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Necrotising enterocolitis

A1
Towards consensus in necrotising enterocolitis: areas of agreement amongst Dutch pediatric surgeons

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Background. Necrotising enterocolitis (NEC) is the most common gastrointestinal emergency in neonates and remains a leading cause of mortality in neonatal intensive care units. Despite this, definitive evidence based literature is lacking in several key areas of diagnosis and treatment. We sought expert consensus amongst pediatric surgeons in the Netherlands concerning the definition, diagnosis, conservative treatment, surgical treatment, and post-operative care of NEC.

Methods. After reviewing the available best evidence, a questionnaire asking about various aspects of the management of NEC was created and distributed electronically to the six specialist pediatric surgical centres in the Netherlands. Results were collated and a consensus meeting subsequently conducted in January 2006. Where available, the evidence-based literature was reviewed; in its absence, expert consensus was reached.

Results. The response rate to the questionnaire was 100% and at least one representative from each centre was present at the consensus meeting. All recommendations were "expert consensus" unless otherwise stated. Consensus could be reached about the definition of NEC, the use of standardized feeding regimes (meta-analysis) and breast milk feeding for prevention (large randomized controlled trials), the combined role of clinical acumen, biochemical and haematological data and radiology in diagnosis, essential principles of conservative management, the appropriate stage for transfer of care, absolute and relative indications for surgery, the choice of procedures at laparotomy with varying degrees of surgical NEC, and the indications for primary peritoneal drainage (retrospective analyses).

Conclusions. The meeting was successful with expert consensus reached and recommendations made on a number of issues. However, the majority of recommendations are based solely on the lowest form of recognized evidence: expert consensus. Indeed, there is a lack of well-designed, high-powered clinical trials upon which recommendations for the surgical management of NEC can be made. The content of the consensus statements will be presented and further discussion invited.

A2
Towards consensus in necrotising enterocolitis: areas of divergence amongst Dutch pediatric surgeons

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Background. Necrotising enterocolitis (NEC) is the most common gastrointestinal emergency in neonates and remains

a leading cause of mortality in neonatal intensive care units. However, definitive evidence based literature is lacking in several key areas of diagnosis and treatment. Despite seeking expert agreement about the definition, diagnosis, conservative treatment and surgical treatment of NEC, consensus could not be reached in a number of key areas of management.

Methods. After reviewing the available best evidence, a questionnaire asking about various aspects of the management of NEC was created and distributed electronically to the six specialist pediatric surgical centres in the Netherlands. Results were collated and a consensus meeting covering these specific topics was subsequently conducted in January 2006. Where available, the evidence-based literature was reviewed.

Results. The response rate to the questionnaire was 100% and at least one representative from each centre was present at the consensus meeting. We were unable to reach consensus about the inclusion of isolated gastro-intestinal perforations under the definition of NEC, reflecting conflicting evidence based literature (level C, prospective trials, and level D, retrospective analyses). While the importance of radiology was evident, the frequency and duration of abdominal x-ray could not be agreed upon despite previous expert recommendations and some limited prospective trial data. Whether clinical deterioration despite maximal medical therapy constituted an absolute indication for surgery remained controversial. Whilst there was agreement about the indications for primary peritoneal drainage in the ELBW neonate with intestinal perforation, consensus could not be reached regarding the need for, timing of or indications for subsequent laparotomy. Most notably, there was considerable divergence in post operative care regarding the duration of antibiotics (no evidence available) and re-initiation of feeding (one small prospective study available) amongst the Dutch centres.

Conclusions. There are many areas of divergence in expert opinion about the best management of NEC. This particular pattern of divergence mirrored a similar audit conducted in the UK. This highlights the paucity of good evidence based literature and suggests areas in need of well-designed clinical research.

A3

Towards consensus in necrotising enterocolitis: areas highlighted for future research

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Background. Despite its prominence within the neonatal intensive care units worldwide, necrotising enterocolitis (NEC) remains incompletely understood. Well-conducted clinical research is lacking in a number of areas, and certain principles of diagnosis and treatment varied even within the small geographical location that is the Netherlands. Here we describe areas in need in of clinical research that were highlighted at the recent Dutch consensus meeting, with particular focus upon those topics where multi-centre co-operation would be advantageous.

Methods. Following a review of the recent literature, a questionnaire about the diagnosis and treatment of NEC was created in late 2005, and distributed electronically amongst the six specialist pediatric services in the Netherlands. Parts of the questionnaire were designed specifically to highlight predicted areas of clinical divergence. The results were collated and discussed at a consensus meeting in January 2006. Areas in need of research were identified when consensus could not be reached, and the evidence was either lacking or insufficient to support one practice over another.

Results. Four areas were identified as being best answered by prospective, multi-centre clinical trials. Firstly there was conflicting opinion and evidence based literature about whether isolated gastrointestinal perforation represents one end of the NEC disease 'spectrum', or indeed if such a distinction necessitates differing clinical management strategies. Secondly, despite two small series advocating the safety of primary anastomoses in the management of NEC, there is a lack of high power evidence to recommend the routine use of this approach in appropriate cases. This is made even more pertinent in light of recent unpublished data showing extremely high morbidity associated with stomas in neonates. The final two topics concerned aspects of post-operative management: the duration of antibiotic cover, and the appropriate timing and strategy for re-initiation of feeding after the diagnosis of NEC. These topics drew the widest range of answers from our questionnaire (5-14+ days for both), and yet have both been poorly researched to date.

Conclusions. There are a relatively small number of trials where well designed, prospective and adequately powered trials on the surgical management of NEC have been conducted. We have identified four areas in need of clinical that which would most benefit from multi-centre clinical research so as to increase sample size, the speed at which a valid clinical answer could be achieved, or both. We invite the audience to seriously consider participation in such research.

A4

Incidence of necrotizing enterocolitis in premature babies < 1500 g at the University Children's Hospital Vienna

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Background. The neonatal regional tertiary care centre of the University of Vienna has been a member of the Vermont Oxford Neonatal Network since 1994. The aim of our study was to investigate whether the introduction of a standardized feeding regimen in 2000 had an influence on the incidence of NEC.

Methods. Between 1994 and 2004, 1650 patients < 1500 g were admitted to our centre with a mean birth weight of 1022 g (range from 967 g to 1076 g) and a mean gestational age of 28 weeks (range 22 to 35 weeks). NEC was defined clinically and radiographically according to the criteria of the Vermont Oxford Neonatal Network. Since 2000 patients with focal gastrointestinal perforation were separately documented in our centre and in the network.

Results. Between 1994 and 1999 the incidence of NEC varied between 3% and 8% in our centre and was comparable in the network (6%). Between 2000 and 2004 NEC incidence declined to 1–4% in our centre while it remained unchanged in the network (5–6%). The incidence of focal intestinal perforation was 2–3% in our centre, similar to the network. Surgical intervention for NEC and gastrointestinal perforation was performed in 2/3 of our patients and in half of the patients in the network.

Conclusions. Introduction of a standardized feeding regimen was associated with a decline in NEC incidence. This needs to be confirmed in a large prospective multi-centre trial.

A5
A 5-year review of surgical procedures and outcome in 37 patients with necrotizing enterocolitis

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Background. Necrotizing enterocolitis is the leading cause of mortality and morbidity in neonatal intensive care units. Indication, timing of surgery and surgical procedures still remains a matter of discussion.

Methods. We performed a retrospective analysis of 37 children with necrotizing enterocolitis who underwent surgical procedures between 2000 and 2005 at our department.

Results. Twenty-eight of the surgical patients presented as very low birth weight infants with less than 1000 g body weight, five children with more than 1000 g and only four children were operated with more than 1500 g (range from 480 to 2830 g; median: 795 g). The median gestational age was 25 weeks (range: 23 to 40 weeks). We performed primary laparotomy and enterostomy in thirty three (89%) infants. Bowel-resection and primary anastomosis because of severe necrosis was carried out in two cases (5%). Severe inflammation and near-perforation bowel were found in only two (5%) patients. Peritoneal drainage and salvage laparotomy was carried out in one child. Seventeen children (45.9%) died in this patients' collective.

Conclusions. In this series most of the operated infants belong to the group of extremely low birth weight patients. The preferred surgical management in these patients is still under discussion. The decision about the adequate surgical procedure was mainly influenced by the degree of intestinal necrosis, preoperative known comorbidity and the clinical state of the patient.

A6
Necrotising enterocolitis – our 5-year experience

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Background. Pathogenesis of necrotising enterocolitis is not yet clear. Although the number of children affected by this condition is still relatively low, necrotising enterocolitis is one of the most common emergencies in neonatal abdominal surgery.

Methods. Treatment of necrotising enterocolitis is probably one of the biggest challenges for every neonatal surgeon. Not all the children require surgery, and on the other hand, there are so many options for surgical treatment ranging from peritoneal lavage to bowel resection and anastomosis.

Results. In a 5 year period (from year 2000–2005), in Children's Hospital Zagreb, we treated 72 children with necrotising enterocolitis (38 female and 34 male). 55 of them (76%) were treated with conservative methods. Average gestational age was 33 weeks, and average birth weight was 1,988 g. The smallest baby weighted 700 g, and the biggest 4,100 g. The average age of diagnosis was 6 days. Ten of the children who were treated conservatively, died (18.1%). In 17 children (23%) we performed surgery. First procedure was peritoneal lavage for 4 children who weighted under 1,000 g. In 3 cases we performed resection and primary anastomosis, and in 10 resection with enterostomy. Four of the operated children died (23.5%).

Conclusions. The number of children who need surgery for necrotising enterocolitis is diminishing, thanks to the modern, conservative treatment options. However, a lot of them treated conservatively end up in a hands of a pediatric surgeon because of the complications. Although the last couple of years mortality is lower than before, it's still quite high (25–40%).The highest is in children who weight under 1,000 g (35–50%). In the terms of mortality and final outcome, our results are similar to those described by other authors.

A7
Neonatal necrotizing enterocolitis in our practice

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Background. Necrotizing enterocolitis (NEC) is one of the most common acquired intraabdominal emergency among newborns and it mainly affects those in intensive care units. The reported incidence is approximately 1–3 cases per 1000 live births and mortality rates in established cases range between 20% and 40%. The aetiology of the disease has been reported to be multifactorial and no single aetiological factor seems to explain neonatal necrotizing enterocolitis but the main risk factors associated with NEC are: low birth weight, low gestational age, low Apgar score, perinatal complications, hyaline membrane disease, and umbilical catheterisation. Most study suggested that NEC is considerably more common in formula-fed newborns than in those receiving human milk.

Methods. This study was introduced to establish some update on epidemiology, etiology, diagnosis and treatment of necrotizing enterocolitis and to determine whether early diet can influence its onset and severity. Data concerning risk factors for NEC were obtained from obstetric and patient records including maternal history and medications, birth history, the infants gestational age, birth weight, sex, feeding history, gastrointestinal symptoms, clinical and radiological finding with NEC, laboratory measurements (blood test included hemoglobin, hematocrit, platelet and leukocyte counts) glucose, sodium and potassium levels, outcome and duration of hospital stay.

Results. Eight cases were reported during the last twelve months in our neonatal intensive care unit. Two of them was been born via cesarean section and four were reconsidered not to be true cases of NEC. The most common presenting clinical features for confirmed cases were abdominal distension (75% in suspected cases), abdominal tenderness (55%), pneumatosis intestinalis (55%), and blood in stool (41%). A complete blood count is included in the standard diagnostic blood test. Anemia is observed concomitant with NEC. Infants with severe NEC develop thrombocytopenia and that often predict the presence of a gangrenous bowel. Hyperglycemia is common in infants with NEC and that is associated with a higher mortality rate. In our study in infants with NEC the most dominant bacterial and fungal microbes cultured from stools were Staphylococcus species, Enterococcus faecalis, Enterococcus species, and Candida Albicans. Most of the infants had low birth weight – median weight 1,395 g (range between 1,211–2,699 g). Overall 62% of reported cases were male (5/8) and 38% female (3/8). Only one of our confirmed cases required surgery. In our study overall mortality rate was 13% (1/8). Treatment includes a regimen of bowel rest gstric decompression by orogastric tube, systemic antibiotics and parenteral nutrition. First line broad spectrum antibiotics consist of ampicillin, gentamycin and clindamycin.

Conclusions. NEC mainly affects infants in ICU and male and female infants have been found to be equally affected. Full term neonates account for only 25% of all cases of NEC (2/8). Age of onset was significantly correlated with gestation, demonstrating that the late onset of NEC was more common in the infants with low gestation. Perhaps this could relate to the earlier use of enteral feeds at higher gestation. Decreasing platelet count and increasing blood glucose level and leukocyte value more than 30×10^9 on several successive days might predict developing NEC. The study provides further support for the protective role of breast milk in the development of NEC, with reduced disease severity in the human milk-fed group.

A8
Prolonged bowel inflammation results in post-NEC intestinal

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Background. Formation of intestinal strictures is a well documented complication after recovery from acute necrotizing enterocolitis (NEC). Strictures result from healing of an area of severe ischemic injury and their extents are determined by the amount of wall involved in the necrotic process and the richness of the blood supply of that segment. The incidence of strictures after NEC is increasing as the mortality rate from the disease decreases.

Methods. We retrospectively analysed two cases of post-NEC intestinal strictures operated last year in our department.

Results. We analysed 2 cases of post-NEC intestinal strictures in neonates born spontaneously in 34 week g.a. (b.w. – f. 2100 g and 91 Apgar; m. 2900 g and 6183 Apg). Clinical signs of suspected NEC appeared after 3–5 days of breast feeding and blood levels of CRP at that moment were normal. Despite treatment, the CRP levels of the boy increased and the vital signs deteriorated – after ventilation for 5 days and improvement the oral feeding was started. After 12 days of conservative treatment the girl was feeded orally; with still increased blood CRP level. The following month both neonates were treated in Pediatric Department, where recurrent periods of poor feeding, distension, subfebrile and higher CRP levels were observed despite broad-spectrum antibiotic. Because of ileus the left colonic and jejunal obstructed segments were resected at the end of 2nd month of life.

Conclusions. We conclude that prolonged and periodically exacerbated intestinal inflammation (in our patients probably induced by too early introduced enteral feeding), coexisting with chronic insufficient blood oxigenation can promote bowel stricture formation. The blood serum CRP levels can be useful for monitoring of bowel inflammation status and helpful in decision of starting enteral feeding in patients suffering from NEC.

A9
Acquired forms of intestinal innervation disorders exemplified in necrotizing enterocolitis

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Background. Histopathologic studies of intestinal innervation disorders have focused mainly on congenital forms (e.g. Hirschsprung's disease), but rarely considered acquired defects of the enteric nervous system (ENS). However, the ENS is susceptible also to postnatal alterations induced by either toxic, inflammatory or ischemic events. The aim of the study was to characterize both structural and functional damages of the ENS subsequent to acute and chronic necrotizing enterocolitis (NEC).

Methods. Cryosections and wholemount preparations were obtained from intestinal fullthickness specimens of patients with acute and post-acute NEC. Histopathologic alterations of the ENS were studied by immunohistochemical methods including the assessment of enteric neurons (PGP 9.5, NF), glial cells (GFAP, S-100) and inhibitory neurotransmitters (VIP, NOS).

Results. In acute NEC the mucosal plexus was completely destroyed, while the submucosal and myenteric plexus layers were still discernible. The remaining ganglia exhibited central lesions displaying degenerated nerve and glial cells. Interestingly, the neuronal and glial loss increased towards the antimesenteric border where the ischemic damage was most evident. Concomitantly, VIP- and NOS-positive neurons were significantly reduced in areas of transmural lesions. Specimens of post-acute NEC displayed similar lesions, but additionally showed reactive hypertrophy of submucosal nerve fibers.

Conclusions. The neuropathologic lesions resemble an acquired form of intestinal innervation disorder best characterized as acquired oligoneuronal hypoganglionosis. The topographic distribution of the lesions speaks in favour of an ischemic event underlying the pathogenesis of NEC. The persistent alterations of the ENS in post-acute NEC may contribute to the development of complications such as intestinal mo-

tor dysfunctions and obstructions frequently observed after an acute episode of NEC.

A10
Prognostic value of standard laboratory parameters in NEC

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Background. Besides clinical and radiological signs further tools for the assessment of neonatal acute abdomen are required. This study was performed to assess the relation between severity of NEC, prematurity of patients, and routine laboratory findings and diagnosis, staging and prognosis in necrotizing enterocolitis (NEC).

Methods. A retrospective cohort study was performed in 128 neonates with NEC born in 1978–2002 at a university hospital. Severity of NEC (Bell stages I-III), degree of prematurity (birth weight, gestational age) and routine blood parameters such as blood counts (leukocytes, platelets), hemoglobin concentration (Hb), lactate, and CRP were evaluated for staging, surgical decision-making, and outcome prediction in the early stage of NEC.

Results. Factors with influence on survival were NEC severity ($p < 0.05$), lactate ($p < 0.005$), BW and GA ($p < 0.005$). CRP was higher in NEC °II-III than in °I ($p = 0.002$) but not prognostic for survival. Blood cell counts and Hb were not useful due to the small difference between groups with increasing severity of NEC, operative and non-operative cases as well as survivors and non-survivors. According to ROC-analysis, the highest predictive potential resulted from a score (1–8 points = lowest to highest probability of poor outcome) combining BW, NEC-grade, lactate and platelet count ($p < 0.001$; AUC 0.83). The ideal cut-off ranged between 4 and 5 (sensitivity 0.71, specificity 0.72).

Conclusions. Increases or drops in other routine laboratory values may be warning signs but only increased lactate was of prognostic value in NEC. A score combining NEC grade, prematurity parameters and laboratory values allows for early prognosis assessment.

Trachea

A11
Tracheal surgery in children: introduction

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Background. Congenital lesions of the trachea are symptomatic early in life and are frequently lifethreatening. They are often associated with esophageal or vascular anomalies. Acquired stenosis, a major problem in the past, is preventable by using small endotracheal tubes.

Methods. Stenotic lesions have been managed with dilatations, local injection of steroids, stenting, cryotherapy or laser vaporisation with doubtful results. We prefer resection of the stenosis as treatment of choice.

Results. Prenataly diagnosed severe congenital tracheal stenosis may be managed by EXIT procedure, until air way is established. Up to one half of the trachea might be resected, longer segments with complete tracheal rings need tracheoplastic procedures usually under cardiopulmonary bypass. Compression by left subclavian artery may cause severe tracheomalacia and will be treated by vascular suspension methods. Vascular rings by double aortic arch will be managed by diversion of the ring. Pulmonary artery sling recommend vascular and tracheal surgery, but have poor prognosis.

Conclusions. Surgical resection of tracheal stenosis has proved to be the most effective treatment and prolonged intubation, tracheostomy and multiple dilatations may be avoided.

A12
A rare case of tracheal agenesis: surgical approach

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Background. Tracheal agenesis represents a rare malformation (1/50.000 live-born) that causes severe respiratory distress frequently associated to others congenital anomalies.

Methods. BG., female, born at 35 wks with prenatal diagnosis of polyhydromnios, duodenal atresia (DA) and complex heart disease. CT scan, performed after birth, showed a tracheal agenesis (typ II sec.Floyd). The trachea was connected to the esophagus by fistula. The larynx appeared with a normal structure.

Results. At birth, the child was admitted to the ICU and at one month of age underwent surgery: by right thoracotomy ligature of the distal oesophagus was performed. An esophagocervicostomy was performed in continuity with pharynx, allowing the saliva's drain. A distal esophagostomy, "pro-trachea", was then realized with a stent to maintain a pervious oesophagus. We obtained the air passage from the esophagostomy through the esophageal tract in the trachea. At the same time the correction of the associated DA and of a Meckel's diverticulum were realized and a gastrostomy was performed too. Later the child underwent cardiac surgery. Twelve months later there was a gradually, constant ponderal and stature growth of the child.

Conclusions. Most of the newborns with severe tracheal agenesis die within early days of life and the survival is related to a sufficient trachea to perform the tracheostomy. Till today tracheal replacement is a challenge and surgical techniques are loaded with life threatening complications. The patient treated in our hospital represented a one case of the survival described in literature.

A13
Successful treatment of a traumatic tracheal disruption in a 6 year old boy

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Background. Tracheal rupture due to blunt chest trauma is rare in the pediatric age group and most cases are managed non-operatively. Therefore major trauma to the trachea and the bronchi is unsuspected and easily overlooked by the attending surgeon.

Methods. To focus the attention to tracheobronchial injuries and their treatment we report the case history of a 6 year old boy who was hit by his grandfather's tractor while riding on his bicycle.

Results. The patient was brought to a district hospital by his parents. At admission he presented with respiratory distress and a large bilateral subcutaneous emphysema. He was intubated and ventilated and the left pleural cavity was drained for tension pneumothorax. Then he was brought to our hospital by road ambulance. Upon arrival the patient was in an unstable condition. Therefore the right pleural cavity was also drained and the chest tube on the left side was replaced. After stabilization of the patient a CT scan of the head, the neck and the thorax was performed. CT scan revealed a dislocated fracture of the left mandible and a fracture of the left clavicle. The trachea was completely ruptured at the level of the 10th cartilage ring and the distal part of the trachea was dislocated at the right side. There was a gap of about 2 cm length only bridged by peritracheal soft tissue. Within 24 hours the trachea was reanastomosed by a transcervical approach. The postoperative course was uneventful, the child went home the fifth postoperative day and tracheobronchoscopy performed 3 months after the trauma showed a normal nonstenotic trachea.

Conclusions. Tracheobronchial rupture by blunt trauma is rare in children. Leading symptom is the subcutaneous emphysema. Dyspnea is uncommon and may be the result of major laceration of the trachea. Therapy includes bed rest, antibiotic prophylaxis and thoracic drainage. Major and complete rupture of the trachea or the bronchi must be treated operatively. The surgical approach depends on the level of the lesion and consists of a right thoracotomy or a transcervical incision. Diagnosis should be made by noninvasive means as CT scan as a first step. Primary tracheobronchoscopy should be avoided and can rapidly worsen the ventilatory state of the patient.

A14
Long term results in endoluminal tracheal stenting

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Background. Different alloplastic materials are used in endoluminal stenting of the airways. We report our experience with this technique in childhood during a period of more than ten years with different stents.

Methods. Between 1993–2005 18 children with airway obstruction were treated by endoluminal endoscopic airway stenting. All children suffered of severe respiratory insufficiency, 10 from 18 had just previous surgery of stenosis of larynx or trachea. In all cases the stent implantation was guided by rigid tracheobronchoscopy. We used the following stents: 2× Steel (Strecker und Palmaz), 4× self expandable metal stents (Gianturco und Wallstent) and 6× „shape memory" nickel titan allow (Nitinol), 6x dacron reinforced silicone stents (Polyflex).

Results. The placement was successful in all cases and thus airway obstruction resolved initially. 2 times the stent positioning was not exactly and it had been corrected immediately. Postoperative dislocation occurred in 4 cases with dacron reinforced silicone stents. Granulation tissue occurred in all stents. These granulations required in 10 children additional therapies, in 2 cases stent removal. Mucus obstruction resulted by the stent surface we saw 6 times. 4 stents are complete submucous integrated. 2 of this had be removed by open surgery because of persistent scarring laryngotracheal stenosis.

Conclusions. An ideal stent does not exist. Complications are notable, so that tracheal stenting should be restricted to single cases. Gold standard of laryngotracheal stenosis remains surgical treatment.

A15
Bronchoplasty for a mainstem bronchial cancer in a child

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Background. A twelve year old boy was admitted for respiratory distress, and cyanosis. He was treated at home the last two weeks for a right lower lobe pneumonia. Eight months before he had an identical infection. At bronchoscopy, the trachea was free. One centimeter after the carina, a bloody mass was seen at the internal wall of the right mainstem bronchus. The intermedial bronchus was obstructed. The right upper bronchus was free. CT confirmed the tumor with an intra and extraluminal involvement of the right mainstem bronchus. The middle and the lower lobes were scarred with air trapping. Scintigraphy (Indium 111) marked on the tumor, led to suspicon of carcinoid tumor. Biological markers were negative.

Results. By a right thoracotomy, we completely mobilized the right mainstem bronchus and the right upper bronchus. The middle, lower lobes and the mainstem bronchus were resected. The right upper bronchus was preserved. Intraoperative frozen section examinations of the right mainstem bronchus and right upper bronchus margins were free of tumor. The carina was closed and an end-to-side anastomosis was performed between the right upper lobe and the anterolateral trachea wall. The postoperative course was uneventful. The final histological diagnosis assessed a 5.3 cm low-grade mucoepidermoid tumor involving the wall of the mainstem bronchus. All six hilar and one carinal lymp nodes were free of tumor. Thirty months after the operation, the child is doing well. The chest x-ray shows a well expanded right upper lobe. There is no anastomotic stenosis at the bronchoscopy.

Conclusions. In childhood mainstem bronchus tumor are rare, usually of good prognosis and of low-grade malignancy (carcinoid, mucoepidermoid tumors). The bronchoplastic surgical technique when possible is suitable in children because

of sparing the pulmonary parenchyma and its minimal morbidity. Finally, the postpneumonectomy syndrome (scoliosis, mediastinal shift) is avoided.

A16
Bronchial sleeve resection performed in a 5-year-old boy with mucoepidermoid carcinoma

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Background. Mucoepidermoid carcinoma of the bronchus in childhood is an extremely rare tumor. We report the successful surgical tumor removal of the proximal left mainstem bronchus performing lung sparing bronchial sleeve resection. We discuss the therapeutical options, surgical techniques and review the literature of these rare bronchial tumors.

Methods. A 5-year-old boy was evaluated for persistent leftsided pneumonia, fever and antibiotic therapy for about 2 months. Bronchoscopy showed an exophytic endobronchial mass occupying the left mainstem bronchus directly at the origin from the trachea. Biopsy showed intermediate-grade mucoepidermoid carcinoma. Staging (thoracic multislice CT, MRI of the cranium) showed no evidence of metastases to local lymph nodes, distal sites or organs. Trying to avoid left side pneumonectomy sleeve resection of the left mainstem bronchus was performed. Right side intubation and sternotomy was performed. The distal trachea and ipsilateral mainstem bronchus were mobilized. The proximal left mainstem bronchus was subsequently transected and the sleeve removed in a length of 2 cm.

Results. Circumferential histologic examination of the proximal and distal margins was tumorfree. Selective intubation of the left and right bronchial system had to be achieved surgically. Broncheotracheal anastomosis was done using vicryl 5.0. The postoperative course was uneventful. Re-staging after 3 and 6 months postoperative showed no evidence of a tumor relapse.

Discussion. Mucoepidermoid carcinoma of the bronchus in childhood is extremely rare. Symptoms, when present, result from the associated airway obstruction, and include recurrent pneumonia, cough, fever, wheezing and dyspnea. These neoplasms are histologically classified as low-grade and high grade carcinomas. Children appear to have more often low-grade tumors. For further diagnostics and evaluation computed tomography is helpful, because it provides excellent visualization of the trachea and mainstem bronchus. For preoperative staging computed tomography is used too. Radical surgical resection is the treatment of choice. Depending on the localisation of the tumor bronchial sleeve resection, lobectomy or pneumectomy is necessary. In our case the tumor was located in the left mainstem bronchus directly at the origin from the trachea. Left side pneumectomy was avoided and sleeve resection of the left mainstem bronchus was performed. Postoperative radiation and chemotherapy is not necessary in low-grade tumors, but in high-grade tumors when surgical extirpation is not possible to perform.

Conclusions. Resection of a mainstem bronchus with pulmonary preservation is a therapeutic option when disease is limited to the mainstem bronchus. It is defined as circumferential resection of either mainstem bronchus. Mucoepidermoid tumors are rare bronchial adenomas comprising 1% of all lung neoplasmas. These tumors arise from the excretory ducts of the submucosal bronchial glands. Location is the mainstem bronchus or the proximal portion of a lobar bronchus. Children are very infrequently affected. They are slightly more common in female patients. Complete surgical resection remains the treatment of choice for mucoepidermoid carcinomas.

A17
Experience with hemangiomas of the larynx and trachea in 56 children

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Background. Hemangiomas of the larynx and trachea are life-threatening diseases in infants. The typical sign is a stridor during the first 2 or 3 months of life, often associated with an increasing size of cutaneous hemangiomas.

Methods. The therapeutic procedure depends on the kind of the angioma. All children were treated by Nd:YAG laser using a 0.4 mm bare fiber with different modes. For capillary angiomas we prefer the non contact technique, interrupted, 15–20 watt. The submucous located cavernous angioma with endoluminal bulging are treated by ablation, 20 watt single impulse. For the transmural angioma we prefer the LITT, 2–3 watt, cw.

Results. In a 20 years period from 1983 to 2003 56 infants underwent endoscopical laser disobliteration of the larynx and trachea in our departments. In 17 cases the endoluminal angioma was associated with additional disturbances like extensive hemangioma of the face or neck. The airway segments most frequently involved were the subglottic area and the larynx (n = 46). The size and expansion were similar to those of cutaneous angiomas: limited to the mucosa (n = 27), infiltrating the submucousa with tumorous endoluminal bulging (n = 21) and transmural spreading in to the extratracheal adherent tissue (n = 8).

Conclusions. Endoluminal laser treatment has been used successfully in all children we have seen. In 50 cases one laser session was sufficient; in six children 2–4 sessions were necessary. In 16 children temporarily tracheostomy was performed. All children are alive.

A18
Laser treatment of newborn and children with cysts of the larynx and trachea

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Background. Congenital cysts of the larynx and trachea in newborns cause an acute obstruction of the upper airways. The cysts are firm and non compressible. In these cases intubation is often impossible and an emergency intervention becomes necessary.

Methods. In our collective of 470 children with stenosis of the airways we only had 15 cases in which the cysts were the main cause of obstruction. Most of the cysts were located in the larynx followed by the subglottic and the thoracic trachea. In 15 cases they become symptomatic in the newborn age.

Results. All children underwent an endoscopic laser therapy. We used a rigid bronchoscope with a channel for the 0.4 mm bare fiber. Large cysts were emptied by puncture followed by fenestration using contact technique, fibertom mode, 20 w cw. Afterwards the inner surface the opened cyst was coagulated by non contact technique, following by shrinkage and scare formation.

Conclusions. All children are alive. In three newborns with congenital cysts, a second session was necessary. Tracheotomy could be avoided in all children.

A19
Surgery for long-term complications of tracheostomy in children

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Background. Improper techniques for tracheostomy in children and a long-term decannulation are the main causes for a resulting tethered tracheostomy scar and tracheal tug appearance. Children with tracheostomy require specialized hospital/home care and when proper attentions are lacking, infection, granulation, dry-skin and tissue retraction may all occur at the trach-site. As a result, persistent tracheocutaneous fistula or a tracheal tug have to be surgically closed for a cosmetic reason as well as for an harmonic and painless deglutition.

Methods. Out of the last 22 long-term decannulated children, 8 presented a tracheocutaneous fistula (TCF) and 6 a depressed scar with a tracheal tug (TrT). In the TCF group, ptyalism, infected secretion and peristomal dermatitis were the most common problems whereas in the TrT group a true dysphagia took place. Global cosmetic appearance was poor and 6 adolescents need for a psychological support. A diamond-shaped incision including a fibrotic-core affecting both skin and soft tissues was done and carried down to the tracheal defect, closed by single nonabsorbable 4/0 stitches. In TrT the fibrotic plates were dissected from the tracheal wall. A final repair included a strap-muscles and platysma suture and a bilateral sternocleidomastoid split (BSS).

Results. All 14 children were cured, without any p.o. infectious complication or recurrence of the TrT or the TCF, 26 months after surgical procedures. No drain were required, no haematomas or syeromas were seen. No residual depression is evident at 2.6 years follow-up and only a small skin scar as a result of the dermal subcuticolar closure is seen. Swallowing disorders or dysphagia completely disappeared in the postoperative period, early after discharge. From a psychological point of view, all children were open to improvement during the first p.o. month and completely recovered in 3 months subsequent to the surgery. From a parental point of view, a lot of advantages were reported and discussed during a postsurgical counselling, such as the absence of the frequent suctioning, a dramatic relief of signs and symptoms (pain, peristomal-dermatitis, persisting wet-dressing, the „neckband" syndrome) as well as the stop of sleep disorders.

Conclusions. A long-term tracheostomy is complicated at least in 45–70% of children and all tissues may be equally affected by fibrosis and scar. If the cicatricial tissue penetrates to a hard structure, such as trachea an undue up-down movement with deglutition is evident, causing discomfort and pain. A definitive and complete repair of TCF and TrT should include a double-muscle flap as well as a BSS. This procedure is recommended for emotional, dysfunctional and cosmetic purposes associated with complicated tracheostomy in children.

A20
Thoracoscopic tracheo-aortopexia for the treatment of life-threatening events in tracheamalacia

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Background. Life-threatening events due to tracheamalacia are a well-known complication in infants with esophageal atresia. As the aortic arch and trachea are closely related, elevation of the aortic arch against the sternum will pull up the trachea too, and thus alleviate the complaints from the collapse of the anterior and posterior wall of the trachea. This procedure is usually performed by a transjugular approach. With the advent of minimal invasive surgery (MIS) the procedure can also be performed by thoracoscopic MIS.

Methods. Between January 2002 and May 2004 five children esophageal atresia were treated by MIS for life-threatening events due to tracheamalacia. Mean age at the time of operation was 5 months (14 d–12 m). Mean weight was 5.5 kg (3–9 kg). There were 3 girls and two boys. The procedure was started with a tracheoscopy. "Kissing" of the ventral and posterior tracheal wall were an indication for thoracoscopic tracheo-aortopexia. After the sutures were placed and pulled against the posterior surface of the sternum, tracheoscopy was repeated to determine the efficacy of the tracho-aortopexia.

Results. All patients tolerated the thoracoscopic procedure well and tracheo-aortopexies could be carried out thoracoscopically. Tracheoscopy at the end of the procedure a clear improvement of the trachial diameter. In two patients recurrence of life-threatening events occurred after two and four weeks respectively, for which a thoracoscopic redo-tracheo-aortopexia was performed with good results. With a follow-up of 27 m (17–45 m) all patients are doing well and have had no more life-threatening events.

Conclusions. Thoracoscopic tracheo-aortopexia for life-threatening events due to tracheamalacia is feasible and safe. In two instances in which symptoms recurred after an interval, thoracoscopic redo-procedure was successful in alleviating tracheamalacia symptoms.

A21

Experimental correction of the Congenital High Airway Obstruction Syndrome (CHAOS): a possible prenatal rescue

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Background. The Congenital High Airway Obstruction Syndrome (CHAOS) is mainly due to a laryngeal atresia or is dependent from a severe form of tracheal stenosis. The prenatal U.S. examination is useful in the diagnosis of complete or near-complete obstruction of the fetal upper airway and may support the severity of the global condition supporting the data concerning the associated fetal abnormalities. Fetal ascites, hydrops, large echogenic lungs, flattened diaphragms and lung hyperechogenicity are all part of this extremely severe condition. However, if the diagnosis is prenatally-made, an EXIT procedure may be life-saving, and a fetal tracheostomy is performed under a laryngo-tracheoscopic guidance. In order to investigate the possibility for a fetal cricotracheal resection (CTR) in selected cases of CHAOS induced by a tracheal stenosis, an experimental model is proposed, using the NZWR as a subject. In fact, in our previous experience we demonstrated that tracheal rings of the young rabbit and child have similar hyaline cartilage, as well as are comparable in the fetal developing period.

Methods. One fetus of NZWR per dam has been operated, during the 28 days of gestation for a total of 5 models; 10 sham-operated fetuses were used as controls. A cricotracheal resection has been accomplished in all, without fetal nor maternal mortality. Data recorded included morphometric analysis, lung hypoplasia investigation and the type of the airway's healing.

Results. The experiments were sacrificed at term (31+/–0.5 days) and all showed a good viability of the CT junction and fibrosis-free line of sutures. As well as in fetal dermal repair, regeneration of the airway cartilage and mucosa were complete and scarless. No evidence for an associated lung hypoplasia is reported.

Conclusions. Children affected by a moderate form of CHAOS supported by stenosis instead of atresia of the high airways, are less prone to be at risk for secondary hydropic syndrome, diaphragmatic paralysis, tracheobronchial malacia and the need for chronic ventilatory support. In those cases, a definitive solution such as a fetal CTR may be taken into account, when we are discussing for a surgical planning of the Maternal/Feto-Neonatal entity.

Thorax

A22

Thoracoscopic drainage of pleural empyema

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Background. Pleural empyema after severe pneumonia is rare, with an incidence of 3.3 per 100,000 children. The incidence is higher in winter and spring and almost all times there is a bacterial infection. The pathogenesis runs from the exudative phase to fibropurulente phase till organisation of the empyema. The treatment was beside premarily iv antibiotics drainage by punction, thoracal drain, use of fibrolytica and eventualy thoracotomy for a decortication. Since 2004 we introduced an early intervention with thoracoscopic drainage.

Methods. Retrospective study of all patients from 2001–2003 (fibrolytica) and 2004–2005 (thoracoscopy) with a pleural empyema.

Results. In both groups 8 patients were included, with an age between 0 and 11 years. The mean hospital stay in the fibrinolytica group was 15 days and in the thoracoscopic group 10 days. There were no complications.

Conclusions. Early treatment of pleural empyema with thoracoscopic drainage reduces morbidity and hospital stay.

A23

Role of VATS (Video Assisted Thoracoscopic Surgery) in empyma management

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Background. Empyema thoracis is defined as collection of pus in the chest. The causes of empyema are: complicated pneumonia, autoimmune disease, cystic fibrosis or trauma. Children with empyema usually present with high grade fever, cough, shortness of breath and pleural effusion on chest X-ray, not responding to I.V antibiotics. Thoracocentesis is one modality of treatment. However thoracotomy previously and VATS nowadays for empyema depridment is the mainstay management.

Methods. 15 children with empyema were reviewed over 13 month (March 2003–May 2004) in RAFH and other hospitals in Riyadh, study of age, sex, site of the empyema, prehospital treatment, radiological investigation pre- and post-op operating time, conventional thoracotomy, hospital stay, all were reviewed.

Results. 14 children with loculated empyema were treated successfully with VATS. Mean operating time was 78.7 min (range: 30 min–180 min). Chest X-ray showed full lung expansion post op. Fever subsided after 48 hours post-op and continues in one case for 7 days. Air leake in two children, disappear, 5 days post-op. chest tube were removed on the 4th day post-op. in 13 patients (86.7%), on 7th day post-op. in one

patient and on the 12th day on the other. Post-op. hospital stay ranged from 5–23 days (average 7 days). No conversion into thoracotomy and no intraop. or post-op. complications. Follow up of the children was conducted in all 15 patients. 13 children showed no recurrence, one child SCID has pleural thickening in the right lower chest. The other child with hyper IGE still has right upper lobe pneumocyst and he is for surgery.

Conclusions. VATS reveal its safety and efficacy in addition to less pain and hospital stay. Ultrasound is operator dependent. CT scan is superior to ultrasound.

A24
Thoracoscopic laser surgery in newborn and children

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Background. Thoracoscopy is a very tissue-preserving technique for the operative therapy of different intrathoracic diseases. It can be performed at any age and offers special advantages in newborn with reduced lung capacity. The use of the laser makes it possible simultaneously the tissue cutting and the hemostasis with sealing of the cutting surface.

Methods. Three point standard thoracoscopy in lateral position, general anaesthesia; artificially Pneumothorax with five to eight millilitres Hg. We used the Nd:YAG laser 1064 nm with an 0.6 mm bare fiber applicating different laser modes and parameters. For cutting 20w, cw, in contact was sufficient. The hemostasis was available with 30w, intermittent in non contact technique.

Results. Since 1981 we operated more than 120 newborn and children with congenital and acquired lesions of the thoracic organs in our hospital. The indications have not been limited by the age or the size of the children. The lowest body weight was lover than 1100 g. We performed lung resections and interventions for tumors and cysts of the mediastinum, thymic gland, pericard and diaphragm. Other indications were chylothorax, empyema, fistulas and so on.

Conclusions. The advantages of laser VATS are the limited surgical trauma, the possibility of reintervention, the bilateral performability in one session, the minimal loss of blood, the negligible number of adhesions, the frequent dispensability of drainages and the uncomplicated postoperative course without any death or other complications. Only in six cases (CCAM, lung sequester, lobar emphysema, chylothorax) conversion to thoracotomy were necessary.

A25
Thoracoscopic procedures in oncological patients

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Background. History of thoracoscopy: in the year 1912 Jacobeus of Sweden performed a therapeutic pneumothorax in a patient with tuberculosis. He published a small number of patients with lung biopsy. 1971 published Klinkowitch a paper about 43 lung biopsies in childhood, in 1976 described Rogers thoracoscopic lung biopsy in 9 children 17 months till 15 years of age. Today thoracoscopy is used for the management of oesophageal atresia in newborns, thoracic trauma and biopsy and resection of intrathoracal masses.

Methods. Our method 3 mm, 5 mm to 12 mm camera port, 2–3 working ports, prone-supine patient's position. All oncological patients were examined by radiographs, CT and MRI before operation.

Results. From 2003–2005 thoracoscopy was performed in 21 patients 2–19 years of age (median 10.5 years). Operation time ranged from 25 to 160 minutes. 3 conversions were necessary because of large tumor, bleeding and pleural adhesions. In 16 oncological patients the Hodgkins lymphoma (4), non Hodgkins lymphoma (2), histiocytoma (2), Ewing's sarcoma (1), cystic thymoma (1), metastatic Schwanoma (1) and testicular cancer (1), mediastinal teratoma (1), soft tissue sarcoma (1), haemangioma (1) and neuroblastoma (1) were reason for the thorascopy. Our non-oncological diagnosis included 4 cases of pleuritis and pleural empyema and 1 patient with traumatic pheumothorax.

Conclusions. In our group of patients we did not notice complications. Thoracoscopy is a useful diagnostic and therapeutic procedure and has secured an important place in pediatric surgery.

A26
Congenital pulmonary cystic lesions: a new etiopathogenetic hypothesis

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Background. Cystic malformations of the lung, bronchogenic cysts (BC), congenital lobar emphysema, cystic (CLE) adenomatoid malformation (CAM) and pulmonary sequestration (PS) can be enclosed in the lung bud anomalies. They are due to the effect of the uncoordinate interaction between embryonal mesenchyma and endoderma during the development of the primitive anterior foregut and its division from the airways.

Methods. In the Pediatric Surgery Department of the University of Bologna from January 1975 to October 2005 have treated been 86 patients (46 males) afflicted by lung malformations. The age ranged from neonatal to 13 yrs old. The histopathologic exams confirmed the following diagnosis: BC = 17, CLE = 18, PS = 14, CCAM = 42. 81 patients showed a solitary lesion. The last five patients (6.3%) had two simultaneous lesions: two cases with CCAM and homolateral intralobar PS (IPS); three patients with CCAM and CLE (2 homolateral and 1 contralateral). Therefore in total there were 91 malformations on 86 patients. In one case of IPS was histologically found ectopic pancreatic tissue. We never found lung lesions with intestinal communication.

Results. All the 86 patients have been surgically treated. 80 lesions have been removed by thoracotomy. In the remaining eleven a thoracoscopy was performed; four of these were removed only by thoracoscopy whereas in 7 cases the operation was thoracoscopic assisted (VAT) and a mini-thoracotomy was performed.

Conclusions. Cystic pulmonary lesions have a high potential risk of malignant transformation, therefore the surgical removal is mandatory. Clinical research should be addressed to find tumor markers in order to identify those lesions that probably would not go to malignant transformation.

A27
Congenital malformations of the lung, experience with 18 cases

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Background. Congenital malformations of the lung are rare and vary widely in their presentation and severity. The purpose of this study is to review our institutional experience of congenital cystic lung disease, with specific reference to diagnosis, treatment, as well as outcome, furthermore, to present some cases with unusual clinical.

Methods. From March 1994 to March 2003, 18 patients were operated for congenital cystic disease of the lung. There were 13 male patients and 5 female, ranging in age from 9 days to 34 years. There were 7 congenital lobar emphysemas, 6 bronchogenic pulmonary cysts, 3 congenital adenomatoid malformations and 2 pulmonary sequestrations.

Results. Surgical procedure included 14 lobectomies, 1 bilobectomy, 2 segmentectomy, and cystectomy in 1 patient. Associated cardiac anomalies included patent ductus arteriosus in 1 patient of congenital lobar emphysema and partial absence of pericardium in 1 subject of bronchogenic cyst.

Conclusions. The most common manifestation of the congenital cystic disease of the lung at newborn and early infancy is respiratory distress. Later on in life, cysts usually lose this compressive character and may remain asymptomatic until infection occurs, while producing cough, dyspnea, and thoracic pain. It seems that congenital adenomatoid malformations and congenital lobar emphysemas usually present in infancy especially in neonatal period with significant respiratory distress that prompts an expeditious workup and treatment. Given the probability of developing infectious complications, the sequestered tissue should excised prophylactically.

A28
A rare case of hepatic pulmonary fusion

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Background. Hepatic Pulmonary Fusion (HPF) is an extremely rare cause of respiratory distress in neonates, that should be differentiated from right side diaphragmatic hernia and needs a plan for definitive management.

Methods. We will present a neonate presented with respiratory distress immediately after birth, intubated and ventilated. Right side diaphragmatic hernia was seen on chest X-ray, echocardiography delineated complex heart disease. After stabilization, laparotomy, plication of eventrated part of diaphragm, pushing the liver to abdomen was down with repair of exomphalous. 1 week later thoracotomy was performed and HPF and additional placation to medial part of diaphragm don.

Results. On chest X-ray: Mass shadow with bowel is seen in right hemithorax, minimal liver opacity in abdomen also noted. Operative partial reduction of liver was performed into the abdominal cavity to provide space for the lungs to expand. No surgical separation between lung and liver was attempted, but was planed to be done after cardiac operation. The patient was extubated and maintaining oxygen saturation on nasal oxygen tube, later he developed chest infection with very limited cardiac compliance, deteriorated, again ventilated and expired at age 6 months.

Conclusions. Hepatic pulmonary fusion is a condition that has no distinction between liver, diaphragm and lung tissue. The liver could not be fully reduced to the abdomen without cutting the fused surface which can be done if life space of the patient expected to be normal.

A29
Therapeutic approach to echinococcosis on lungs in children

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Background. Diagnosing and surgical treatment of echinococcosis as well as treatment of echinococcosis with unusual localization in children.

Methods. A total number of 52 children aged between 5 and 17 years with lungs echinococcosis were operated at the Clinic of Pediatric Surgery in a 6-year period 1995–2003. In all patients, plain-x-ray of the lung was postoperatively done. In this way, regression of the disease was also followed-up.

Results. Percutaneus drainage of the contents of residual pericystic cavities (lungs – left lobe) was performed helped by US. Only in three children re-intervention was necessary due to hydatid cyst relapse in the existing pericystic cavities.

Conclusions. The exact diagnosis is a key moment in therapeutic treatment of hydatid disease in children. Surgical treatment is the first choice in handling with this disease. Adjuvant therapy minimizes the risk of recidivism and is the principal therapy only in cysts smaller than 2 cm.

A30
Impact of ECMO on the survival of congenital diaphragmatic hernia in Mannheim, Germany (1990–2004)

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Background. Although extracorporeal membrane oxygenation (ECMO) is a lifesaving intervention, the role of ECMO on Congenital Diaphragmatic Hernia (CDH) survival is still controversial. In this study, we examined the relationship between increased ECMO utilization and its effect on CDH survival in Mannheim, Germany over the last 15 years (1989–2004).

Methods. Over the last 15 years, Mannheim neonatal and neonatal surgical unit has received 244 patients with diaphragmatic hernias. For the purpose of the study and because of different management protocols, this period was divided into 3 5-years periods. From 1990–1994, the unit received 33 patients with CDH, ECMO was employed in 19 patients. In the period 1995–1999, 45 patients were admitted in the unit and ECMO was utilized in 22 patients. During the period 1999–2004, 166 babies with diaphragmatic hernias were admitted to the unit and ECMO was indicated in 86 patients.

Results. The number of CDH referred to the unit during the last five years was five times the number of patients referred during the first 5 years (166 against 33). The number of patients requiring ECMO was more or less the same (about 50%). However the number of patients who died after ECMO was reduced form more than 55% in the first 5 years to less than 30% during the last 5 years. The overall mortality in CDH was reduced from 50% in the period (90–94) to 30% in (2000–2004). Most of the patients who died in the last 5 years did not have surgery because, according to the selection criteria and the protocol of management, it was clear that they were not going to survive.

Conclusions. A strong association exists between employment of ECMO and improved survival in CDH. The study also shows the importance of referral to a specialised unit in order to develop experience and improve survival. The study showed that although 50% of patients with CDH required ECMO during the different time periods, the mortality under ECMO was reduced from 57% earlier, down to 30% during the last five years.

A31
Late presenting congenital diaphragmatic defects – experiences of the last 26 years

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Background. Beside "classic" diaphragmatic defects, which are usually diagnosed intrauterine or immediately after birth, there exist forms of congenital herniation, which present with different features later in infancy or at any time of life, or stay asymptomatic. In contrast to diaphragmatic hernias, diagnosed after birth because of pulmonary complications, later presenting diaphragmatic hernias are associated with unspecific pulmonary or gastrointestinal symptoms. Occasionally they appear with acute symptoms (like an ileus), sometimes they stay asymptomatic or are recognized by accident due to another disease.

Methods. Within the last 26 years 15 children were treated because of delayed diagnosed diaphragmatic defects at our department (5 female, 10 male). 4 presented with acute symptoms (acute respiratory distress [3 children], ileus [one child]), 8 with chronic complaints (repeated pulmonary infections [5 children], dyspnoe [1 child], repeated abdominal pain [2 children]) and 3 stayed asymptomatic. Diagnosis could be performed with standard investigation modalities. In 13 patients the diaphragmatic defect could be closed with direct sutures, 2 defects required an implantation of a patch. Apart from one patient with a partial postoperative diaphragmatic relaxation, which could be snatched up, the postoperative courses were free of complications.

Conclusions. Late presented diaphragmatic hernias show a better prognosis than prenatal diagnosed defects, most likely because of the fact, that this kind of diaphragmatic malformation is rarely associated with pulmonary anomalies.

Rare tumours I

A32
IPSO – rare tumor registry

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Abstract not available.

A33
Pancreatic tumors in children – pathological aspects and treatment approaches

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Background. Pancreatic neoplasms are rare in children and adolescents. There exist three relevant tumor entities: pancreaticoblastoma, solid pseudopapillary tumors and pancreatic carcinoma. According to the dignity different therapeutic strategies may become necessary.

Methods. The authors present a series of pancreatic tumors in children in cooperation with the German Tumor Registry of the Paidopathological Institute in Kiel (Pancreaticoblastomas n = 5; pancreatic carcinomas n = 22; solid pseudopapillary tumors n = 35). Morphological aspects and histopathological markers of the three tumor entities are described. The relevance of chemotherapy and the surgical treatment according to the staging system are analysed based on our own data and a literature review.

Results. The most common pancreatic tumors in childhood and adolescence are pancreaticoblastomas and solid pseudopapillary tumors. Pancreaticoblastomas contain pluripotent cells capable of differentiating along the pathways of all three pancreatic cell types. The pathological distinction between pancreaticoblastomas and carcinomas can be challenging. The main goal in the treatment of all mentioned tumor entities is the complete tumor resection. The Whipple procedure is the surgical treatment of choice in most cases. An open question is the relevance of segmental resection of the portal vein for survival. Generally, children with pancreaticoblastoma and pseudopapillary tumors have a good prognosis after complete tumor resection. A central problem in children with pancreatic carcinomas is the metastatic disease. The conventional chemotherapy with gemcitabine, folinic acid or 5- FU represents only a palliative treatment option. Possible new treatment strategies such as high dosis chemotherapy (VP 16/

Carboplatin) with stem cell rescue are not evaluated sufficiently yet. The world wide experience in these rare cases is only based on singular case reports.

Conclusions. Diagnostic procedures and treatment strategies of children with pancreatic tumors remains a challenge. Depending on the dignity, a radical surgical approach seems justified. A common international trial for these rare tumors seems the only way to prove the efficiency of new chemotherapeutic strategies.

A34
Management of solid-pseudopapillary tumours of the pancreas

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Background. Solid-pseudopapillary tumours (SPT) of the pancreas are increasingly diagnosed, but the exact surgical management needs to be standardized.

Methods. Twelve female patients with a median age of 21 years (range 13–54), underwent resection for SPT with the aim of a tumour free margin of at least 1 centimetre. Preoperative, operative, clinicopathological and survival data were analyzed.

Results. Abdominal pain was the most common presenting symptom and occurred in eight patients (67%). No patient presented with jaundice. Of the two patients with weight loss, liver metastasis was found in one. Five patients had SPT of the pancreatic head and all underwent pylorus preserving pancreatoduodenectomy. Of the seven patients with distal tumours, one underwent a central pancreatectomy and the remaining six patients a distal pancreatectomy. Spleen preservation was possible in 2/6 of these patients while splenectomy was necessary in the remaining 4. One patient with a distal tumour had concomitant liver metastasis and underwent staged distal pancreatectomy and right hemihepatectomy five months later. The median postoperative stay was 9 (range 6–18) days. There was no in-hospital mortality and the morbidity was extremely low. Only one patient developed cholangitis postoperatively which could be managed conservatively. The resection was microscopically radical in all patients. All patients are disease free at a median follow-up of 3.8 (standard deviation 3.8) years.

Conclusions. Solid-pseudopapillary tumours of the pancreas can be adequately managed by complete resection through pylorus preserving pancreatoduodenectomy, central pancreatectomy, or distal pancreatectomy with good early and long-term results.

A35
Nesidioblastosis – case report

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Background. Pancreatic hypoglycemia is a problem in childhood and occurs in 4 groups of patients: Sy. Becwith-Wideman, nesidioblastosis, islet cell adenoma, glucagons disturbance. Nesidioblastosis is an alteration of a normal ductal architecture of pancreas, which leads to hypoglycemia.

Methods. A patient (child, 4.5 years of age) treated many times because of hypoglycemia and convulsions is presented. At two years of age she was hospitalized with convulsions, hypoglycemia and high level of insulin. The therapy with Diazoksid improved the condition shortly, but 2000 ml 10% Dextrose i.v. daily, day after day, implies the need for surgical treatment. It was decided to permormed subtotal pancreatectomy. Also partial ileal resection was done because of existence of hamartoma on small intestine. Ex tempore material was sent from small intestine mucosa and pancreatic tissue.

Results. After operation the child did not show the need of the administration of the pancreatic enzymes or the insulin.

Conclusions. The combination of nesidioblastosis and hamartoma of small intestine is very rare. The patient was cured by subtotal pancreatectomy and ileal resection.

A36
PET-CT guided surgical therapy of congenital hyperinsulinism (CHI) in infancy

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Background. One of the challenges of congenital hyperinsulinism (previously called nesidioblastosis) is the identification of the patients with focal disease. These can be cured for ever by selective surgical therapy with complete removal of the focus. The differentiation of focal and diffuse disease is not possible by clinical or conventional imaging data. With the 18F-DOPA-PET, however, a new innovative tool has become available. It allows a high definition imaging of the actual hyperactive metabolism of focal hyperinsulinism.

Methods. Among sixteen infants with the clinical signs of CHI, six patients were classified to have focal disease by PET-CT. At surgery, selective partial resection of the pancreas was performed guided by the PET-CT report. The cutting edges of the specimens were examined intraoperatively by a specialized pathologist. The surgical findings were compared with the preoperative results of the PET-CT.

Results. In all six surgical cases the PET-CT showed a distinct focus with increased activity of the dopamine decarboxylase. In three cases the focus was found by the surgeon in the pancreatic head, corpus or tail, respectively, exactly at the place where the PET-CT had localized it. Removal of the focus resulted in cure of the infants without any complications. In one case, however, the focus was palpated at another site as the PET-CT had suggested and it was successfully removed. In two cases the PET-CT localized the focus correctly but in the histological examination of the cutting edges conspicuous islet cells were found. The PET-CT report, therefore, underestimated the extension of the disease. After further extended pancreatic resection the course of one infant was uneventful. In the other case the intermediate form of CHI (mosaic-like) was diagnosed finally by the pathologist. Despite a hemipancreatectomy in this infant hpyoglycemia persisted and necessitated further medical therapy.

Conclusions. PET-CT is a new, innovative imaging tool in CHI which gives valuable information to the surgeon for the search of the pancreatic focus. For the final evaluation of the resection, however, the intraoperative histological examination of the specimen by an experienced pathologist is still mandatory.

A37

Intrathoracic tumor as a rare cause of respiratory depression in a neonate

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Background. A boy of healthy parents, after a pregnancy without pathological findings, delivered by cesarean section due to breech presentation in the 38 and 2/7 week of gestation. Initially good adaptation, 8 minutes postpartum however increasing alinasal flapping and inspiratoric groaning. In the context of the diagnostics the following findings attract attention: chemical blood analysis shows an increment of NSE to 28.2 ng/ml (usual < 15 ng/ml), of HCG to 2.62 mU/ml (norm < 2 mU/ml) as well as of IL6 to 81.6 pg/ml (norm < 15 pg/ml). The other routine parameters as well as AFP and CEA are within the normal range according to age.

Results. Sonography reveales a plain-circumscribed tumor of a diameter of 37 × 24 × 23 mm with a connection to the left thoracic wall and parietal pleura. In the chest X-ray the 7th rip seems swollen. In the MRI and the CT of the thorax a marginal calcified tumor with mixed solide and cystic parts and with a diameter of approximately 3 × 4 cm is seen. It is responsible for the destruction of the 7th rip and the extrusion of the inferior lobe of the left lung. Evidence of metastases is not found. Taking the differential diagnosis of teratoma, PNET, neuroblastoma or osteosarkoma into account the surgical exploration is carried out on the 9th day of life. Intraoperatively, after access through the 4th intercostal space, a tumor covered by pleura reveals, which fills the entire lower portion of the hemithorax. The inferior lobe of the lung is partly atelectatic and completely displaced, however it does not seems to be infiltrated. The tumor which apparently seems to be resectable is extirpated completely and without complications and with it the affected parts of the 7th rip. After a smooth course of progression dismission follows on the 8th postoperative day. A last ambulatory control is accomplished in the 12th postoperative week. Thereby, besides an altogether good prosperousness, a dysfunction of the innervation of the abdominal wall attracts attention, which impresses clinically as a rectus diastasis. Furthermore a thoracic right convex scoliosis is evident.

Conclusions. Histologically a mesenchymal chest wall hamartoma (WHO-classification), extirpated in healthy area, is diagnosed and is confirmed by the center of reference in Hamburg, Germany. In literature only about 60 cases of this rare sort of tumor have been described so far.

A38

Rare malignant and benign tumors of the kidney in childhood

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Background. The most common malignant tumor of the kidney in childhood is nephroblastoma. We present other rare renal tumor entities of our surgical patient population from 1996 to 2004. Renal cell carcinoma (RCC) in childhood is a very rare tumor and is estimated to be under 2% of all malignant renal tumors. Clear cell sarcoma (CCS) of the kidney has been classified as high risk tumour in the previous International Wilms tumor studies and is estimated to be under 2 to 5% of all malignant renal tumors. Ewing sarcoma is frequently seen in childhood as bone tumor. Primary manifestation as renal lesion is extremely rare. On the other hand rare benign lesions of the kidney are metanephritic adenoma and angiomyolipoma.

Methods. Retrospective analysis of patient charts from 1994 to 2005.

Results. Three cases of RCC (7 years old male T4M1, 14 years old female pT1G2, 3 years old male pT1G2), three cases of CCS of the kidney, one case of malignant rhabdoid tumor in a 2 years old girl and one case of primary extraossary Ewing sarcoma of the kidney are presented. Moreover, as benign lesion one case of metanephritic adenoma in a 12 years old girl and an angiomyolipoma in a 13 months old girl are presented.

Conclusions. On the opposite of adults, experience with RCC in childhood is very limited. Based on the clinical course of three patients the updated literature of RCC is reviewed. Therapeutical strategies including immunotherapy are presented. CCS, rhabdoid tumor and renal manifestation of Ewing sarcoma do rarely occur. No consensus strategies may be applied. The current literature is reviewed and current therapeutical modalities are presented. Benign lesions of the kidney may be diagnosed by radiological techniques. In most cases diagnosis will be achieved by surgical biopsy. Diagnostic and preoperative strategies in differentiation of benign and malignant lesions are discussed.

A39

Rare tumors of the adrenal gland

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Rare tumors of the adrenal gland

Background. Except for neuroblastomas the adrenal gland is a rare localisation for tumors and present with a very different spectrum of clinical features.

Methods. The medical records of 5 children with adrenal tumors between 1995–2005 were reviewed. Clinical data, operative details, histology and follow up were recorded. 2 patients had adrenocortical carcinoma (ACC), 1 presented as acute abdomen the other as pubertas praecox, 1 with Conn

syndrome, 1 with Cushing syndrome and 1 with severe hemorrhage in an adrenocortical cyst.

Results. One patient with ACC had advanced tumor stage and died of progressive disease, the patients with hormone producing tumors were cured by surgical resection, as well as the patient with the hemorrhage into the cyst.

Conclusions. Adrenal gland tumors constitute less than 1% of pediatric neoplasmas, clinical picture depends on the hormon production of the tumor. Surgery offers cure in benign cases, but outcome is poor in ACC.

A40
Renal tumour with extensive intracardiac tumour thrombus

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Background. Tumour thrombus in Wilms' occurs in 5–10% of the cases. Extension into the atrium is very rar and extension into the right ventricle and pulmonary arteries has not been reported.

Methods. A case report and review of the literature.

Results. SND, a 3-year old girl presented with a 2-week history of progressive abdominal distension and decreased effort tolerance. A large firm mass was palpable in the left hypochondrium which extended 14 cm to the suprapubic area. There was no aniridia or hemihypertrophy. There was no pallor or bruises to suggest bone marrow infiltration. Computed tomography of the abdomen showed a large heterogenous mass occupying the left supra renal region extending into the left kidney.There was no pulmonary metastasis or lymphadenopathy. A tumour thrombus was seen within the IVC with extension into the right atrium, and entering the right ventricle. An echocardiography revealed obstruction of flow into the pulmonary artery. A trucut biopsy of the abdominal mass and this yielded mainly necrotic tissue and a focus of malignant round cells. A second trucut biopsy was performed and this was also inconclusive. A provisional diagnosis of left Wilms tumour Stage 3 was made. Chemotherapy was instituted following the National Wilms Tumour Study Group, NWTS-3 protocol. Due to raised LFTs only 30% of the recommended dose of doxorubicin was given. Two days later she became increasingly breathless, with poor peripheral perfusion. Echocardiography then showed near-total obstruction of right ventricular output by the tumour thrombus. She underwent an emergency thoracotomy on cardiac bypass. Removing the thrombus did not improve her condition and she succumbed during the surgery.

Conclusions. Histopathological examination of the tumour thrombus revealed loosely arranged spindle and polygonal cells in a myxoid stroma. Immuno-histochemistry for vimentin was strongly positive while factor 8 was weakly positive and other stains namely CD34, LCA, MNF116, PASD and mucicarmine were all negative. There were no elements to suggest Wilms tumour. The most likely diagnosis was a poorly-differentiated sarcoma probably arising from the soft tissue in the left renal fossa or left kidney. It is important to recognise that tumours apart from Wilms' can metastasize via the venacava and combined with accurate histopathological diagnosis aids in instituting the appropriate chemotherapeutic regime.

A41
Spinal canal involvement of Wilms' tumour in children

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Background. Metastases are common in Wilms' tumour. However spinal canal involvement is not a common presentation of Wilms' tumour in childhood. Early diagnosis of the tumour is therefore essential to improve the neurological status of the patient.

Methods. A case report and review of the literature is undertaken.

Results. A 3 year old Chinese boy presented with abdominal distension and intermittent fever of four months duration. Physical examination revealed a large left renal mass and computerized tomography scan of his abdomen (CT), confirmed the presence of a left renal mass extending to the paravertebral gutter, with no obvious lymphadenopathy. The cava was free of any tumour and there were no metastatic pulmonary nodules on a chest CT. Urinary VMA was negative. A Fine Needle Aspiration Cytology (FNAC) was performed and over the next few days whilst awaiting the results he developed acute urinary retention and paraplegia with a muscle power of 0/5. The urgent CT of his abdomen revealed a dumb bell intraspinal tumour extending from T11 to L1 with cord compression. He was therefore treated along the N6 protocol for Neuroblastoma with Vincristine, Adriamycin and Cyclophosphamide. His muscle power improved from grade 0/5 to 2/5, but he still suffered urinary retention. The FNAC depicted spindle shaped cells and chemotherapy was continued. A repeat MRI two weeks following chemotherapy revealed complete resolution of the intra spinal tumour. A month following chemotherapy he underwent a radical left nephrectomy. The histopathology of the kidney revealed a predominantly necrotic tumour with spindle shaped cells. There was no anaplasia and it was reported as a Wilms' of favourable histology. He has since been commenced on the SIOP-WT 2001 protocol.

Conclusions. Early histopathological diagnosis of these tumours with intra spinal spread and neurological dysfunction is essential to institute appropriate chemotherapy. As intraspinal spread in Wilms' is rare the question that arises is whether one should classify these tumours as Wilms' or assign them a separate category.

A42
Three children with rare bladder tumors

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Background. Bladder masses in children are extremely rare pathologic lesions and may be of benign or malignant origin. The symptoms of tumor manifestation vary considerably. They are often unspecific and include abdominal pain, urinary voiding abnormalities, sterile hematuria, dysuria, and a palpable mass.

Methods. Retrospective analysis of patient's charts.

Results. We present three illustrative cases: The first is a four year-old boy with a benign inflammatory myofibroblastic tumor of the bladder. The other two cases are children with malignant lesions including a nine year-old boy with an inflammatory fibrosarcoma and a four year old boy with an embryonic rhabdomyosarcoma. In the first case, the lesion was diagnosed using ultrasound, MRI, and both cystoscopic and ultrasound guided transcutaneous needle biopsy. Only the transcutaneous biopsy allowed for conclusive diagnosis. The boy was then treated conservatively by steroids and cotrimoxacol. With this therapy, the tumor shrank significantly from 4.5 × 4.5 × 3 cm to actually 1.7 × 1.4 × 0.4 cm. Diagnosis of both malignant tumors was obtained by ultrasound, MRI, and transcutaneous needle biopsy. In the fibrosarcoma case, cystoscopic biopsy was also performed, but not diagnostic. The patient with the inflammatory fibrosarcoma was treated by preoperative chemotherapy followed by complete surgical tumor resection (R0). The rhabdomyosarcoma patient had preoperative chemotherapy and radiotherapy, then a complete surgical tumor resection (R0), followed by chemotherapy. Up to date both cases are recurrence free.

Conclusions. A stepwise diagnostic approach is mandatory, includes ultrasound, MRI or CT, cystoscopy and biopsy, and usually allows prompt and conclusive diagnosis. Therapy varies depending on histological findings. Ultrasound guided transcutaneous needle biopsy is a safe and reliable method to obtain a conclusive histology.

Rare tumours II

A43
Unusual case of prostatic rhabdomyosarcoma

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Background. Urogenital rhabdomyosarcomas (RMS) constitute from 4% to 8% of the total malignant solid tumors of childhood, second only to lymphomas, central nervous system tumors, neuroblastomas, and Wilms tumors. Usual symptoms are urinary obstruction, hematuria, or both or as a large pelvic mass that causes abdominal pain and intestinal obstruction. We present a case of prostatic rhabdomyosarcoma with an unusual presentation.

Methods. An 11 years old boy was referred to the pediatric Surgery Clinic in Novi Sad by the family physician due to penile and scrotal edema. Patient was treated initially treated ambulatory as an allergic edema with antihistaminics. As there was no response he was referred to the Pediatric Surgery Clinic.

Results. Antihistaminic therapy was continued. Urinanalysis was normal. Voiding and stooling was normal. An allergic, vascular, infective or dermatological abnormality was excluded. Rectal examination revealed presence of soft tissue mass at the region of prosthetic urethra. An ultrasound and CT examination showed presence of a soft tissue mass 8 × 6 cm in diameter at the pelvic floor. Parailiac and retroperitoneal lymph nodes were enlarged. Kidneys, liver spleen, adrenal glands and pancreas were normal. Transrectal biopsy of prostate was performed and the diagnosis of prostatic rhabdomyosarcoma was confirmed. Cytostatic therapy (vincristine, actinomycin D, cyclophosphamide) was introduced. After an initial good response (short remission) metastases in the lymph nodes of the neck appeared. Another cycle of chemotherapy was introduced but without success. Patient died within 6 months from the diagnosis.

Conclusions. Although usual symptoms of urogenital rhabdomyosarcomas are urinary obstruction, hematuria, abdominal pain and intestinal obstruction, one must bear in mind that the disease might present with unusual symptoms.

A44
Lipoblastoma and lipoblastomatosis: A report of three cases and review of the literature

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Background. Lipoblastoma and Lipoblastomatosis are rare benign tumours of white fat tissue, which occur almost exclusively in infants and children. Some 100 cases have been described in the literature. 80 to 90% are occurring in children younger than 3 years. The exact histogenesis remains uncertain, and hamartoma or persistent embryonic tissue have both been discussed. Recent cytogenetic studies have shown clonal chromosomal aberrations, thus suggesting a neoplastic origin. This could make lipoblastomatosis more distinguishable from myxoid liposarcoma.

Methods. Three cases of our hospital, an almost one-year-old girl, a full-term male infant and an one-year-old boy, are reported and a review of the literature is given.

Results. Nine month after total excision of the lipoblastoma, the little girl developed tumour recurrence, with unexpected histopathological maturation. In the case of the neonate, lipoblastomatosis with infiltration of the surrounding muscles and involvement of nerves was found. In the one-year old boy the resection of the lipoblastoma has not been performed completely, thus radiologic routine controls are still necessary.

Conclusions. Lipoblastoma and lipoblastomatosis are benign neoplasms, but especially the lipoblastomatosis may recur more often after surgical excision. The diagnosis of this tumour was made by the pathologist, but the histopathological picture may be indistinguishable from myxoid lipocarcoma. Recent studies describe rearrangements of chromosome 8q11-q13 as a new discriminative marker.

A45
What predicts recurrence of deep fibromatosis in children?

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Background. Fibrous tumors of infancy and childhood vary from benign to malignant. The most important subgroup of these tumors is deep fibromatosis. Surgery is the corner-

stone of its treatment. Aim: to identify clinical and pathological features predictive of recurrence.

Methods. Between 1988–2004, 37 children were treated at our Center for fibrous tumors. Histopathology showed 22 deep fibromatosis (aggressive fibromatosis and infantile fibromatosis), 6 myofibromatosis, 3 digital fibromatosis, 3 fibrous hamartoma, 2 fibromatosis colli, and 1 fibromatosis plantaris. Only deep fibromatosis was included in this retrospective analysis. Histopathology was re-evaluated. Data regarding age, sex, localization, primary treatment, collagen content, cellularity, mitotic index, necrosis, inflammation, myxoid changes, macro- and microscopical margins were correlated with the risk of local recurrence (LR).

Results. Surgery was the only treatment in 17 (7 LR), surgery followed by chemotherapy in 2 (2 LR), chemotherapy alone in 2 (both have stable disease), and surgery followed by radiotherapy in 1 (no LR). After a mean follow up of 41/4 yrs (range 0–16 yrs), 17 were in complete remission (CR). LR developed in 9/22 (40%). Of all LR, the male:female ratio was 7/16:2/6, site: limbs in 8/12, and trunk in 1/7. The macroscopical margin had been classified incomplete in 5, and marginal in 4. In all LR, microscopy of the primary resection margin was positive, whereas all excisions with negative margins were in CR. There was no correlation between histopathological aspects of the tumor, and LR.

Conclusions. Resection with negative microscopic margins is the goal of treatment. Microscopic evaluation of the resection margin is mandatory. All other parameters i.e. sex, age, site, histopathological features, etcetera, had no predictive value for LR in this series.

A46
Infantile fibromatosis of the tongue

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Background. Fibromatoses are tumour-like proliferations, which can be categorized in terms of their biological behaviour between benign fibrous tumours and fibrosarcomas. They show typically aggressive local infiltration, do not metastasize and have a high tendency for recurrence. Spontaneous regression is documented. They might appear in any age. The most common sites of appearance are the dermis or the subcutaneous tissues of the head, the neck and the trunk.

Results. Case report: We report the case of a 3 month old boy with a large tumour of the tongue. Measuring 2 cm in diameter and overreaching the tongues midline the tumour was excised macroscopically in toto. Histological examination suggested the tumour reaching the resection margins. It showed diffuse infiltration of spindle-cell-shaped cells into submucous stroma and the interlacing fascicles. Immunohistochemistry revealed a strong expression of Vimentin and expression of CD34. Based on the histological and immunohistochemical features the diagnosis of an infantile fibromatosis of the tongue was made. On follow-up no recurrence of the fibromatosis neither on physical examination nor on MRI scans was found. The clinical appearance, treatment and outcome are described and compared to the literature.

Conclusions. Although very rare infantile fibromatosis should be taken into consideration in solid tumours of the oral cavity and the tongue. Characteristic features enable to distinguish from other fibrous lesions and from benign and malignant smooth muscle tumours.

A47
Extra abdominal desmoid tumor. Fibromatosis aggressiva of the glutei minor muscle

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Background. Fibromatosis aggressiva (desmoid tumor) is a very rare nonmetastazing soft tissue tumor, which accounts less then 0.03% of all neoplasm. In some author's opinion desmoid tumor represents grade I fibro-sarcoma. Extra abdominal desmoid tumors are solitary, painless, poorly circumscribed, and deep soft tissue masses. They are infiltrating muscle, which results in a loss of function in involved muscles. If nerve structures are involved, pain occurs.

Methods. Eight years old boy, who is a dance player, had intermittent pain during dancing. Pain had started ten days before, located in the right knee region. His parents noticed some limping during the gait. On the clinical examination, there was swelling over the right upper gluteal region. The right Trendelenburg's sign was positive. Leg length inequality on orthoroentgenography was 12 mm. Sedimentation rate was 13. Other laboratories value C-reactive protein, LD (lactate dehydrogenase), CK (creatinin kinase) and ALP (alkaline phosphatase), were normal. Ultrasound hip examination showed tumor of the glutei minor muscle. On MRI examination we found soft tissue tumor of glutei minor muscle, which was 80 × 50 mm in a diameter.

Results. We performed radical surgical excision of the neoplasm. Histopathologically, desmoid tumor was confirmed. Over one year follow up period there were no sign for the recurrence.

Conclusions. Unusual feature of this case was leg length inequality. On our opinion desmoid tumor was a local factor increasing growth rate.

A48
Epithelioid hemangioendothelioma of external iliac vein with multiple metachronous localizations

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Background. Epithelioid hemangioendothelioma (EH) with metachronous liver and lung localizations are rare. The therapeutic concepts are controversial: from lung-liver transplantation to a wait and see concept. This case illustrates the difficulty and pitfalls of making the correct diagnosis and treatment.

Methods. 11-year old girl was presented with left leg swelling since 3 weeks. She had signs of ileo-femoral venous obstruction, because of lymph nodes in the left groin (US and CT). Bartonella IgG was positive and she was treated with Doxycyclin. 6 weeks later no change of the size of lymph nodes in CT. Laparoscopy and exploration of the left groin detected tumor infiltration into the external inguinal vein. Pri-

mary histology was juvenile myofibrosis and extramural expert consultation give diagnosis of EH. At this time bilateral lung metastases were found in CT and map like structures in the liver was seen. Thoracoscopic biopsy of lung metastases and total resection of the tumor in the left groin with interposition of the vein was performed.

Results. CT of thorax and MRI of the liver 4 weeks later shows no change of lung metastases. In the follow-up diffuse liver infiltration was again seen in MRI, but never in US. Laparoscopic biopsy of liver lesions confirm EH. Lesions in the thorax and liver were stable for 2 years in the follow up, and afterwards new lesions in thorax and progress of liver lesions were recognized. The girl was without any symptoms. In this case we did not perform any systemic therapy over 2 years, because there was no deterioration, but in the follow up a slightly deterioration was recognized and interferon therapy was finally started.

Conclusions. In the literature approximately 50 cases are reported. The overall mortality is less 20% at 3 to 5 years. Spontaneous regression of pulmonary EH is described in asymptomatic patients after 5, 13 and 15 years. Patients with pleural effusion and clinical symptoms died within 1 year. Adjuvant chemotherapy, radiation therapy or both have no proven benefit. Interferon is described, but with severe complications in children.

A49
Kaposiform hemangioendothelioma: a rare vascular tumor which requires multidisciplinary therapy

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Background. Kaposiform hemangioendothelioma (KHE) is a rare neoplasm of the skin, deep soft tissue and bone with aggressive behaviour to surrounding tissues. Surgical intervention is almost limited to biopsies and casual reduction of the tumor mass. Though application of cortisone and interferone maybe followed by chemotherapy (CWS) and could contribute to tumor reduction or prevent further aggravation.

Methods. In 6 patients with KHE the tumor was located on thorax, shoulder, femoral, face, neck and sternum. Diagnostic biopsy was done in all patients. In 2 patients surgical therapy was possible; we performed complete resection of the sternum and extensive femoral tissue resection with following skin transplantation. Therapy with Cortisone and Interferone was applied to 2 resp. 4 patients. Due to failure of regression, chemotherapy was installed (CWS).

Results. Tumor reduction could be achieved in 2 patients. 1 patient showed primary regression with then alternating phases of increase and restitution. 2 patients showed a very little reduction of the tumor mass, but they obtained better clinical constitution.

Conclusions. Complete surgical therapy of KHE without mutilation is restrained to rare cases, mostly a multidisciplinary proceeding is recommended including chemotherapy, cortisone and interferone.

A50
Kasabach-Merrit syndrome in giant hemangioma of the lower limb – case report

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Background. Kasabach-Merrit syndrome is an association of vascular lesion and consumptive coagulopathy. We report a case of a male infant with a large vascular tumor of the left lower limb, accompanied by consumptive coagulopathy, revealed on the very first day of life and severe episodes of bleeding.

Methods. A newborn male, 39 GW, weighing 3500 g, was admitted at the neonatal surgery department, in the first day of life, with diagnosis of giant hemangioma, deforming whole thigh, knee region and the upper part of the left calf, showing signs of consumptive coagulopathy with progressive thrombocytopenia, moderate anemia and no infection. Steroid therapy with Prednisone 5 mg/kg, was administered, with no benefit for two weeks. Suddenly, in spite of therapy, the patient suffered of severe, life threatening bleeding into the tumor and surrounding tissue, so the whole region from the navel, anterior abdominal wall, both inguinal regions, external genitalia and the whole left leg, including ankle and foot, were dark blue, with enormous hematomatous swelling.

Results. Alpha 2a Interferon was administered on the very first day of bleeding episode, in a dose of 1 million iu/m2 sc, as well as ciclokapron, dipiridamol and a great deal of resuscitation means, blood and plasma derivates. The neonate survived the crisis. Next month, another episode of bleeding was much less severe and afterwords, not only the hemangioma locally, started shrinking every day, but also the general condition was more stable, although with continuous thrombocytopenia and other signs of consumptive coagulopathy for several months of the next follow up, but with gradual improvement. The therapy was successful, continued for almost a year. Several months after completion of treatment, hemangioma got reduced completely, the infant started to walk and grow normally.

Conclusions. Due to our experience in a particular case, our opinion is that Alpha 2a Interferon (Roferon) is extremely useful in treatment of large hemangiomas complicated with Kasabach-Merrit syndrome. Disadvantage of such treatment is its long duration and expensiveness of interferon. We did not observe any serious side effect during the treatment.

A51
The multi-step therapy – experiences in treatment of lymphangioma of infancy

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Background. During the last two decades therapy of lymphangioma is discussed. Radical exstirpation further more is golden standard. Morphology and localization of many lym-

phangioma prevent a successful surgical approach or are followed by severe damages on important surrounding structures. In the departments of pediatric surgery in the University hospital Benjamin Franklin and the St. Joseph pediatric hospital, a multi-step therapy was developed completely integrating the laser therapy. Above all this kind of therapy the special requirements of localization and morphology of every single lymphangioma is considered.

Methods. From 1984 to the end of 2004 we treated 200 children with lymphangioma, in 155 children laser therapy was used. Different forms of application were used, most frequently the ITT and the excision with laser. The multi-step therapy is using 4 steps, which take place within a time framework depending on the different findings of every lymphangioma. Step 1 contains the volume reduction and the therapy of complications caused by the lymphangioma, such as restriction of trachea due to compression. Step 2 is for the treatment of the microcystic parts of the lymphangioma. In a 3rd step the aim is the reestablishement of the anatomical conditions. The last step includes the fibrosis of the remaining microcystic parts of the lymphangioma after a treatment free interval.

Results. The median number of treatment was 2 in our patients, in individual cases up to 19 sessions were necessary. Most frequent postoperative complication was the seroma, which however never needed a treatment. 2 children suffered of spontaneously reversible paresis. Three children received postoperative tracheostoma which all could be closed by now. In 15 % of the children another treatment was needed after end of therapy. Only 8% were recurrences, the rest aroese by filling of still existing cysts of the lymphangioma. End of 2004 145 children were free of complaints and had no recurrences.

Conclusions. The multi-step therapy permits an individual therapy with consideration of crucial factors and the free combination of all treatment options especially the laser treatment. Thus is offered a therapeutic concept which enables even the treatment of inoperable regarded lymphangioma. Additionally the advantages of the minimally invasive treatment are used with the ITT.

A52
Metastasis of testicular teratoma – a rare differential diagnosis of retroperitoneal tumor in children

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Background. Case report: In 2005 a 17-year-old boy from Kosovo presented to us with a huge abdominal tumour. He had been suffering from right-sided flank and abdominal pain for two months. Furthermore he reported sweating at night and a total weight loss of 6 kilograms within four months. Abdominal ultrasound and CT in Kosovo revealed a retroperitoneal mass and the patient was transferred to our hospital for surgery.

Methods. The boy presented in a fair general condition. A solid, painless tumour in the right hemiabdomen was palpable. The spleen was not enlarged. Testes were both descended, with the right testicle being slightly bigger than the left one. No pain on palpation. Further physical examination – especially lymph nodes – did not reveal any abnormalities. On the abdominal CT-scan the tumour was localized in the retroperitoneum, measuring 17 × 10 × 30 cm in diameter. No infiltration of blood vessels, which were partially displaced or other organs was seen. The right kidney was compressed by the tumour but appeared otherwise normal. No pulmonary or mediastinal metastases were detected on the CT of the thorax. Surgery was performed and biopsies taken as the tumour was not resectable due to dissection from the abdominal blood vessels being impossible. Histological examination showed a malignant germ cell tumour (90% embryonal carcinoma, appr. 5% differentiated teratoma, appr. 1% yolk sac carcinoma).

Results. After orchiectomy of the right testicle a small cystic teratoma was diagnosed – without evidence of undifferentiated cells. Thus the patient was staged as pT1, N3, M0, S2. He is currently undergoing chemotherapy (3 courses BEP, 3 courses PEI), which lead to a considerable decrease in tumour seize. Tumour-related neuronal pain has vanished completely. After completion of chemotherapy resection of tumour-remnants is planned.

Conclusions. Although testicular multicystic teratomas lead to metastases only in a small percentage of patients it should always be taken into account in young men presenting with a retroperitoneal tumour. Actual therapeutic guidelines suggest: 1. inguinal orchiectomy, 2. chemotherapy, 3. second-look-surgery to detect viable tumour-cells and complete resection of tumour if possible and 4. additional chemotherapy if histopathology reveals vital tumour remnants. In longterm-follow-up LDH, AFP and β-HCG should be monitored as sensitive tumour-markers. Additionally imaging studies as MRI or CT scans should be performed regularly. As differential diagnosis lymphoma, soft-tissue sarcoma, myosarcoma, myoblastoma, ganglioneuroma, neuroblastoma and PNET should also be considered.

A53
Ultraradical resection of thyroid carcinoma: surgical heroism or mutilation?

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Background. In medullary thyroid carcinoma, surgery is the only validated therapeutic option. We report the case of a 12 year old girl with a metastatic and locally advanced medullary thyroid carcinoma. At the time of presentation, the patient had severe obstructive respiratory failure due to a thyroid tumor which had grossly infiltrated a long tracheal segment, both recurrent nerves and the esophagus. Furthermore, large bilateral cervical lymph node metastases reached up to the base of the skull.

Methods. In July 2004 we performed a multivisceral resection of the thyroid including a 4 cm segment of the trachea, the recurrent nerves, the anterior esophageal wall and an extended radical bilateral neck dissection. Additionally, a tracheostomy and a gastrostomy had to be created. Macroscopically, the tumor could be completely resected. Histopathological staging revealed a medullary thyroid carcinoma UICC IV (pT4, pN1 [56/82], L1, V0, M0, R1). The girl was included in

an experimental study protocol of a combined high dose radio-chemotherapy.

Results. She has been followed up in 3 monthly intervals and has no evidence of residual disease or recurrence today, 18 months after surgery.

Conclusions. With view to her permanent functional impairments we will discuss whether this ultra-radical surgical approach is justifiable.

A55
Tumors of the ovaries as late extramedullary recurrence of acute leukemia

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Background. Isolated extramedullary relapse of acute leukemia is rare. We report on two girls with this type of recurrence by presenting the case histories, and we will discuss the role of surgery in these cases.

Methods. Case 1: Pre-B-ALL (acute lymphoblastic leukemia) was diagnosed in this girl at the age of three years. She was immunologically typed as "low risk of recurrence". Treatment was done. Methods: according to the CoALL 82-protocol, which is one of the two usually used treatment protocols for ALL in Germany. Bone marrow relapses occurred at the age of 7 and 9 years, respectively. They were re-treated by similar and even more aggressive chemotherapy at the age of 11 years and eight years after the initial ALL diagnosis. She presented with a huge abdominal tumor of undeterminable organ origin. By open biopsy of the mass, lymphoblastic cells were diagnosed and an abdominal tumor-like leukemia recurrence was ascertained. Chemotherapy and low-dose radiotherapy succeeded in shrinking the tumor mass, unless it became operable. Surgery was done on a small, but not further shrinking tumor, which finally revealed the mass to have originated from one of the ovaries. Salpingo-oophorectomy was done, followed by short term chemotherapy after the operation. 12 years after this event and 20 years after the first diagnosis of ALL, the young women is in complete continuous hematologic remission.

Case 2: This 14 year old girl, in whom pre-B-ALL was diagnosed, and who therefore also had a low overall probability of recurrence, was treated according to the successor treatment protocol CoALL 06-97. After having achieved complete hematologic remission in the bone marrow after 4 weeks, she stayed in remission for 18 months. She then developed signs of an abdominal tumor: painless swelling. Ultrasound and CT showed masses originating from both ovaries, the larger extending over 5 through 8 cm. We performed a laparoscopy, laparoscopic lymph node staging from the parailiacal to the bifurcation area, and we took biopsies from one ovary and from a lymph node. Contamination with tumor cells by biopsy could be neglected in these suspected recurrences of systemic to be treated hematologic diseases: cytologic examination of the smears revealed ovarian relapse of ALL. During operation, acute findings were interdisciplinary discussed and decided on; further, bone marrow puncture was done in the operation room. Postoperatively, long-term chemotherapy was started following BFM protocol for ALL-recurrence. Regression of tumor masses were monitored by ultrasound, and after 6 months control laparoscopy was done.

Conclusions. By this intensive monitoring additional radiation as a potential treatment modality was avoided and fertility probably preserved. Laparoscopy enables to explore abdominal tumors, to do staging procedures, and gives a very good visualisation of intraabdominal tumors. In selected cases biopsies can be done; the indication can intraoperatively be discussed with pediatric oncologists. Laparoscopic exploration of abdominal gives safer results of histologic specimen compared with percutaneous biopsy, and is it less invasive compared with open biopsy.

A56
A case of mucinous tumor of borderline malignancy in a 14 year old girl

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Background. Mucinous ovarian tumors are among the most difficult ovarian neoplasm for surgical pathologists to interpret. 20% of primary ovarian mucinous tumors are borderline tumors, non invasive carcinomas or invasive carcinomas; the reminder are cystoadenomas. Mucinous borderline tumors (MBT) are subdivided into two categories: intestinal type and endocervical-like. The intestinal type is by far the more common and the prototypical borderline mucinous tumor. These tumors present from 80 to 90 % as stage 1.

Case report. A 14 year old girl presented with a history of fever up to 39° C (5 days) with no initial abdominal complain. The clinical examination showed an abdominal distension and a painful tender mass was palpated. At that point the patient revealed that she had already noted the swollen abdomen but she had hidden it, fearing a pregnancy. The gynaecological history was characterized by the administration of "the day after" pill 4 months ago followed by 1 month of conventional estrogenic anticonceptional treatment. Menstruation had disappeared two months before. Ultrasound showed a pluriconcamerated cystic mass, of about 19 cm in diameter of ovarian origin. Tumor marker (a feto protein, βHCG, CA 19.9, CA 125, CEA) were measured with a rise in CA125 (245 U/ml) and CEA (17.95 ng/ml). The girl was operated and a huge mass of about 25 × 20 × 10 cm, weighting 2600 g and originating from the right ovary was removed, with the omolateral salpinx, the adherent omentum and peritoneal liquid. Biopsies were performed on the controlateral ovary. The final histology was "mucinous tumor of the ovary of borderline malignancy". The peritoneal fluid showed the presence of few atypical cells, while the other biopsies were negative for malignant cells. The tumor markers declined to normal. Few weeks later, a laparocele was noted and the girl was reoperated: in that occasion a second look showed residual ascite and few granulation on the interstinale serosa, but the histological findings were those of inflammatory reaction and no more atypical cells were found in the peritoneal fluid. At 4 months follow up the girl is free of disease. All tumor markers are negative.

Conclusions. Mucinous ovarian tumors of borderline malignancy are rare in the 2nd decade of life, as in our case: the

peak incidence is between the fourth and seventh decades and they are usually low stage tumors with a good prognosis after surgery alone. The problem with these neoplasms is to obtain a correct histological diagnosis, based on a wide sampling of the mass, which is the guide for eventual adjuvant therapy. While in women who already had children the administration of chemo or radio therapy can be proposed with minor worries about fertility, the same approach in case of a nullipara adolescent girl, is always a major issue of concern.

A57
Extraspinal myxopapillary ependymoma

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Background. Sacrococcygeal tumors outside the newborn period are uncommon. We report an extraspinal myxopalillary ependymoma in a schoolchild.

Methods. The 9-year-old boy was admitted with a circumscript subcutaneous tumor of the sacral region. There was no pain or neurological sensations. Primary we suspected an organised hematoma. Serum alpha-fetoprotein was not elevated. The sonographical examination showed an echogene, inhomogene and sharp bordered solid RF. The additional MRI presented a homogeneous tumor without contrast medium enhancement. The Lesion was excised en block including the top of the os coccygis. Histology revealed a myxopapillary ependymoma without bone involvement. Metastases were excluded.

Results. Although the subcutane EEP is a possible neoplasm of the sacrococcygeal region in children, the primary diagnosis failed often in case of rarity. In a metaanalysis of 22 children in the literature pilonidal cysts, epidermoid cysts or lipoms were the preoperative diagnosis. As possible localisations are described presacral ventral, dorsal of the sacrum in the subcutis, paraovarial and the mediastinum. The EEP have no connection with the spinal channel. These tumors usually grow slowly. Histogenetical the EEP develop from glia cells. The origin are coccygeal medullare remnants. The therapy is the complete resection of the lesion taking care of the rules of oncological surgery. Extraspinal myxopapillary ependymoma have the potential risk to develop metastases and local relaps. 11 children in a metaanalysis were affected by local relaps or metastases.

Conclusions. In danger of metachronic local relapse and or metastases a long term follow-up is recommended.

A58
Selected cases of rare benign but still life-threatening intracranial tumors in children

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Background. Primary tumors of the central nervous system are the most common type of solid neoplasm in children. Among different specific tumor types, gliomas, primitive neuroectodermal tumors, and ependymomas are the most common pediatric brain tumors and account for about 75%. But there are also rare tumors which may increase intracranial pressure when obstructing cerebrospinal pathways. In these life-threatening instances, immediate neurosurgical interventions are necessary.

Methods. We are presenting three different types of rare benign intracranial tumors: neurozytoma, pineocytoma, and choroid plexus papilloma. They account together for about 5% of pediatric brain tumors. Due to their sites in or very close to the ventricular system, they occur more likely with signs of increased intracranial pressure caused by obstructive acute hydrocephalus. The variability of associated magnetic resonance signal characteristics, MR multiplanar slices, 3 dimensional reconstructions and different surgical approaches by substitution of neuronavigation as well as surgical strategies for the different tumors are demonstrated and discussed.

Results. Five children, ranging in age from 1 to 18 years, were operated on neurozytomas (two cases), pineozytomas (two cases), and choroid plexus papilloma (one case). Both neurozytomas were located in the frontal horn of right lateral ventricle and were extirpated through a transfrontal approach. Both pineozytomas were resected through a supracerebellar-infratentorial approach, and the choroid plexus papilloma was removed through a transparietal approach. All tumors were totally resection. Operative complications were minimal. One patient after resection of a neurozytoma had a mild transient hemi paresis and one patient after resection of a pineozytoma had a transient parinaud syndrome. Both children with neurozytomas and one with pineozytoma needed after tumor resection a shunt implantation because of persistent hydrocephalus. None of the patients had a recurrence during the follow-up period, which ranged from 4 year to 9 years in duration.

Conclusions. The goal of treatment of all these tumors is complete excision. Once a total resection is achieved, the prognosis of the patients is excellent and no additional tumor specific therapy is necessary. Malignant transformation or recurrence is almost nonexistent. Despite total resection, hydrocephalus may recurr or persist and needs special attentive long-term care.

A59
Stereotactic neurosurgery, 3D imaging and neuronavigation in brain operations

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Background. Stereotacic surgery is a method in neurosurgery and neurological research for locating points within the brain using an external, three-dimensional frame of reference usually based on the Cartesian coordinate system. Image-guided surgery has been employed in neurosurgery since the mid-1990s. This new technique relies on a powerful computer system, which assists the surgeon in precisely localizing a lesion, in planning each step of the procedure via a 3D model and in calculating the ideal access to the tumor before the operation.

Methods. In our department we use the Radionics CRW (Cosman-Roberts-Wells) stereotactic system, which is the most widely used frame-based system. There are over 1,500 CRW systems placed in neurosurgical institutions worldwide. The system enables neurosurgeons to position fine instruments in the brain with sub-millimeter accuracy, making the CRW a versatile tool

for a variety of neurosurgical applications. For neuronavigation the Medtronic stealth station is used. Data acquisition runs via MRI and a special workstation directly to the OR.

Results. Stereotactic techniques were originally used to create accurate maps of the human brain. Its first clinical application occurred at the end of World War II. Stereotactic guidance was used to increase the accuracy of brain operations. Despite the initial attempts, stereotactic neurosurgery did not become popularized until the late 1970's when dramatic improvements in neuroimaging occurred. At this time computed tomography, CT, redefined the ability of a physician to identify diseases within the brain. In the last 10 years 114 stereotactic biopsies were performed in our departement. Neuronavigation enables the surgeon to plan smaller sized and better centered skin incisions and craniotomies and to approach the target lesion with less dissection of intact brain tissue. Despite more radical removal of lesions the overall invasiveness of the operation was decreased. The tumor and its surroundings can be viewed from different angles and in relation to landmark structures. During the operating procedure, the movement of the instruments in use inside the brain can be tracked on the monitor with a precision of 1–2 millimeters, through which damage to healthy tissue and to critical areas can be avoided as much as possible. In our department 714 computer navigated operations were performed in the past ten years, including gliomas, menigeomas, metastases, AVM, cavernomas, aneurysm and pituitary adenomas.

Conclusions. Stereotactic biopsies and computer assisted navigated brain operations are routine in our department. Image-guided surgery assists the surgeon during all phases of the operation. This allows the surgeon to keep the operation minimally invasive and to avoid critical structures of the brain. Thus, the risks of paralysis or other impairments after surgery are minimized and recovery time is considerably reduced.

Urology

A60
Role of primary health services in detection of cryptorchidism: quality improvement through a multifunctional system

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Background. Cryptorchidism represents one of the most common congenital malformations in males which carry various complications. Considering significant decreases in gastroenteritis and pneumonia after WHO protocols in recent years, we made our minds to design similar protocols in countries which have referral systems in their health services. The aim of present study is to accurately assess prevalence of cryptorchidism and the role of mid-wives and general practitioners in detection and referral of boys with undescending testes for surgery.

Methods. Of 9019 consecutive male live births, 6237 (69.1%) were examined for cryptorchidism at primary health services by mid-wives thereafter subgroup of cases with the persistence of cryptorchidism was re-assessed during a follow-up examination first by a general practitioner in the second level and then was referred to a surgeon if undescending testes was suspected in the third.

Results. 84 cases (35%) of diagnosed once in primary health services which was carried out by mid wives was accurate, further more GPs had reconfirmed 186 of referred cases. It could demonstrate acceptable efficiency (45% correct detection) in this level. Prevalence of undescending testes was 3% in open population.

Conclusions. In order to establish reliable information on the detection of cryptorchidism, all male births in the open population have to be examined systematically. We have shown that measurable improvements in early detection of cryptorchidism can be achieved through a combination of education of primary care, GPs and expert specialists team and agreed local standards for referral.

A61
From boy to man: long term fertility results of cryptorchism

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Background. Delayed orchidopexy for undescended testis has been associated with abnormal testicular histology, but its functional effect on later fertility is unknown.

Methods. All patients (now > 18 years old) who underwent orchidopexy and testicular biopsy during their first two years of life at our department were contacted. Until now 18 patients responded and were assessed using a sexual history, physical examination, hormonal – and sperm – analysis. We matched histological findings of the testicular biopsy (present or absent of Ad spermatogonia) with their total number of sperm.

Results. Six patients had bilateral and 12 patients had unilateral cryptorchism. If Ad spermatogonia were present (8/18) in testicular biopsy, 6 (75%) had a total sperm count of 40 × 106/ejaculate or greater. By contrast, if Ad spermatogonia were absent (10/18), all patients had abnormal spermiograms. There was no correlation between the total number of sperm and the levels of follicle-stimulating-hormone (FSH) or the testicular localisation.

Conclusions. Our results showed that only in 33% of all cases a normal spermiogram could be found. The transformation of gonocytes into Ad spermatogonia might to be crucial for the male development and seems to be the best indicator of future fertility.

A62
First case report of a patient with Denys-Drash syndrome and prostatic utricle

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Background. We report about a boy with diagnosed Denys-Drash syndrome. Denys-Drash syndrome is a rare symp-

tom complex associated with nephrotic syndrome, male pseudohermaphroditism, and extra Wilms tumor risk. It is ascribed to a mutation of the WT-1 gene.

Results. In 7/02 a mature born boy attracts attention postpartum because of a genital malformation in terms of a male pseudohermaphroditism with a bilateral abdominal retention of the testis' as well as penoscrotal hypospadias with micropenis. From 4/03 to 10/03 gradual mobilisation of the testis' and displacement into the upper scrotal compartment as well as an erection and reconstruction of the penis is accomplished. 4/04 subsequent to a viral infection progressive oedemas and faintness occur. Admission into hospital due to terminal renal failure. Beginning of renal substitution therapy because of total anuria by peritoneal dialysis. After exclusion of inflammatory or immunological causes molecular genetic assurance of Denys-Drash syndrome. In 6/05 a bilateral nephrectomy and an allogenic kidney transplantation with antireflux-ureterocystoneostomy using the Gregoire procedure on the right side is carried out. Primary functioning graft. Two month after transplantation first pyelonephritis caused by Pseudomonas aeruginosa. Sonographical evidence of a retrovesical, plain-circumscribed structure of low signal intensity with a diameter of 18×12 mm and a length of 20 mm. In the context of an urethrocystoscopy demonstration of a prostatic utricle which empties in the area of the colliculus seminalis into the urethra. In the micturating cystourethrogram no contrastation of the utricle during miction, demonstation of a vesicoureteral reflux III°–IV° into the ureter of the renal transplant.

Conclusions. Despite a reinfection prophylaxis two urinary tract infections were seen (Pseudomonas and Proteus), whereas the prostatic utricle just like the vesicoureteral reflux have to be discussed as germ pools. In case of a possible resection of the utricle the problem of finding an advantageous access way for the operation occurs. It can't be operated from ventral transperitonal as the renal translant is located retroperitoneal on the right side of the small pelvis. A ventral transvesical or a perineal access would be possible. This is the first report of a patient with Denys-Drash syndrome and prostatic utricle.

A63
Renal transplantation in children with severe malformations of the lower urinary tract – own experience

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Background. Patients with chronic renal insufficiency due to severe malformations of the lower urinary tract such as bladder agenesia and neuropathic bladder require specific surgical preparation of the bladder (augmentation cystoplasty or ileal conduit construction). The first kidney transplantation to neuropathic bladder was performed in February 2001 in our hospital.

Methods. The kidney was transplanted into an ileal conduit in two cases: one with bladder and right kidney agenesia and other operated by reason of imperforate anus in newborn period (with non-compliant high pressure neuropathic bladder due to abnormalities of sacrum and lumbal segment of spinal column). Ileal conduit was constructed before or during transplantation. The kidney was transplanted into a augmented neuropathic bladder in 3 cases. Bladder was emptied by using CIC. In one case we undergone ileocystoplasty and simultaneously performed construction continent appendico-caecostomy using Malone's technique (MACE procedure), in one colocystoplasty with construction continent catheterizable channel urinary and faecal and the other ureterocystoplasty using both ureters.

Results. Very good results of transplantation in children with bladder agenesia and neurogenic bladder were achieved. The most common complication was urinary tract infection, mostly asymptomatic bacteriuria with no impact on renal graft function. Good emptying of low-pressure bladder or urine reservoir is the most necessary condition for good graft function.

Conclusions. 1. Neuropathic bladder with abnormal high intravesical pressure requires preparation before transplantation surgery for its change for low pressure reservoir with adequate capacity usable for CIC. 2. Function of renal transplant is normal (correct) in all patients in spite of stable or recurrent infections of the urinary tract. 3. These infections have often asymptomatic character. 4. It seems that graft's good function is provided by keeping an efficient urine outflow without dilatation of its collecting system.

A64
Continence rate and spontaneous voiding after neonatal reconstruction of bladder exstrophy

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Background. To determine continence rate and spontaneous voiding after neonatal reconstruction of bladder exstrophy (BE) without formal bladder neck reconstruction (BNR) in primary reconstructed patients treated with clean intermittent catheterization (CIC) following closure.

Methods. Between 1987 and 2003, 15 consecutive cases (8 boys, 7 girls) with BE were reconstructed neonatally. Reconstruction focussed on bringing bladder neck and proximal urethra intra-abdominally and meticulous closure of pelvic floor muscles around the urethra. Three weeks postoperatively, CIC was started until potty-training age. Bladder capacity, continence status, renal anatomy and function, and additional urologic surgical procedures during follow-up were analyzed.

Results. Nine patients (60%) became socially continent and voiding on primary closure without any additional bladder neck surgery. 12/15 patients (80%) were continent and voiding when endoscopic bulking injection was included. One patient became socially continent after BNR and one patient was dry and on CIC after BNR and ileocystoplasty. One patient remained incontinent because parent's refusal for surgery. Ultimately, of 15 patients, 14 are dry (93%); 10 completely continent and voiding, 3 partially continent (dry intervals 1–3 hours) and one dry and on CIC. Bladder capacity was adequate for age in 80% of patients. Febrile urinary tract infection occurred in 33% of patients and 67% needed endoscopy for urethral stenosis. Upper tract dilatation and loss of renal function was not seen.

Conclusions. Primary repair of BE followed by CIC has encouraging continence and bladder capacity rates with limited need for additional bladder neck surgery and preservation of the upper urinary tract.

Experimental Surgery / Varia

A65
L1 expression in neuroblastoma is correlated with a better clinical outcome – preliminary results

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Background. Neuroblastoma (NBL) is the most common solid tumor in childhood. L1 is a cell adhesion molecule which is expressed by many malignant tumors of neuroectodermal origin. In this study we assessed L1 expression on a NBL microtissue-array and analyzed the impact on clinical outcome and prognosis.

Methods. 62 specimens of neuroblastomas were stained for L1 on tissue arrays by immunohistochemistry. Statistical analysis was performed for patients' clinical data. Furthermore, Kaplan Meier analysis for event free survival and overall survival of the patients was used to correlate the L1 status of the NBLs with the outcome.

Results. L1 positive tumors were significantly ($p = 0.020$; F-test) more common in patients without an event ($N = 48$; 90.6%) than in patients with an event (relapse or death; $N = 5$; 55.6%). The Kaplan Meier survival curves were significantly better for patients with L1 positive vs. L1 negative NBL (log-rank test: $p = 0.0001$ for OS; $p = 0.0022$ for EFS).

Conclusions. The data suggest that L1-positive NBL patients belong to a better clinical risk group and have a significant longer survival compared to L1-negative NBL patients. Thus, L1 seems to be an useful marker with a highly specific prognostic value.

A66
Submucosal plexus disappear in dilatated bowels after experimental gut ligation in chicken embryos

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Background. Dilatation and impaired function of the gut is a condition often seen in newborns with enteric atresia, bowel obstruction or gut stenosis. In a previous experimental study in chicken embryos, we observed a rarification of the submucosal plexus and ultrastructural changes of the myenteric plexus at the end of gestation after small bowel ligation. Aim of this study was to investigate the timely changes of the general morphology and of enteric nervous system (ENS) after gut ligation.

Methods. 56 chicken embryos were investigated. In the operation group fertilized eggs and chorion allantoic membrane were opened and the small bowel was ligated at day 11. The controls were sham-operated. The gut was prepared and harvested for analysis at day 12, 13, 14, 15, 16, 17 and 18 of fertilization. Silver staining or a staining for acetyl-cholinesterase (AcH) was performed of the specimens.

Results. A marked dilatation of the bowel was observed three days after operation (day 14). The submucosal and myenteric plexus appeared normal until the fifth day post OP (day 16). Then the submucosal plexus diminished in the dilated segment, whereas the myenteric plexus remained normal until day 18.

Conclusions. The data suggest that dilatation of the gut results in a rarification/loss of the submucosal plexus, whereas myenteric plexus seems not to be affected until day 18. Further studies have to show whether these changes of the ENS are reversible.

A67
Supra-umbilical ventral body wall malformations – a new model in the chicken embryo

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Background. Supra-umbilical ventral body wall malformations are rare anomalies. In humans, omphaloceles and ectopia cordis belong to this spectrum. Up to now, the embryological background of these malformations is unknown. Recently, an animal model was developed using suramin and trypan blue as teratogens in chick embryos. In this study, we modified this model in order to produce higher numbers of supra-umbilical malformations.

Methods. In one series, suramin was applied in ovo to normal fertilized chick embryos at day 2.5 of incubation (stage 15 according to Hamburger/Hamilton). In a second series, trypan blue was used in an identical fashion. After the application of the agents the eggs were re-incubated for 5 additional days (stage 30-32 Hamburger/ Hamilton). At this time point, the embryos were removed from the eggs, fixed and prepared for scanning electron microscopy.

Results. At day 8 of incubation, 60 embryos of the suramin treated group and 60 embryos of trypan blue treated group were still alive. In the survivors, both substances induced a spectrum of malformations including head and eye defects (16.6 %), abnormal body size (49.9%), abnormal pelvis (10%) and legs (41.6%), and mild forms of cloacal extrophies (1.6%). Additionally, a high number of supra-umbilical ventral body wall malformations were observed in both series: 3 embryos showed an isolated ectopia cordis (5%), 24 embryos (40%) developed a supra-umbilical body wall defect, half of them associated with ectopia cordis.

Conclusions. In our model, high numbers of supra-umbilical body wall malformations could be observed using suramin or trypan blue as teratogenetic substances. In ongoing studies we want to test whether this model is useful to elucidate the embryogenesis of these malformations.

A68
The PAUL-Procedure: A new technique for congenital abdominal wall defects

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Background. Congenital abdominal wall defects (AWD) are impressive and dramatic malformations. Common surgical therapy for AWD is to place the herniated viscera back into the abdomen and to close the fascia. Small defects can be closed directly by surgical treatment. In large defects resorbable and non-resorbable artificial materials are necessary to close the fascia. The aim of this study is to create a new technique for the treatment of large abdominal wall defects.

Methods. A median laparotomy was performed in a small (SAM) and a large (LAM) animal model (SAM: Wistar-WU-Rats with a body weight of 75–100 g, n = 6; LAM: Goettinger Minipigs with a body weight of 7.0–10.0 kg, n = 10). AWD was created in SAM and LAM by excising a full thickness segment including fascia, muscles and peritoneum. These defects were then closed by using the PAUL-Procedure (PAUL = postnatal – abdomino – umbilical –Lyoplant®-Procedure).

Results. (1) The PAUL-Procedure could be performed technical easily in SAM and LAM. (2) No wound infection could be observed throughout the experiment. (3) No animal died during the time of observation. (4) Compared to the untreated control, all animals showed physiological grow and a normal bodyweight curve. (5) No abdominal hernia developed in either group.

Conclusions. Our experimental results indicate that the PAUL-Procedure can be used easily for the therapy of congenital abdominal wall defects.

A69
Treatment of gastroschisis – Why change a running horse?

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Background. Gastroschisis is a rare embryological defect of the anterior abdominal wall of unknown etiology. In literature several therapeutical concepts compete: primary surgical closure, patches, silos, delayed midgut reduction without general anesthesia and others. As a center of "conservative" surgical management, i.e. primary surgical closure whenever achievable, we are more and more on the opposite of the mainstream in literature. We present the results of our institution from 1994–2005 and compare with the current literature.

Methods. Retrospective (from 2005 prospective) study from 1994 to 2005 using patient charts. Statistics performed using SPSS 12.0.

Results. 54 children with gastroschisis (29 to 39 weeks of gestation) with a rate 92% of caesarian section and a weight from 1200 g to 3850 g were treated at our institution from 1994 to 2005. In contrast to the literature we had a relatively prevalence of related malformations. In more than 80% primary surgical closure was the therapeutical strategy of choice. 20 children developed infection / sepsis / pneumonia (38.5%) and 18 children developed ileus / perforation / vovulus / NEC / patch infection (36.5%). 32 children had single gastroschisis related surgery (62.7%), secondary surgery made up to 7 operations. We had to regret 1 gastroschisis related death (compartment). The shortest hospital stay was 26d.

Conclusions. The Vienna experience from 1994 to 2005 shows that gastroschisis is a disease regularly combined with surgical complications like infection, ileus and perforation. The end results with mortality of 2% are quite satisfactory. Several therapeutical strategies are discussed and compared to the Vienna experience.

A70
A technique to improve vascularity in colon replacement of the oesophagus

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Objective. To present a simple method to improve the results of colon replacement of the oesophagus in children with post-corrosive oesphageal stricture or long gap oesophageal atresia.

Technique. At the operation to establish a gastrostomy, the abdomen and colon are explored and the segment of colon to be used for replacement is chosen. The trunk of the middle colic artery supplying the transverse colon is ligated and divided proximal to the marginal artery or if another segment of the colon is chosen, the corresponding vessel is ligated.

Methods. Between November 1999 and October 2002, 11 children had the middle colic vessels ligated during the gastrostomy operation. They were 6 boys and 5 girls. Five neonates had long gap esophageal atresia with or without fistula. The other 6 had long segment oesophageal stricture due to swallowing caustic potash. Their ages ranged from one day to 40 months. The hospital stay ranged from 10 to 14 days. The interval between vascular ligation and the replacement was one to 3 months depending on the general condition of the patient. The follow up period is 21 to 56 months.

Results. After the definitive operation of colon replacement of the oesophagus, the children resumed feeding through the gastrostomy on day 5. They were fed by mouth from day 8 and all were home by day 15.

Complications. There was no wound infection, no fistula, or chest complications. One patient developed stricture at the colo-oesophageal anastomosis two months after surgery. The oesophageal anastomosis was excised six months after the colon interposition surgery and reanastomosis performed through the same neck incision and the patient was sent home 7 days later.

Conclusions. Ligation of the middle colic vessels during the gastrostomy operation increases the blood supply to the transverse colon through the left upper colic and marginal vessels. This adds extra ten minutes to the operation of gastrostomy. This technique has increased the success rate in colonic replacement and minimised morbidity in our unit. Although the principle is commonly used in plastic surgery in pedicled flap reconstruction and in pediatric surgery in high abdominal testis (Fowler Stephens procedure), to our knowledge, this has never been applied in intestinal surgery before.

A71
Partial external bile diversion for Alagille

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Background. New approaches to reduce drug-resistant pruritus in Alagille syndrome (AS) are partial external bile diversion (PEBD) and extracorporal albumin dialysis (MARS). PEBD and MARS aim to deplete the body from bile salts (BS) and pruritogenic substances. We aimed to quantify the effects of PEBD and MARS on BS pool size and pruritus in three patients with AS.

Methods. PEBD was performed by a separated jejunal loop in isoperistaltic direction between gallbladder and the skin of the right upper abdomen. Pool sizes and synthesis rates of the two major primary BS, cholate and chenodeoxycholate, were determined by stable isotope methodology in 2 AS patients (female, 10 yr) before and at 4 weeks after PEBD through cholecystojejunocutaneostomy, and in 1 AS patient (female, 12 yr) before and at 6 weeks after MARS. At each time point, severity of pruritus was scored (Yerushalmi, JPGN 1999), and urinary, fecal and biliary (PEBD patients) excretion rates of endogenous BS were quantified for 3 days. All patients received ursodeoxycholate treatment.

Results. Prior to intervention, all three patients had severely debiliating pruritus (grade 3 of 3 point scale). The patients had high total serum BS concentrations (~380 μM), and BS excretion occurred predominantly via urine. In two patients a reoperation had to be performed: one for intra-abdominal adhesions and one for porlaps of the stoma. PEBD completely resolved the pruritus (each to 0 of 3 point scale). After PEBD (n = 2), BS excretion via the biliary diversion accounted for ~90% of total BS excretion from the body, and urinary BS excretion was reduced by ~65%. PEBD strongly reduced the pool sizes of cholate (by ~66%) and chenodeoxycholate (by ~51%) and increased their synthesis rates between 2 and 5-fold. PEBD strongly increased total BS excretion (stoma + urine + feces; by 340% and 960%) and decreased serum BS concentrations (by ~80%). MARS treatment only decreased pruritus for 2–3 weeks, and did not significantly affect serum BS concentration, nor the pool sizes or synthesis rates of cholate and chenodeoxycholate at 6 weeks after treatment.

Conclusions. Present data indicate that the antipruritogenic effect of partial external bile diversion in Alagille syndrome is related to persistent, profound decreases in BS pool sizes and urinary BS excretion, and to an increase in BS synthesis.

A72
Laparoscopic pull-through for anorectal malformations

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Background. Anorectal malformations comprise a broad spectrum of anomalies for which several different treatment modes have been developed over the years. For the high and intermediate types of malformations for the past 25 years the PSARP procedure has been the treatment of choice. Although the principal of anatomical dissection was sound, the results, however, remained variable. The statement the higher the malformation, the poorer the result remained. The idea is that with the dissection innervation of the pelvic floor and sphincter complex is disrupted. With the advent of laparoscopic surgery it was suggested that blunt dissection of the pelvic floor and sphincter complex would cause less detrimental damage to the innervation. The first preliminary results of the laparoscopic pull-through technique are described.

Methods. Between July 2000 and November 2005 12 patients with anorectal malformation were treated by laparoscopic pull-through. Of these two children had a cloacal malformation, and one child had a rectal atresia. One patient was referred to our center for the laparoscopic pull-through. The level of the fistula was at the urethra (2), prostate (4), and bladder (2). One patient had no fistula. Four patients had a total of eight concomitant abnormalities. Eleven patients received a diverting colostomy within the first 48 hours post partum. One patient was given a colostomy at the time of pull-through on day 2. Mean age at the time of pull-through was 31/2 months. Colostomy closure was carried out after a mean of 4 months.

Results. One patient with a cloaca is still awaiting definitive reconstruction. In the other cloaca, as well as the child with the rectal atresia, the rectum was mobilized laparoscopically, after which a PSARP was performed. In all other children the laparoscopic pull-through could be performed successfully. The postoperative course of a patient with tethered cord was complicated by rectal and stomal prolaps, for which revision was necessary. This child later needed a rectopexy and still has his colostomy. In one patient anastomotic leakage occurred after closure of the colostomy for which a temporary colostomy was given back for three months. All patients underwent postoperative dilatations with climbing Hegars 6–12 or 14. In two patients dilatations were initially difficult, because they had difficulties to relax their pelvic floor. After due time the difficulties subsided. One of them is still on laxatives. Follow-up of nine patients has been from 1–5 years. Two children (cloaca, high ARM bladder roof) only have defecation on rectal washout. Four patients have been doing well up to five years without laxatives and regular defecation. One patient has been without medication for four years, but has recently been put on laxatives.

Conclusions. Anorectal malformation is a complex disease and outcome has sofar not been outstanding. In a large series Pena decribes 41% of his patients to have voluntary bowel movements and no soiling (totally continent). In this preliminary series of laparoscopic pull-through procedures 4/8 patients are totally continent, with one other just recently on, hopefully temporary, laxatives. The preliminary outcome therefore seems at least to be comparable with the PSARP. Further studies are necessary to determinate more accurately the outcome of laparoscopic pull-through technique.

A73
Laparoscopic repair of Morgagni hernia in children. Experience at the Royal Medical Services

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Background. The study was designed to prove the feasibility of endoscopic repair of Morgagnis hernia.

Methods. Between 1998–2003, eleven children, seven girls and four boys, with Morgagni hernia were treated laparoscopically at Queen Alia Military Hospital. Age of the children was between 4 months and 4 years. All patient presented with respiratory distress and recurrent chest infection. Diagnosis was made either by plain films, barium meal or barium enema. One patient had bilateral disease only diagnosed at surgery.

Results. Three trocars were used: two 5 mm and one 10 mm. The sac first was incised and then the defect was closed by mesh which was fixed by clips, sutures or tissue Tacher. The operative time was between 30 minutes to 60 minutes. There was no conversion to open surgery and no operative or postoperative complications. Pain was minimal and controlled by paracetamol suppositories. All patients were discharged from hospital in less than 48 hours. Follow up 6 months to 7 years showed no recurrence.

Conclusions. We conclude that laparoscopic repair of Morgagni hernia in infants and children is safe, effective and should be considered as the primary treatment of Morgagni hernia in children.

A74
Extreme Medicine® (ExM®) – Are there any limits in treatment of life-threatening burns in children?

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Background. Extreme Medicine® (ExM®) is absolutely new, just established medical term we reserved for periodical treatment of some usually life-threatening health conditions in which certain medical procedures are carried out in so extensive or aggressive way, even recognized by physician, endangering patient by itself. Severe burned child is one of most serious surgical patients at all.

Methods. This paper will present the course of the disease of 87% TBS, mostly IIIrd and IVth degree, burned 12 years young boy admitted in our Children's Hospital after explosion of propane-butane gas and air mixture at his home. Multidisciplinary approach was carried out. We will discuss about how we entered in EM during treatment.

Results. The boy survived! Dismissed from our Department of Burns, Plastic and Reconstructive Surgery after: 1 goal: to live, 2 tracheotomies, 3 central venous catheters placed, 6 types of anesthetic agents used, 9 types of analgetics for pain relief used, 12 bronchoscopies, 13 autologous, split-thickness skin graft transplantations, 14 pathogenic microorganisms detected, 20 different antibiotics used, 30 other drugs administered, 203 days in PICU, 326 days in post-intensive and nursing care at our department, 453 changing of dressings in OR under general anesthesia, 529 days in total in Children's Hospital Zagreb, 2370 ml of blood transfusion, 7790 ml of packed RBCs, 8260 ml of fresh frozen plasma, burn shock, several resuscitations, pneumonias, pulmonary oedema, lymphoedema of lower extremities, ostheoporosis, several prolongated sepsis, atelectasis, anxiety, etc.

Conclusions. Mayor burn injury, especially in children, evokes strong emotional response in most health professionals who are confronted by the spectrum of pain, deformity and potential death. Severe pain and repeated episodes of sepsis followed by predictable outcomes of either death or survival encumbered by pronounced disfigurement and disability has been the expected pattern of sequelae to serious burn injury. Even burn injury is still intensely painful and sad, the probability of resultant death has been significantly diminished. At the present time, most children, even young children and children with severe burns, should survive their injuries. For that goal, sometimes it is really necessary to carry out Extreme Medicine procedures that could be justified.

A75
A comparison of Mepilex transfer and Aquacel Ag® for the treatment of intermediate thickness burns

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Background. No consensus exists on treatment of intermediate thickness burns in children.

Methods. In a prospective clinical trial, we therefore compared the efficacy of two burn dressings (Mepilex transfer® [M], Aquacel Ag® [A]) in children under 10 years of age with primarily assumed partial thickness burns.

Results. 16 patients were treated with M and 13 with A. Values are expressed as means (min – max). There were no significant differences according to age (M and A: 3.5 y [7 m–9.5 y]), total body surface area (M: 5% [2–10], A: 8% [3–15]), number of hospitalisations (M: 7/16, A: 9/13) and length of hospital stay (M: 14d [4–25], A: 8 d [2–22]). Despite a longer healing time in A treatment (A 13d [9–17], M 8 [4–11]) ($p < 0.05$), complications and discomfort were significantly higher in M treatment with higher need for autologous grafting (M: 3, A: 0), more dislocation of dressings, more pain during changes of dressings and one case of toxic shock syndrome due to staphylococcus aureus infection.

Conclusions. We conclude that, even with longer healing times, the silver containing wound dressing Aquacel Ag® saves from discomfort and complications.

Poster

P1
Gastrointestinal stroma tumor in a 12 year old female (GIST)

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Background. GIST is the most common mesenchymal tumor of the gastrointestinal tract in the adult. In children it is extremely rare. Some 50 pediatric cases have been published so far. GIST a potentially malignant entity and should be considered under oncologic standards.

Methods. We present a 12 years old girl who presented with upper gastrointestinal bleeding (Hb 5.5 g/l). Gastroscopy was performed. Histology reaveled GIST. Staging showed no signs of metastatic disease. Preoperative chemotherapy with GLIVEC was administered.

Results. She underwent partial gastrectomy with gastroduodenostomy (BI). The postoperative course was uneventful. Under regular restaging she is tumorfree for 15 months. Histology showed GIST in two nodules (spindlecell-like/epitheloidcell-like), malignant according to Newman criteria, 100% positive for C-Kit, CD34 positive.

Conclusions. In children GIST is a rarity. Although is should always be considered as reason for gastrointestinal bleeding or other gastrointestinal symptoms. GIST a potentially malignant entity and should be considered under oncologic standards. GLIVEC (imatinib, specific Kit-inhibitor) is the first orally administered chemotherapy available with significant improvement in the clinical outcome.

P2
Imperforat anus with multiple jejunal and ileal atresias – a case report

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Background. Anal atresia is one of the most common forms of congenital intestinal atresias. The incidence is approximately 1 : 5000 live births. The combination of anal atresia with other intestinal atresias is very rare. There are only a few reports in the literature describing anal atresias combined with any other intestinal atresia.

Methods. We report about an unusual case of imperforated anus associated with jejunal atresia and double ileal atresia, diagnosis, management and follow-up. It was very difficult to classify the anorectal malformation due to the presence of other associated intestinal atresias. Two atresiae were detected during the first procedure.

Results. The plan was to perform a double barrel colostomy but the operation ended up as double barrel jejunostomy and jejuno-jejunal anstomosis. During the second operation, the imperforate anus was corrected through a posterior sagittal approach (intermediate type). During the third operation for closure of the double barrel jejuonsotomy, the third atresia was detected (type intestinal atresia, a complete ileal membrane with normal looking ileum from outside). Such combination of the anomalies is very rare and has not been described in the literature to date.

Conclusions. Although very rare, the possibilities of other gastrointestinal obstructions or atresias should be considered when dealing with anorectal anomalies.

P3
Paralytic ileus due to traditional medicine – a diagnostic and therapeutic problem in Malawi

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Background. The majority of people in rural and even urban Malawi in case of an acute health issue still attends in the first instance a traditional healer. Patients, who later seek help in a hospital may present symptoms of intoxication after the use of traditional drugs. These symptoms may cope signs of the underlying disease, challenge the diagnostic skills of the surgeon and complicate the identification of appropriate treatment.

Methods. We prospectively recorded 12 cases of suspected paralytic ileus due to intoxication with herbal medicine. After interviewing the patients, all involved relatives and village authorities and contacting a traditional healer we identified in co-operation with the local botanical garden the toxigenic plants and developed a strategy of clinical and pharmacological management.

Results. Out of the observed 12 cases only the history of four cases could be documented appropriately. Non-operative treatment was successful in all cases, consisting of nothing per os, i.v. fluids and the administration of the parasympaticomimetic drug Distigmin (Ubretid). In cases where under the initial assumption of obstruction or peritonitis a laparotomy was performed, post-operatively the restitution of normal bowel motion was delayed, contributing in two cases to the development of a burst abdomen. The ingredients of the traditional medicine given to patients with diarrhoea were identified as Psidium guajava and Lantana camara. Both plants contain a wide range of biologically active and potentially toxic substances. Querecetin is suspected to be the relevant component to cause paralysis of the gut.

Conclusions. Patients with diarrhoea may present after administration of herbal medicine at a local healer with paralytic ileus in the hospital as a symptom of intoxication. In such cases operative treatment is obsolete, parasympathicomimetics may break the papalysis. The possibility of intoxication should always be taken into consideration and carefully explored.

P4
An external device for fecal incontinence

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Background. A simple external device that enables the incontinent patient to control the time, frequency and place of defecation is described. The device is based on the principle of the „ball & socket“ valve. The „ball“ is an inflatable silas-

tic balloon whereas the „socket“ is the anorectal junction. The device can be used with minor modification in patients with terminal colostomy to make them continent and avoid the need for colostomy bags.

Methods. The inflatable plug has been used successfully in eighteen incontinent children for a period ranging from six months to six years. The child decides the amount of air inside the inflatable plug that is comfortable and yet adequate to prevent soiling. This usually ranged between 10 to 15 cm of air.

Results. To date, the device has been manufactured manually. All the children tolerated the plug without discomfort. Deflation of the balloon occurred after 3–5 days of use due to defective manufacturing. None of the patients developed ischemia of the bowel or skin excoriations.

Conclusions. This conservative, simple inflatable plug enhances both qualitative and quantitative faecal continence in children with fecal soiling. An improvement in quality of life was also perceived by the patients and their parents. Further proper manufacturing may improve the results.

P5
Operative management in two cases of apple peel atresia (Bland-Sutton type IIIb)

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Background. Apple-peel-atresia is a rare malformation of the intestinal mesenterium and the small bowel. The jejunum and the ileum distal the atresia turn around a central artery, like a apple which is peeled. The distal part of the small bowel is very small and underdeveloped. Mostly it is combined with an intestinal malrotation.

Methods. We want to present the operative management of two cases with fetal ileus of the small bowel, where we found an apple-peel-atresia.

Results. In case one a fetal ileus was diagnosed in the 25th week of pregnancy. The baby was born in the 37th week via c-section. After confirmation of the prenatal diagnoses, the neonate was undergone laparotomy. We found atresia of the small bowel after 45 cm of the jejunum. Distal the atresia we saw an apple-peel-malformation. The distal part of the small bowel was underdeveloped and turned around a central artery. The proximal part of the jejunum was dilatated. There was such a big difference in calibre, that there was no chance to make a primary anastomosis. A stoma of the proximal and the distal part of the atresia was performed. Three days after operation, we start with oral nutrition and the distal bowel was filled via the stoma with the intestinal fluid from the proximal stoma. After 4 weeks an end-to-end anastomosis was performed. No postoperative complications have been observed. In case two a fetal ileus was diagnosed in the 29th week of pregnancy. The preoperative management was identical. Intraoperative we saw an atresia after 25 cm of the jejunum. The apple-peel-malformation was much longer than in case two and the blood circulation of this part was critical. We chose the same operative way. The postoperative management was identical; the end-to-end anastomosis was performed after three weeks. No complications have been observed postoperatively. Both children show a good development till now; there are no signs of a short bowel syndrome, malnutrition or subileus.

Conclusions. We believe, that it is the best way to operate this malformation in two steps, to avoid short bowel syndrome. The recovery time of both children was very short; the postoperative development of the patients after end-to-end anastomosis is close to normal. No signs of subileus have been observed. At the second operation, we saw a good development of the distal small bowel and it was much easier to build a functional end-to-end anastomosis, cause the difference of the calibre was not so much anymore.

P6
Recurrence of colonic perforation in a 2-month-old girl, and management with use of an abdominal zipper

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Methods. 2-month-old girl, since 10 days has abdominal distention, bilious vomiting and fever. No prenatal problem, normal vaginal delivery at term, birth weight 3100 g, weight: 5500 g (0.50 percentile). Mildly hypotonic, dehydratation P: 142 / min, BP: 70 / 30 mm Hg marked abdominal distention and pneumoperitoneum (erect x-ray). No salmonellosis (typhoid fever), no amibiasis and bacteria in the blood culture, and no bacteria intraperitoneal region. Ganglion cells + (colon, rectum and appendix). Transvers laparatomy, isolated perforation (3 × 1 mm) in descending colon (5 cm distal of splenic flexure). Primary repair and drainage PO # day 8, fecaloid drainage from the drains. After resuscitation 2nd operation: Repaired site was normal, 2 × 1 mm cecum perforation was detected. Primary repair was made and 7 days after 2nd operation again pneumoperitoneum and at the 3rd operation all repaired perforation sites were normal but a new perforation from the ascending colon about 2 × 1 mm was detected. Again primary repair, ileostomy and abdominal zipper applied (10 × 6 × 0.1 cm).

Results. 3 × 2/day abdominal irrigation with warm %0.9 NaCl via the zipper, 3 × 1/day abdominal irrigation with warm %0.9 NaCl-rifampicine via the zipper (7 days), TPN, antibiotics. 4th operation: PO # 7th day removal of abdominal zipper, oral feeding (PO # 3rd day), PO# 25 day colon enema: normal passage (+). All clinical and laboratory findings were normal 2 months after administration: Discharged with ileostomy. 5th operation (closure of ileostomy) PC: normal physical and motor development, no gastrointestinal complain and good bowel function. Abdominal USG, CT: normal.

Conclusions. The use of abdominal zipper can be life saving method in recurrent intestinal perforation or recurrent peritonitis.

P7
Assault with intent and thoracic trauma

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Background. The frequency of thoracic penetrating trauma due to assault with intent in pediatric surgery is low.

Methods. This report concerns 2 cases with penetrating thoracic trauma with the flink-knife and the special finger spike.

Results. Diagnosis was based on history, clinical examination and relevant investigation like radiography and ultrasonography. The patient with the pnemomothorax (finger spike) was treated with intercostal tube thoracostomy. The patient with small laceration of the left lung (flink-knife), without pneumothorax, underwent thoracotomy and intercostal tube thoracostomy. Both patients were treated by general supportive measures and without complications.

Conclusions. The authors describe our experience with the successfully treatment for two penetrating thoracic injuries due to the special stabbing.

P8
Cat Scratch Disease as a rare cause for multiple granulomas or suspected abscesses

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Background. We present Cat Scratch Disease as a rare cause for abdominal pain, undulating fever and multiple granulomas or suspected abscesses in a five year old boy. Cat Scratch Disease, caused by Bartonella henselae, typically presents with a localized lymphadenopathy, a brief period of fever and general symptoms. However, there are atypical cases with a wider spectrum of clinical manifestations.

Methods. Our patient, a five year old boy, was presented after suffering from fever and abdominal pain for five weeks. Prior to admission to our department he underwent an "open" appendectomy. The histological evaluation of the appendix showed secondary inflammatory signs of the appendix wall. As fever and pain continued, the boy was transferred to our medical center. Ultrasound evaluation showed an abscess-like formation in the spleen and multiple foci of low density in the liver. MRI, blood cultures and bone marrow samples did not lead to a diagnosis. Consecutively, diagnostic laparoscopy and liver biopsy were performed.

Results. The pathological examination revealed typical Cat Scratch Disease associated granulomas.

Conclusions. The presented case illustrates the diagnostic value of laparoscopy and proves that representative samples can be taken safely. In the case mentioned above it led to the unexpected final diagnosis.

P9
The triphalangeal thumbs in a 4 years old girl

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Background. The triphalangeal thumb is characterized by the interposition of an extra phalanx between the two normal phalanges, with the incidence of about 1 in 25,000 births. The extra phalanx may be fully developed, rectangular, or wedge shaped. It is often associated with many anomalies. The classification is based on a modified Wassel system.

Methods. In our case, the girl, operatively treated at the age of 4 years, had the delta phalanx on her left, and the rectangular phalanx on her right thumb, both causing the thumbs to bend toward the index finger.

Results. The left thumb was treated with removal of the delta shaped middle phalanx and collateral ligament reconstruction from the soft tissues and periosteum surrounding the delta phalanx. Newly formed joint was fixed with the K-wire. The right thumb was shortened and deangulated with the complete excision of the distal growth plate and the transverse osteotomy of the middle phalanx. The bone ends were fixed with the K-wire, collateral ligaments also reconstructed. Postoperative X-ray and clinical findings were satisfactory. To ensure ligamentous healing, K-wires were removed after 4 weeks with some limitation of the movement of the thumbs, suggesting the parents to allow the child to use the thumbs freely and to keep a night time splint on for another 2 weeks. Following this, the child resumed full activities.

Conclusions. Until the future research into the exact etiology of the triphalangeal thumb become reality, careful preoperative planning and initial correction of all forces will lead to mostly satisfactory results.

P10
Penile incarceration – the rare case of recurrent incarated inguinal hernia

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Background. Inguinal hernia repair is one of the most common elective procedures in paediatric surgery. Recurrent inguinal hernia is rare and the operation is more technically advanced, sometimes anatomy of recurrent hernia is very unusual.

Methods. Case report from the Department of Paediatric Surgery of Kaunas Medical University.

Results. 8 years old boy was admitted to the Department with acute pain and local swelling in the lower part of the abdomen and penis, nausea and vomiting. During the examination we found elastic and painful mass in the lower middle part of the abdomen descending to the proximal part of penis, the postoperative scar in left groin area and tenderness and swelling there. The left part of the scrotum was empty; there was no testis in it. Ultrasound revealed intestinal loop with gas and air in the proximal part of the penis. It was known from anamnesis that 5 years ago the patient was operated on due to congenital inguinal hernia then 1 year later – due to recurrent inguinal hernia in the district hospital. Unusual incarceration of recurrent inguinal hernia was diagnosed and the patient was operated on. During the operation we found inguinal hernial sac set in the penis with the incarceration of the small bowel loop. Surprisingly the testis was also found in the subskin layer of the penis. Funiculolysis, orchidopexy, hernioplasty was performed. The cause of such unusual anatomy of recurrent inguinal hernia could be the mistake during the reoperation 4 years ago when the testis was not placed in the scrotum and the recurrent hernia incarcerated via the iatrogenically ectopic spermatic cord into the penis.

Conclusions. The anatomy of recurrent inguinal hernia sometimes is very unusual and the penile ectopy of the testis can be iatrogenic.

P11
A new instrument in the diagnosis of Hirschsprung disease: triple rectal suction biopsy and its advantages

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Background. Several rectal suction biopsy devices are available but none of them full fills the requirements needed a clinical practice.

Methods. The biopsy device is made of stainless steel and easy to sterilize. It has two cylinders passing through each other and containing three 0.5 cm diameter holes within a 2 cm distant from each other. The holes are positioned facing to the posterior rectal wall. While a negative pressure with 50 cm H_2O is being applied via the canula located at the bottom of inner cylinder, mucosa – submucosa are sucked. By pulling the inner cylinder 1 cm back out of the outer cylinder, specimen are cut. After removing the device from the rectum, the biopsy specimen are taken out of the holes separately. With this new instrument , firstly, 36 biopsies in total, 9 from each subject in the clockwise of 5, 6 and 7, were taken from the rectum of 4 New Zealand rabbits and investigated with Crossman's modified triple staining. And then, 12 biopsies in total, 3 from each with a dimension of 0.5×0.5 cm, were taken from 4 children from the posterior rectal wall in the clockwise of 6, and examined in terms of mucosa, submucosa and the existence of ganglion cells with the same methods.

Results. In the 36 and 12 biopsy materials taken from the Rabbits and children, it was found that mucosa and submucosa were sufficient and in all of the cross sections, parasympathetic nerve fibers and ganglion cells did exist.

Conclusions. Suction biopsy was first defined by Noblett in 1960s and for several years single biopsy was applied. However, new instrument (triple rectal suction biopsy) can be applied in out patients without general anesthesia and risks. Taking more than one biopsy with a single shot provides easiness for the patient and the doctor in the diagnosis of Hirschsprung disease. This instrument is also economic, safe and easy to use.

P12
Dislocation of the elbow: caveat medial epicondylar fracture

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Background. In children, dislocation of the elbow is combined with a medial epicondylar fracture (MEF) in 30–55% of cases. If one is not familiar with this combined injury, the fracture may be missed. Aim of this study: (1) to analyze in which percentage the combined injury was present, (2) how often the medial epicondylar fracture had been missed at primary diagnosis, and (3) to assess the results of treatment.

Methods. Retrospective analysis of all consecutive patients, who were treated for dislocation of the elbow and/or medial epicondylar fracture between 1990–2004 at our center. The following data were collected and analyzed: gender, age, cause and side of injury, missed fracture, therapy, follow up, elbow function at the end of treatment.

Results. The group was comprised of 39 children (18 boys, 21 girls) with a median age of 10 yrs (range 5–16 yrs); 28 had a dislocation of the elbow, of whom 19 (68%) a combined MEF; 11 had an isolated MEF. In 2 (10.5%) children with a dislocated elbow, the concomitant MEF had been missed. The occurrence of the fracture was independent of age. The combined injury occurred more frequently on the left side (13/19). The cause of the injury was in and around the house in 20, sports and games in 17, and traffic in 2. Closed reduction was possible in 8/9 dislocations, 4/19 combined injuries, and 1/11 epicondylar fractures. The medial epicondyl was fixed with either vicryl or K-wire. All elbows were after treated with plaster of Paris during 3 weeks. After a median follow up of 11.5 weeks (range 5–201 wks) the elbow function was normal in 7/9 dislocations, 16/19 combined injuries, and 11/11 medial epicondylar fractures.

Conclusions. (1) A combined injury was present in 68% of cases, (2) the concomitant medial epicondylar fracture had been missed at primary diagnosis in 10.5% of cases, and (3) the function of the elbow is less at risk after a medial epicondylar fracture (extra-articular), than after a dislocation, whether the dislocation is combined with a fracture, or not.

P13
The results of intramedullary nail fixation of radial neck fractures in children

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Background. We evaluated the results of radial neck fractures treated with different methods of reduction and internal fixation with intramedullar nails.

Methods. 23 patients (10 male, 13 female; mean age 8.3 years; range 6.1 to 10.5 years) underwent closed reduction (n = 20) or K-wire assisted reduction (n = 3), both followed by intramedullar fixation. According to the Judet classification system, fractures were classified as type III in 9 patients, type IV in 14 patients. Fractures of type I or II according to Judet are not included in this study. Accompanying fractures involved the ulnar metaphysis (n = 1), the olecranon (n = 1) and the avulsion of the ulnar epicondylus (n = 1). Clinical and radiographic evaluations involved the use of a modified Morrey functional evaluation index. The mean follow-up period was 38.4 months (range 3 to 67 months).

Results. An anatomical reduction was assessed in 13 patients, good reduction was observed in 10 patients. No secondary dislocation, necrosis or pseudarthrosis of the radial head appeared. Functional results were generally excellent or good. Only one patient complained about continuous pain and decrease in range of movement.

Conclusions. The review of our 23 patients seems to indicate that atraumatic reduction plus intramedullary nailing provides good results with a low complication rate.

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