicated ward facility, where teenagers can be cared for with their peer group and all were treated with younger children in the paediatric ward. This discussion will address the need for dedicated space to allow adolescents to be cared for with peer group.

OUTCOMES OF CHILDREN WITH MALIGNANCY WHO REQUIRE INTENSIVE CARE IN SOUTH AFRICA: A RETROSPECTIVE STUDY

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Despite dramatic improvement in outcomes for children with cancer the intensive and sustained use of chemotherapy presents a great challenge for doctors who are faced with managing children suffering from severe myelo and immune-suppression developing intractable sepsis. In the west, there is a growing acceptance of an improvement of survival for children with malignancy who require admission to paediatric intensive care (51% mortality reduced to 34%) There are, however no published data on children from Africa with malignancy who require intensive care.

Methods: Our aim was to establish the outcomes of children treated in our paediatric intensive care unit with an underlying diagnosis of malignancy. Retrospective note review followed by prospective database collation. Results: From Jan 2007-2008 we identified 17 children. 58% were older than 6 years. There were an equal spread of solid tumours, lymphomas and leukaemias (30% each) and 12% CNS tumours. 29% presented after relapse but the majority were within the first month from diagnosis (41%). The commonest reason requiring admission was septic shock (40%), followed by post-operative cases (24%), tumour mass effect (12%) and neurological (12%). There was a 100% mortality with leukaemia either during their admission (80%) or in the months since (20%). 60% mortality in the solid tumours group, 40% for the lymphomas with a further child dying since admission. There was a 100% cumulative mortality for children in respiratory failure and tumour mass effects. There was 70% mortality for children in septic shock and a 75% survival post-operatively. Overall mortality was 53% with cumulative mortality of 71%.

Conclusion: There is a high mortality rate in children with malignancy presenting to intensive care. The results of our ongoing study will enable us to build a picture of the burden of oncology disease in the intensive care unit and modify strategies for improving survival of such patients.

AUDIT OF CHILDREN PRESENTING WITH ACUTE LEUKAEMIA AT GEORGE MUKHARI HOSPITAL BETWEEN 2000 AND 2007

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Aim: To review the outcome of children presenting to Dr George Mukhari paediatric oncology unit with acute leukaemia.

Method: A retrospective analysis was done of patient records of children diagnosed with acute leukaemia at Dr George Mukhari Hospital between January 2000 and December 2007.

Results: There were 101 patients diagnosed with leukaemia. The majority (60.3%) had acute lymphoblastic leukaemia (ALL) versus 39.6% with acute myeloid leukaemia (AML), with an ALL:AML ratio of 1.5:1. The overall male:female ratio was 1.2:1, while it was 1.8:1 for ALL and 1:1.4 for AML. The overall median age was 8.35 years with a range of 1.17 years to 13.67 years. The median ages at diagnosis for ALL and AML were similar, namely 8.30 years and 8.40 years respectively. The patients were treated with a Modified BFM protocol for ALL. For AML, the ANLL modified Denver protocol was used until mid-2002, followed by the BFM-98-modified protocol. The overall mortality rate was 56.4% with a higher mortality rate for AML (61.5%) than ALL (53.2%). However 38.7% of survivors have subsequently been lost to follow-up.

Conclusion: There were more AML patients than expected, which can be explained by the higher incidence of AML in African patients, as in this study population. The major problems in this unit were poor outcome for both ALL and AML patients, and loss to follow-up. There was not a dedicated paediatric oncologist since 2001. The plan is to re-establish a formal paediatric oncology unit, which will be twinned with the unit at Kalafong Hospital for support. The treatment protocols are currently being reviewed and updated.

ACUTE MYELOID LEUKAEMIA (AML) – THE KALAFONG EXPERIENCE

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Aim: The aim was to audit the patient files of all patients diagnosed with AML from 1993 to 2007 at Kalafong Hospital.

Methods: All patients diagnosed with AML were included in the audit. Data was collected retrospectively from patient records. Data analysis included age, sex, type of AML and outcome.

Results: There were 64 patients diagnosed with AML. The mean age was 6 years 9 months (range: 1 month – 15 years 10 months) and the male: female ratio was 1:1. The FAB classifications were as follows: M0 – 8%; M1- 16%; M2 - 14%; M3 – 22%; M4 – 16%; M5 – 14%; M6 – 3%; and M7 – 3%, unknown – 5%. Overall survival was 25% with a mortality of 64%, while 11% were lost to follow-up. Sepsis was the major cause of death (34%), followed by relapse (19%), and disease progression (6%). Another 6% died within 24 hours of admission and before treatment was initiated. Disseminated intravascular clotting was the cause of death in 6% and 2% died at home of unknown reasons. Patients with acute promyelocytic leukaemia (APL; M3) had the best outcome (70% overall survival), while patients with AML (M6, M7 and M0) had the worst outcome (100% mortality).

Conclusion: The major modifiable reasons identified for poor outcome were the high incidence of sepsis and patients lost to follow-up. Patients with APL had a good outcome, which is well comparable to outcome reported in the literature. Supportive care of AML patients will receive more attention in the future.

HIV ASSOCIATED NON-HODGKIN'S LYMPHOMAS: WHAT IS THE MOST OPTIMAL THERAPY IN A RESOURCE LIMITED SETTING?

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Introduction: Lymphomas are a late manifestation of HIV. There is little literature guidance as to the most optimal therapy in a resource limited setting.

Method: All Non-Hodgkin's lymphomas were reviewed at the Johannesburg Hospital from January 1997 to December 2007. With HAART available after 2006 HIV positive patients were treated with chemotherapy. These patients were reviewed in detail.

Results: 108 patients with NHL were diagnosed. Of these 55 (51%) were diagnosed with Burkitt's/Diffuse Large B Cell NHL. 17 of the 55 (31%) were HIV positive. 11/15 HIV positive patients with NHL were treated at CH-Baragwanath Hospital with the Berlin B cell protocol with 5g/m^2 of Methotrexate. This resulted in significant morbidity with increased supportive care needed and mortality. Patients at the Johannesburg Hospital were treated with only 500mg/m^2 of Methotrexate. 8 cases were seen. Mean age at diagnosis was 7,7 years and male to female ratio was 7 to 1. There were 5 Burkitt's Lymphoma, 2 Diffuse Large B Cell NHL and 1 Plasmablastic NHL. The sites of the tumour were: 3 maxilla, 1 axilla, 3 cervical and 1 spine. 1 patient had been on HAART for 1 year prior to diagnosis and the rest were diagnosed on admission. Mean CD4 count was 20,4% and mean viral load 66668 copies/ml. HAART was started approximately 1 month from diagnosis after stabilization with chemotherapy. 1 patient had palliative radiation only. 7 patients were treated and tolerated the chemotherapy with similar morbidity to the non-HIV positive patients. All had complete response. 1 died of fulminant Hepatitis A.

Conclusion: Treatment of HIV positive patients with B cell/Burkitt's NHL with a protocol with a lower dose Methotrexate results in good response with acceptable morbidity. HAART may be started as soon as the patient has stabilized without prejudice to outcome.

HOW MUCH DOES IT COST TO TREAT HODGKIN LYMPHOMA IN AFRICA?

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Aim: To assess the cost of treatment of Hodgkin disease in a series of 138 children treated at two hospitals in South Africa.

Method: The records of the children with Hodgkin disease treated from 1986 to 2007 were analyzed retrospectively with regard to age, sex, stage, method of treatment, protocol, survival according to protocol, complications and relapses. A price list for chemotherapy was obtained from the hospital's pharmacy and for investigations from NHLS. The duration of hospitalization was not taken in consideration.

Results: The mean age at diagnosis was 8,9 years. There were 76% males and 24% females. Stage 2 was the most common stage at diagnosis and there was no difference in the incidence of A and B symptoms. The protocols used were CLVPP 11%, ABVD (alone and in combination) 50%, MOPP/COPP 21%, OEPA/OPPA 14% and unknown 6%.The survival rate for stage 1 and 2 treated with ABVD was 100%.

The cost of the protocols varied between R984.80 for OEPA (stage 1 and 2) and R3985.92 for ABVD (4 cycles, stage 1 and 2) for first line treatment. The most expensive protocol was the third line salvage, CEP, at R18 213.42 and it was more expensive to treat girls than boys.

Conclusion: Early diagnosis reduces the cost dramatically whilst contributing to the best results. We suggest that a more aggressive diagnostic approach to persistent lymphadenopathy might result in earlier detection of a large proportion of Hodgkin cases.

CORRELATION BETWEEN ETHNICITY AND PROGNOSIS IN HODGKIN DISEASE IN TWO SOUTH AFRICAN HOSPITALS

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Aim: To review a series of 138 children with Hodgkin lymphoma treated at two centres in South Africa, from 1986 to 2007, with the aim to analyze and compare the clinical presentation, age, sex, stage at diagnosis and survival in different ethnic groups.

Method: The records of children with Hodgkin disease treated during the study period were analyzed retrospectively.

Results: White children presented on average 30 months later than black children (133 months versus 103 months). Girls presented later than boys (127 months versus 107 months). Of the total patients 56% were black, 28% coloured while only 16% were white. Black patients represented 56%, coloured children 28% and whites 16% of the total patients. White children presented with earlier stages of disease compared to black patients (p=0,02). Black children had predominantly a mixed cellularity histology (50%) compared to whites who presented predominantly with nodular sclerosing subtype (50%)with nodular sclerosis (50%). The coloured children had an almost equal number of mixed cellularity end nodular sclerosing subtypes group had almost equal mixed cellularity (42%) and nodular sclerosis (45%).

Survival according to stage was the highest in the coloured group for stage 1 and 2 (96%) and lowest for the whites (82%) and highest for the black group (72%) in stage 3 and 4 and still lowest for the whites (20%) Survival for stage 1 and 2 was highest in the coloured group (96%) and lowest in the white patients (82%). In stage 3 and 4 the black patients had the best survival (72%) while the white patients only had a survival rate of 20%

Conclusion: Hodgkin disease remains a major disease within the spectrum of childhood cancers. There are prognostic factors associated with ethnic groups. Larger studies could bring valuable information in order to modify the management for best results.

TUBERCULOSIS IN CHILDREN WITH CANCER: IS SCREENING NECESSARY?

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There is a dearth of studies addressing the incidence and clinical presentation of tuberculosis in children with cancer. This study evaluates the incidence of TB in paediatric oncology patients at Tygerberg Hospital, located in a Cape Town area with high TB prevalence, and describes the clinical characteristics of the disease in this particular group of patients, whose treatment typically suppresses their immune response.

We reviewed the records of 625 paediatric oncology patients admitted from 01.01.1991 to 31.12.2005. Out of these, 87 received treatment for TB; however, only 57 cases had sufficient data to support a diagnosis of TB and only these were used for further analysis. In this group, acute lymphatic leukaemia (ALL) was the most common malignancy (13/57; 22.8%). The incidence of TB in the study group was 9117/100 000/year, which is 22 times higher than the overall TB incidence reported in children from a similar background. Importantly, 47% of the active infections appeared in the first 5 months of chemotherapy, suggesting reactivation of latent TB.

Identifying latent TB in our patients and providing prophylactic treatment during the first months of chemotherapy may have prevented disease progression in these cases. In order to verify this assumption, a prospective study was initiated in July 2007, consisting of routine screening of paediatric oncology patients for latent TB infection, by means of tuberculosis skin test, in conjunction with Quantiferon and Elispot tests, as well as screening for active disease, prior to the initiation of cancer therapy. Study participants found to have latent tuberculosis were treated

preventively, simultaneously with the management of their malignancies. The preliminary results of this study will also be presented here.

A SURVEY OF THE MANAGEMENT OF IDIOPATHIC THROMBOCYTOPENIC PURPURA IN CHILDREN IN SOUTH AFRICA: DO WE NEED GUIDELINES?

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Aim: To describe the management of idiopathic thrombocytopenic purpura (ITP) in children in South Africa.

Methods: A prospective survey was sent to 410 doctors in the country, including 4 different scenarios in children newly diagnosed with ITP, concerning the diagnosis and management.

Results: Steroids were the first line of choice in treating ITP. Anti-D immunoglobulin was not considered in the management and most of the practitioners would perform a bone marrow aspiration even if no treatment with steroids will be given. The vast majority of the patients will be treated in the hospital.

Conclusions: This study is the first study done in South Africa (and in Africa) and it shows a great variation in the management practice of children with newly diagnosed ITP. Prospective studies in developing countries where various constraints to health care delivery exist are required to produce evidence based recommendations for this patient group.

WHAT COLOUR IS YOUR CANCER?

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A 7 year old child was referred to Tygerberg Hospital with a short history of dizziness, loss of appetite and tiredness. A thrombocytopenia, anaemia and leuco-erythroblastic reaction with ±90% blasts were detected on blood examination. The rest of the clinical examination was normal. Chest radiograph showed a large (wide) mediastinum. CT scan chest report showed the presence of the thymus. A blood diluted bone marrow aspirate (no particles) showed 90% blasts, varying in size and amounts of cytoplasm. Some had fine eosinophilic granules but no Auer rods.

Immunophenotypic analysis:

An atypical immunophenotype with more than 90% blasts expressing CD117+ /CD33+ /CD19+ (dim) /CD34+ (bright) /CD22+ (dim) /CD7+.

The cCD3, cCD22, cCD79a, MPO, TdT, CD1a and cμ were negative.

In addition there was partial expression of cCD13, CD13, HLA-DR and CD11b (dim) (35-60% of blasts). CD4 and CD8 represented a small group of mature T-lymphocytes. A panel of predominantly cytoplasmic markers was repeated at a neighbouring academic centre with similar findings. Overall there were insufficient points to classify this as a biphenotypic leukaemia when using the EGIL classification. Cytogenetic studies produced no metaphases due to low cell counts. FISH analysis was negative for t(8;21) and 11q23.

The initial diagnosis was ALL (most likely precursor B) or possibly a biphenotypic leukaemia.

The patient was started on ALL BFM protocol SR. With the flow cytometry findings suggestive of AML the protocol was changed to AML BFM 2004 and the child received the 8 days induction. He became severely neutropenic and febrile for more than 3 weeks.

Is it ALL or AML? How would you treat?

HIV AND LYMPHOMA: THE EXPERIENCE OF THE PAEDIATRIC ONCOLOGY UNIT AT THE BLOEMFONTEIN ACADEMIC HOSPITAL COMPLEX

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A retrospective analysis of all patients with a diagnosis of HIV and lymphoma was undertaken to determine the patient profile, types of lymphoma, the outcome and survival rates. A total of 14 patients were seen over a 7-year period with 40% being seen in the last two years. There were more males than female and the ages ranged from 3 to 10 years. Nine patients had B cell lymphomas, the majority Burkitt lymphoma, 3 had nodular sclerosing Hodgkin Disease (HD) while 2 had T cell lymphoma. Those with HD had nodal disease while both those with T cell lymphoma appeared to be extranodal. The disease in those with Burkitt lymphoma appeared to be nodal and was stage 3 or 4 with three patients presenting with paraplegia. Of the Non-Burkitt lymphoma two were nodal and two extranodal. Chemotherapy was offered to all except one patient. The protocol most often used was a modified LMB 89. Of the 7 patients still alive 5 are off treatment for periods ranging from 1 month to 47 months while those who died survived for periods ranging from 12 days to 36 months. Only three patients died of tumour progression.

It appears from this limited study that it is possible to treat children who are HIV positive with lymphoma with chemotherapy without major mortality associated with the chemotherapy.

PROFILE AND OUTCOME OF PAEDIATRIC PATIENTS WITH BURKITT LYMPHOMA SEEN IN BLOEMFONTEIN DURING THE LAST 20 YEARS

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The aim of this retrospective study was to document the profile and outcome of patients with Burkitt lymphoma (BL) and compare our results to the literature. All patients were seen between January 1986 and December 2006 by the Paediatric Oncology division.

Out of a total of 123 patients with Non-Hodgkin lymphoma 52 were diagnosed with BL. There were more than twice as many males as females and the average age was 70 months with a range from 20 to 171 months. The most common primary site was the abdomen and more than 90% had advanced stage disease. Bone marrow was the most common site of spread. A variety of protocols were used but most patients received the LMB 89 protocol. The survival rate of all patients was just less than 70% with CHOP protocol giving the best survival rate of 80%. Males did better than females but the difference was not statistically significant. Fifty percent of the deaths occurred within one month of presentation with sepsis and metabolic complications as the most common causes while relapse accounted for almost 60% of the late deaths.

In conclusion, our incidence, age range, sex ratio and primary site is similar to the literature but we appear to have more stage 3 and 4 disease than is reported. Our survival rate with the LMB protocol is less than expected. We have too many early deaths and need to aggressively treat these very ill patients for sepsis and metabolic derangements.

HODGKIN'S DISEASE IN KWAZULU-NATAL

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Nodular sclerosing Hodgkin's disease (HD) is rare under 5 years of age. There is a male predominance below 10 years of age. In developing countries there is an early peak in HD incidence. Advanced disease and poorer histological types have been reported from many centres.

Aim: To document the pattern of HD in our centre.

Method: Chart review of children with HD under the age of 12 years. HIV infected children with HD were included.

Results: 146 patient charts were evaluated. There were 121 African and 25 Indian patients. HIV infection was documented in 4 African patients. Overall M:F ratio was 6.3:1 with the M:F distribution for African patients being 105:16 and for Indian patients 21:4. Median ages were African 7 yrs (3 to 12 yrs) and Indian 5 yrs (2 yr 3 mo to 11 yrs). There were 24 patients (8 Indian) less than 5 years old.

Histological types and stages were (HIV patients in brackets):

Type	Unspec. HD	LD	LP	MC	NS
	20 (1)	6	10	74 (2)	36 (1)
STAGE	**I**	**II**	**III**		**IV**
	2	44	58		41 (4)

The commonest presentation was peripheral lymphadenopathy (mainly cervical) for periods up to a few years. All HIV patients had stage IV disease with 'B' symptoms and lack of access to ARVs was a major constraint in the management. Severe hepatic dysfunction was an adverse factor. There were 20 deaths during the period of follow up.

Conclusions: Mixed cellularity HD and advanced stage disease was common. Liver and marrow involvement was common in patients with HIV and HD.

Sincere acknowledgement to Dr J A Naidoo.

B CELL NON-HODGKIN LYMPHOMA

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B lineage lymphomas including Burkitt lymphoma (BL) and other high grade B cell lymphomas are the commonest lymphomas seen at our centre. Certain lymphomas are AIDS defining and together with Kaposi sarcoma represent the 2 commonest cancers seen with HIV infection.

Aim: To document our experience with B cell lymphomas and evaluate the impact of HIV infection in our paediatric population.

Method: A retrospective chart review of children under 12 years seen over the past 25 years. Histology included high grade B cell lymphoma, plasmablastic lymphoma, immunoblastic lymphoma, B lymphoblastic lymphoma, large cell lymphoma, T cell rich B cell lymphoma, BL and Burkitt-like lymphomas. HIV infected children were included.

Results: There were 128 patients of whom 31 were HIV positive. Racial distribution was African 116, Indian 9, White 2 and Coloured 1 patient. There were 88 male and 40 female patients. The overall median age was 4 years 10 months (range: 7 months – 11 years) and for HIV infected patients was 4 yrs. Stages were (HIV patients in brackets):

STAGE	I	II	III	IV
		12 (5)	73(12*)	39 (13)

(* I patient incomplete – possibly a stage IV)

Abd. involvement prominent – mass (80); hepatomegaly or HSM (15). Of the HIV+ pts 15 of the 31 patients had an abdominal mass at presentation. Bone lesions – 30 pts (25 inv. maxilla +/or mandible; 5 proptosis) of whom 13 had an abdominal mass. Other manifestations included skull mass, scalp mass, sternal mass, parotid swelling and testicular swelling. BM was involved in 26 pts and CSF positive in 23pts. 7 pts had paraparesis (1 Rx related).

Conclusions: Abdominal involvement is prominent and BL/BLL is common. Patients with HIV have a similar presentation apart from the primary CNS tumour and MALT lymphoma of the parotid and lung.

REVIEW OF HODGKIN'S LYMPHOMA AT CHRIS HANI-BARAGWANATH HOSPITAL

R D Wainwright, S Poyiadjis, G Naidu, D Mackinnon

A review of Hodgkin's lymphoma cases treated at CHBH from 2000 to 2007 was performed. Twenty-two cases were found – four were excluded - incorrect diagnosis, death prior to treatment, 2 were relapsed patients. The remaining eighteen patients' files were analysed.

There were 11 males and 7 females. The mean age at presentation was 9 years, range 4 years 6 months to 14 years. Disease stage – 10 with stage II, 4 stage III, 4 stage IV. Thirteen cases were still alive, two relapsed, and five died.

Three of the deaths were stage IV patients with complications. One stage III patient died with fulminant chickenpox, and one stage II patient died after surgery for removal of a renal mass – histology, metanephric adenoma.

Two protocols were used – ABVD on 7 patients, OEPA/COPP on 10 (1 died pre-treatment). Eight cases received radiation therapy – one developed radiation pneumonitis. There was an overall survival of 70 %.

A late presentation in the cases with advanced disease, and existing co-morbidities, contributed to their poor outcome. No comparison between the two protocols is possible owing to the small number of patients.

HIV RELATED LYMPHOMAS AT CHRIS HANI-BARAGWANATH HOSPITAL

R D Wainwright, S Poyiadjis, G Naidu, D Mackinnon

A review of cases diagnosed with lymphoma and HIV infection was made from 2003 to 2006. Seventeen children had been assessed for possible treatment. Poor Lansky performance status in 3 cases changed the treatment plan to palliative care. One patient died prior to commencement of therapy. Thirteen cases received treatment – 7 died, 3 from progressive disease, 4 from septic deaths, and 6 are currently alive and well. Fourteen were male and three female. Ages ranged from 3 years to 16 years, mean 5 years 3 months.

Treatment was started concurrent with anti-retroviral therapy (HAART). Difficulties encountered – late presentation, advanced disease, co-morbidities complicated management – prior organ damage, concurrent tuberculosis, HAART interactions, toxicity from high-dose chemotherapy – methotrexate encephalopathy, prolonged pancytopenia, recurrent sepsis – nosocomial, viral, fungal. Supportive care was essential –neupogen, platelet and blood products, antibiotics, antiviral and antifungals were required repeatedly, incurring high costs. Limited life support was available – neither ICU nor High Care, so a balance needs to be reached as regards the intensity of the treatment and available support. Survival of 46% is acceptable for this complex group of patients.

Future suggestions – in order to obtain statistically significant numbers, co-operation between oncology units to use uniform protocols adapted to the South African situation, is needed and to share data. The number of new cases per year is steadily increasing so cost effective treatment is essential. Patients with CNS disease did poorly.

THE SACCSG TUMOUR REGISTRY: FIGURES FOR 2005 AND AN 11-YEAR RETROSPECTIVE SURVEY

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Introduction: The SACCSG registry records all reported cases of childhood cancer that occur in South African (SA) children and are treated in the Republic of South Africa (RSA). Since 1995 the data have been collected by one institution, resulting in the inclusion of an 11-year survey (1995-2005).

Results: In 2005 620 cases were reported to the registry, an incidence rate of 40.6/106 for all SA compared with 33.8/106 for the 11 years. The Western Cape province had the highest recorded incidence rate (76.9) for this period, followed by Gauteng (56.3/106). Four of the 9 provinces recorded higher rates for the 11 years from 1995 to 2005 than for 2005.They are indicated below with *.

The incidence rates per million children for all the provinces were:

	1995-2005		2005
Limpopo*	9.8	and	16.6
North West Province*	24.4	and	25.8
Mpumalanga	29.0	and	25.4
Gauteng	56.3	and	49.4
Free State*	45.0	and	48.7
KZN	45.2	and	28.5
Northern Cape*	45.1	and	48.2
Western Cape	76 9	and	68.5
Eastern Cape	28.6	and	22.0

Conclusions: In 2005 we reported rates of 32.7/106 children for all of the RSA. There does, therefore, appear to be some improvement in the incidence rates.

The rates above are perhaps optimistic as, they depend on the officially recorded children according to official census figures for SA. Also we are concerned about the provinces reporting higher rates for the 11-year period than for 2005.

Data contributed by: WCP = P S Hartley, KZN = R Thejpal and G P Hadley, Pretoria = M Kruger, Jhb General = J Poole, Chris Hani-Baragwanath = L Wainwright, East London = M Painter and R Mathew, Northern Province = V Naidoo.

JOINT South African Paediatric Association (SAPA) Union of National African Paediatric Societies and Associations (UNAPSA)

FEATURES OF BACTERIAL MENINGITIS IN THE CHILD LESS THAN 5 YEARS IN THE CHU OF YOPOUGON (ABIDJAN, CÔTE D'IVOIRE)

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Introduction: Meningitis remains burdened with a high mortality and morbidity rate. To improve curative care and preventive measures periodic epidemiological and bacteriological assessment is required. Our objective is to describe the epidemiological and bacteriological profile of bacterial meningitis in children less than 5 years old in CHU of Yopougon.

Methodology: It is a descriptive retrospective study, which took place in the Medical Pediatrics Department CHU of Yopougon over a period of 5 years (2002 to 2006). It focused on children from 1 month to 59 months, hospitalized for bacterial meningitis confirmed by the bacteriological investigation of the cerebrospinal fluid: identification of an organism by culture or presence of a soluble antigen. Eighty-nine cases were selected.

Results: The median age was 18 months. We noted a male predominance. The main organisms found were: *Streptococcus pneumoniae* 47.2% (42/89), *Haemophilus influenzae* b 39.3% (35/89), *Neisseria meningitidis* 7.9% (7/89), *Salmonella* sp 4.5% (4/89) and *Flavobacterium* sp 1.1% (1/89). *Haemophilus influenzae* b and pneumococ meningitis were more common in infants: *Haemophilus influenzae* found in 31/35 cases (88.6%), with a clear predominance in infants under 12 months (20/35) and *Streptococcus pneumoniae* in 33 / 42 cases (75.6%). The fatality rate was 31.5% (28/89). The distribution of this was as follow: *Salmonella* sp 100% of deaths (4/4), *Streptococcus pneumoniae* deaths 42.9% (15/35), *Neisseria meningitidis* 28.6% of deaths (2 / 7), and *Haemophilus influenzae* b deaths 16.7% (7 / 42).

Conclusion: In our study, *Haemophilus influenzae* b and *Streptococcus pneumoniae* bacteria remain the most frequently found organisms in purulent meningitis in children less than 5 years. The introduction of the vaccine against *Haemophilus influenzae* b in infants less than 12 months would help reduce morbidity from purulent meningitis in children.

CLINICAL AND BIOLOGICAL FEATURES OF SEVERE MALARIA WITH HYPERPARASITAEMIA IN THE PAEDIATRIC DEPARTEMENT OF UTH OF YOPOUGON IN ABIDJAN, CÔTE D'IVOIRE

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Introduction: Severe malaria is an important cause of death in children in tropical countries. In order to improve the diagnosis and the management of severe malaria and to prevent death, 15 criteria of severe malaria was proposed by the WHO (since 2000), one of which is hyperparasitaemia. Because the density of parasites is rarely appreciated with microscopy, only a few studies mention these aspects of malaria.

Objective: The aim of this study was to describe the characteristics of severe malaria with hyperparasitaemia in children.

Methodology: It is a retrospective study performed in the paediatric department of the University Teaching Hospital (UTH) of Yopougon, in Abidjan. All children aged 1 month to 15 years who were hospitalised from January to December 2007 with severe malaria with hyperparasitemia were included in the study. Hyperparasitaemia was defined as more than 20% of the red cells on the blood smear infected with *Plasmodium*. It means a parasitaemia of more than 250 000 trophozoans of *Plasmodium falciparum*/mm^3.

Results: Thirty-two children were included. Malaria with hyperparasitaemia represented 2.96% of all cases with severe malaria seen in the paediatric department. The sex-ratio was 1.9. There were 66% boys and 34% girls. Children aged 5 years to 9 years were more affected (14 children representing 43.7%). The three main symptoms were fever, anaemia and seizures found respectively in 65.6%, 47,8% and 31.,2%. Hyperparasitaemia was associated with several other criteria of severe malaria: 21.9% simultaneously had cerebral malaria and severe anemia, 15.6% had both cerebral malaria and hemoglobinuria and 12.5% had 3 criteria of severe malaria : cerebral malaria, severe anemia, and hemoglobinuria.

A density of parasites between 250 000 and 500 000 was present in 46.8% and 18.8% had a density of > 1 000 000. Severe anemia with haemoglobin < 5g/dl was found in 18.8% of children, and thrombocytopaenia in 87.6%.

Artemeter was used in 56.3% and quinine salt in 31.3%. Blood transfusion was given in 14 patients (43.8%).

Twenty-eight (87.5%) children recoved without any sequelae. Two patients (6.2%) died and 2 children had severe neurological sequelae. The mean duration of hospitalisation was 5 days, but 40.62% were hospitalised between 5 to 10 days.

Conclusion: Severe malaria with hyperparasitaemia was more common in older children. Children with severe malaria can have several of the criteria of severe malaria simultaneously. The outcome of treatment is good, but a few children had severe neurological sequelae.

PEADIATRIC HEALTH STAFF KNOWLEDGE, ATTITUDE AND PRACTICE CONCERNING CHILDREN WITH UNCOMPLICATED MALARIA IN YOPOUGON (ABIDJAN, CÔTE D'IVOIRE)

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Introduction: To counter the threat of resistance of *Plasmodium falciparum* to monotherapies and to improve treatment outcome, WHO recommends combination of antimalarials for the treatment of falciparum malaria (artemisinin-based combination therapy: ACT). These recommendations have been accepted by the national programme for malaria control in Côte d'Ivoire from 2002 onwards.

Objective: The aim of this study was to evaluate the knowledge of paediatric health staff with regard to the WHO recent recommendations and to assess their practice and their attitude with regard to the treatment of uncomplicated malaria in the paediatric population.

Methodology: A prospective and cross-sectional study was performed from March to April 2007. The medical staff's knowledge was assessed by means of an interview (16 in UTH, 10 in a health center and 10 in two pharmacies in the same area). Management of patients at 384 medical consultations (184 visits in UTH, 100 in the health center, and 100 in the 2 pharmacies) were observed. These consultations concerned all children aged 1 month to 15 years diagnosed with uncomplicated malaria. The children with severe malaria were excluded.

Results: Seventy-seven percent of paediatric health staff was aware of ACT (artemisinin). Information was obtained form pharmaceutical companies in 58.3%. WHO recommendations were known by 91.7%, but only 50% were aware of the national guidelines for the treatment of malaria.

Despite the fact that artesunate/amodiaquine was recommended as the first-line treatment in Côte d'Ivoire, 77.8% of health staff mentioned artemeter/luméfantrine as the first line.

ACT was not prescribed by 52.8%, because according to them these combination medications had complicated dosages in children and they were also afraid of ACT side-effects in the paediatric population.

With regard to medical consultations, 54.2% were by paediatricians. Of all the children seen, 71.1% were less than 5 years old. The diagnosis of malaria was proven in only 10.93% cases (parasitological diagnosis was made in only 42 out of a total of 384 patients), in spite of the presence of a parasitological laboratory in UTH and in the health center. ACT was prescribed in 70.8% and monotherapies in 29.2%. ACT was more likely to be prescribed in the health center.

Conclusion: The paediatric health staff who managed children with uncomplicated malaria in Yopougon is aware of ACT, but they are hesitant about prescribing it due to perceived ACT side-effects in children. Monotherapies continued to be prescribed in 29.2. The diagnosis of malaria continued to be presumptive in most cases despite the availability of parasitological laboratories.

This study demonstrated that the national programme for malaria control should intensify training of paediatric health staff with regard to the management of malaria in children. It also showed that more studies must be done to assess the prevalence of ACT side-effects in the paediatric population.

NT-PROBNP AS A MARKER OF NECESSITY OF IMPLANTATION A PACEMAKER INTO NEWBORNS DIAGNOSED WITH CONGENITAL ATRIOVENTRICULAR BLOCK (CAVB)

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Introduction: Autoimmune AV block occurs in approximately 1 per 14,000-20,000 live births. The foetal mortality rate of isolated CAVB may be up to 30-50%, new newborns – about 5%.

Beta-mimetic, immunoglobulins and sometimes immunosuppressive treatment are used. Pacemaker implantation is frequently a necessary procedure and timing the decision for the procedure is vital, particularly with the youngest patients.

Case study: A newborn, from G1 mum, at term, with a body weight of 3080g was admitted in serious condition, with heart rate of 50-70/min and good ventricular contractility. He was treated with izoprenaline and immunoglobulins. On the 6 day after delivery his general condition deteriorated, with an increasing cardiac insufficiency. Heart rate remained at 50-70/min; however, contractility deteriorated (EF=30%). On day 7 a pacemaker was implanted and the patient's condition

improved considerably. 6 months later the baby remains healthy and neurodevelopmentally appropriate for age.

Prior to the procedure the level of NT-probeBNP was monitored. On the 2 day the high level – 34421 p/ml – decreased, reaching 32421 pg/ml on day 3. The level of NT-probBNP increased rapidly on day 6, up to 59 133pg/ml and 74 536 pg/ml immediately before the pacemaker was implanted. Desaturation and progressive cardiac failure was noticed at the same time.

On the 2 day after the operation a considerably drop of NT-probeBNP was recorded, down to 42 410 pg/ml, with further normalization of the marker level.

Increasing level of NT- pro BNP preceded the signs of cardiac failure obtained in ECHO and ECG.

Conclusion: BNP as a sensitive marker of cardiac insufficiency may play a significant role in deciding on timing of pacemaker implantation into newborns with CAVB.

THE EFFECT OF HIGHLY ACTIVE ANTI-RETROVIRAL THERAPY (HAART) ON HIV ASSOCIATED ARTHRITIS IN CHILDREN

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Background: HIV infection is associated with chronic arthritis, causing significant morbidity and disability. Conventional disease modifying anti-rheumatic drugs are either ineffective or are contraindicated in children with HIV infection. HAART is effective in treating HIV infection.

Objective: To investigate the effect of HAART on HIV associated arthritis in children attending the Paediatric Rheumatology Clinic.

Methods: An analysis of the clinical records of all children with HIV associated arthritis who were started on HAART from July 2005 to July 2007.

Results: Fifteen children received HAART The male:female ratio was 12:3. The mean age at onset of arthritis was 5.8 years [range 2-9 years]. The mean duration of follow up on HAART was 13.6 months [range 6-24 months]. The clinical stage of HIV infection significantly improved in 12 [80%] children from stage 3 and 4 to stage 1. Nine [60%] of children on HAART showed complete remission; 3 [20%] had greater than 70% improvement in their joint count; 2 [13%] had greater than 50% improvement and 1 child had greater than 20% response. The erythrocyte sedimentation rate returned to normal in 11 [73%] of patients.

Conclusion: HAART significantly improved the outcome of arthritis in children with HIV associated arthritis.

THE INCIDENCE AND SEASONALITY OF INFANTILE ROTAVIRUS DIARRHOEA IN SIERRA LEONE

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Background: Rotaviruses occur throughout the world, and are important causes of gastroenteritis. Because of the urgent need to develop an effective vaccine to protect against severe rotavirus disease, there is a need to provide data everywhere, also in Africa, on the incidence of rotavirus infections, as well as the antigenic and genomic diversity of the rotavirus required for the design of an effective vaccine strategy. No rotavirus studies have been done in Sierra Leone so far. We endeavoured to determine the epidemiology of rotavirus infection in Sierra Leone.

Methods: This was a descriptive study with prospective enrolment of children, between the ages of three months and three years, presenting with gastroenteritis, at the Freetown Children's Hospital in Sierra Leone. Physical examination findings were recorded. Stool specimens were screened for the presence of rotavirus antigen. The samples were tested in South Africa using polyacrylamide gel electrophoresis to confirm rotavirus infection. Informed written consent was obtained.

Results: Over a five-month period 143 children presenting with gastroenteritis were recruited. Stool samples obtained from study subjects (128), were tested for the presence of rotavirus. There were more males (56%) than females. Almost half of the subjects (45%) were aged between three and nine months and the mean age was 10.85 months. About a third of the 128 stool samples (48; 38%) tested positive for rotavirus; 80 (62%) tested negative. Rotavirus positive incidence peaked in August (64%), during the rainy season. Disease severity was worse in rotavirus positive patients; this difference was statistically significant.

Conclusions: The incidence of rotavirus infection is 20% higher in boys; a gender difference which is confirmed elsewhere in West Africa. The rotavirus season in Sierra Leone takes place between July and September, and coincides with the rainy season.

CHEST RADIOGRAPHIC DIAGNOSIS OF SUSPECTED PULMONARY TUBERCULOSIS AMONG CHILDREN IN A COMMUNITY SETTING

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Background: Definitive radiographic diagnosis of pulmonary tuberculosis (PTB) may be difficult in ambulant children with mild disease.

Aims: To compare agreement on radiographic diagnosis among children with suspected tuberculosis in a community setting.

Methods: Records of 1869 children investigated for PTB during a South African tuberculosis vaccine trial (2001-2006) were used. Chest radiographs were reviewed blind by 3 experienced paediatricians and classified into 6 categories of likelihood of diagnosis (PTB = highly likely/likely/suspicious; NOT PTB = inconclusive/other abnormality/normal). Radiographs of 1354 children (72.4%), mean age 15 months, were reviewed by all 3 reviewers for purposes of this comparison.

Results: 67% of these children reported TB contacts, 40% had cough > 2 weeks, and 22% loss of weight. 1.9% were HIV-infected, 28% had a Mantoux test > 15 mm, and 107 children (7.9%) cultured *Mycobacterium tuberculosis* (sputum / gastric lavage).

Radiographic diagnosis of PTB was made by each of 3 reviewers in 12%, 20%, and 66% of suspected cases. Percentage agreement between reviewer pairs for binary categories (PTB / NOT PTB) was 43%, 51%, and 81%, with kappa 0.09, 0.18, and 0.29. Percentage agreement for hierarchical categories was 51%, 57%, and 84%, with weighted kappa 0.06, 013, and 0.28.

Conclusions: Among young children in the community with suspected PTB, the percentage with radiographic diagnosis of PTB ranged from 12 to 66% of cases investigated. Agreement between reviewers was poor for binary and hierarchical diagnostic categories.

THE EFFECTS OF PROBIOTICS ON FEEDING TOLERANCE, BOWEL HABITS AND GASTROINTESTINAL MOTILITY IN PRETERM NEWBORNS

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Objective: The aim was to investigate the effect of dietary supplementation with a probiotic on feeding tolerance and gastrointestinal motility in healthy formula-fed preterm infants.

Study design: Thirty preterm newborns were enrolled in the study. Ten were exclusively breast-fed whilst the remaining 20 were randomly assigned in a double-blind manner to receive either *Lactobacillus reuteri* ATCC 55730 (at dose of $1x10^8$ colony forming units a day) or placebo for 30 days. Clinical symptoms of gastro-intestinal function (regurgitation, vomiting, inconsolable crying and evacuation) and physiological parameters (gastric electrical activity and emptying) were recorded before and after the dietary intervention.

Results: Body weight gain per day was similar in the three groups and no adverse events were recorded during the study. Newborns receiving probiotics showed a significant decrease in regurgitation and mean daily crying time, and a larger number of stools compared to those given placebo. Gastric emptying rate was significantly increased and fasting antral area was significantly reduced in both the newborns receiving *Lactobacillus reuteri* and breast-fed newborns compared to placebo.

Conclusions: Our results suggest a useful role for *Lactobacillus reuteri* supplementation in improving feeding tolerance and gut function in formula-fed preterm newborns.

CLOSING THE AUDIT LOOP: USING CHILD PIP DATA TO DECREASE MORTALITY DUE TO ACUTE GASTRO-ENTERITIS IN WITBANK HOSPITAL

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Background: Witbank Hospital Paediatrics department has been using the CHIP programme (Child Healthcare Problem Identification Programme),

which aims to save lives through death auditing, since 2004. We aim to show how, by using our CHIP data and closing the audit loop, we managed to significantly decrease deaths due to acute diarhoea in our institution.

Methods: Every child death since 2004 was audited and coded on the CHIP-program. One of the main problems identified during 2006 was a rising number of deaths due to acute diarhoea. By using the CHIP program's data analysis function a number of modifiable factors were identified and certain interventions were implemented in 2007. CHIP data for 2007 were then analyzed and compared with those of 2006.

Results: During 2006 there were 605 admissions for acute gastro-enteritis, with 14 deaths, giving a case fatality rate of 2.3%. Of the deaths 84% occurred during the first 24 hours of admission and 74% occurred after hours. The most common modifiable factors found were: a) assessment of shock/dehydration insufficient, b) shock not treated appropriately (both on initial presentation), and c) monitoring of hydration level in ward inadequate. The interventions were geared towards solving these problems.

A 52% reduction in the case fatality rate of acute gastro deaths were achieved.

	2006	2007
Total acute gastro admission	605	540
Total deaths	14	6
Case fatality rate: (%)	2,3	1,1

Full results and methods of interventions will be presented at the conference.

Conclusion: Interventions borne out of Child PIP data analysis and closing of the audit loop led to a significant reduction in mortality due to gastro-enteritis at Witbank hospital.

CLINICAL AND IMMUNOLOGICAL CHARACTERISTICS OF HIV INFECTED INFANTS LESS THAN 60 DAYS OLD IN SOUTH AFRICA

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Background: There are few descriptions of HIV-infected and -exposed infants in the first 10 weeks of life. Such data may guide diagnostic algorithms.

Aims: 1. To examine the predictive value of clinical characteristics for HIV-infection in very young infants comparing HIV-infected, HIV-exposed uninfected (EU), and HIV unexposed uninfected (UU) infants. 2. To identify clinical characteristics associated with severe immune suppression (CD4 ≤25%).

Methods: Infants were identified through the PMTCT programs in Cape Town and Soweto. HIV-infected (n=540), HIV-exposed uninfected (EU, n=125) and HIV-unexposed uninfected (n=125) infants were identified. Clinical data was collected in a standardized format. Characteristics of infants in the 3 groups were compared using Kruskal-Wallis and Fisher's exact tests.

Results: The median age of all infants was 44 days (range 28-78). HIV-infected infants had significantly lower weight and were significantly more likely to have generalized lymphadenopathy (OR 6.4 95%CI 2.8-14.9), oral candida (OR 3.7 95%CI 2.0-6.8), and hepatomegaly (18.5 95%CI 2.5-134.3) than EU infants. Compared with HIV unexposed infants, both HIV infected (OR 26.6 95%CI 6.5-108.8) and EU (OR 7.1, 95%CI 1.6-32.3) infants were more likely to have oral candida. Clinical gastroesophageal reflux disease and splenomegaly were found in 3 and 6% of the HIV-infected infants respectively, and none of the uninfected infants. All associations persisted when adjusted for infant age. The sensitivity of each of these symptoms for HIV infection was low (<20%) but specificity was high (98-100%) in predicting absence of HIV infection. Infection/exposure status was not associated with a BCG local adverse event, sepsis, or encephalopathy. Low weight and pneumonia were the only age-adjusted findings having strong associations with CD4%<25%.

Conclusions: This is the one of the first clinical descriptions of a large cohort of very young HIV-infected and exposed infants from South Africa. When evaluating children at risk for HIV whose status is unknown, the lack of certain findings may be useful for identifying those who are less likely to be HIV infected when HIV PCR is not possible, and in identifying those without severe immunosuppression when CD4 percentage is unavailable.

EPIDEMIOLOGICAL USES OF CHILD HEALTH CARE PROBLEM IDENTIFICATION PROGRAMME DATA

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Background: Child health care problem identification programme (Child PIP) has been used as a dependable in-hospital mortality auditing tool to improve quality of care children receive within South African health system. Facility-based Child PIP data may be useful in estimating population-based indices of mortality and thus provide another method to triangulate and validate population-based mortality data.

Aim: The aims of this study were 1) to demonstrate the utility of hospital admission and mortality data to assess the adequacy of pre-hospital healthcare among children aged < 5 years within a health district; 2) to develop a conceptual model of the use facility-based mortality data in enhancing our understanding of population-based indices of child mortality.

Study design: Period-based cross-sectional study.

Methods: We analysed admission and mortality data from 8 participating hospitals from all levels of care (district, regional and tertiary) in KwaZulu-Natal in 2006. Total admissions, characteristics of admissions and deaths, all-cause, cause- and age-specific in-hospital mortality rates (IHMR) among children < 5 years were computed.

Results: Of all admissions < 5 years (N=8621), 62% were infants and 63% were malnourished. All-cause IHMR was 9.2 per 100 admissions (range: 6.5-10.8), with highest rates among infants. Cause-specific IHMR among children <5 years were: 23.9 for severe malnutrition, 14.9 for weight < 3rd centile, 11.0 for respiratory infections, and 9.0 for diarrheal diseases. More than 2/3rd of all deaths occurred within 72 hours of admission. At least 67% of all preventable causes of in-hospital deaths were identified as being outside the hospital (Home, Clinic, and Accident & Emergency). Using mathematical formulae developed by Murray *et al*. we proposed a child mortality conceptual model.

Interpretation: Despite hospital mortality statistics being inherently biased toward children who were either very ill or those who had access to facility level healthcare, indices from Child PIP data could reliably inform about the quality of care outside the hospital and may be used as sentinel indices for monitoring and evaluation of pre-hospital child health programmes. Child PIP data may provide valuable complementary data on overall measures of child mortality.

RECURRENT JUVENILE LARYNGEAL PAPILLOMATOSIS AN IMPORTANT CAUSE OF AIRWAY OBSTRUCTION IN YOUNG CHILDREN: A CASE PRESENTATION

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Background: Juvenile recurrent laryngeal papillomata (JRLP) are due to infection with human papillomavirus (HPV) type 6 and 11. In children less than 5 years of age the infection is thought to be related to intrauterine acquisition from an infected mother. The clinical presentation is that of stridor and respiratory distress and thus misdiagnosed as laryngomalacia, asthma and croup.

Case presentation: We present a 26 month old who presented with respiratory distress secondary to stridor at age 17 months. He was admitted to a primary hospital for two weeks with a diagnosis of croup, for which he was treated with adrenaline nebulisations. Due to deterioration, he was referred to a regional hospital where he was treated for a further week for grade 3 to 4 croup. He continued to deteriorate and therefore was intubated. A direct laryngoscopy was performed and diagnosis of papillomata involving the true and false vocal cords was confirmed. He subsequently required seven debulking procedures for recurrences over a period of eleven months.

Conclusion: JRLP is also associated with primary immunedeficiencies in patients with HLA Class II polymorphisms, transporter proteins of these polymorphisms as well as abnormalities of CD4 cells. The co-infection of sexually transmitted infections as well as HIV is well documented. Whether HIV infection poses a further risk for young children to acquire RJLP is still not described in the literature.

The management of JRLP is a very costly because of the need for repeated surgical or laser debulking which are the gold standard therapies. Thus, this creates a significant financial burden and impacts on the quality of life in affected patients. Adjuvant therapies are expensive and not always successful. The use of anti-viral vaccines which include HPV 6 and HPV11 provides hope for the reduction in the incidence of these lesions in children.

PREVALENCE OF BACTERIAL CONTAMINATION OF MILK FEEDS IN THE PMTCT CLINIC AT THE DR GEORGE MUKHARI HOSPITAL

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The risks of using commercial infant feeds have been described.

The aim of this study was to determine the bacterial contamination of formula milk feeds obtained randomly from bottles of babies who were in the outpatient department at the DGMH to have their babies tested for HIV at the PMTCT clinic. All milk samples were prepared at home.

One hundred samples were collected into sterile containers which were transported in a cooler box to the microbiology laboratory within one hour of collection for incubation. All positive culture colonies were gram stained and analysed.

The quantitative levels of bacterial contamination were determined and expressed as colony forming units per millilitre of sample (CFU/ml). Contamination was defined as any positive culture from the milk samples.

Bacterial contamination was found in 76% of the milk samples. The commonest organisms found were *Enterococcus* species (67%), *Staphylococcus aureus* (37%) and *Klebsiella pneumonia* (33%). 17 samples had up to three colony types of bacteria. The CFU/ml in all samples ranged from 18 to over 300 CFU/ml. Only 24% of the milk samples were not contaminated in this study.

Conclusion: Although HIV mothers are supposedly counselled on how to prepare infant formula prior to discharge from hospital, there was an unacceptable high prevalence of bacterial contamination of milk feeds prepared at home which poses a health risk for these babies.

VENTILATOR-ASSOCIATED PNEUMONIA IN A SOUTH AFRICAN PAEDIATRIC INTENSIVE CARE UNIT WITH HIGH HIV PREVALENCE: A RETROSPECTIVE SURVEY

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Aim: To obtain preliminary prevalence, aetiological and outcome data on South African paediatric patients with ventilator associated pneumonia (VAP).

Methods: Non-bronchoscopic bronchoalveolar lavage (BAL) specimens taken between January 2004 and September 2005 were prospectively recorded and related clinical data were retrospectively reviewed. VAP was defined as a new isolate on BAL and a modified Clinical Pulmonary Infection Score ≥5.

Results: 230 patients aged 3.9 (2.2-9.1) months [median (IQR)] underwent 309 BALs during 244 PICU admissions. Most patients (84%) were admitted with acute infectious diseases; with a 70% incidence of comorbidity. 76 patients (33%) were HIV-exposed and 56 (24%) HIV- infected.

Of 172 BALs taken ≥48 hours after intubation; 74 specimens from 55 patients fulfilled VAP criteria. *A. baumannii* was the most common VAP pathogen, followed by *K. pneumoniae*, *S. aureus* and *P. aeruginosa.* Antibiotic resistance patterns differed in patients with and without VAP.

Patients who developed VAP had a higher proportion of comorbid conditions (76% vs. 55%, p = 0.01) and reintubations (39% vs. 12%, $p < 0.0001$) when compared to non-VAP patients. Median (IQR) length of PICU stay was 12.5 (5-21) days vs. 8 (5-14) days (p = 0.03); total mortality was 44% vs. 33% (p= 0.04); and the risk adjusted PICU mortality was 1.38 vs. 0.79 (p = 0.002) in VAP vs. non-VAP patients respectively.

Conclusions: VAP is associated with significant morbidity and mortality and may relate to the high incidence of comorbid conditions in this population. Primary VAP pathogens differ from developed countries.

SAVING CHILDREN 2005-2006: THE HIV STORY FROM SOUTH AFRICAN CHILDREN

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Introduction: Child health care Problem Identification Programme (Child PIP) aims to improve quality of care in the health system of South Africa by death auditing.

Methods: An audit of death of children less than 18 yrs from January 2005 to Dec 2006 was done at 26 hospitals in South Africa using Child PIP. This report focuses on the impact of HIV. Data have been analysed using the Child PIP software

Results: There were 3848 death audited from 56 146 admissions with In-hospital case fatality rate of 6.2%. Regarding the ages 60% were less than a year, 29% were underweight for age and 35% had severe malnutrition. The major causes of death were pneumonia (18%), septicaemia (15%) diarrhoea (12%), tuberculosis (9.2%) and pnuemocystis jeroveci pnuemonia (8.3%). Only 12% received single dose nevirapine at birth and in 52% it was unknown. Co-trimoxazole prophylaxis was given to only 22% and in 39% it was unknown. There were 33% with HIV World Health Organization clinical stage IV and 16% stage III. Only 4% were on HAART at time of death.

Conclusion: The data show that half of children dying in hospitals have symptomatic HIV disease while less than 20% had documented proof of benefit from the PMTCT and Comprehensive Care Management and treatment (CCMT) programme for HIV. We therefore recommend documentation on the Road To Health Card for better health care provision of children and for the data to be used to advocate for more resources for PMTCT and paediatric ARV programme. The data also show that Child PIP can be used to assist in monitoring PMTCT and CCMT programme.

ASSESSMENT OF THE IMPACT OF ROLL BACK MALARIA IN NORTHERN NIGERIA: STUDY OF A LOCAL GOVERNMENT AREA

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Introduction: Malaria causes about a million deaths annually in Africa, mostly in under-five children and significantly slows continental economic growth. The Abuja Declaration on Roll Back Malaria (RBM) in 2000 set as a goal the access of at least 60% of under-five children and pregnant women to benefits of malaria protective measures, including the use of insecticide-treated nets (ITNs) by 2010. More than halfway into the timeframe of the initiative there is concern that the goals may not be met.

Materials and methodology: In 2006 a survey was carried out in Zaria Local Government Area of Kaduna State, Nigeria. The survey was 3-phased: an evaluation baseline study, an intervention period, and a post-intervention assessment survey. The baseline involved 300 caregivers from randomly selected households and 706 children including 300 (42.5%) febrile ones. The interventional phase included public enlightenment programmes and educational activities to healthcare providers at different levels of health system, and distribution of ITNs. One year after the intervention an impact assessment was carried out.

Results: The proportion of caregivers opting to treat at home increased (15.1 - 24.3%). Attendance at primary health facilities increased while there was a reduction in patronage of patent medicine stores. Paracetamol and Chloroquine® remained the two commonest drugs for initiation of treatment while artemisinin-based combination therapy was rarely used. ITN use increased from 0.6% to 21.1% among U5 while intermittent presumptive treatment of malaria in pregnant women improved from 3.2% to 69.8%. The U5 mortality reported increased from 10.0% to 11.7%.

Conclusion/recommendation: The risk of not achieving the goals of the RBM initiative is high. A lot needs to be done to empower major stakeholders. With adequate scale-up of antimalarial activities malaria burden in Africa may be significantly reduced by 2010. This may boost achievement of the millennium development goals on the continent.

EVERY DEATH COUNTS: SAVING MOTHERS, SAVING BABIES, SAVING CHILDREN CONSENSUS DOCUMENT

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Aim: To establish the best estimates of maternal, neonatal and child mortality rates, determine the major causes of deaths, the coverage of effective interventions, and harmonise the recommendations of the Saving Mothers, Saving Babies and Saving Children reports.

Methods: Representatives of the Saving Mothers, Babies, and Children groups, the MRC Maternal and Infant Health Care Strategies, Burden of Disease and Health System units, the national Department of Health Maternal, Child and Women's Health and Nutrition Cluster and Saving Newborn Lives, met and reached consensus on the above aims

Results: The maternal, neonatal and child deaths and the prevalence of HIV are all increasing, indicating South Africa is moving in the opposite direction to achieve the millennium development goals (MDGs) 4, 5 and 6. The big 5 causes of maternal and child deaths are HIV/AIDS, complications of pregnancy and childbirth, newborn illness, childhood illnesses and malnutrition. The big 5 health system causes of death are lack of use of the health care facilities by communities, inadequate transport between health institutions, inadequate facilities, inadequate skills and lack of caring attitude by health care workers. There were serious gaps in data to determine coverage of effective interventions, although effective interventions are readily available.

Conclusion: Saving lives is dependent on coverage of effective interventions and the quality of care with which these effective interventions are practiced. Coverage of the interventions depends on specific activities namely adequate knowledge and usage by the community and having accessible facilities. Quality of care is dependent on adequate staffing, drugs and equipment, skills and a caring attitude. Achieving these specific activities is the responsibility of different distinct groupings of people: the community; the enablers, i.e. the health care administrators and policy makers and the doers, i.e. health care providers and teachers. There are recommendations common to all three reports which targeted these groups and also specific recommendations directly related to the big 5 causes of maternal, neonatal and child death, also grouped into community, enablers and doers.

To reduce the number of deaths and move towards achieving the MDGs, these integrated recommendations must be implemented by those responsible.

THE VON DATABASE AND MEDI-CLINIC GROUP POOLED DATA

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Medi-Clinic Group

The **POOLED** data of **SIX** neonatal units in the Medi-Clinic Group are compared to the Vermont Oxford Network (VON) for all babies 400 to 1500 grams OR 22 to 29 weeks 6 days gestation.

Differences are noted between the Medi-Clinic Group (MCG) units as to

1. Antenatal steroid use
2. Use of conventional ventilation
3. Late bacterial sepsis
4. Severe retinopathy of prematurity (ROP)
5. Caesarean section rates.

These are reported on a no name basis. There may be an association between increased oxygen use, use of conventional ventilation and ROP in the limited numbers reported.

For the period of 01-01-2006 to 31-12-2006:

- Average birth weight of neonates was: 1155g at MCG compared with 1044g for VON
- Average gestational age was 29 weeks at MCG compared with 28 weeks at VON
- Discharge weight was 2312 g at MCG compared with 2417 g at VON
- Average length of stay to discharge was 58 days at MCG compared with 62 days at VON
- The mortality rate was 8% at MCG compared with 16% at VON

Other markers for comparison include infant characteristics, caesarian section rate, ante- and post-natal steroids and key performance indicators.

Overall MCG figures compare favorably to the VON.

Future goal: To pool National Neonatal data to assist with drawing up appropriate guidelines for Neonatal Care in South Africa.

IMPROVING QUALITY OF CARE FOR CHILDREN IN THE SOUTH AFRICAN HEALTH SYSTEM: CHILD PIP 2005-2007

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Introduction: The quality of care children receive in the South African health system is experienced by providers and receivers as suboptimal. The mortality review process for children who die in hospital enables detailed assessment of the health profile of children who die and the quality of care they received. The Child Healthcare Problem Identification Programme (Child PIP) provides structure for the mortality review process. This paper reports on Child PIP findings from 2005-2007.

Methods: Since 2005, over 30 hospitals in South Africa have used Child PIP, and submitted data to the national database. At each site the study population included all admissions (0-18 yrs), and detailed data were collected on each death. Health profile information was gathered for each child (social, nutritional and HIV context, and cause of death). Quality of care information was obtained by determining modifiable factors. Data were analysed using Child PIP software.

Results: From monthly tally data, there were over 3,490 deaths from 54,000 admissions, and 4,287 deaths were reviewed in detail. The in-hospital mortality rate was 6.5 deaths per 100 admissions. For each death there were 2.0 modifiable factors. As a proportion of all deaths 65% were UWFA and 52% were HIV exposed or infected. The most frequently recorded diagnoses in children who died were ARI (18%), septicaemia (15%), diarrhoeal disease (12%), TB (9%) and PCP (8%). Sixty-six % of modifiable factors occurred in the health system (14% in clinics, 23% at admission, and 29% in children's wards) and 34% at home. For each death, clinical personnel were responsible for 1.1 modifiable factors, administrators for 0.4 and caregivers for 0.8.

Conclusion: The Child PIP audit provides information showing the health profile of a paediatric population ravaged by HIV and poverty, and dying of preventable conditions. It also indicates that the quality of care they receive throughout the health system can be improved.

It is the responsibility of health care workers and managers to respond to the challenges posed, and to create solutions.

CARE OF THE SICK CHILDREN IN THE HEALTH CENTERS OF REFERENCE TO BAMAKO

Sylla Mariam

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Our study aimed to estimate the quality of the care of sick children in the reference health centres of Bamako.

The following was evaluated: the availability of equipment and medicine, the description of the profile of the providers, the appreciation of the knowledge and the attitudes and practices of providers with regard to the main symptoms of the IMCI.

There was good availability of medicine and medical equipment in the centres.

The evaluation of the children by the providers of care was insufficient: danger signs were looked for in only 32.1 % of the cases. The nutritional state and the vaccine state were rarely estimated.

Less than half of the parents were informed about the diagnosis. The relationship between diagnosis and treatment was correct in 55.7% of the cases.

The care of the children in the structures of 2nd level is not in accordance with the standards and the procedures although these standards are known to the staff. A formation/recycling is necessary for quality services.

INVASIVE BACTERIAL INFECTIONS IN THE PAEDIATRIC UNIT OF THE CHU GABRIEL TOURÉ

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It is a retrospective study on invasive bacterial infections performed in the paediatric department of CHU Gabriel Toure (Bamako). Its aim was to study the confirmed invasive bacterial infections in this department during 2004.

Invasive bacterial infections accounted for 5.2% of the hospitalizations and 30.5% of the invasive infections were due to septicemia. Invasive bacterial infections were most common in the age group 1 to 11 months. No season was incriminated but the greatest number of patients was observed in April.

Streptococcus pneumoniae, *Haemophilus influenzae B*, *Salmonella* spp, *Staphilococcus aureus*, *Escherichia coli*, *Salmonella typhi*, and *Nesseria meningitidis* A were the organisms most frequently observed in invasive bacterial infections. Combination therapy with Ceftriaxone and Gentamicin was the most commonly used antibiotic regimen. The sensitivity patterns of the cultured organisms were as follows: *E coli* (ceftriaxone 57.1%), Hib (100% with ciprofloxacin), *Neisseria meningitidis A* (chloramphenicol 100%, ampicillin 88%), *S aureus* (oxacillin 88%), *S typhi* (ceftriaxone and ciprofloxane 100%).

We obtained a cure rate of 69.5%, and 21.7% of children died. The majority of the deaths were due to pneumococcus, Hib and E coli.

MAKING A DIAGNOSIS OF CYSTIC FIBROSIS IN SOUTH AFRICA

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Introduction: Cystic fibrosis (CF) occurs in **all** South African populations and is one of the commoner serious inherited conditions in this country. CF has multiple manifestations ('typical' and 'atypical') from the antenatal to the adult phases of life. Early diagnosis of cystic fibrosis is important as it leads to a better prognosis.

Methodology: A literature review, and consensus of an expert panel led to the development of a proposed algorithm for CF diagnosis. This has been tailored to make applicability to the South African health environment possible.

Results: The algorithm is based on a tabular categorisation of 3 groups of patients based on the likelihood of CF:

1. Typical cystic fibrosis probable (e.g. Poor growth from an early age, and/or Persistent diarrhoea or smelly stools, and/or Persistent or recurrent chest problems, and/or Other **typical** features such as meconium ileus)
2. Typical cystic fibrosis possible (symptoms/signs that have other likely causes, e.g. chronic diarrhoea)
3. Atypical cystic fibrosis possible (e.g. milder versions of CF respiratory patterns, male infertility).

Using these entry points the clinician is guided to sweat testing, faecal pancreatic elastase and/or genetic testing in a logical fashion.

Conclusion: The MSAC of the Cystic Fibrosis Association are seeking to improve the diagnosis of this condition and prompt practitioners to entertain the diagnosis of CF early in the life of an affected child, thereby promoting continued good health and longevity.

NUTRITIONAL PROGRAMMING: HAS INFANT FORMULA FEEDING CONTRIBUTED TO THE OBESITY EPIDEMIC?

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Accumulating evidence from many different parts of the world has linked the developing fetus' and young infant's nutritional environment to epigenetic programming that may predispose the organism to specific identifiable health risks in later life. After initial predominant attention to insufficiency or excess of nutrition during gestation and early infancy, increasing evidence now also links the actual composition and balance of nutrients during critical periods of adaptation to nutritional programming.

Epigenetics refers to changes in gene function that occur without a change in gene sequence. DNA methylation contributes to conformational change in chromatin during periods of enhanced epigenetic lability that can repress transcription activation and "silence" the gene.

The thrifty phenotype hypothesis links a mismatch between programming and later nutritional environment to the development of obesity, the metabolic syndrome and cardiovascular risk.

While the obesity epidemic is undoubtedly shaped by numerous interrelated associations and multiple complex causations, we ask whether nutritional epigenetic programming through exposure to non-human protein instead of breastfeeding in the first few months of postnatal life is contributing to this epidemic.

The development of infant formulae and satisfactory bottles and teats for infant feeding was associated with a massive decline in breastfeeding from around the middle of the 20th century, starting in and spreading from the industrialised countries. In this paper, we point out an epidemiological and temporal association between the decline in breastfeeding and the rise of the obesity epidemic and review the evidence for a protective effect of breastfeeding concerning the prevention of later obesity.

LAUNCHING A PAEDIATRIC 'WIKI' IN SOUTH AFRICA

David Woods

Eduhealthcare

In order to maintain a high standard of care for sick children, it is essential to provide easy access to continuing education in child health to all health workers involved with paediatric services.

The Perinatal Education Programme (PEP) has provided decentralised, self-help learning courses in maternal and newborn care to nurses and doctors for many years. Over 50 000 participants have used this opportunity in South Africa to take responsibility for their own professional growth and continuing education.

Recently the book *Child Health Care: a learning programme* for professionals has expanded the range of PEP topics to include child health. Twenty three specialists have contributed to Child Health Care which uses a learner-based, problem-solving layout to challenge most of the common and important clinical problems encountered in primary care paediatric practice.

In addition to a print format, this distance learning course is also available on a free website so that colleagues, especially in poor countries beyond Southern Africa, can also have access to the education material. This enables health care workers to manage their own training programmes.

Through a partnership with Electric Book Works, the web-based Child Health Care course has now been converted into a 'wiki' format. Using a similar concept to the hugely successful Wikipedia, both local and international contributors are invited to comment on the learning material. All contributions will be formally reviewed and recognised to maintain a high academic standard. Comments will be added to the web text as drop down boxes. In this way it is anticipated that the core content of all these courses will be enriched and expanded through shared comment based on personal experience and evidence-based trials

An end of course examination will also be made available on the website to enable individuals, groups or institutions to manage their own course assessments.

Access to the 'wiki' format and end of course examination is available on www.ebwhealthcare.com.

It is hoped as many paediatric colleagues as possible in South Africa will join this innovative project by both using and contributing to these learning programmes.

South African Children's Cancer Study Group (SACCSG) South African Association of Paediatric Surgeons (SAAPS) combined session

WHO SHOULD OPERATE ON NEPHROBLASTOMAS? QUESTION STILL NOT SOLVED

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In 2004 we showed that the urologists' patients in our institution did best when compared with outside surgeons and academic, non-paediatric general surgeons. We revisited this scenario and looked at 36 new patients operated between 2003 and 2007. No outside surgeons performed any operations this time round. The paediatric surgeons performed 31 nephrectomies and the urologists 5. The spread between stages 1/2 and 3/4 was according to average for the surgeons (68/32%), but favoured the urologists (80/20%). Only 1 primary operation was done for a bleeding tumour (urology). The urologists had 1 relapse (20%) and no death (0%), while the surgeons had 2 relapses (6%) and 8 deaths (26%). Is it merely a question of who does most?

RESULTS OF THE TREATMENT OF CYSTIC PARTIALLY DIFFERENTIATED NEPHROBLASTOMA

A G Maharaj, G P Hadley

Background: Cystic partially differentiated nephroblastoma (CPDN) represents the benign end of the spectrum of Wilms tumour. It is a rare tumour with a good outcome. We have reviewed our management, with emphasis on the changing pattern thereof, and outcome in our environment.

Methods: We retrospectively analyzed the departmental database of solid tumours and identified the charts of patients treated for histologically proven CPDN.

Results and discussion: 13 patients operated on from 1986 to 2007 were entered into the study. They ranged in age from 10 to 48 months with an average of 22 months. There were five females and 8 males. Tumour weight ranged from 293g to 2500g with a mean of 1372g. Follow ranged from 1 month to 156 months with an average of 46 months. There were no documented recurrences. Three patients did not receive either chemotherapy or radiotherapy. These were treated after 1994 when awareness of the benign nature of the disease together with preoperative identification of the radiological features resulted in omission of preoperative or post operative chemo radiotherapy. These radiological features were those of a well defined cystic mass arising from the kidney with intervening septae and little other solid material.

9 patients had recorded morbidities. These ranged from leucopenia related to chemotherapy to sepsis and radiation enteritis resulting in death. Morbidities also included adhesive obstruction requiring a laparotomy as well as Chemotherapy drip infiltration requiring a skin flap. There were two mortalities prior to 1988 and these were thought to be related to chemo radiation.

Conclusion: CPDN is a rare variant of Wilms tumour that has low potential for recurrence and in our series a male preponderance. Morbidity and mortality related to chemo radiation which is now considered unnecessary in most patients.

NON-WILMS RENAL TUMOURS: AN IMPORTANT UNDERSTUDIED GROUP OF TUMOURS

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Wilms tumours remain one of the commonest tumours in South Africa but are characterized by considerable histological diversity in terms of cell types, organizational patterns and cellular differentiation. Non-Wilms renal tumours may have been under diagnosed in the past

Non Wilms tumours of the kidney are an uncommon but important subgroup of renal tumours in childhood [<10% of renal tumours and <1% of tumours of childhood] and are diagnosed histologically by exclusion. The most frequently encountered include renal clear cell sarcoma, rhabdoid tumor of the kidney, renal cell carcinoma, mesoblastic nephroma, and multilocular cystic nephroma.

We present 20 non-Wilms of 126 renal tumours (16%) <14 yrs presenting to Tygerberg Hospital. These included renal clear cell sarcoma (2), rhabdoid tumor of the kidney (1), renal cell carcinoma (2), mesoblastic nephroma (9; 3 atypical), multilocular cystic nephroma (3) ,1 highly undifferentiated renal tumour and 1 intra-renal neuroblastoma. Age ranged from 1 to 144 months and all were at atypical ages for Wilms tumour. The age of presentation appeared important in terms of prognosis. More malignant tumours occurred in older children (>5 yrs). All were treated surgically and by standard oncologic protocols.

Of the 10 malignant tumours 2 patients (20%) died (1 renal cell carcinoma; 1 rhabdoid tumour). 3 atypical mesoblastic nephroma did well on observation despite incomplete excision in 1.

This study draws attention to the non-Wilms tumours occurring in childhood and their special characteristics in terms of diagnosis and management and tumour response are evaluated.

PROFILE AND SURVIVAL RATES FOR PATIENTS WITH NEPHROBLASTOMA: A COMPARISON BETWEEN SIOP AND NWTS PROTOCOLS

D K Stones, J du Plessis, S Stannard

Introduction: As South Africa is a developing country the tumours we see are large compared to those in the developed world. In 1999 we decided to change from a primary operation based protocol (NWTS) to a protocol were most patients receive pre-operative chemotherapy (SIOP). We were already treating selected patients in the NWTS group with pre-operative chemotherapy.

Aim: To compare the profile and survival rates of patients treated on these protocols.

Method: It was an ongoing study of patients under 15 years of age diagnosed with nephroblastoma between January 1986 and June 2004. Patients were followed up for at least 6 months.

Results: A total of 163 patients were seen, 52 in the SIOP and 111 in the NWTS group. Sixteen patients, 6 in the SIOP, 10 in the NWTS group were excluded because they died soon after admission. The sex ratio and the median age were similar. In the SIOP group 44/46 received preoperative chemotherapy while 57/101 in NWTS group received pre-operative chemotherapy because of large tumours. There was a significant difference in the number of stage 1 and 3 patients between the two groups. There was no significant difference in the survival rates.

Conclusions: The patient profile was similar. There was a significant difference in the number of stage 1 patient; more in SIOP group (down staging) more stage 3 in NWTS group (bias because of size). Both protocols gave an event free survival of approximately 60% which is less than the protocols report. Both protocols can be used successfully in a developing country.

IS IT WORTHWHILE TREATING PATIENTS WITH STAGE 4 NEPHROBLASTOMA?

D K Stones, J du Plessis, S Stannard

Introduction: South Africa has limited resources and we need to select which cancer patients to treat. Nephroblastoma is common and generally has a good prognosis but stage 4 patients have a statistically poorer survival than the rest of the patients – is it worth while to treat them?

Aim: To determine the patient profile and survival of our stage 4 nephroblastoma patients and compare it to the literature and the rest of our patients with nephroblastoma.

Method: It was an ongoing study of patients under 15 years of age diagnosed with nephroblastoma between January 1986 and March 2006. Patients were followed up for at least 3 months.

Results: There were a total of 177 patients and stage 4 nephroblastoma accounted for 26% of them and for almost 40% of the deaths. There was no difference in the sex incidence, the laterality or the protocol used between the two groups. Metastases of the lung were seen in 20, liver in 13 while 7 patients had metastases in both organs. There was a statistically significant difference in event free as well as the overall survival rates but there was no significant difference between the different sites of the metastases.

Conclusions: The patient profile between the two groups is similar and as in other studies lung or liver metastases were most common. We have a higher incidence of stage 4 patients as well as a lower survival rate than other large overseas studies but our incidence and survival rates compare favourably to other South African centres.

Recommendations: Despite the significant difference in survival rates compared to overseas studies and the rest of our patients, the overall survival of stage 4 patients is almost 50%. Thus it is reasonable to treat our patients with stage 4 disease aggressively and to the best of our ability.

Conclusion: It appears that the stage of HIV disease does not influence the development or expression of allergy. The dermatitis and chronic rhinitis that is prevalent among the HIV infected patients is probably due to some to the factor as opposed to immune dysregulation.

QUALITY OF LIFE AND SYMPTOMS ASSESSMENT IN SUBLINGUAL IMMUNOTHERAPY FOR PATIENTS WITH HOUSE DUST MITE RELATED PERENNIAL RHINITIS: DEFINITION OF A RESPONDER PROFILE

Potter Paul, Nurse Barbara, Hawarden Diane, Combebias Anne, Fadel Riad, Baker Sheila, Fenemore Bartha, Terblanche Lindi

Allergy Diagnostic and Clinical Research Unit, University of Cape Town Lung Institute

Background: The efficacy and safety of sublingual immunotherapy (SLIT) is well-established in seasonal allergic rhinitis. However fewer data were available regarding perennial allergic rhinitis. The aim of this study was to assess the relationship between the effect of SLIT on symptoms and quality of life changes in patients with perennial rhinitis.

Methods: This exploratory phase IIIb, double-blind, placebo-controlled trial was conducted during years 2003 to 2005 in the Cape Town area (South Africa). Sixty patients with house dust mite induced allergic rhinitis were enrolled. Patients were randomized to receive, during 2 years, Dermatophagoides pteronyssinus SLIT solution (Stallergenes, Antony, France), with a maintenance dose of 300 IR (n=39), or placebo (n=21). The primary efficacy endpoint was the mean T5SS (Total Symptom Score for five symptoms: sneezing, runny nose, nasal congestion, ocular redness/itching/tearing, itchy nose/throat/ears. The limit percentage of improvement was fixed at 60% for the 'responders'. Rescue medication intake, individual symptom scores and quality of life (QoL-RQLQ) were assessed as secondary endpoints.

Results: The intention-to-treat (ITT) population included 55 patients (mean age: 32.93 yrs ±11.31). The mean T5SS change was lower for the 300 IR (-6.57) group compared to placebo (-5.02), but the difference did not reach significance. The mean percentage of days with rescue medication, as well as each individual symptom score, were lower for the 300 IR group compared to placebo. The percentage of good responders is significantly higher in the active group (p=0.0405). For the QoL, the percentage of good responders the ocular score is higher in the active group (p=0.0512), reaching significance. The association between good responders to QoL and good responders to T5SS gives an odds ration = 15 (p<0.0001) for all patients (OR = 8.75, p=0.0061) in active group) showing a good correlation. The most frequent adverse events were oral pruiritis and throat irritation. No related SAE occurred throughout the study.

Conclusions: This exploratory study gives promising results with a two-year treatment using Dermatophagoides pteronyssinus SLIT solution at a maintenance dose of 300 IR three times a week. A population of 'responders' may be defined as patients with an improvement of 60% or more of the clinical symptoms. QoL is strongly correlated to clinical symptoms.

Paul Potter and Di Hawarden served as investigators on the clinical trial for this report which was funded by Stallergenes, Antony, France.

Paediatric Neurology and Developmental Association of Southern Africa (PANDA)

A HEARING SCREENING PROGRAMME FOR NICU INFANTS AT KALAFONG HOSPITAL

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Universal newborn hearing screening is part of routine neonatal care in developed countries like the USA and UK where more than 95% of all babies born are screened before discharge from the hospital. Prevalence rates of permanent sensory and neural losses vary between 2 and 6 per 1000 live births. In countries where resources are limited a risk-based screening approach can serve an important intermediate step toward a universal programme. NICU babies comprise a significantly at risk population for both sensory and neural hearing loss. The prevalence of hearing loss in NICU babies is reported to be 20 to 30 times higher than for well babies and includes perinatal, progressive and late-onset losses. This study will report on the hearing screening programme for NICU infants at Kalafong hospital documented over a 4-year period (2004 - 2007). The screening protocol included both Automated Auditory Brainstem Response (AABR) and Oto-Acoustic Emission (OAE) tests and regular follow-up appointments. This report will present the incidence of hearing loss and compare the efficacy and cost-efficiency of different screening techniques and protocols employed. Recommendations will be made regarding optimal test and protocol selection for identifying hearing loss as soon as possible in this vulnerable group of infants.

INFANT-PARENT PSYCHOTHERAPY: A SHORT-TERM PSYCHOLOGICAL INTERVENTION MODEL FOR THE PSYCHODIAGNOSIS AND TREATMENT OF PAEDIATRIC SLEEP DISORDERS

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Paediatricians are for many mothers a first port-of-call when behavioural difficulties cause stress and anxiety in the families of infants and young children. Paediatricians who are knowledgeable about the more unconscious, psychological aspects of feeding, sleeping, excessive crying and other emotional issues can begin treatment in their consulting rooms. An effective, short-term, psychotherapeutic model employed by child psychologists for identifying and treating infant and young child sleep disorders is described. The five-session parent-infant or parent-young child psychotherapy model can inform paediatric interventions, both in terms of its emphasis on the history-taking process as a psychodiagnostic and therapeutic tool, and as a referral source in more stubborn or severe cases. This paper uses sleep disorders, which occur frequently in the paediatric population and represent the most common behavioural problem facing parents of infants and preschool children, to illustrate how parents, paediatricians and child psychologists can be partners in conceptualising and addressing the more psychological aspects of a presenting symptom. The effects of disordered sleep on a family can be profound; interfering with the quality of sleep and frightening and frustrating parents. There is a link between sleeping disorders in early childhood and later hyperactivity, conduct problems and learning difficulties. Sleep disturbances are strongly associated with difficulties in the parent-infant relationship. Such difficulties include: poor attachment and separation issues; feeding problems; excessive crying; absent or inconsistent limit-setting and poor maternal mental health, particularly postnatal depression which is strongly correlated with disturbed infant sleep. Routine investigation of sleep behaviour can contribute to an early identification of adverse psychosocial factors and can lead to an early and preventative intervention, focusing on the infant-parent relationship. The paper includes vignettes from case material that are used to illustrate key aspects of the model and the underlying psychological theory.

THE SILENT EPIDEMIC OF INFANT HEARING LOSS: CURRENT STATUS OF EARLY INTERVENTION IN SOUTH AFRICA

Peter Friedland, *Dept ENT Wits University; Donald Gordon;* **De Wet Swanepoel,** *Dept Communication Pathology, University of Pretoria;* **Claudine Storbeck,** *Centre for Deaf Studies, University of Witwatersrand;* **Suzanne Delport,** *Dept Paediatrics, University of Pretoria and Kalafong Hospital*

Infant hearing loss is the most common occurring congenital sensory disorder with a reported prevalence of between 2 to 6 per 1000 live births which doubles by 9 years of age. It has been referred to as the silent epidemic of developing countries where a higher incidence is expected due to poorer health and socioeconomic conditions.

Late detection of infant hearing loss severely compromises development of speech, language and cognitive skills essential for optimal development in stark contrast to the established benefits of initiating intervention before 6 months of age. Benefits include linguistic, speech and cognitive development that is comparable to normal hearing peers affording children the opportunity to develop to their maximum potential and to become active participants and contributing members of their communities.

These facts have led to universal newborn hearing screening and intervention programmes becoming standard of care in developed countries, where countries like the USA and UK screen 95% of all newborns using highly accurate physiological techniques.

Unfortunately initial detection of hearing loss in South Africa is still primarily passive as a result of parental concern about observed speech and language delays, unusual behaviour or the complications of otitis media. This report will present prevalence data for newborn hearing loss in South Africa and survey the current status of early detection, diagnosis and intervention services in the country.

The importance of a multidisciplinary approach for promoting the development of widespread early detection and intervention programmes will also be presented as the way forward.

THE CHANGING PROFILE OF AUTISM IN A CLINIC FOR CHILDREN WITH DEVELOPMENTAL DELAY: A TEN YEAR SURVEY

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Introduction: The purpose of the study was to test the hypothesis that there is an increasing number of children with features of the Autistic Spectrum disorder presenting to clinicians. A secondary objective was to further examine the characteristics of such children and compare the characteristics of this cohort to that of a control group and to studies done in the developed world.

Method: The study was done in a clinic for children with developmental delay and physical disabilities. All the files for children seen between 1996 and 2005 were selected. The first 1218 (47%) of a potential 2598 files were examined and the information inserted into a database. The information on children with ASD was extracted and further examined. The children seen over the same period without ASD were used as a control group. The study group was compared to both the control group and to studies done elsewhere.

Results: The study showed an 8.2% increase in the numbers of children presenting with featured of ASD over the past 10-year period. The characteristics of the cohort with autism were further explored and show an interesting trend which will be explored.

Conclusion: Resources need to be allocated for early intervention and education of these children.

CEREBRAL PALSY AS A CONSEQUENCE OF SHAKEN BABY SYNDROME

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Shaken Baby Syndrome (SBS) has been recognised in literature since Caffey in 1946 and results from the violent shaking of infants by caregivers. Intracranial injuries may include haemorrhages, oedema, increased intracranial pressure, anoxia, diffuse axonal injury and atrophy. In addition to the intracranial injuries, diagnosis may include retinal bleeding and a range of fractures in the infant. Despite a vast amount of literature on SBS, major documented controversies exist. These range from disputing the existence of the syndrome and the mechanism of injury to the severity and number of symptoms that need to be present for a definitive diagnosis. While there are various differential diagnoses for the injuries in isolation, SBS is seen as a syndrome resulting in a wide spectrum of acute and chronic symptoms, injuries and prognoses. A review of the literature shows that SBS exists but its true prevalence is unknown, as it is often misdiagnosed, over or under diagnosed. The result is perilous as SBS may cause varying degrees of disability or death. The aim of this paper is to describe the injuries and their mechanism.

DOWN SYNDROME: A JOURNEY NOT A DESTINATION!

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Introduction and objective: Down syndrome is synonym with different constraints, physical, sensory, learning, cognitive, language, social and environmental. The reality is that a child with Down syndrome may experience all of these constraints. As with any journey, especially in South Africa, there are a lot of potholes, but fortunately there could be 'hole in ones' too as part of the solution.

The value of early intervention and appropriate stimulation is universally accepted. Developmentally delayed and handicapped children and their parents need help, starting as soon as possible after the diagnosis is made.

The START (Strive Towards Achieving Results Together) and PLAY FUN stimulation programs are home-based, early intervention therapies designed, to utilize aids/utensils commonly available in a household in order to stimulate normal neuro-developmental skills.

The aim of this study is to access the effectiveness of these specific programs and the sustainability in children with Down syndrome.

Materials and methods: All infants and children identified as having Down syndrome at the Bloemfontein clinical genetics clinics are referred for therapy, including the START and PLAY FUN stimulation programs, at the time of first contact.

Results: The value of early identification, intervention and parent co-operation will be demonstrated. Early referral and diagnosis were integral with onset of the appropriate stimulation. Infants who initiate these programs before 6 weeks of age demonstrate a markedly better neuro-developmental outcome than the children starting after 3 months of age.

Conclusion: Working in collaboration with parents, referring doctors and the Down Syndrome Association is a pre-requisite for intervention in children with Down syndrome. These programs are beneficial, but the onset of the journey with the devoted parents should begin before 6 weeks of age.

ATTENTION DEFICIT HYPERACTIVITY AND OPPOSITIONAL DEFIANCE DISORDER IN HIV-INFECTED SOUTH AFRICAN CHILDREN

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Objective: To determine the prevalence of attention deficit-hyperactivity disorder (ADHD) and oppositional defiance disorder (ODD) in HIV-infected South African children.

Methods: The medical records of 100 consecutive HIV infected children attending the infectious disease clinic at Tygerberg hospital were retrospectively reviewed over a 3-month period. Parent and teacher Swanson, Nolan and Pelham (SNAP) IV questionnaires were used to determine ADHD and ODD severity and the Good-enough-draw-a- man test was used to estimate non-verbal intelligence. Associations between behavioural subtypes, non verbal intelligence, demographic and health variables were investigated.

Results: The SNAP IV parent questionnaires showed a 26% prevalence of ADHD inattentive type, 38% hyperactive type and 24% combined type. In contrast the teacher questionnaires demonstrated a much lower prevalence; 4.8% for both inattentive and hyperactive ADHD and 7.1% for the combined type. The prevalence of ODD was 12% on parent questionnaires and 9.5% on teacher's questionnaires. Combined parent-teacher ADHD and ODD scores were less than 5% in all categories due to poor parent-teacher correlation and low teacher scores. The non-verbal mean developmental quotient (DQ) of the study population was 87 and 18% of the patients were intellectually disabled (DQ less than 70). No association was noted between behavioural problems and immune markers such as viral load, CD4 count and percentage. Children with low CD4 counts and percentage generally had lower non-verbal intelligence scores. The severity of inattentive- and combined ADHD increased with duration of antiretroviral therapy according to parental questionnaires.

Conclusions: Parents-only SNAP IV questionnaires indicate a high prevalence of significant ADHD (all subtypes) and ODD in HIV-infected children. Non verbal intelligence scores were lower in children with low CD4 percentages and counts. The SNAP IV questionnaires and Good-enough-draw-a-man test were easy to perform and may prove valuable screening tools in HIV children with behavioural and/or scholastic concerns.

South African Association of Paediatric Surgeons (SAAPS)

NEONATAL PYOGENIC LIVER ABSCESS: A RARE BUT IMPORTANT DIAGNOSIS IN PERSISTENT SEPTICAEMIA

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Introduction: Pyogenic liver abscess is a rare entity, although some significant large series have been reported locally. In neonates, prematurity, persistent septicaemia proven on blood cultures but not responding to antibiotic treatment, ascending infection from umbilical vein catheterization, portal vein spread from the gut in NEC, central parenteral nutrition catheter lines and hypertonic glucose infusions are all associated with this rare entity. Due to diagnostic difficulties presented by this condition, we would like to highlight the importance of the condition especially in premature neonates with misplaced umbilical lines and those receiving parenteral nutrition by presenting the clinical and radiological findings in 5 neonates.

Methods: Retrospective chart review of 6 patients diagnosed with hepatic abscesses between 2000-2006 including clinical and radiological features and evaluation of potential risk factors.

Results: All 5 neonates had a low birth weight and gestational age (range 30-34 weeks). Raised CRP levels and positive blood cultures persisted in

all cases. The causative organisms were Klebsiella (3) and staphlococcus (3) – one a MRSA. Enterobacter was also cultured from 1. In one patient the diagnosis of tuberculosis was eventually made. In 2 patients there was an association with a misplaced umbilical line and a high concentration glucose infusion. Ultrasound (US) proved a reliable method of diagnosis and 4 were right sided and 1 central in position. Some difficulty in interpreting resolving abscesses was encountered on US follow-up. Trans-diaphragmatic spread occurred in one necessitating thoracoscopic decortication. Interventional aspirational drainage was required in 4/5 patients which proceeded to open drainage in 2.

Conclusion: While rare in neonates, pyogenic hepatic abscesses should be considered in premature neonates with ongoing septicaemia, especially in the context of misplaced central and umbilical catheters. This review draws attention to the risk factors pointing to this infrequent complication and discusses diagnosis and management.

THE LONG-TERM OUTCOME IN SURGICALLY-MANAGED NEC IN A DEVELOPING COUNTRY – A HIGH MORBIDITY

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Introduction: Necrotizing enterocolitis (NEC) is a condition affecting premature babies which has a high early morbidity and mortality. Recent studies have also shown an increased risk of long-term complications in surgically-managed NEC compared to other premature neonates, including a higher rate of overall infections with a high associated morbidity and mortality and impaired growth and neurodevelopmental outcomes.

Methods: Retrospective review of records of 108 premature neonates with surgical NEC treated at Tygerberg Hospital from 1992 to 1995 and analysis of problems at follow-up.

Results: In 128 premature babies with surgically treated NEC, mean gestational age was 32 weeks and average birth weight was 1413g. Early (>30 day) survival was 69% (n=88). Amongst those <1500g birth weight (n=68; 53%) there was a 64% early survival rate. Deaths mostly occurred from overwhelming sepsis or disseminated intravascular coagulopathy during post-operative course (n=16) or pan-intestinal necrosis (n=18). There were 10 late deaths (> 30 days) of which 4 were due to short bowel syndrome, 2 cardiac disease and 4 from sepsis. 30 patients were lost to follow-up and 48 were followed-up, of which 29 for more than 2 years. Late surgical complications included late colonic strictures (9), incisional hernias (2) and adhesive obstruction (3). Of the long term survivors, 8 (27.5%) had documented or suspected neurological damage, of which 50%with suspected hypotonia improved by 2 years. 3 had documented severe auditory deficits.

Conclusions: The impact of NEC on premature infants is considerable in terms of morbidity and mortality. A number do well but septic complications may be ongoing and recurrent. Due to high risk of neurodevelopmental and other problems, patients should be "flagged" on discharge for careful follow up.

VIDEO PRESENTATION: LAPAROSCOPIC RESECTION OF UPPER POLE IN RENAL DUPLICATION

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Introduction: Excision of an upper pole moiety in a duplex kidney is widely reported in the literature. Fairly large flank incisions are required for it complete excision and this can be reduced in size considerably if minimally invasive techniques are employed. A video of this particular approach is presented.

Case report: A 13-year old boy with intermittent left flank pain and known left renal duplication on CT-IVP was admitted to Johannesburg General Hospital for an elective upper pole hemi-nephrectomy. A retroperitoneal space was created by blunt dissection by a 5 mm trocar inserted just below the 12th rib. A further two 5 mm trocars were inserted in a standard triangle manner. Dissection was carried out by a curve Ligature forceps, the upper pole opened and once visualisation of the upper pole pelvis was possible, this was excised with the surrounding parenchyma. This was removed after enlarging the first port site to about 15mm. The remnant renal tissue was approximated with 4-0 monofilament sutured and the wound closed with a drain in-situ. The child recovered rapidly and discharged the next day.

VIDEO PRESENTATION: LAPAROSCOPIC TREATMENT OF SPONTANEOUS COMMON BILE DUCT PERFORATION

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Introduction: Spontaneous common bile duct (CBD) perforation in infants is a rare but well described condition. Although no consensus exists as to the etiology of the condition, there is evidence as to mal-union of the CBD and pancreatic similar to that described for choledochal cyst. Indeed this condition may be a perforation of type II choledochal cyst.

However the treatment is somewhat easier than choledochal cyst variant, as most cases will resolve with simple drainage alone. If subsequent study confirms a distal obstruction further surgery may be necessary but this accounts for only 10% of cases.

Laparoscopic treatment for this condition has not been described to our knowledge, and we would like to present a video of this procedure.

Material: A 4-month-old boy with obstructive jaundiced was referred to our unit with radiological diagnosis of CBD perforation. He underwent a 3-port laparoscopy, bile- stained ascites was washout and after gentle blunt dissection and mobilization of the gall bladder, a small perforation of the CBD distal to cystic duct identified. A rigid drain was placed near the perforation and abdomen closed. The procedure time was 72 minutes. Parental nutrition was commenced immediately and after 8 days he passed pigmented stool. The drain was withdrawn and feeding started. He was discharged post-operative day 14. He had a subsequent MRCP 3 months later that did not show a CBD stricture. He is now 6 months post surgery and growing well with no complications.

Conclusion: Spontaneous CBD perforation is a rare but easily treatable condition by an experienced laparoscopist.

VIDEO PRESENTATION: LAPAROSCOPIC MANAGEMENT OF PEG RELATED COLO-CUTANEOUS FISTULA

B Banieghbal

Department of Pediatric Surgery, Johannesburg General Hospital, Johannesburg

Introduction: Gastro-colic and colo-cutaneous fistulae are rare but well described complication of percutaneous endoscopic gastrostomy (PEG) placement.

Traditional treatment consists of PEG removal to allow for delayed spontaneous closure and a further PEG placement at a later stage. A one stage laparoscopic treatment is not reported in literature to our knowledge.

Case report: A 6-year old boy with cerebral palsy underwent PEG placement in Nov 2007. He was re-admitted 2 weeks later with signs of sepsis but responded to intravenous antibiotics. A contrast study, at that time through the PEG, did not show any fistulae. Three months later, he was re-admitted for an obvious colo-cutaneous fistula which was confirmed on Ba-enema. He was taken to operating theatre and a 3-port laparoscopic exploration performed. A colo-cutanous and a small gastro-colic fistulae was identified. Colonic injuries were oversewn and covered with an omental patch. The existing gastrostomy was used for a placement of Foley- type gastrostomy tube. He was given parental nutrition for 5 days and he recovered uneventfully and discharged home 7 days later.

VIDEO PRESENTATION: THORACOSCOPIC MANAGEMENT OF SYMPTOMATIC BILATERAL HYDATID CYSTS

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Introduction: Hydatid disease of the lung is relatively rare in most regions of South Africa. Surgical resection is the common practice for cure. Bilateral disease is more difficult to manage as bilateral thoracotomies on different occasions are needed or the resections are done by median sternotomy. We would like to present a bilateral hydatid case which was managed by thoracoscopic approach.

Case report: A 9-year old boy was admitted to Johannesburg General Hospital with possible diagnosis of hydatid cyst affecting both upper lobes. Both cysts had air-fluid level suggestive of rupture into upper lobe bronchi. CT of liver did not show any liver cysts. On the left side, erroneously, an attempt had been made at percutaneous intercostals drainage by the doctors from the referring hospital. A sternotomy and surgical excision were planned for him but after further discussion, a bilateral thorcospic injection of absolute alcohol into the cysts was made. 10 minutes later the cysts were opened and the daughter cyst removed. Chest drains were placed and hypertonic saline was left in both thoracic

cavities for one hour after surgery. He was kept overnight in an intensive care unit, he continued to have significant broncho- pleural fistulae with a further admission to ICU 3 days later, for a further 2 days. His intercostals drainage eventually ceased by day 12 postoperatively. He was discharged day 14. He continues to have a small right- sided residual cyst, still visible on the CXR, on the 6-month follow-up.

VIDEO PRESENTATION: LAPAROSCOPIC MANAGEMENT OF SYMPTOMATIC MECKEL'S DIVERTICULUM

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Introduction: Meckel's diverticulum is relatively uncommon condition, but it is a well recognized cause of rectal bleeding in children under 2 years of age. Laparoscopic excision has now replaced open surgery as the gold standard. A video of this operation is presented here.

Case report: An 18-month-old boy was admitted to Johannesburg General Hospital with 2 episodes of significant rectal bleeding requiring blood transfusions. A colonoscopy by an adult surgeon elsewhere contributed this to a pan-colitis. However, in view of massive rectal bleeding, a Meckel isotope scan was requested and performed by radiologists. This was conclusive of gastric mucosa in right iliac fossa, strongly suggestive of a Meckel's diverticulum.

A standard 3-port laparoscopic exploration was carried out, and a large Meckel diverticulum was found with severe thickening of its base, making it unsuitable for staple excision. The diverticulum was mobilized with diathermy dissection and delivered outside the abdomen, through an enlarged umbilical port site.

Standard small bowel resection and anastomosis were carried out and the bowel returned to abdominal cavity. The child recovered rapidly and was discharged 3 days later.

RAPID CORRECTION OF SEVERE METABOLIC ALKALOSIS IN HYPERTROPHIC PYLORIC STENOSIS WITH INTRA-VENOUS CIMETIDINE

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Aim of study: The definite treatment of hypertrophic pyloric stenosis (HPS) is pyloromyotomy as described by Ramstedt. In most instances of HPS, there is mild metabolic alkalosis, requiring intravenous fluid resuscitation with 5% dextrose/saline for 1-2 days. However, in some instances there is a delay in diagnosis and as the result the alkalosis is severe. In these cases a much longer resuscitation period of 4-6 days is needed to normalize the serum pH.

Metabolic alkalosis of HPS is due to excessive vomiting of HCl and therefore if it is possible to reduce HCl production, serum pH may normalized faster.

We would like to report on use of intravenous cimetidine (CM) in these cases. To our knowledge, there is no previous report of this technique in the medical literature.

Material: Over a 2-year period; 23 cases of HPS were managed by the author. 12 children (aged 5-8 weeks) with HPS had arterial pH over 7.65. In this sub-group, 4 were treated with standard resuscitation protocols for 3 days prior to intravenous CM, and 8 cases had immediate use of CM. CM was given at 15mg/kg twice daily for 24 hours.

Result: In all 12 cases, pH reduced below 7.5 after 24 hours of CM treatment, thus allowing for pyloromyotomy the next day. The patients were commence on oral feeds the following day and were discharged at 1-2 post-operative days. There was no complication due to use of CM or as the result of surgery.

Conclusion: In this small series of patients with HPS, it appears that a severe metabolic alkalosis can be normalised rapidly with the usage of intra-venous CM. Pyloromyotomy can be performed earlier, thus saving hospital costs by allowing earlier discharge. Proton-pump inhibitors are an alternative but their higher cost takes away some of the cost advantage of CM use.

LIPOMATOUS BENIGN TUMOURS IN CHILDHOOD

P Beale, D Vieten

The presentation of these mass lesions gives rise to alarm. Their radiological appearance and ultimate histology however are reassuring as to their benign nature.

5 cases of lipoblastoma in childhood presenting in a variety of anatomical sites are presented and the histology and benign nature discussed.

Two other cases of intrathoracic lipomatous lesions mimicking more ominous conditions

- Mediastinal lipoma
- Pleural based lipoma

are described.

NEONATAL + INFANT PRESACRAL LESIONS – NOT ALL SACROCOCCYGEAL TERATOMAS

P Beale

Although the foremost diagnostic consideration in a neonate or small infant with a presacral cystic mass would be an Altmen type IV, sacro-coccygeal teratoma, there are other possibilities.

These lesions may present with – pelvic outlet obstruction.

- Anorectal compression and obstruction
- Anal stenosis or absence as in the Curarino syndrome
- Distal urinary obstruction either at the urethral level or the distal ureteric level.

The surgeon may be faced with the decision whether to operate either transabdominally or by a sacrococcygeal access or both.

The histology may reveal unexpected revelation.

As in our experience:

1 x presacral meningocoele

1 x cystic neuroblastoma

Clinical presentation and pre op radiology may differentiate retrorectal cystic hamartoma as in 2 cases.

PAROTIDECTOMY IN THE PAEDIATRIC AGE GROUP

P Beale, V Gentilino

Parotidectomy may be indicated for both benign and malignant disease in childhood. The indications for this procedure in our experience are reviewed.

These included:

- lymangioma = cystic hygroma
- Parotid malignancies

  Adenocystic carcinoma

  MUCO epidermoid – carcinoma
- Chronic inflammatory parotitis

Including Lymphoepithelial proliferative parotitis associated with HIV infection with breakdown and with large cystic cavitation.

Uni or bilateral parotid enlargement is the most easily recognizable stigma of HIV infection and that these young patients endure the embarrassment of repeated questioning as to the nature of their disfigurement.

This resulted in school refusal by one adolescent. These patients were very grateful for superficial parotidectomy. Irrespective of the indication for parotidectomy, it is obligatory to identify and preserve the main trunk and radicles of the facial nerve.

OPEN EXTRAPLEURAL OESOPHAGEAL ATRESIA REPAIR – STILL THE GOLD STANDARD

P Beale

Review of an individual experience of oesophageal atresia repair reveals results and benefits not easily exceeded by thoracoscopic OA TOF repair.

These include a skin incision of 2.5 - 3.5cms in the skin lines below the angle of the right scapula. A non muscle cutting extra pleural thoracotomy. Routine use of a head light and surgical loupes.

- The option of utilizing techniques for approximating the oesophagus in wide gap cases. A leak rate under 2%. A recurrent fistula rate of less than 1%.
- A recent consecutive run of 130 cases without leak.
- The option of early feeding as soon as the baby swallows its own saliva – often from the post op third day limiting NNICU and in hospital stay.
- Operating time of 60 to 90 minutes.

Intrapleural thorascopic repair introduces a risk factor and prolongs operating time without any benefits above the described technique which results in a very inconspicuous limited scar.