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Oral Presentations

Iatrogenic disorders

Secondary tumours: iatrogenic or genetic?

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All cancers may be genetically determined with or without other initiating factors. This predisposition may be inherited, occur by mutation or possibly be caused by chemical agents or radiation used in the treatment of tumours. In the treatment of a primary cancer, chemotherapy and radiation are frequently used either separately or together. These agents may be responsible for the development of a second cancer. Primary genetic abnormalities are known to predispose towards more than one cancer. The Regional Paediatric Oncology Data Bank was searched for patients with a second tumour: 27 were found. The age at presentation varied from 1 year to 17 years, with a mean of 6.9 years. The second tumour occurred between 1 and 23 years, the average being 7.9 years. The primary tumours were Hodgkin's disease, lymphosarcoma, rhabdomyosarcoma, retinoblastoma, Ewing's, Wilms', intracranial tumours and leukaemia. The secondary tumours were osteosarcoma, cerebral neoplasms, skin tumours, rhabdomyosarcoma and leukaemias. The cases were reviewed in relation to the treatment with chemotherapy or radiation and the site of the tumour in relation to any radiotherapy given. There were few distinguishing factors that might help to differentiate between a treatment-induced tumour and a second primary tumour. Detailed results of the survey and illustrated examples will be given. In conclusion, whether there is genetic predisposition to a second tumour or not, treatment is known to increase the risk of developing a second tumour, and this should be taken into consideration in order to reduce the adverse effects as much as possible.

Reference

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Iatrogenic cardiovascular disorders

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Purpose of study: To review all the iatrogenic cardiovascular disorders that occurred during the past 25 years in our institution.

Methods: Some 8000 catheterizations have been performed during the past 25 years. Incidents related to the procedures were reviewed, and we identified 305 iatrogenic lesions.

Results: The main groups are as follows: cardiac disorders: lesions of the walls of the chambers of the heart (23), pericardial effusions (14), cardiac foreign bodies (8), aneurysms (2); vascular disorders: hematoma (12), arterial thrombosis (53), venous thrombosis (102), vascular foreign bodies (6); miscellaneous disorders: deaths (48), pulmonary disorders (31), cerebral disorders (6). The most frequent disorders were venous (33%) and arterial (17%) thrombosis.

Conclusion: Complications cannot be avoided but should be reduced as much as possible. Risk factors are young age, small size of vessels, complexity of procedure and severity of heart disease.

Radiologic evaluation of pulmonary complications occurring during respiratory assistance of the newborn with high-frequency oscillatory ventilation (HFOV)

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Purpose: Evaluation of the possible reduction of pulmonary damage occurring during the treatment of RDS and ARDS of the newborn by means of HFOV.

Material and methods: Comparison between results obtained in 1990 with conventional ventilation (CV) and those of 1993 with HFOV.

	CV	HFOV
No. of patients	60	40
Mean gestational age (weeks)	31.6	31
Mean body weight (g)	1700	1500

Results:

	CV	HFOV
Mortality	13 (22%)	2 (5%)
Interstitial emphysema	37 (62%)	6 (15%)
Pneumothorax	20 (33%)	11 (28%)
Chronic lung disease	9/47 (19%)	8/38 (21%)
Type I	1	8
Type II	8	0

Conclusions: The major benefit of HFOV lies in the reduction of prolonged sequelae (chronic lung disease). Severe bronchopulmonary dysplasia and large bullous lesions were not present in our series.

Conventional radiology vs US, vs CT and vs MRI

Conventional radiography vs MR imaging in evaluating epiphyseal trauma

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Purpose: Fractures with epiphyseal plate involvement may interfere with normal epiphyseal growth due to bone bridge formation and partial epiphyseal closure. Posttraumatic growth disturbances can be observed even in the absence of radiologic abnormalities. MR imaging was performed in order to evaluate the exact involvement of the epiphyseal plate.

Materials and methods: Forty-two children aged 1.5-14 years with a history of recent trauma were prospectively examined with conventional radiography and MR imaging. In total 17 knee and 25 ankle joints were evaluated.

Results: In 14/25 patients with ankle joint trauma, MR imaging demonstrated additional injuries, including six Salter-Harris 3 fractures, four flake-fractures and four ligament injuries. In 8/20, MRI showed the extent of the injury to better advantage. The therapeutic regimen was changed in 11/25 patients because of MR findings. In 17 patients with knee trauma, MR imaging revealed eight pre-

viously undiagnosed fractures. Additional findings were anterior cruciate ligament rupture and osteochondrosis dissecans. Trauma was ruled out in 6/17 patients, and therapy was modified in 15/17.

Conclusions: MR imaging is useful in children with suspected epiphyseal plate trauma of the knee and ankle and demonstrates the exact extent of the fracture and the involvement of the epiphyseal plate to better advantage than do conventional radiographs.

Comparison of interictal ^{99m}Tc HMPAO brain SPECT, MRI, CT and EEG findings in children with epilepsy and hemiplegia

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We have reviewed the interictal brain ^{99m}Tc HMPAO SPECT scans performed on 53 children from 1990 to 1994 and compared their studies with findings on EEG, CT and MR scans. The ages ranged from 1 month to 17 years, with a mean of 8.5 years. Forty-five children had seizures, seven had hemiplegia or hemiparesis and one suffered haemorrhagic shock encephalopathy. Five SPECT studies were of poor diagnostic quality and one was a failure. In 18 patients with seizures there was good correlation between the EEG, MR and SPECT findings, and in general the CT study contributed little. However, in three of these infants the SPECT study showed a more extensive abnormality than MR. In the other 21 patients with seizures there was poor correlation between the MR and SPECT findings. In five, the SPECT was normal with positive MR findings. The MR correlated with the EEG study in only two patients. Of nine patients with a normal MR and an abnormal SPECT, the EEG and SPECT findings were consistent in seven. There were seven patients where there was an abnormality on both the MR and SPECT studies but the findings did not correlate in the two examinations, and the SPECT study was consistent with the EEG findings in only two of these. There were seven infants who presented with hemiplegia or hemiparesis. Six of these had MR studies. In three there was good correlation between the MR and SPECT studies, and in a further three the SPECT examination showed a more extensive abnormality. In the seventh patient there was good correlation between the CT scan, EEG and SPECT study. In the patient with haemorrhagic shock encephalopathy, the SPECT and MR findings were similar. In conclusion: (1) MR has become the investigation of choice in childhood epilepsy. (2) The CT study is of little value in determining the abnormality, and a HMPAO study is useful if MR is not available. (3) The interictal HMPAO SPECT study is of value if the MR examination is normal and there is an abnormality evident on EEG. (4) Ictal SPECT examinations are not practical in our department but they may provide more accurate information.

The evaluation of malignant abdominal masses in children: a comparison between ultrasonography and CT or MRI

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In making the diagnosis of malignant diseases of the abdomen in children, ultrasonography represents diseases of the abdomen in children, ultrasonography represents the first approach to abdominal masses and offers a high amount of diagnostic information. This study aimed to find out whether CT or MR as additional methods obtain further essential information, and therefore are indispensable, justifying their routine use. In the years 1987–1994, 60 children with malignant masses of the abdomen were treated in our hospital. All of them had initial ultrasonography followed by CT and/or MRI. We compared the reports from sonography with those from the additional CT or MRI and from the histological examination. In determining the organ of origin, we found a co-

incidence in 55 cases out of 60 (90%), mostly supported by excretory urography. Only in 32 patients (53%) did we find agreement regarding tumor size. In 47% of the cases (28 children) there was no agreement between ultrasound and CT or MRI concerning the relationship to other organs. For adenopathy and metastases, found in 20 cases (33%), sonography turned out to be insufficient. The inability to detect them by ultrasound was due in most of these cases to artifacts. Nearly all reported cases (95%) were investigated by the same physician, who had many years of experience. Comparing the reports made by less experience, we found a great number of differences in all parameters, so that an interindividual comparison is of less value in this context. Our conclusions are: The accuracy of ultrasonography depends on the investigator's experience. Ultrasound is a method with a high specificity for localization of the organ of origin of abdominal masses and sufficient information about the character of a tumor. In certain cases excretory urography helps to determine the relation of the tumor to neighboring organs. The gaining of information about the relationship to other organs and vessels and the depiction of adenopathy and metastases depends on the penetration of disturbing artifacts. In such cases additional imaging is necessary. We recommend that in larger centers the decision for imaging in addition ultrasonography should be made in those cases in which the investigator is not able to obtain precise information on relation to other organs, size, and adenopathy and metastases.

Role of ultrasound and magnetic resonance imaging as reflected by publications in pediatric radiology

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The considerable changes that ultrasound and MRI have undergone as imaging procedures in pediatric radiology are reflected in the literature published during the last 15 years. The sensitivity and specificity of ultrasound compared to CT and the diagnostic accuracy of ultrasound information in the evaluation of the parenchymal organs and soft tissue structures are well established. A change in imaging strategies occurred with the use of ultrasound for assessing the heart and brain as well as the hip in children. More sophisticated scanning techniques are used only in cases where additional diagnostic information is required. In this respect, MRI is more and more replacing CT in pediatric radiology. Imaging techniques not requiring ionizing radiation are in particular used in pediatric oncology. New diagnostic possibilities in the evaluation of the bone marrow have been introduced with the use of MRI, which yields morphologic images of medullary changes. The latter is of utmost importance in the diagnostic assessment of tumors. The change in imaging strategies, which is especially evident in pediatric radiology, is demonstrated by a statistical analysis of the international literature.

Renal atrophy after treated neuro- and nephroblastoma

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Purpose: To describe late sequelae of combined chemotherapy and surgery in patients with neuroblastoma and Wilms' tumor.

Material and method: Four children (age range 2–4 years) had follow-up sonography, intravenous urography and CT of the kidneys after medical and surgical treatment of suprarenal neuroblastoma ($n = 2$) and Wilms' tumor; in both children (one with bilateral tumor, the other with involvement of the caudal duplex system) the kidneys were preserved.

Results: Doppler sonography 1–4 months after surgery revealed normally sized kidneys with regular intrarenal flow pattern. Follow-up studies (sonography, urography, CT) demonstrated pro-

gressive atrophy of the organs; after 8–12 months, in three patients no ipsilateral kidney could be identified, while one child had a very small kidney-like structure without significant function. In no patient was arterial hypertension present; there was no evidence of recurrent neoplastic disease.

Conclusion: Aggressive pre- and postsurgical chemotherapy and tumor excision may eradicate the neoplasm, but the combined treatment may damage the ipsilateral kidney and induce atrophy.

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Urological imaging

Spiral twist in testicular torsion, a reliable US sign: 16 cases

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The purpose of this paper is to emphasize the importance of detection of spiral twist of spermatic vessels in the diagnosis of testicular torsion. High-resolution sonography combined with color Doppler was done in 16 patients (aged 11–24 years) with acute scrotal swelling. Examination was performed with an ATL UM9 HDI machine and a linear 5–10 MHz transducer. Surgical correlation was available in all cases. Color Doppler demonstrated lack of intratesticular flow in only seven of the 14 patients with proved torsion; in five, the testis was swollen and hyperechogenic, associated with venous infarction; in two, peritesticular tissues were thickened and hyperemic and the testis was heterogeneous, associated with necrosis. In seven cases, the testis appeared normal and intratesticular flow was present (symmetrical in five, discretely decreased in two). But in all these cases, we could detect spiral twist of the spermatic cord in the inguinal canal and/or near the testis. Presence of blood flow within the twist was correlated with the degree of testicular vascularization. In two cases, testicular blood flow was increased, with peritesticular hyperemia in one and cord hyperemia in the other, but no spiral twist was detectable. Surgical exploration revealed an appendix torsion. So, even in the case of partial or intermittent torsion, visualization of the spiral twist allows a reliable diagnosis, whatever the color Doppler and morphological findings.

Magnetic resonance angiography of pediatric renal transplants with quantitation of allograft blood flow

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Purpose: To evaluate phase contrast magnetic resonance angiography (PC-MRA) for: producing renal transplant arteriogram; measuring renal blood flow in children.

Materials and methods: PC-MRA was performed at 1.5 T in 12 children (mean age 10 years) with normal allograft function and normal color Doppler ultrasonography. Venous renal blood flow (RBF) was determined using the PC technique by the product of the velocity of protons and vessel cross-sectional area. Results were compared to the standard *p*-aminohippuric acid (PAH) clearance technique.

Results: Three-dimensional PC-MRA produces consistently renal arteriogram with a good visualization of the iliac artery, anastomosis, main renal artery and proximal arterial branches. Comparison of blood flow revealed the MRA and PAH techniques to be signif-

icantly related ($r = 0.75$, $P < 0.05$). The average renal blood flow was similar by each method, 395 ± 114 ml/min by PAH clearance and 385 ± 153 ml/min by MRA, but the agreement among individuals between the two methods was only modest, with a 95 % confidence interval of -168 to $+148$ ml/min.

Conclusion: MRA can provide, noninvasively, both an accurate arteriogram and fairly accurate quantitation of blood flow rate in the transplanted kidney in children.

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Balloon dilatation of the pelviureteric junction in children: early experience and pitfalls

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Although balloon dilatation of the pelviureteric junction (PUJ) is a well-established technique in adults, there is little published experience in the paediatric age group. We have performed ten procedures in children aged from 5 months to 10 years of age. Six procedures were performed for PUJ obstruction and four for strictures of the pelviureteric anastomosis following pyeloplasty. In all cases PUJ obstruction was confirmed using ultrasound, nuclear medicine or intravenous urogram. In none of the cases were stents inserted following the procedure. Balloons of 8 mm or 10 mm diameter were used. In three cases the procedure was performed by an antegrade approach following nephrostomy and in seven cases the procedure was performed by the retrograde approach following cystoscopy. In four cases the procedure was successful, with relief of symptoms and/or improvement in radiology; in one case the radiology was unchanged; and in five cases the procedure resulted in complete obstruction of the PUJ requiring surgical intervention. Two of the retrograde procedures resulted in vesicoureteric junction obstruction. Some individual problems will be illustrated. Our experience suggests: (1) Smaller balloons (4 mm or 6 mm) should be used in babies. (2) The vesicoureteric junction can be damaged by balloon withdrawal and procedures should be performed using the antegrade approach. (3) Stenting may be required to prevent early obstruction of the PUJ. (4) The procedure can be used in selective cases as an alternative to surgical pyeloplasty.

The paediatric retrograde: an obsolete investigation?

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The retrograde pyelogram is still regularly used in adult urological practice, but its use in paediatrics is seldom, if ever, mentioned. Is the technique obsolete, being replaced by other imaging modalities, or does it still have a role? In the 5 years from January 1990 to December 1994, 66 retrograde examinations were performed in this hospital in 65 children. There were 34 boys and 31 girls, and the children's ages ranged from 7 months to 15 years. Twenty-eight children had bilateral examinations, 20 children had a left retrograde examination, 16 children had a right retrograde, and one child had a right retrograde on two occasions. The examination was usually carried out for suspected obstruction, of either pelviureteric or vesicoureteric junction, but other indications included haematuria infection, renal calculi and abnormal renal anatomy. Many of the cases had not been discussed with the radiologist prior to the examination, the child appearing in the department with the catheter already in situ. In five cases the catheter was in the bladder or urethra and the examination could not be performed. In 27 cases (40 %) the examination was of value, enabling a definitive

diagnosis to be made. In 14 cases (22 %) the examination was unnecessary and should not have been performed. In 25 cases (38 %) the information from the retrograde could have been obtained by less invasive investigations – nuclear medicine scans or IVU. The examination was most helpful in children with dilated renal tracts and reduced renal function; in children with a thickened bladder wall; and in those with abnormal renal anatomy. The retrograde pyelogram is of limited value in paediatric practice and should only be performed in particular cases following consultation between paediatric surgeons and radiologists with a specific interest in renal disease.

The role of magnetic resonance imaging in the assessment of paediatric pelvic rhabdomyosarcoma

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The use of magnetic resonance imaging (MRI) in the management of adult pelvic tumours has been demonstrated and discussed in the literature. A retrospective review of MRI in pelvic rhabdomyosarcoma was carried out in 13 children in order to assess the role of MRI in the management of this tumour. Twenty-eight scans were available, four at diagnosis and 24 during treatment. All patients had a combination of axial, coronal and sagittal images with T1-weighted (4 with intravenous gadolinium) and either T2-weighted or inversion recovery (IR) sequences. The scans were assessed for tumour signal, site of origin, extent and optimal imaging sequences and planes. Tumour signal was of muscle signal intensity in 28/28 T1-weighted scans, enhanced in 4/4 with gadolinium and high on all T2-weighted and IR sequences. Tumour site and tumour extent were correctly identified in 10/10 patients and 5/6 patients, respectively, where surgical correlation was available. Sagittal and axial imaging were useful in intravesical, prostatic and vaginal tumours, whereas coronal imaging was more useful in lesions involving the pelvic side walls. Extension into the spinal canal and muscle involvement were best seen on coronal T2-weighted and IR sequences. T1-weighted sequences were useful for prostatic tumours. We conclude that MRI is of value in the management of pelvic rhabdomyosarcoma in children, with the added benefit of the lack of ionising radiation.

The relationship between tuberous sclerosis complex and renal cell carcinoma: four cases and meta-analysis

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Introduction: A number of cases have been reported in the world literature which attest to the occurrence of renal cell carcinoma in tuberous sclerosis complex (TSC) patients. Though the accumulated literature suggests that patients with TSC are at increased risk of renal neoplasia, the association of renal cell carcinoma and angiomyolipoma with or without TSC may lead to confusion, particularly since angiomyolipomas can be locally invasive and demonstrate tumor-like neovascularity on angiography. We evaluate whether this confusion can be alleviated.

Methods: Four pediatric cases of TSC/angiomyolipomas were collected, one of which had metastatic renal cell carcinoma diagnosed. Reviews and case reports of TSC and renal cell carcinoma and related imaging findings were identified by searching the MEDLINE database. Manual searches of the “bibliography of reviews” in Index Medicus, of personal files, and of the reference lists from all identified reviews were performed. Cases found suitable for review in the available literature from 1922 through 1993 comprised the data analyzed.

Results: A cumulative summary of the literature amassed a total of 89 cases from 61 manuscripts. This case is the 17th case of angiomyolipoma associated with renal cell carcinoma in TSC. Analysis of the distribution of characteristics and incidence of renal cysts, renal cell carcinoma, and lymph node involvement demonstrate a decreased risk of renal cell carcinoma in patients with TSC ($P < 0.01$) despite their presentation an average of 20 years younger than patients without TSC.

Conclusion: Despite convincing anecdotal evidence over the last 70 years suggesting a possible relationship between TSC and renal cell carcinoma, a meta-analysis of a majority of reported cases and cases of coincident angiomyolipoma and renal cell carcinoma without TSC implies a lower risk of malignant degeneration in TSC patients. Statistical analysis of the data indicates that at least 46 more cases of TSC with coincident renal cell carcinoma need to be documented to refute the conclusion of this meta-analysis.

Color Doppler ultrasound versus enhanced CT: a comparative study in acute pyelonephritis

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Aim: To demonstrate, by comparison to enhanced CT, the ability of color Doppler imaging (CDI) to diagnose acute pyelonephritis.

Patients and methods: Fifteen patients aged 4–15 years presenting with typical symptoms of acute pyelonephritis (fever $> 39^{\circ}\text{C}$, lumbar fossa pain, positive urine culture) were evaluated. Ultrasound evaluations were performed with HDI equipment (ATL). They included a standard study of the urinary tract and a CDI vascular mapping of both kidneys using the so-called DPI software (highly sensitive for intra-renal vasculature). Ten-millimeter non-interleaved CT slices were obtained immediately after IV injection of 1 ml/kg of iodinated contrast. All patients underwent both studies within 24 h.

Results: On CDI, intra-renal vasculature disappeared in lobes with acute pyelonephritis. Wedge-shaped intra-renal zones involving cortex and medulla were delineated within the normal remaining kidney. They were compared to the triangular hypodense lesions demonstrated by enhanced CT. Our preliminary results showed a 90 % correlation rate between the two techniques. Contrary to previous studies [1], focal areas of hyperperfusion were not identified.

Discussion: Making an accurate distinction between lower and upper urinary tract infections is mandatory for immediate treatment (intra-venous or oral antibiotherapy) as well as for scheduling radiological evaluation or surgical management. Clinical and biological findings may or may not be sufficient. Up to now, two imaging modalities (^{99m}Tc DMSA scintigraphy and enhanced CT) were available in difficult cases. Both studies have about the same sensitivity with regard to acute pyelonephritis [2], but both require transportation of sick patients and deliver radiation. CDI seems able to yield comparable results. We could expect this result, since the basic phenomenon (delayed/decreased vascularization of the infected lobe) is shared by all imaging modalities.

Conclusion: New CDI softwares such as DPI are capable of diagnosing acute pyelonephritis in the most cost-effective and non-invasive conditions. CDI could replace CT or ^{99m}Tc DMSA scintigraphy in the near future.

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Technology

Pediatric digital radiology: spatial resolution requirements

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Background: Digital imaging techniques offer the potential advantages of image manipulation, image transfer and radiation dose reduction. Performance proficiency must be equal to or better than that permitted by the film system. To define radiological needs will influence the price, duration of transmission and archiving.

Purpose: To define the minimum spatial resolution (pixel size, or pl/mm) required for each type of pediatric digital X-ray examination.

Material and methods: The authors made an exhaustive survey of the radiological literature of the past 10 years. Conventional screen-film combination can reach a spatial resolution of 15 pl/mm. Digital fluoroscopy: spatial resolution depends on amplifier and matrix size. A 1024 × 1024 matrix size and 20 cm amplifier diameter correspond to 2.5 pl/mm. Phosphor plates have a spatial resolution of 0.1 mm or 0.2 mm (5 or 2.5 pl/mm).

Results: According to the literature, the minimal spatial resolution needed for the thorax varies from 0.4 mm for nodules to 0.2 mm for pneumothorax, interstitial lesions, and mediastinum. Concerning bones, 0.2 mm suffices for routine examinations, 0.1 mm is required for special procedures. GI and UT examinations can be performed with a spatial resolution of 0.2 mm.

Conclusions: A totally digital pediatric radiology department is now possible. It needs the use of both digital fluoroscopy and computed radiology.

RETAIN project: advanced multimedia pediatric teleradiology on ATM networks

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Teleradiology is a tool to acquire and transmit digital images over public networks, allowing radiology expertise to be extended, to distant sites by remote reporting, consultation between radiologists and clinicians, and better distribution of resources. Multimedia is an enhancement by providing videophony – audio and video transmission in real time – allowing collaborative work and better human communication as well as synchronization of the systems, enabling the same images, cursors, or image annotation and drawings on both sites. These user requirements require high-speed communication networks, such as ATM, a new standard network technology with speeds up to 155 Mbit/s per user. In the framework of the TEN-IBC program of the EU, the RETAIN project has implemented a multimedia teleradiology work station that has been tested on pediatric radiology between Hospital Vall d'Hebron in Barcelona and the University Hospital in Rennes. Using standard computer components, the system provides a multimedia interface: the computer monitor acts as an electronic view-box, with a small window in one corner of the screen providing a moving image of the remote reporting room. A loudspeaker and a microphone are used for audio communication. In a second phase, an ATM multimedia terminal was added to the system, providing high-quality audio and video over a second TV monitor. The bandwidth of the network was limited to 10 Mbit/s on both phases of the tests. For a period of 3 months scientific medical sessions involved radiologists of both centers. Images acquired by any modality, connected to the PACS, were exchanged between workstations. Con-

sultations were done using the multimedia workstations. The system is described and the medical and technical results of the experiments are presented.

A closer look at the European survey of the variation in dose and image quality of common X-ray examinations of the 5-year-old child

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A survey using questionnaires and thermoluminescence dosimeters for nine frequent X-ray examinations in the 5-year-old child was carried out in 106 European children's clinics during 1993–1994. Entrance surface dose (ESD) measurements were taken, and the original radiographs were evaluated for image quality by a panel of nine pediatric radiologists. A brief report on a study on the evaluation of radiographic technique image quality and ESD was given at this year's European Congress of Radiology. This report will deal in more detail with selected aspects of this study, in particular with the impact of good radiographic technique on patient dose and image quality as recommended by the "Quality Criteria for Diagnostic Radiographic Images in Paediatrics", i. e. the Working Document of the Commission of European Union. As reported, wide variations of ESD values and radiographic technique were again found. The ratios between the maximum and minimum ESD values were as high as 71 : 1 (standard chest pa film). As was shown in a previous survey in infants, significant patient dose reduction was reached by those departments who used the recommended guidelines without any accompanying loss in image quality. This survey again emphasizes the need for quality assurance measures in all age groups in pediatric radiology. A large collective dose reduction could be achieved if these guidelines are implemented.

Study of relationship between cardiac function and carotid perfusion in an animal model, using quantitative color velocity imaging

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Background: Quantitative color velocity imaging (CVI-Q) is a new ultrasonographic technique to quantify volume flow (VF) in blood vessels [1–3].

Purpose: To use CVI-Q VF measurements to study the relationship between cardiac function and carotid blood flow (CBF).

Methods: In seven swine, an arteriovenous fistula was created between the common carotid artery (CCA) and the jugular vein. CBF was measured externally with CVI-Q and compared with continuous VF readings from a Transonic probe clamped around the CCA, cardiac output (CO; through Swan-Ganz catheter thermomodulation method), heart rate, and mean arterial blood pressure. CO and CBF were varied by (1) opening or closing the AV fistula, (2) hyperventilation, (3) withdrawing or infusing intravascular fluids, (4) infusing medications (dopamine, β -blocker).

Results: We produced a wide range of hemodynamic conditions (high- and low-output cardiac failure, hypovolemic shock, vasopressor overdose). CVI-Q VF measurements correlated well with Transonic readings, but were less reproducible. CBF autoregulation compensated for changes in CO. CVI-Q VF waveform analysis revealed marked variability of CBF resistance in different hemodynamic states.

Conclusions: CVI-Q is a valuable noninvasive technique to study the relationship between CBF autoregulation and cardiac function. These results are applicable to children undergoing complex cardiovascular interventions.

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Carotid volume flow measurements in children by CVI flowmetry

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Purpose: To study carotid blood volume flow (VF) in normal pediatric subjects using the color velocity imaging flowmetry (CVIF) technique. CVIF is a new ultrasound method to quantify VF in different vessels. Some 15% variability of VF measurements was reported in prior CVIF studies in adults. No prior data are available regarding carotid VF in children using this new non-invasive method.

Patients and method: In 32 normal children (13 male, 19 female; age range 5–17 years), 64 carotid VF measurements were performed. Patients with a history of cardiovascular disease, hypertension, diabetes, trauma or neurological disorder were excluded. The method was standardized according to the prior studies performed in adults. Before the VF study, 2D imaging and Doppler analysis were performed on both sides. Diameter, peak systolic and end-diastolic velocities were determined. A Philips P700 US system was used with CVIQ software. A minimum of three VF measurements were done on each side. All studies were documented on video prints and on video tape.

Results: Carotid VF in our population varied between 272 and 515 ml/min. Acceptable intraobserver variability was achieved. Age, side and weight correlations were calculated.

Conclusion: CVIF has a clinical potential to study normal and abnormal carotid VF in the pediatric population.

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Intraarterial calcium injection for localization of hyperinsulinism in children

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In our experience pancreatic venous sampling (PVS) is the method of choice for preoperative localization of hyperinsulinism in children. However, in a few patients general anesthesia induces normoglycemia, probably by inhibition of insulin secretion. PVS becomes impossible. In two such cases, selective intraarterial injection of calcium in the splenic and gastroduodenal artery, in association with insulin sampling in the hepatic vein, allowed us to make the diagnosis of diffuse form in one case and focal form in the

other. In conclusion, in certain cases selective arterial injection of calcium in children allows localization of insulin secretion in hyperinsulinism.

Intussusception-type antireflux valve in the Roux-en-Y loop in hepatoporto- or biliojejunostomy: ultrasonographic pitfalls

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Between 1991 and 1994, 82 children were treated with an intussusception-type antireflux valve (ITARV) in the Roux-en-Y loop, as described by Nakajo et al. [1], in order to prevent ascending cholangitis after hepatic portojejunostomy or bilio-digestive anastomosis. The patients included 68 cases of orthotopic liver transplantation (OLT), 10 of biliary atresia, 3 of congenital bile duct dilatation (CBDD), and 1 of biliary tree trauma. We report on 5 patients (2 boys, 3 girls) displaying an uncommon ultrasonographic (US) aspect (4 cases) or presenting complications (1 case) due to this surgical procedure. The first case is a 16-month-old girl complaining of abdominal pain and vomiting 7 days after OLT. Abdominal US showed an intussusception but contrast enema was normal. Discussion with the surgeon led to the conclusion that the US intussusception aspect corresponded to the ITARV. The second case is a 5-year-old girl with OLT (2 years 3 months) presenting with gastroenteritis. Abdominal US performed in another institution showed an intussusception aspect, which was thought to be due to the ITARV. The third case is a 13-year-old girl showing an transitory dilatation of the Roux-en-Y limb after creation of an ITARV for CBDD. Finally, in two boys aged 17 and 10 months, systematic US of the liver (respectively 2 and 1 month after OLT) showed dilatation of the Roux-en-Y limb proximally to the ITARV. The first boy was asymptomatic and dilatation subsequently decreased. The second one developed transitory biliary tree dilatation and one episode of cholangitis. A second-look laparotomy failed to show any mechanical obstacle at the level of the valve. We conclude that ITARV in the Roux-en-Y loop may lead to US diagnostic errors, such as intussusception or intestinal obstruction, related to the recent of this surgical technique and/or the lack of experience of the radiologist. Careful analysis of the "pseudointussusception" picture and knowledge of this surgical technique may help to avoid such pitfalls.

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Gastrointestinal imaging

Meconium plug-like syndrome in premature infants of low and very low birth weight

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Purpose: To understand why we are observing an increasing number of cases of functional intestinal obstruction among low and very low birth weight premature infants within their 2 first weeks of life. To determine the role of radiological examinations for the diagnosis and the treatment of this condition.

Material and methods: The study included 11 premature infants, mean gestational age 29 weeks and mean birth weight 1055 g, admitted to our hospital during the past 2 years, and presenting clinical and radiological suspicion of functional intestinal obstruction. We

retrospectively analysed the respiratory conditions at birth and their treatment, the timing of meconium passage, the type of feeding and the medications received. Radiological features suggesting intestinal obstruction and/or excluding other diseases were reviewed, as well as the results of contrast enema when applied. The clinical outcome was analysed focusing on intestinal evolution.

Results: At birth, all the 11 prematures presented with hyaline membrane disease and were treated by mechanical ventilation, fluid restriction and morphinomimetics. They were fed by total parenteral nutrition. (Partial) meconium passage was observed within the first 2 days of life in six patients and was markedly delayed in five. Obvious intestinal distension was observed on plain abdominal films as soon as day 4 and as late as day 13 (mean day 8). Hypertonic water-soluble contrast enema was performed at bedside in nine of the patients, using 30–60 ml of Gastrografin, without complication. It had to be repeated in six patients, from 2 to 4 times; thick meconium plugs were evacuated and intestinal outcome was favourable in all nine patients. The two infants who did not have contrast enema underwent surgery respectively at days 4 and 13. In the first, no perforation had occurred, but thick meconium at the ileocaecal junction was removed preoperatively. The other presented with an ileal loop perforation, and thick meconium plugs were observed.

Conclusion: Several factors contribute to the genesis of meconium plug-like syndrome in low and very low birth weight prematures (colon immaturity, restriction, morphinomimetics and the lack of enteral nutrition are the most important factors). This condition is likely to occur at the end of the first week of life, even if previous (partial) meconium passage has been observed. It has to be differentiated from other meconium diseases and from necrotising enterocolitis. The clinical condition, along with plain film of the abdomen, should suggest the diagnosis, and the treatment should remain conservative, using hypertonic water-soluble contrast enema at the bedside. Unrecognized, this entity can lead to intestinal perforation.

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Diagnostic accuracy of CT for GI perforation following blunt abdominal trauma in children

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Object: Gastrointestinal (GI) perforation in pediatric blunt trauma victims is often clinically occult, yet, unlike solid organ injury, requires early surgical management. We assessed the role of CT in identifying these patients.

Materials: All surgically confirmed post-traumatic GI perforations over a 10-year period were retrospectively reviewed. Forty-one patients were identified, with 20 having had preoperative CT evaluation. In addition, the CT scans of a further 10 patients were identified with suspected bowel injury who underwent laparotomy or post-mortem without perforation evident. All CT studies were evaluated for extraluminal air, free fluid, bowel wall thickening or enhancement and bowel dilatation.

Results: Extraluminal air was seen in 10/30 cases, bowel wall enhancement in 24/30, bowel wall thickening in 24/30, bowel dilation in 15/30 and free fluid in 27/30. Associated solid organ injury was present in five patients, all of whom had perforation. When all five of the evaluated CT findings were present, there was 100% sensitivity and specificity for GI perforation. However, this was the case in only 5/20 patients with perforation. Eleven studies had 4/5 findings, nine had 3/5 findings, three had 2/5 findings and two had 1/5 findings. The 10 false positives included four hypovolemic shock bowel syndromes and six mesenteric hematomas with serosal tears of bowel, of which two involved additional retroperitoneal hemato-

ma. No CT of a patient with perforation was normal, and if four or more of the above CT findings were present specificity was 75%.

Conclusion: CT is sensitive for detection of GI perforation and, in a small group, highly specific. Directed CT evaluation in conjunction with repeated clinical assessment by experienced surgical staff will improve early detection of GI perforations in the remaining patients.

Enteroclysis in children: a report on 213 examinations

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Introduction: Enteroclysis is the term for contrast administration by means of duodenal intubation. We have elected to pursue this method because contrast material administered orally is altered by fluid in the stomach and small bowel, which leads to segmentation and flocculation. As a result, mucosal folds are not properly visualized. The enteroclysis provides a more constant and more rapid filling of the jejunum and ileum. The contents of these portions of the bowel are pushed ahead of the barium column, resulting in better images.

Objectives: To evaluate the role of enteroclysis in the diagnosis and management of children with various abdominal problems; to elucidate its indications and reliability.

Materials and methods: We reviewed the clinical data, indications, imaging material, and outcome of the children who underwent enteroclysis in the Emma Kinderziekenhuis in the period from 1978 to 1993. All children had duodenal intubation one or more times, with informed consent and full knowledge of the procedure by the child and parents. After careful introduction of the tube, preferably by the oral route (mean duration about 15 min), the barium suspension, specific weight 1.15–1.20, depending on age and weight of the child, was infused at a rate of 25–100 ml/min. During the examination, graded compression was applied by the radiologist under fluoroscopic control and appropriate spot films exposed. In most cases diagnosis could be correlated with other clinical data, operation, and follow-up.

Results: Indications for the study of the small bowel included Crohn's disease, obstruction (stenosis/adhesions), Meckel's diverticulum, and malabsorption. Diagnostic findings included normalcy in 40% of cases, Crohn's disease in 35%, obstruction and/or adhesions in 10%, atrophy in 5%, and fistula in 3%. The diagnosis of a giant Meckel's diverticulum could be made in retrospect only.

Conclusions: (1) The enteroclysis method is especially suitable for the demonstration of normalcy, for the assessment of the exact nature and extent of disease, and for pre-operative delineation of stenoses, fistulae, and infiltrations of the bowel wall. (2) Given adequate information and positive understanding of the young patient, the procedure is tolerated well. (3) The advantages of enteroclysis for examination of the small bowel are: reliable findings, radiographs of superior quality, and a relatively short examination time.

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An exceptional echographic aspect of pyloric stenosis

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Ultrasound (US) has become the standard diagnostic procedure in hypertrophic pyloric stenosis, but in some cases it can also give specific etiological information. The authors present the case of a 3-week-old infant with characteristic clinical symptoms of outlet gastric obstruction. Diagnosis of pyloric stenosis was confirmed by ultrasound; nevertheless, US showed that muscular pyloric thickening was unusual in that it was hyperechogenic, with fading of different layers. Moreover, there was associated abnormality of pancreatic echostructure and hyperemic increase of gallbladder wall thickness. The familial history of this infant revealed hyperlipoproteinemia and allowed the understanding of the US findings: the intramural hyperechogenicity was an obstructive fatty infiltration of the pyloric wall. This was confirmed by surgery and by US follow-up under medical treatment. With this exceptional case, the authors emphasize the interest of ultrasound in differentiating rare causes of pyloric stenosis. This can change the surgical management.

Enteric fistula formation following necrotising enterocolitis: an important although uncommon complication

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We report five cases of enteric fistula formation following necrotising enterocolitis (NEC), diagnosed in our unit over a 10-year period from 1984. In four of the infants, (mean gestational age 30 weeks) the NEC was very severe and extensive, requiring early surgical intervention. Two of these infants also had Down's syndrome, but both had structurally normal hearts and no evidence of Hirschsprung's disease. The fifth baby developed NEC at 1 week following surgery for Fallot's tetralogy. In this infant the NEC was less severe and responded to medical treatment alone. All these infants were investigated, 2–6 weeks following resolution of the acute episode, for continuing gastrointestinal symptoms. The fistulae (colo-colic, jejuno-colic and ileo-colic) were demonstrated by either upper or lower gastrointestinal contrast studies. All fistulae were associated with bowel strictures, and in some cases there was evidence of previous localized perforation. Four of the strictures were confirmed surgically: one patient died from a septicemic episode in the pre-operative period. Our findings confirm that fistulae formation does occur in NEC but is an unusual complication. The possibility of a fistula in addition to strictures should be considered in infants who develop relatively non-specific gastrointestinal symptoms several weeks following the initial episode of NEC [1]. In recent years, balloon dilatation of colonic strictures has become feasible [2], and was considered in one of our cases. The procedure was cancelled following the diagnosis of the fistula on the pre-procedural contrast study. This emphasizes the importance of considering the possibility of an enteric fistula, in addition to a bowel stricture, as the cause of continuing gastrointestinal symptoms.

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Natural history of gallstones in children

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Purpose: To determine the natural history of cholelithiasis in children and the most appropriate follow-up.

Methods: The clinical, sonographic and surgical findings were reviewed at diagnosis and follow-up in children and adolescents who had a sonographic diagnosis of cholelithiasis in the 14-year period 1979–1993.

Results: Cholelithiasis was found in 276 children (149 female, 127 male; from in utero to 18 years of age). Follow-up ranged up to 11 years.

	Asymptomatic (n = 115)	Symptomatic (n = 161)
Stones went away	6 (5%)	0 (0%)
Emergency surgery	0 (0%)	16 (10%)
Elective surgery	13 (11%)	74 (46%)
Conservative management	96 (84%)	71 (44%)

Of the 96 initially asymptomatic patients treated conservatively, three became symptomatic (one choledocholithiasis, two dilatation of intrahepatic ducts). No surgery was required. Of the 71 initially symptomatic patients treated and conservatively, one developed cholangitis requiring emergency surgery and 24 required elective surgery after 6 months. *Choledocholithiasis* was noted in 13 patients: present at the time of initial diagnosis in eight (all symptomatic) and developing up to 4 years later in five (two of whom were initially asymptomatic, three initially symptomatic). *Inflammatory complications* developed in only one patient (? mild cholangitis) who was initially asymptomatic. However, in the symptomatic group, 15 had cholecystitis, four pancreatitis and two cholangitis; most within 4 years following initial diagnosis.

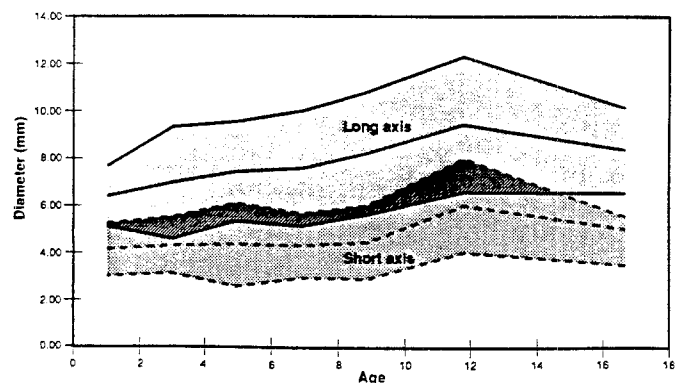
Conclusion: Asymptomatic children with gallstones require clinical follow-up only. Complications developed in a small number of patients who were initially asymptomatic. Sonographic follow-up is, therefore, only required at times of acute symptomatology in order to exclude ductal dilatation.

Hepatoduodenal lymph nodes in children: normal values

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It is commonly accepted that abdominal lymph nodes are difficult to visualize and infrequently visualized in children. Therefore any visible lymph node is thought to be probably abnormal. This is grossly inaccurate, as in the region of the hepatoduodenal ligament and the mesentery normal lymph nodes are visible in virtually every child. This presentation concentrates on the nodes around the hepatic artery which we have nicknamed "the daisy chain". If the hepatic artery can be demonstrated just medial to the take-off of the gastroduodenal branch, nodes are visualized in this region in 96% of normal children. The major reason for non-visualization is interposed gastric gas. In 200 subjects from birth to age 20 years in whom the anatomical location could be visualized and in whom the nodes were visualized, the long axis and short axis of the largest node were measured in order to establish the normal range for this region in children. Appreciation of the sensitivity of ultrasound in demonstrating normal nodes during routine abdominal echography and of the expected range of normal size should increase the sensitivity of detection of abnormal lymph nodes.

Mean diameters \pm 1 SD



An area of increased echogenicity of the peripheral portal veins in the liver: a sign of focal intrahepatic pylephlebitis?

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Purpose: Echogenicity of portal veins in the liver is normally homogeneous in intensity throughout the liver parenchyma. We present three children in whom we detected an area of locally increased echogenicity of the peripheral portal veins in the liver. We suspected that this ultrasound finding represented an area of focal peripheral intrahepatic pylephlebitis. The purpose of the study was to establish whether the increased echogenicity was related to an inflammatory process in the abdomen which causes focal intrahepatic pylephlebitis in these children.

Methods: We reviewed and analyzed the findings of abdominal ultrasound and the data of medical history, physical examination, laboratory tests, antibiotic therapy, and surgery in all children.

Results: The children were three boys, age 9–12 years. In all, the area of increased intrahepatic echogenicity of the portal veins was located in the caudal peripheral part of the right lobe of the liver and measured several centimeters in diameter. The walls of the veins were hyperechogenic, and the diameter of the veins was slightly increased; in two children the veins contained some internal echoes. The surrounding liver parenchyma was not evidently altered, nor was the liver enlarged. The rest of the intrahepatic portal veins and the extrahepatic portal vein were normal. The hyperechogenicity of the walls disappeared in a few days; none of the children developed abscess. All children were referred for abdominal symptoms and had an abdominal inflammatory process prior to and at the time of the detection of hyperechogenic portal veins. All had acute appendicitis and the following complications: (1) an acute gangrenous appendicitis of a retrocecal appendix complicated with a post-operative abscess, (2) a perityphlitic abscess treated surgically and complicated with a recurrent abscess, and (3) appendicitis with a retroperitoneal and subphrenic abscess. All children had fever, elevated white blood cell count, SR and CRP. Blood cultures were negative. In all, *Escherichia coli* was isolated at the time of surgery. All received antibiotic therapy prior to and after the demonstration of the hyperechoic portal veins.

Conclusions: In three children in whom we detected a focal area of hyperechogenicity of peripheral intrahepatic portal veins and suspected the finding to represent focal peripheral intrahepatic pylephlebitis related to an inflammatory process in the abdomen, we found complicated appendiceal inflammatory disease and concluded that the infection spread to the peripheral intrahepatic portal veins from the abdominal inflammatory process, causing focal pylephlebitis in the liver. The ultrasound sign suggestive of focal intrahepatic pylephlebitis can be an important clue in detecting an inflammatory abdominal process, may imply a change in antibiotic therapy, and when detected should be followed up to ensure the recognition of any liver abscess.

Cystic presentation of biliary atresia: US findings in a series of 21 patients

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A retrospective review of 21 infants presenting preoperatively with a cystic structure in the hepatic hilum on US and the final diagnosis of biliary atresia (BA) was undertaken to assess the imaging and surgical findings and the clinical evolution of these patients. They were 11 girls and 10 boys, aged from 15 days to 4.5 months, referred to us with the antenatal diagnosis of choledochal cyst in three, abdominal situs inversus in one and cholestasis in the other cases. In six of the most recent cases US was complemented by color Doppler imaging. Opacification of the cyst and/or the gallbladder (GB) was performed preoperatively in three

cases and preoperatively in 19. On US the size of the cyst ranged from 0.2 to 4 cm (mean 1.3 cm), and the GB was present in 14 cases. In none of our patients were dilated or even normal intrahepatic bile ducts (BD) seen. The opacification of the cyst showed communication in 15 cases, with a network of narrowed and irregular intrahepatic channels typical of the BD of BA. In five of these cases, there was also communication of the cyst with the GB. In none of our cases was there progression of the contrast medium from the cyst into the duodenum. Success of the biliary drainage procedure with regression of cholestasis was obtained in 16 patients. The US finding of a cyst in the porta hepatis without visible communication with dilated or normal BD allows early positive diagnosis of BA. The main differential diagnosis of the largest cysts is choledochal cyst with patent BD, which is a distinct entity and is thought to be due to anomalous pancreatobiliary union. Color Doppler imaging can be very helpful on one hand to differentiate the smallest cysts from branches of the hepatic artery, and on the other hand in the largest cysts to differentiate, even antenatally, intrahepatic vessels from dilated BD.

Subhepatic cyst in the perinatal period (three cases)

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Purpose: Subhepatic cyst (SHC) is a very rarely reported entity in the perinatal period. Its diagnoses may be difficult, because of confusion with choledochal cyst, duodenal duplication and ovarian cyst. We report three recent cases, including imaging features and percutaneous treatment in two of them.

Material and method: Three cases were investigated (two females and one male) using US with antenatal detection of a subhepatic cyst in two patients (at 34 and 38 weeks of gestation) and after birth in the third patient. Percutaneous opacification of the cyst was performed in each case in the neonatal period. Two patients had a barium meal, and in one patient a transhepatic cholangiography (THC) was performed.

Results: In the three cases, US showed an isolated cyst located on the medial aspect of the gallbladder fossa. Barium meal demonstrated an extrinsic compression. Percutaneous opacifications showed no communication with the biliary tract, which was confirmed in one case by THC. In the first patient, the cyst was associated with congenital hepatic fibrosis; it was a simple exophytic hepatic cyst confirmed by surgery and by pathological examination. In the two other patients, the cyst was isolated and the only treatment was percutaneous aspiration, with complete resolution after a 6-month follow-up.

Conclusion: In the perinatal period, the diagnosis and the treatment of a rare SHC may be carried out using US and fine-needle percutaneous opacification and aspiration.

Primary calcification in lymphoproliferative disease in pediatric orthotopic liver transplantation: report of a case

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Between January 1984 and December 1994, 430 orthotopic liver transplantations (OLT) have been performed in 362 children (age 96 days to 14.5 years) in our institution. We observed lymphoproliferative disorders with masses in the transplanted liver detected by radiological studies in three patients. In one child, one of the nodules initially appeared calcified. In another, two liver calcifications developed during recurrence of the disease at the site of a previous surgical biopsy. In the first child, a 18-month-old boy, rou-

tine ultrasound (US) of the hepatic graft 4 months after OLT disclosed nine masses, among which one spontaneously calcified. Two months earlier, US study of the liver was normal. The child did well, but blood tests and liver biopsy revealed lymphoproliferative disease. Evolution under treatment revealed decrease in size of the masses, but the calcification remained unchanged. Another 15-month-old boy was investigated by US 4 months after OLT for poor general condition and abnormal liver enzymes. US showed multiple hepatic masses and abdominal nodes. The diagnosis was established by liver biopsy. Several weeks after discontinuation of immunosuppressive therapy, the nodules disappeared, and subsequently recurred with two calcifications at the location of a previous surgical biopsy. So far, spontaneously calcified liver masses in lymphoproliferative disease have not been described in children with OLT. The etiopathogenesis of the calcified masses is discussed in light of the data from the literature concerning lymphomas.

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Gastric emptying evaluation by ultrasound: physiological and pathological aspects

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Background: The pathogenesis of gastroesophageal reflux is not yet fully understood, and there have been reports suggesting that delayed gastric emptying (GE) increases propensity to reflux. GE examination techniques rely currently on radionuclide equipment, which is expensive and delivers radiation. After having validated a US method by comparison to scintigraphy, a twofold study was undertaken in order to investigate the physiological and pathological aspects of GE.

Population and methods: Physiology of GE was determined in 200 non-vomiting babies aged 2 months to 2 years by varying feeding formula quantitatively and qualitatively. GE was determined in 300 children with variable patterns of symptoms; among 133 vomiting babies, 30 had esophagitis, 48 had recurrent or long-standing respiratory diseases, 34 had apneic spells and 85 were siblings of near-miss infants.

Results: Tremendous physiological variability in the GE could be observed, depending on the volume intake and the feeding formula, which have to be carefully taken into account before defining the “normal”. The relation between GE and reflux was as follows: normal emptying inducing no reflux or a limited number of events (< 3/10 min), whereas delayed emptying favored refluxes. However, reflux rate was not exclusively dependent on the emptying delay, but also on the associated length of abdominal esophagus, GE delay and extreme esophageal shortening inducing iterative reflux. In the case of symptomatic refluxes, emptying delay was present in 100% of those with esophagitis and in 42% of those with reflux-related cardio-respiratory symptoms. Different specific patterns could be defined: (1) In the case of esophagitis, delay was specific by its long-standing “en plateau” pattern extending over the 1 h. On the other hand, association with a patent shortening of the abdominal esophagus, leading to a severe dysfunction with repetitive refluxes, was pathognomonic. In these cases the impaired balance between emptying delay and esophageal continence was a far more reliable finding than the increased rate of refluxes, which could disappear in some circumstances, especially under

treatment. (2) In the case of cardiorespiratory pathology, two patterns became apparent: (a) A considerable emptying delay specifically associated with esophageal and pyloric dysmotility leading to esophageal retention. This aspect was found in 16/98 cases, including low birth weight babies with apneic spells, repaired hiatal hernia and mentally disabled children with recurrent pulmonary symptoms. (b) A moderately non-specific delayed emptying associated with hiatal and pyloric spasticity and constricted esophagus leading to unexpected massive and explosive refluxes. This feature was found in 41/98 children in association with pharyngolaryngeal abnormalities – gaping larynx (16 cases), laryngospasm (12 cases) proven by barium swallow, or reflux-induced cough (20 cases).

Conclusion: GE after standardized meal can easily become a routine part of studying reflux. Evaluation of gastric emptying together with esophageal length and gastroesophageal motility makes examination a rather pathophysiological process, defining specific patterns of “pathological reflux”.

Neurological imaging

Functional MRI in the pretreatment work-up of cerebral AVMs in children

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Aim: To establish the usefulness of functional MRI in the pretreatment work-up of cerebral vascular malformations in children.

Material and methods: Four children (three boys, one girl), right handed, from 7 to 11 years of age, with AVM of the brain in the frontal and rolandic region, were studied with functional MRI to ascertain the location of the AVM and the motor area. The study was performed on a Signa 1.5-T device (GEMS). SPGR sequences were obtained during the performance of a motor paradigm with the contralateral hand. The signal analysis was performed in the time domain thanks to software developed by C. D. and Y. C.

Results: In one case of frontal AVM the motor area was localized in the usual precentral region, far from the AVM. In two cases the motor area was seen to be localized at the periphery of the AVM. In one case, ipsilateral activation of the motor cortex was seen, with no activation of the contralateral hemisphere. After embolization no motor deficit was observed in three cases. In one case, a transient motor deficit was observed 48 h after embolization.

Conclusion: Functional MRI appears to be a useful tool to help plan the embolization in the treatment of cerebral AVMs in children.

Comparison of the parenchymal lesions in grade IV intraventricular hemorrhage (IVH4) and cystic periventricular leukomalacia (CPVL) and their neurodevelopmental sequelae

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Purpose: Premature infants with parenchymal lesions are at risk of significant neurodevelopmental impairment. This study evaluated and compared infants with such lesions (IVH4 and CPVL) and their outcomes.

Patients and methods: Ninety-one infants (< 32 weeks gestational age) born in the period 1988–1993 were diagnosed with CPVL ($n = 51$) or IVH4 ($n = 40$). Of those who survived at least 1 year, it was possible to follow up 44 of 48 (92%) with CPVL and 15 of 18 (83%) with IVH4. Their lesions were analyzed by a single so-

nologist and all underwent a neurological examination and a Bayley cognitive assessment (MDI).

Results:

Lesions	Survivors	Followed ≥ 1 year	Cerebral palsy (CP)	MDI < 70	CP and/or MDI < 70
CPVL (n=51)	48	44	93 %	71 %	98 %
IVH (n=40)	18	15	40 %	43 %	64 %

Conclusions: The sequelae of these lesions are significantly different. There is higher initial mortality from IVH4, but CPVL predicts greater neurodevelopmental impairment in long-term survivors.

Computed tomography in sensorineural hearing loss in children: results of 121 consecutive cases

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Purpose: To evaluate the contribution of computed tomography in sensorineural hearing loss (SNHL) in children, as this method has recently been questioned [1].

Materials and methods: Between October 1988 and December 1994, 121 temporal and cerebral CT examinations were performed in children with SNHL (57 unilateral, 64 bilateral). All CT were retrospectively compared to clinical, audiometric data and findings of surgery (when performed).

Results: In our study, a high proportion (40 %) of abnormalities were detected by CT: 21 cases (17.5 %) of inner ear malformation (one unilateral, 20 bilateral); nine cases (7.5 %) of ossified labyrinth; eight cases (7 %) of asymmetrical internal auditory canal (IAC) (two enlarged, six stenosed); six cases (5 %) of cerebral anomalies; two cases of otosclerosis; and two cases of abnormal oval window.

Conclusion: These results support the idea that CT still remains the first investigation to perform in SNHL in children, since acoustic tumors are rare among them. However, CT is known to overlook membranous labyrinth abnormalities as well as non-ossified labyrinthitis, which may account, at least in part, for our 60 % negative CT. On the other hand, MR imaging, as sole method, would fail to demonstrate the bony inner ear, footplate and IAC abnormalities.

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Changes of cerebral blood flow in asphyxiated newborns

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One of the great problems of today's neonatology is the so-called postasphyxia syndrome which follows intrapartum asphyxia. There are two main reasons to detect the cerebral blood flow (CBF) in that disease: (1) The CBF plays an important role in the pathogenesis of the disease, so it affects the diagnosis and therapy. (2) The potential neuroprotective agents have severe side effects, the therapeutic window is very narrow (the first 24 h), and the disease itself or the cerebral oedema caused by it cannot be detected either clinically or with sonography or CT at the newborn age. MR examina-

tion is difficult in patients needing intensive care. Color Doppler measurement can be a helpful tool both in diagnosis and in therapy, especially with the advanced technology used in the measurements. We divided our patients into three groups according to their birthweight (below 1250 g, 1250–2500 g, above 2500 g) and performed the examination with ATL UM9 HDI equipment with color Doppler technique using computed spectral analysis. We calculated the mean flow velocity (MFV). Griese et al. [1] found a correlation of 0.72 between the measurements of CBF by the MFV and the ¹³³xenon clearance method. Resistance index, used previously, had a correlation of 0.56 only. We formed two groups concerning asphyxia: the newborn was classified as asphyxiated if the Apgar score was less than 1/5, and as a normal control with an Apgar score more than 8/9. Altogether, 18 children were examined repeatedly (every 6 h) within the first 72 h. Parameters which might alter the CBF were also checked, like Hgb, Hct, pH, pCO₂, pO₂, RR, P, and respiration parameters. We conclude: (1) There is a linear increase of MFV and CBF with gestational and postnatal age. (2) There is an increase of flow within the first 24 h after asphyxia. (3) The detected hyperaemia is suitable to support the diagnosis of postasphyxia syndrome, allowing choice of adequate therapy. Changes in the flow have prognostic value. (4) With this method a more specific therapy can be achieved in order to decrease the hyperaemia with more efficacy, thus improving the cost-benefit ratio. (5) The CBF changes promptly in association with the above-mentioned parameters, so close control is needed when decreasing the hyperaemia. Summarizing, we think that measurement of the CBF can be helpful in the diagnosis and therapy of asphyxiated newborns.

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Cranio-facial low-dose CT in children

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Purpose: CT produces relatively high levels of radiation in comparison to other radiologic examinations. Because of the higher sensitivity of their tissues to radiation, and because of their long life expectancy, infants and children are more prone to suffer from radiation-induced pathologies than adults. This retrospective study was conducted to evaluate the feasibility, limits, and indications of low-dose CT (≤ 100 mAs) of the cranio-facial area in children.

Material and methods: Between October 1991 and January 1995, more than 100 low-dose CT examinations of the head and face were performed at the University Children's Hospital, Zürich, Switzerland. All studies were done using a fourth-generation scanner (Picker PQ 2000). The areas examined included the mandible, maxilla, paranasal sinuses, and orbits. The studies were performed for evaluation in trauma, infection, and malformation. Examination parameters were as follows: 45–100 mAs, 1.5–4 mm slice thickness. Both high-resolution and standard reconstruction modes were used.

Results: All studies were considered to be diagnostically adequate. Low-dose CT appeared particularly suited for areas with inherently high tissue contrast (air to soft tissues, and air to bone); however, contrast discrimination for characterization of associated soft tissue lesions was not adequate.

Conclusion: Low-dose CT is an important adjunct for evaluation of trauma, infection, and malformation of the cranio-facial area in children. Low-dose CT may be used alone, or in addition to normal-dose CT in cases where soft tissue characterization is required.

Periventricular signal abnormalities shown on MRI of the term and near-term infant following hypoxic-ischemic events: radiologic-pathologic correlation

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Purpose: PVL is a ischemic-hypoxic insult characterized by its manifestation in preterm infants. We observed, however, a high incidence of signal changes in the periventricular white matter in term infants with complex congenital heart disease. To investigate the nature of these lesions, postmortem MR images of uncut brains were compared to matched histological sections.

Materials and method: Patients with complex congenital heart disease or respiratory failure requiring extracorporeal membrane oxygenation (ECMO) who had died between 1 August and December 1993 were selected from the records of the Department of Pathology. Five infants aged 3–60 days, mean 24 days, with ECMO or hypoplastic left heart syndrome following major surgery in hypothermia and heart arrest were included in this study. Postmortem MR imaging of the entire fixed brain were performed in the five infants on a 1.5-T supraconducting unit. Short TR (700 ms), short TE (14 ms) spin-echo and long TR (3000 ms), short (22 ms) and long (120 ms) TE were used to obtain T1, proton and T2-weighted series. Slice thickness was 4 mm, number of acquisitions 1 and matrix size was 192 × 256. A gradient-echo sequence for increased susceptibility was also obtained. Signal abnormalities seen in the postmortem MRI were drawn into schematic coronal sections. In addition to obtaining standard serial macroscopic and histologic sections of the brain, lesions seen on MR were labeled and multiple histologic sections obtained from these areas.

Results: Among 43 lesions, 37 were in the periventricular and six in the subcortical white matter. Histologically, 30 of 43 areas of signal alteration were due to acute ($n = 6$), subacute ($n = 19$) or chronic PVL ($n = 5$), eight lesions were due to congestion or focal edema, and in four areas of abnormal appearance on MR no abnormality was found on histological sections. Typical signal changes were increased signal on T1- and decreased signal on T2-weighted sequences. Signal abnormalities were related to microcalcification (10), increased cellularity (10), coagulation necrosis (4) and white matter gliosis (2).

Conclusion: A large proportion of periventricular white matter lesions in term infants at risk for hypoxic-ischemic events are due to PVL. Microcalcification is a major but not the only factor underlying abnormal signal on MRI.

The role of colour Doppler sonography in the differentiation between benign subarachnoid space enlargement and subdural effusions

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Extracerebral fluid collections in infancy are a diagnostic problem because noninvasive neuroimaging has permitted no definite differentiation between two distinct pathological conditions. An enlargement of the subarachnoid spaces in children with macrocephaly is a frequent observation of mostly unknown etiology, but is known to be associated with a good prognosis. If surgery is necessary in these patients, ventricular shunting is required. On the other hand, subdural effusions are often of traumatic origin and frequently require neurosurgical intervention (subdural shunting). Most reports on extracerebral fluid collections in infancy have not differentiated between these two pathological conditions and therefore reveal confusing results. Recent studies using magnetic resonance imaging (MRI) have shown that vascular flow phenomena in the arachnoid space can be used for reliable diagnosis, whereas previous neu-

roimaging attempts, including high-resolution computerized tomography (CT) have been useless [1]. From 1992 to 1994 we investigated a cohort of 20 patients (five female, 15 male) aged 4–30 months (mean 10.5 ± 6.6 months), 16 with the history of macrocephaly and normal neurological development and four patients after head trauma and the symptoms of elevated intracranial pressure. In all 16 patients with the clinical diagnosis of benign subarachnoid space enlargement, colour-coded Doppler sonography detected arachnoid vessels within the fluid collection. Furthermore, high-resolution ultrasound demonstrated the dural border of the arachnoides as an echogenic membrane. This can be used as a further sign of the subarachnoid location of the fluid collection. In the four patients with subdural hematoma, the fluid collection showed an increased echogenicity, no vascular structures and no surrounding border. From these observations we conclude that high-resolution ultrasound and colour Doppler sonography are able to reliably differentiate between a subdural and a subarachnoid fluid collection. MRI, with its higher risks (sedation, anesthesia), focused only on this target, seems therefore unnecessary in these patients.

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Noninvasive evaluation of intracranial pressure in children by means of orbital sonography

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The presence of elevated intracranial pressure (ICP) is suggested by CT findings and monitored by invasive methods like intracranial catheters. A new tool for noninvasive estimation of ICP is measurement of optic nerve sheath diameter (ONSD). With B-scan sonography the perineural subarachnoid space can be depicted and changes recorded at the bedside. This phenomenon depends on pressure transfer from the subarachnoid intracranial compartments. Our **experimental** study with 40 postmortem human optic nerves showed that pressure induced bulging predominantly in anterior regions (mean 1.54 mm, range 0.9–2.2 mm). Effects of probe orientation, sound direction and different examiners proved negligible. On the basis of these results, a **clinical** investigation of 24 children under treatment for increased ICP was performed. With transbulbar sound direction the ONSD was measured repeatedly in axial views. Parallel changes of ONSD and ICP were recorded, especially when ICP increased. Age-matched controls never exceeded an ONSD of 4 mm, but all patients except two did. The peak ONSD was significantly increased in patients compared to controls (5.3 vs 3.0 mm; paired t -test, $P = 0.007$). In **conclusion**, findings of increased ONSD in children demonstrate elevated ICP, provided local orbital disease can be excluded. This noninvasive method is helpful for therapy planning and for selection of patients for invasive catheter monitoring.

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Accelerated myelination in early sturge Weber syndrome (SWS): MR – SPECT correlations

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The prognosis of SWS is partly related to early occurrence of seizures, but the diagnosis of this phakomatosis may be difficult during the first year of life. Neurological signs can be absent, and the usual neuroradiological abnormalities (cerebral atrophy, cortical calcifications, enlargement of the choroid plexus, perfusion abnormalities) may be delayed. We have performed a retrospective study of seven patients with confirmed SWS (7 days to 3 months old). None of the patients presented with seizures. They all underwent MRI (T1, T2 sequences) and SPECT (single-photon emission computed tomography) at the same time. Regional cerebral blood flow was measured using tomographic ^{133}Xe 564 × xenon-133. In all cases, myelination appeared to be accelerated in the abnormal cerebral hemispheres on both MR sequences. This was particularly obvious in the areas underlying the leptomeningeal angioma. In five cases, SPECT showed hyperperfusion in the damaged hemisphere. In one case, SPECT was symmetric; in one case SPECT showed an area of hypoperfusion in the damaged hemisphere which was already atrophied. These data suggest that the accelerated myelination is not related to ischemia but to transient hyperperfusion. This MR pattern can be helpful for the early diagnosis of SWS, which is of utmost importance because of the preventive antiepileptic treatment.

Quantitative color velocity imaging of carotid blood flow in children with cardio-cerebrovascular disease

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Background: Study of the interrelationship between cardiac function and cerebral blood flow is important in children following surgery for congenital heart disease (CHD) or embolization of cranial arteriovenous malformations (CAVM). Quantitative color velocity imaging (CVI-Q) is a new ultrasonographic technique to characterize and quantify volume flow (VF) in blood vessels [1–3]. Semiautomatically, a VF curve (in ml/min) versus time is generated, analogous to a Doppler spectral waveform of linear flow velocity.

Purpose: To determine the value of CVI-Q VF waveform analysis of carotid blood flow in children with CHD or CAVM.

Methods: We performed 65 carotid CVI-Q studies in 40 children with CHD, four with CAVM and 38 normals. Correlation was made with echocardiography, carotid spectral Doppler and clinical parameters of cardiac function.

Results: Normal carotid artery VF waveforms were characterized by antegrade flow during systole and diastole. Abnormal findings were: presystolic flow reversal in aortic regurgitation and significant PDA; postsystolic flow reversal in left ventricular output failure; holodiastolic flow reversal in large surgical L-R shunts; and increased VF with decreased pulsatility in high-output failure due to CAVM.

Conclusion: Analysis of carotid CVI-Q waveforms provides, non-invasively, useful hemodynamic information in children with CHD or CAVM.

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Reversal sign in neonatal hypothermia

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Newborn infants are particularly prone to hypothermia because of unsatisfactory heat production due to lack of movement, increased heat loss due to impaired vasomotor reflexes, large surface to body volume ratio and poorly developed subcutaneous fat. Our medical center serves a large semi-desert area with poor socioeconomic conditions compared to the rest of the country. Six out of 11 infants admitted with neonatal hypothermia (the majority being Bedouins) presented with multiple organ failure, seizures and coma. The CT scan demonstrated diffuse cerebral edema with “reversal sign”, a reversal of the normal density relationships of gray and white matter and a relatively increased density of the thalami, brainstem and cerebellum. The follow-up of five infants demonstrates severe developmental delay with brain atrophy and multicystic encephalomalacia. One infant expired after 30 days. The “reversal sign” is not specific. These patterns have been previously described in abused children with head injuries [1], preterm ischemia [2] and anoxia due to drowning [3]. However, it has never been described in hypothermia.

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Miscellaneous

Diagnosis of subglottic hemangiomas by laryngeal ultrasonography

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A laryngeal US was performed in 20 infants (aged 2–15 months) suffering from stridor and laryngeal dyspnea. Seven of them had a cutaneous facial hemangioma. The results of US were compared to the endoscopic findings. In 16 cases, the radiologist was unaware of the endoscopic findings at the time of the US examination. In nine cases, the subglottic area appeared normal at US as well as at the endoscopic examination. In 10 cases, US diagnosed and located a hemangioma with good accuracy. In one case, the hemangioma was initially overlooked by endoscopy because of an associated edema and was found retrospectively in US as a hyperechoic spot within a subglottic edema. This hemangioma was followed up by US and progressively increased in size. The hemangioma was left-sided ($n = 4$), right-sided ($n = 3$), posterior ($n = 2$) or circumferential and predominantly posterior ($n = 2$). The smallest lesion had a 3 mm diameter. Compared to muscles, the hemangioma was isoechoic in six cases and hyperechoic in five cases. The main differential US diagnoses of subglottic hemangiomas are briefly discussed. In conclusion, subglottic hemangioma are the most frequent benign laryngeal tumor in infants. Their great volume variability with time explains the false-negative results on endoscopy. US is easily reproducible and appears to be a valuable tool in the diagno-

sis of subglottic hemangiomas. Moreover, it could be more reliable than endoscopy in the valuation of the modifications in size of the hemangioma after treatment, since it is performed in physiological conditions.

Burkitt's lymphoma of the jaws revisited: radiology of pre- and post-chemotherapy changes

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Methods: A prospective study of 14 children (six males, eight females, age range 3–8 years) admitted to the ENT wards of Kenyatta National Hospital, Kenya with a proven diagnosis of Burkitt's lymphoma of the jaws was carried out. Standard views of the facial bones and the mandible were obtained in all patients prior to chemotherapy. Post-treatment radiographs were acquired in six patients. The clinical and radiological distribution of the tumours was tabulated according to whether one or more jaw quadrants were involved.

Results: Clinically 29% of patients and radiologically 92% of patients had involvement of more than one jaw quadrant. Some 42% of patients had involvement of all quadrants. The clinical and radiological presentations were tabulated. All cases had a history of painless loosening of teeth, distortion of intraoral ridge and an ulcerated intraoral mass. Radiologically, one patient had early signs of the tumour, i. e., loss of lamina dura around the affected first mandibular molar surrounded by small discrete lucencies and an enlarged crypt shadow. The rest of the patients showed late signs. Mandibular tumours showed an ill-defined lucency, displaced teeth and loss of lamina dura. Maxillary tumours showed antral opacification, bony destruction, nasal mass plus dental changes. Post-chemotherapy films in three patients showed grossly displaced teeth returning to their normal sites. Bony regeneration without regeneration of the lamina dura occurred in all six cases.

Conclusion: (1) Radiographs show tumour formation in jaw quadrants not clinically involved. (2) In endemic areas it would be good practice to take jaw radiographs in children 3–8 years old with loose molars for which no local cause can be found. (3) After therapy, changes do not include regeneration of the lamina dura. It remains to be seen whether this has any effect on future dental development.

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Hypothalamic-pituitary vascularization in pituitary stalk transection syndrome: is the pituitary stalk really transected?

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Background: Pituitary stalk transection syndrome is defined as anterior pituitary hypoplasia, ectopic posterior pituitary at the hypothalamic level and “transected” pituitary stalk, affecting 30–45% of patients with “idiopathic” hypopituitarism.

Purpose: (1) To verify whether the pituitary stalk is not identified because of limitations of MRI technique or because of its real absence; (2) to examine the vascularization of ectopic posterior pituitary, which is normally supplied by the inferior hypophyseal arteries, and that of residual anterior pituitary, normally supplied by the superior hypophyseal arteries and portal system.

Patients and methods: Thirteen patients with hypopituitarism ranging in age from 11 to 24 years were studied. Eight had isolated growth hormone deficiency (IGHD) and five had multiple pituitary hormone deficiency (MPHD). MRI revealed anterior pituitary hypoplasia, unidentified pituitary stalk and posterior pituitary ectopia in all patients except for four with very thin pituitary stalk. Conventional and dynamic MRI studies were performed before and after Gd-DTPA. The times to enhancement of the straight sinus (temporal reference point) and the initial posterior and anterior pituitary were recorded and compared to those in normal subjects.

Results: Pituitary stalk was identified in eight patients after Gd-DTPA injection, six with IGHD and two with MPHD. Initial posterior pituitary enhancement occurred simultaneously with that of the straight sinus in all but four patients with IGHD (delay 3.7 s). The time to enhancement of the straight sinus and of the posterior pituitary was 7.4–14.8 s, as in normal controls. The initial anterior pituitary enhancement was 11.1–18.5 s and did not differ significantly from the controls.

Comment: A very thin vascular pituitary stalk was identifiable after Gd-DTPA in 60% of cases, while neural stalk disconnection was documented in all these patients. Ectopic posterior pituitary is rapidly vascularized, as is normally located posterior pituitary. The vascular system which supplies the ectopic posterior pituitary is arterial but it has not been recognized with certainty. The vascularization of the residual anterior pituitary occurs normally both in the presence and absence of vascular pituitary stalk. Pituitary stalk transection is an inappropriate term to define the syndrome of congenital hypopituitarism with anterior pituitary hypoplasia and posterior pituitary ectopia.

Left adrenal hemorrhage and calcified inferior vena cava thrombus: fortuitous association?

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Purpose: Presentation of a hemodynamic hypothesis to explain the association of a calcified inferior vena cava (IVC) thrombus and a left adrenal hemorrhage occurring in the perinatal period.

Material and method: We observed three boys with calcified IVC thrombus detected at birth on plain film of the abdomen and confirmed by ultrasonography. The thrombus was located at the level of T11–T12 with a “bullet-shaped” appearance in all cases. A left adrenal hemorrhage was associated, detected in the prenatal period in two patients (38 and 38 weeks) and at 2 days of life in the last one. Inferior cavography was performed in one patient, demonstrating the exact position of the thrombus between the level of the right adrenal vein, above, and the left renal vein, below.

Discussion: The similarity on location and shape of calcified IVC may suggest stationary flow in the IVC, caused either by an anatomical variant or, more likely, by an increasing flow in the ductus venosus. This last hemodynamic change have been demonstrated in perinatal fetuses when prolonged hypoxia occurred. The stationary flow may result in increased pressure in the IVC, the renal veins and therefore the left adrenal vein, resulting in rupture of fragile adrenal vessels and subsequent hemorrhage.

Conclusion: Anatomical and physiological anomalies in fetal venous blood flow may explain the likely nonfortuitous association of antenatal IVC thrombus and left adrenal hemorrhage.

Serial imaging of the liver in patients with glycogen storage disease

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Purpose: Children with glycogen storage disease (GSD) are at risk for developing hepatocellular adenoma and carcinoma. The pur-

pose of this project is to review imaging in children with GSD to help guide screening.

Materials and methods: Serial ultrasound (US) images of 35 patients with GSD were obtained. Each patient was followed up for an average of 6.6 years. Six children with focal liver lesions (FLL) on US had magnetic resonance (MR) studies. The US and MR studies were retrospectively reviewed to assess for focal lesions.

Results: FLL on US showed three patterns: hypoechoic, hyperechoic or target-like. On MR, FLL had a low T1 and high T2 signal. While most FLL were seen on both MR and US, some were seen on only one study. Of the 13 patients (37%) that developed FLL, the average age at recognition was 13 years (range 6–27 years).

Conclusion: Based on clinical assessment, none of our group have developed hepatocellular carcinoma, but 37% have at least one presumed adenoma, with the youngest patient being 6 years old. Based on our and other's data, FLL are rare below the age of 6 years, and yearly screening before that age may not be necessary. It is not clear whether US or MR is the better screening modality.

Displacement of the posterior rim apophysis of lumbar vertebra in adolescents: indications for CT and MR

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Purpose: The purpose of this retrospective study was to determine the value of CT and MR in adolescents with displacement of the posterior rim apophysis of lumbar vertebra.

Patients and methods: Over a 8-year period, seven children in this series (age range 10–15 years, mean 12 years) were referred to the orthopedic department for lumbar and sciatic pain. Four patients had an initial acute trauma. In all cases, conventional X-rays were performed. Six underwent CT, and four had a MR study with T1- and T2-weighted images.

Results: Plain X-rays showed avulsion of the posterior rim apophysis in three cases. Two patients had associated isthmic lysis. CT and MR studies showed avulsion in all cases. MR T2-weighted images demonstrated an abnormal intervertebral disc signal in two cases. It also showed in all cases disc material herniated between the vertebral body and the posterior rim apophysis. In one case MR demonstrated disc herniation at the same level.

Discussion: In the literature two etiologies are suggested: true fracture or lumbar posterior marginal node. According to Laredo et al. [1] and Takata et al. [2], true fracture is usually associated with acute trauma and lumbar posterior marginal node with irregularities of the end plate. Our series does not correlate with such findings.

Conclusion: CT is valuable for the diagnosis. MR (sagittal T2-weighted images) shows the lumbar posterior marginal node and the posterior rim apophysis. Anatomy and pathophysiology of these lesions remain controversial. MR performed early in the course of the disease may lead to a better understanding.

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Autosomal recessive osteopetrosis: MRI and scintigraphy after bone marrow transplantation

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The outcome of bone marrow transplantation (BMT) in two patients with autosomal recessive (AR) osteopetrosis was analyzed, performing MRI and technetium-99m sulfur colloid scintigraphy. The patients were enrolled in a protocol after BMT involving an extensive metabolic, clinical, and imaging workup. The imaging component of this protocol included sequential bone marrow MRI and scintigraphy and routine skeletal radiography. Scintigraphic images were evaluated by a radiologist, who was blinded to the clinical status of each patient as well as to the findings at MRI and conventional skeletal radiography. A general assessment of the marrow stores after BMT was performed, and non-blinded correlations were made with the simultaneously acquired imaging data. The recovery of osteoclast function after BMT allows an improvement of bone marrow stores. Bone marrow scintigraphy provides a direct, whole-body method for visualizing the marrow spaces. These marrow spaces are less opaque than the surrounding sclerotic bone on conventional radiographs and may be of higher signal intensity than sclerotic bone on T2-weighted MR images. MRI may be helpful in evaluating marrow stores in the pelvis and spine, where scintigraphy is limited by the exuberant activities of the liver and spleen. Knowledge of bone marrow activity can be useful in assessing the success of BMT, which remains the only curative treatment for the majority of patients with AR osteopetrosis.

Pyogenic thyroiditis in children due to a fourth branchial pouch fistula

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Acute suppurative thyroiditis is a rare disease in childhood. It is usually due to a sinus tract extending from the tip of the piriform sinus to the thyroid region. Embryologically it arises from a fourth branchial pouch fistula. It often presents as an abscess in the lower lateral neck, usually on the left side. The diagnosis can be suggested by US, radionuclide scintigraphy, or CT, but the diagnosis can only be established by barium swallow, demonstrating the sinus tract. However, the barium swallow can be a very difficult procedure with false-negative results. Demonstration of an air bubble in the abscess by plain film or CT is very suggestive of the diagnosis. Demonstration of the fistulous tract by air while drinking a carbonated beverage can be an alternative diagnostic tool replacing the barium swallow. Complete surgical excision of the sinus is essential. Five cases of pyogenic thyroiditis due to a piriform sinus fistula are presented, four of them on the left and one on the right. The patients ranged in age from 1 to 15 years. The value of various diagnostic modalities will be shown and discussed.

MR imaging of normal age-related bone marrow: shoulder, thoracic wall and extremities

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To evaluate the normal age-related bone marrow change of thoracic wall, shoulder and extremities, we retrospectively reviewed 122 T1 weighted MR examinations of thoracic wall and shoulder and 128 examinations of upper and lower extremities. The patients had no history of bone marrow disease. Concerning thoracic wall and shoulder, we evaluated the bone marrow changes in three anatomic segments of the proximal humerus (epiphysis, metaphysis, proximal diaphysis) and three segments of the thoracic wall (sternal manubrium, scapular body and ribs). The patients were divided into six age groups: 0–1, 1–3, 4–6, 7–10, 11–15, and 16–24 years. The signal intensity of all anatomic segments was as low as or lower than that of muscle in all patients younger than 3 months, reflecting an underlying hematopoietic marrow. The first segments to become hyperintense were the humeral epiphyseal ossification cen-

ter, the diaphysis and the scapula. While in the humeral epiphysis and diaphysis bone marrow reaches a diffuse increased signal intensity with age, the sternum, ribs, scapulae and humeral metaphysis maintain varying percentage of intermediate signal intensity. Regarding upper and lower extremities, we considered the metaphysis and diaphyses of major long bones. Among round bones, the capitata, calcaneus, talus and patella were considered. Proximal femurs were excluded because previously evaluated. Five age groups were studied: 0–1, 2–5, 6–10, 11–15, and 16–24 years. The signal intensity of all anatomic segments was as low as or lower than that of muscle in all patients younger than 2 months. After this age, the first anatomic segments to become hyperintense were epiphyseal ossification centers, metatarsal bones and phalanges, followed by diaphyses and metaphyses. In the round bones, age-related T1-weighted MR images follow a pattern similar to the epiphyses. The pattern of physiologic conversion from red to yellow marrow in different anatomic segments, related to the age groups, is reported. Knowledge of the normal age-related MR appearance of red and yellow marrow is necessary for the recognition of abnormal patterns as well as of marrow infiltration by tumors or other pathologic processes.

The natural history of deferoxamine-induced platyspondyly in hypertransfused thalassemic patients: its similarity to post-radiation platyspondyly

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Reports have appeared in the past 5 years of skeletal dysplasia (both long bone and spine) resembling spondyloepiphyseal dysplasia in patients with thalassemia treated with hypertransfusional therapy and deferoxamine chelation to reduce iron overload. The affected patients had long-duration therapy, often starting in the first 2–3 years of life and at a dose level considered high by today's standards. We report the natural history of spine changes in two patients whose spine films were available from infancy through childhood until adult life. Each went through a radiographic sequence of normal spine appearances at onset to bulbous vertebrae to eventual flattening identical to the changes reported by Neuhauser and colleagues in the early 1950s in survivors of childhood abdominal tumors treated with radiation. Radiation therapy has also been noted to cause long bone rickets-like metaphyseal irregularities similar to those seen with chelation therapy. The common pathway in both (post-deferoxamine and post-radiation therapy) is probably a primary effect on cartilaginous, not bone, growth and development with an effect on enchondral ossification.

MR findings in four children with "idiopathic" panhypopituitarism

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Purpose: Attempt to define (a) the patterns of hypothalamo-hypophyseal abnormalities identified by MR imaging in children with „idiopathic panhypopituitarism” and (b) any relationship between MR findings and patient's clinical history.

Patients and methods: MR imaging was performed in three girls and one boy (4–11 years old) with panhypopituitarism and no diabetes insipidus. The studies were performed on a 2.0-T MR unit (Bruker, Germany). T1-weighted images (500/20/4, TR/TE/NEX), before and after contrast were acquired in the sagittal and coronal planes. The scans were evaluated for anterior and posterior pituitary size and location as well as pituitary stalk integrity.

Results: Three patterns of pituitary lesions were identified: (1) aplasia of adenohypophysis and absent stalk in one case, (2) hypoplasia of adenohypophysis and absent stalk in two cases, (3) hy-

poplasia of adenohypophysis and stalk in one case. In addition ectopy of neurohypophysis at the median eminence of hypothalamus was noted in all children. Size of adenohypophysis did not correspond to plasma levels of anterior pituitary hormones. All children had a history of perinatal complications: asphyxia in two, severe hyperbilirubinemia in one and complicated breech delivery in one. **Conclusion:** We hypothesize that the three combinations of findings represent three different degrees of perinatal, traumatic and/or ischemic injury to the pituitary stalk. The partial or complete disruption of the hypothalamic-hypophyseal portal system would explain the hormonal deficiency of the adenohypophysis. Absence of diabetes insipidus might be due to a spontaneous regeneration of an ectopic neurohypophysis at the proximal end of the transected neurohypophyseal tract.

Gastric emptying: validation of a US method by comparison to scintigraphy

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Purpose: Emptying appears very important in the assessment of gastroesophageal reflux when considering scintigraphic studies. The latter are very precise methods, but are based on expensive equipment and deliver radiation. US, which is non-invasive and more physiological, has been proposed as an alternative. In this study, US and scintigraphy (Sci*) were compared in order to validate the US method and to make it acceptable in clinical practice.

Population and methods: Ten vomiting children aged 2 months to 1 year were examined in a double-blind manner by simultaneous US and Sci* examination, after 25 ml/kg milk feeding added with 200 Ci ^{99m}Tc. Five scintigraphic measurements were made at 0, 30, 60, 120 and 180 min. US evaluation took place, each time, immediately before and after the Sci* measures. Two US examinations according to Lambrecht's technique were performed in two orthogonal cross sections of the gastric antrum with the patient, in the right lateral decubitus position. The first section was cranio-caudal, passing through the vena cava and the mesenteric vein, the second transverse, passing through the pylorus and the gallbladder. A gastric filling index was determined in a simplified way, considering the value obtained just after the meal as 100 % and expressing following determinations as percentages of this value.

Results: The patterns of gastric emptying as expressed by the Sci* time-activity curve were in good accordance with the US emptying in all except four of 50 instances. In two of these cases, the Sci* measures were determined from posterior and the overlying duodenum made evaluation of emptying inaccurate. In the other two, the residual gastric content was small (< 30 %) and became masked by air.

Conclusion: We conclude that this kind of US study of the antropyloric region allows accurate, non-invasive determination of gastric emptying and can easily be included in each study of gastroesophageal reflux.

Chest imaging

Childhood pneumonia: risk factors for delayed resolution of parenchymal infiltrates

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We studied the influence of some supposed clinical and radiological risk factors on the time of radiological healing of childhood pneumonia. A total of 597 children with clinical signs of acute

pneumonia were included. Pulmonary infiltrates were verified at radiography. Chest X-ray was repeated after 4–6 weeks. Children with residual infiltrates had a third chest examination another 3–6 weeks later. One hundred and seventy-eight children had residual changes at the second chest X-ray, and among these, 57 patients still had radiological changes at the third X-ray. The history of the children and the size and type of the radiological findings at the first X-ray were analyzed for possible predisposing factors for delayed healing. The results were:

	A (n = 597)	B (n = 121)	C (n = 57)
Chronic disease	6 %	7 %	18 %
Recurrent pneumonia	11 %	17 %	23 %
VOC	3 %	2 %	11 %
History of neonatal disease	3 %	5 %	5 %
Asthma	14 %	21 %	9 %
Large infiltrates	20 %	17 %	44 %
Atelectasis	17 %	19 %	40 %

Group **A**, the total study group; group **B**, children with infiltrates on the second but not on the third X-ray; group **C**, children with residual infiltrates at the third chest film.

Chronic disease such as Down's syndrome, congenital heart disease, immunological deficiency and cerebral palsy, and a history of repeated pneumonia, were significant factors associated with delayed resolution of parenchymal infiltrates. Among the children with residual parenchymal changes at the third X-ray control, we also found one child with a foreign body in the airways, one tumor case, and a previously unknown case of congenital heart disease. Asthma was not associated with delayed healing of pneumonia. The presence of large infiltrates and atelectasis on the first chest examination was associated with delayed radiological healing. The likeness between groups A and B in these respects seems to indicate that the optimal time for control X-ray may be more than 6 weeks after the initial examination in cases without known predisposing factors.

Right aortic arch: can mediastinal density help detection?

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Introduction: The diagnosis of a right aortic arch on chest roentgenograms (CXR) is problematic, particularly in neonates and infants. Nevertheless, identification is important since there is a high association of right arch with congenital heart disease and vascular rings. Identification of tracheal indentation, tracheal deviation, position of aortic knob or aortic descent have been used to determine the side of the aortic arch. As the arch is an additive water density on the CXR, it increases the overall mediastinal density adjacent to the trachea. A retrospective study was carried out to determine which sign or signs are most useful in diagnosing a right aortic arch on CXR.

Material and methods: The initial CXRs of 37 children with a right aortic arch, confirmed by angiography, were independently reviewed by three pediatric radiologists. Each radiologist determined which of the following signs were present on each CXR: aortic knob, aortic descent, tracheal indentation, tracheal deviation, and mediastinal density sign. Differing observations were resolved by consensus. The original CXR report was reviewed to determine the number of patients initially diagnosed as having a right arch.

Results: There were 17 females and 20 males ranging in age from 1 day to 68 months (mean 8 months). Of the five radiographic features, the mediastinal density sign was observed most frequently (81 %). Identification of a portion of the right descending aorta was the next most frequent finding (70 %). These two radiographic observations, alone or in combination, identified over 90 % (34/37)

of the right arches. Tracheal indentation (40 %) or deviation (27 %) were relatively infrequent and were more subjective, being dependent on visualization of the airway and patient rotation. Identification of the aortic knob was least frequent (16 %). The initial CXR interpretation correctly identified the right arch in only 51 % (19/37) of patients. A left arch was reported incorrectly in two patients, the arch position was stated to be indeterminate in five patients, and in 11 patients the position of the arch was not mentioned.

Conclusions: A right arch is often missed on the initial CXR. The roentgenographic diagnosis of a right aortic arch is best made using the mediastinal density sign and observation of right aortic descent. Identification of tracheal indentation, deviation, or the aortic knob may be helpful but are rarely isolated findings.

Musculoskeletal imaging I

Normal craniovertebral bone and ligament anatomy, and the diagnosis of occipital condyle fracture and ligament injury using fine-slice CT with reconstructions

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Purpose: To demonstrate normal craniovertebral bone and ligament anatomy and to detect fractures of the occipital condyles and ligament injury.

Materials and methods: Seven children and young adults were studied (age 4.5–21 years, median 11 years) after decelerating head injury with components of either high velocity or axial compression. Glasgow Coma Scale score on admission ranged from 3 to 15, median 7. With appropriate cervical protection and in the supine position, patients underwent fine-slice (1.2 mm × 1 mm), contiguous, axial computed tomography of the region between the second cervical vertebra and the foramen magnum, either at initial presentation or during hospital admission. Coronal, sagittal and curvilinear reconstructions with bone and soft tissue windows were performed. Normal bone and ligament anatomy was identified, including occipital condyles, first and second cervical vertebrae, transverse ligament of atlas, alar ligaments, and tectorial membrane, as well as the posterior atlanto-occipital membrane and apical ligament of dens. Occipital condyle fractures were classified as type I, II, or III according to Anderson and Montesano [1] and displacement and hematoma were described. Ligament injury included disruption (discontinuity) or asymmetry (ipsilateral shortening or contralateral stretching) of the alar ligaments.

Results: Four occipital condyle fractures were identified in three patients (one bilateral type I and two unilateral type III). Ligament injury was seen in one case and hematoma in two. Normal bone and ligament anatomy was clearly demonstrated in four cases.

Conclusions: Craniovertebral bone and ligament anatomy, fractures, and ligament injury can be clearly demonstrated using fine-slice axial computed tomography with reconstructions. The severely injured patient need not be endangered by a coronal scanning angle, and delineation of pathology may guide the therapeutic approach.

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Computer-assisted assessment of osteopenia

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Purpose: The term *osteopenia* is used to indicate a reduction of calcified bone, which may be the result of many skeletal disorders frequently indistinguishable by conventional radiography. This study assesses the value of digital radiographic techniques in determining the cause of osteopenia.

Methods: The amount of cortical bone and its degree of mineralization (bone density) were measured with special CT software in the mid-shaft of the femurs of 102 children. Skeletal radiographs of 51 children (24 girls, 27 boys, ages 4–19 years) manifested osteopenia; 31 had osteoporosis, nine had osteogenesis imperfecta, two had primary hyperparathyroidism, and eight had familial hypophosphatemic rickets. For comparison, a control group of 51 children of the same age, sex, weight and height were studied.

Results: Neither gender, age, height nor weight influenced cortical bone density, and values were similar for the 51 healthy children. Compared to controls, children with osteoporosis and osteogenesis imperfecta had reduced bone mass, but similar bone density. In contrast, cortical bone density was reduced in children with hyperparathyroidism and rickets (1150 ± 30 vs 910 ± 120 mg/cc; $P < 0.001$).

	Osteoporosis	Osteogenesis imperfecta	Hyperparathyroidism	Rickets
Bone mass	↓	↓	↓	Normal
Bone density	Normal	Normal	↓	↓

Conclusion: Unlike conventional radiography, CT aids in distinguishing among the various disorders that reduce bone mineral content.

Complementarity of [¹²³I]MIBG scintigraphy and MRI in the detection of bone marrow metastasis of neuroblastoma

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Purpose: To compare [¹²³I]metaiodobenzylguanidine scintigraphy (MIBG) and magnetic resonance imaging (MRI) in the diagnosis and follow-up of bone marrow metastasis in children with neuroblastoma.

Patients and methods: Nine patients, 8 boys and 1 girl, aged from 2 months to 11 years, (mean 5 years) were studied by MIBG and MRI. Surgical biopsy and bone marrow biopsies were performed in all patients. A negative bone marrow biopsy in the presence of a positive MIBG and/or MRI was followed in two patients by a vertebral bone biopsy. Histologic examination revealed neuroblastoma in seven patients and ganglioneuroblastoma in two patients.

Results: At diagnosis, six of nine patients were negative for all three methods and remained negative on follow-up examinations. Three patients showed signs of bone marrow infiltration on MIBG and MRI, which were confirmed by bone marrow biopsy. Two patients, a case of normalisation and a case of relapse, had concordant findings on MIBG and MRI at follow-up examinations. One patient showed complete healing of the bone marrow infiltration on an MIBG scan performed after four courses of chemotherapy. However, a persistent hypointense signal of the bone marrow on MRI was suggestive of marrow infiltration until 15 months after diagnosis. In this patient, marrow and vertebral body biopsies showed the absence of tumour.

Conclusions: These results suggest a good concordance between MIBG and MRI findings for bone marrow infiltration. Further studies are needed to understand the differences of kinetics in bone marrow response to treatment. It is essential to combine functional data given by MIBG and morphological data given by MRI for a better staging and treatment follow-up of neuroblastomas in children.

Value of MRI in the early diagnosis of growing skull fracture

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Growing skull fracture (GSF) is a progressive enlargement of fracture due to an underlying tear of the dura. It is a rare complication of severe head injury mainly reported in young children. Classically, the diagnosis is made on clinical follow-up weeks or months after the original injury on the basis of a palpable skull defect or a bulging mass. Initial skull X-rays show a large (3–4 mm) fracture developing to a large bone defect with irregular edges when diagnosis of GSF is made. CT is useful to show evidence of brain damage always present beneath the fracture. We present the MRI findings of GSF in a series of eight children. They were 1 month to 8.5 years old. Seven children had a severe head injury and the last one probably a birth trauma. All children underwent skull X-rays and cerebral CT at the time of initial injury. MRI was performed for six patients in the first 3 months after trauma (day 8 to 3 months). Two children underwent MRI with a delay of 8 months and 6 years. All children had initially a large linear fracture and on CT underlying brain damage. In all cases the MRI showed a zone of the same intensity as the brain contusion or CSF advancing through the bone margins of the fracture to the subcutaneous plane. This finding was interpreted as an indirect sign of the dural tear. Six of our eight children were operated on, with surgical confirmation and repair of GSF, ranging from 2 to 14 months after the initial injury. One child has only undergone a shunting procedure because of severe hydrocephalus. In one case the parents refused surgery. We think that MRI can yield an early diagnosis of GSF, even at the time of the initial injury. Surgical repair of the lesion which consists mainly of closure of the dura, could be proposed earlier, before the increase of the dural tear underlying the GSF. Moreover, some authors believe that progressive brain damage could happen in injured cerebral tissue unprotected by dura.

Intervention for lung abscesses

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Purpose: To determine whether percutaneous aspiration and/or catheter drainage aids in the diagnosis and treatment of lung abscesses.

Methods: From 1987 to 1995, 13 children with lung abscesses were treated with either percutaneous therapeutic needle aspiration (eight cases), percutaneous catheter drainage of the abscess (four cases) or both (one case). Guidance for interventional therapy was by ultrasound (10 cases), fluoroscopy (three cases), and/or CT (two cases).

Results: Eleven of 13 bacteriologic samples had positive Gram stains despite prior antibiotics. Ten patients became afebrile within 24 h of intervention. Three of four patients demonstrated bronchial communication on follow-up abscessogram. Duration of abscess catheter drainage was 4–20 days (mean 12 days). Five patients developed pleural effusion treated by percutaneous chest tube drainage. No patient developed a bronchopleural fistula; there were no foreign bodies identified. Only the patients with a pre-existing cyst or sequestration required a lobectomy; the other 11 healed without surgery.

Conclusion: Percutaneous sampling determines the organism and its sensitivity to assure proper antibiotic therapy. Percutaneous needle aspiration is therapeutic in abscesses with 15 ml of pus or less. Percutaneous catheter drainage is therapeutic for abscess collections as large as 100 ml. In our experience, bronchoscopy was unnecessary.

Musculoskeletal imaging II

The radiological spectrum of the Shwachmann syndrome

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Purpose: To describe the varied and diverse skeletal manifestations of a cohort of 22 patients with clinical diagnosis of Shwachmann syndrome (metaphyseal chondrodysplasia with exocrine pancreatic insufficiency and cyclic neutropenia), their spectrum of radiological abnormalities, the natural history of the disorder and the impact of therapy.

Materials and methods: The serial radiographs of 22 children with Shwachmann syndrome [mean age 4 years 2 months, range 2 months–11 years] were retrospectively reviewed by two radiologists and a consensus obtained for the presence and distribution of skeletal abnormalities, including: delayed skeletal maturation, the distribution of metaphyseal chondrodysplasia, the presence of coxa vara, slipped capital femoral epiphyses, thoracic cage deformity and configuration of the costochondral junctions, the presence of vertebral anomalies, scoliosis, abnormal long bone tubulation, clinodactyly and phalangeal hypoplasia. Imaging of the pancreas, if performed, was analysed.

Results: Skeletal maturation was normal in 9/22 patients and delayed in 13/22 (59%). Mean delay was 24 months [range 6 months (at the knee) to 4 years (at the wrist)]. Metaphyseal chondrodysplasia was classed as minimal in 4/22 patients. The more severely affected regions included the hips in the vast majority (20/22 patients), the knees in 1, and the ankles in another patient. In order of severity, the other affected metaphyses were at the knees (15/22), shoulders 8/22, wrists (6/22) and ankles (3/22). Coxa vara was present in 7/22 patients with marked shortening of the femoral necks, requiring femoral osteotomies in two patients. Two children developed slipped capital femoral epiphyses (bilateral asymmetrical). The sacroscliac notch was narrow in 3/22 patients, and in a further three patients the pubic symphysis was wide with hypoplasia of the inferior pubic rami. The thoracic cage was elongated and narrow in 14/22 patients, with a coat-hanger configuration of the ribs, with shortening and gross expansion of their distal ends in 8/22 and less marked metaphyseal splaying and cupping in 6/22 patients. A thoracolumbar scoliosis was present in 5/22 patients, increased coronal height and biconvex irregular end plates in the upper lumbar vertebral bodies in 8/22 patients, and marked lumbar lordosis and sacral tilt in 4/22 patients. Abnormal long bone tubulation was present in 11/22 children, manifested as flaring of the distal femoral metaphysis and gracile bowing of the diaphysis, with rhizomelic shortening in 3/22, and a pseudochevron deformity at the knee in 2/22 children. Clinodactyly was seen in 11/22 patients, with hypoplasia of the middle phalanges of the fifth fingers, and 9/22 showed the formation of pseudoepiphyses at the bases of the first, second and fifth metacarpals. One child developed acute leukaemia, with osteopenia and further metaphyseal lucencies, another chronic lung disease secondary to scoliosis. Two children had pancreatic CT with gross fatty change, duct ectasia and calcification. Two further children developed nephrolithiasis.

Conclusion: The spectrum of skeletal abnormalities in Shwachmann's syndrome is wide.

Ultrasound imaging of the hip joint synovial-capsular complex in mucopolysaccharidosis

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The purpose of this study was to document abnormalities of the hip joint synovial-capsular complex (SCC) in mucopolysaccharidosis (MPS) and to seek a possible simple method for assessing response to enzyme replacement therapy in this condition. Ultra-

sound imaging of the hip was performed prospectively in ten patients with MPS. The study group consisted of 1 Hurler, 3 Sanfilippo, 4 Morquio and 2 Maroteaux-Lamy syndrome patients. Using a standard technique, the ultrasonographic characteristics of the hip joint SCC were evaluated and assessed quantitatively. MRI correlation was available in three cases. Pathologic correlation was obtained in one case of Morquio and one of Maroteaux-Lamy syndrome. In a normal population of 65 patients, a logarithmic relationship was demonstrated between the hip joint SCC distance (SCCD) and patient age. No significant difference ($P < 0.01$) was shown between the sexes or sides. In all MPS patients there was significant thickening ($P < 0.001$) of the hip SCCD when compared to normals. In addition, the Maroteaux-Lamy patients showed a significantly ($P < 0.05$) greater involvement than the remainder of the MPS types. SCC thickening was related to soft tissue infiltration, which was isoechoic or slightly hyperechoic to muscle and of homogeneous echotexture. Most deposition occurred in the soft tissues superficial to the joint space, MRI confirmed the ultrasound findings, demonstrating the abnormally thickened capsule to be isointense to fibrous tissue and the synovium to be isointense to muscle on T1, T2 and FLASH sequences. Histopathologic examination revealed abnormal thickening of the fibrous hip joint capsule with infiltration of both the superficial and deep layers of hypertrophied synovium by mucopolysaccharide storage cells, shown by immunoelectronmicroscopy to contain glycosaminoglycans. The degree of SCC thickening correlates mainly with the amount of mucopolysaccharide deposition in hip synovium. It probably represents a major contributing factor in causing joint stiffness, which was present in all of our cases. Ultrasound is a simple, noninvasive technique allowing quantification of mucopolysaccharide deposition in the hip and may have a role in evaluating response to treatment, particularly with current developments in recombinant enzyme replacement therapy [1, 2].

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Autosomal recessive osteopetrosis before and after allogeneic bone marrow transplantation: bone marrow MRI at 1.5 T

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Purpose: To document the MRI features of autosomal recessive osteopetrosis and define the changes observed following allogeneic bone marrow transplantation (BMT).

Materials and methods: Six children with autosomal recessive osteopetrosis (four severely affected, two intermediate form – one in association with carbonic anhydrase deficiency) were imaged with a 1.5-T super-conducting magnet (Siemens), using spin echo technique with two pulse sequences. Multiplanar images of the spine and femora were acquired with T1 and T2 weighting.

Results: Eight scans were performed on six patients – four before and four after BMT (one patient was imaged both before and after BMT, another on two separate occasions after BMT). In all severely affected patients, MRI of the spine and abdomen prior to BMT showed complete lack of signal from the bone marrow alternating with signal intensity equivalent to intervertebral discs, resulting in a step-ladder appearance of the spine. Hepato-splenomegaly with low signal relative to the kidneys was present on short-TR/TE images due to extra-medullary haemopoiesis. One patient imaged 18 months after BMT had central low signal with intermediate signal peripherally in the vertebral bodies, the humeral and femoral

medullary cavities appearing of normal signal, consistent with active haemopoietic marrow and successful engraftment with functioning donor osteoclasts. The size of and signal from liver and spleen were normal. A subsequent scan at 20 months showed regression of the marrow spaces to the pre-treatment status coincident with late graft failure. Another patient imaged before and 6 weeks after BMT showed little significant change, with minimal increased signal peripherally at the vertebral bodies and persistent hepato-splenomegaly, coinciding with failure to engraft donor osteoclasts and reconstitution with autologous marrow. A further patient showed linear metaphyseal high signal at the knee after BMT, consistent with early osteoclastic activity confirmed clinically by the detection of donor DNA in the peripheral blood using molecular probes.

Conclusion: MRI is useful before and after BMT in patients with osteopetrosis, as successful engraftment or reconstitution by native marrow can be differentiated.

Fibromatoses in children: review of 17 cases

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Fibromatoses are mesenchymal tumors of fibroblastic and/or myofibroblastic derivation. They comprise approximately 12% of soft tissue neoplasms in childhood and differ in many respects from their counterparts in adults [1]. Various subtypes have been described exhibiting benign, indeterminate, and malignant biologic behavior. Few case reports exist on the imaging characteristics of fibromatoses. We reviewed the radiologic findings of 17 pathology proven cases with fibromatosis. Infantile myofibromatosis, fibromatosis not otherwise specified, and aggressive fibromatosis were the most frequent diagnoses. Others included fibrous hamartoma of infancy and fibrosarcoma. We analyzed the features on US, CT, and MRI, which allowed us to define the location, size, contour, and architecture of the lesions within the soft tissues. Radiography in addition allowed characterization of osseous involvement. Imaging findings of all entities will be presented. Both specific and nonspecific features of the different entities were noted. In conclusion, we found plain radiography and MRI particularly useful in preoperative assessment and staging of fibromatoses. Specific features in some of the lesions may obviate biopsy.

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Bone fractures in children undergoing orthotopic liver transplantation

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Orthotopic liver transplantation (OLT) is an established form of treatment for children with a variety of liver disorders. It is well recognized that these children are at risk of developing complications due to the underlying liver disease or as a result of OLT itself. There are a number of factors which may be important in the aetiology of bone fractures in such children, including trauma, metabolic bone disease, drugs and immobilisation. The purpose of this study is to document the incidence and distribution of fractures in children with liver disease, both before and after OLT, and to attempt to identify which children are at risk of developing fractures. One hundred and seventeen children (60 male, 57 female) underwent OLT (141 grafts) over a 6-year period (1987–1993). Ninety-nine children (84.6%) were transplanted for chronic liver disease

and 18 (15.4%) for acute liver failure. All plain-film radiology, both before and after OLT, was retrospectively reviewed. In those children in whom fractures were found a detailed analysis of clinical data, including history of trauma and drug history, was carried out. Nineteen children (16.2%) sustained a total of 69 fractures. Thirteen children (11.1%) were pre-OLT and six children (5.1%) were post-OLT when the fractures occurred. Age at time of fracture ranged from 3 months to 9 years 8 months (median 13.5 months). The reason for OLT in the children with fractures was extrahepatic biliary atresia ($n = 12$), neonatal hepatitis ($n = 2$), Alagille's syndrome ($n = 1$), tyrosinaemia ($n = 2$) and primary hyperoxaluria ($n = 2$). Metabolic bone disease, assessed qualitatively on plain radiographs, was present in 17/19 children at the time of fracture. This took the form of rickets ($n = 3$), osteopenia ($n = 12$) and osteosclerosis ($n = 2$). There was no documented trauma in 14/19 cases. Children in this group were aged from 3 months to 9 years 8 months (median 7.75 months) at the time of fracture, and in six cases the fractures occurred during the hospital stay in which the OLT took place. Common factors in these six children were young age, generalized skeletal demineralisation due to chronic cholestatic liver disease, prolonged hospitalisation and in the post-OLT phase, the use of drugs, including steroids, cyclosporin and, in one case, heparin. The findings demonstrate that children undergoing OLT are susceptible to fractures and that this risk is probably related to end-stage chronic liver disease. The need for clinical and radiological awareness of the fracture risk, particularly during the peri-transplant period, is emphasized.

Contribution of ultrasound to limb lengthening

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Osteotomy followed by limb lengthening is a new orthopedic procedure in the treatment of asymmetric limb development. Starting the distraction at the proper time is important. Conventional radiography shows the developing callus too late and until now distraction between the two osseous fragments has been begun on a theoretical base. We believe that ultrasonography has potential value in accurately measuring the distraction gap and in imaging the new bone formation earlier than radiography. Limb lengthening in a small series ($n = 5$) was monitored by ultrasound scans. The distraction gap appeared as a sonolucent area within which echogenic foci developed and became aligned in the longitudinal plane. We propose an echographic staging of new bone formation to enable the orthopedic surgeon to monitor the distraction rate in each particular child. Since overly fast distraction inhibits bone formation and overly slow distraction leads to premature consolidation, ultrasound may serve a useful role in the qualitative evaluation of new bone in limb lengthening.

A vascular necrosis in children with chronic renal disease

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Purpose: To determine the incidence of avascular necrosis (AVN) of the femoral head in children with chronic renal failure.

Materials and methods: We reviewed the AP radiographs of the hips of 205 children (159 boys, 47 girls, ages 6 months to 16 years, mean 5.7 ± 3.5 years) with chronic renal failure; six children had metabolic, 21 acquired and 178 structural renal lesions. All patients participated in multicenter studies evaluating the effect of recombinant growth hormone in skeletal growth. Radiographs were obtained every 6 months for periods extending from 1 to 7 years (mean 3 ± 2 years)

Results: Radiographic findings of AVN were seen in 14 children. There was no gender difference in the incidence of this complica-

tion, which occurred in 11 boys (6.9%) and three girls (6.4%). Affected children had short stature, markedly delayed bone development, and were frequently asymptomatic. In two instances, AVN developed while the children were on corticosteroid therapy.

Conclusions: The incidence of AVN in children with chronic renal failure is approximately 70 times that of idiopathic AVN. Unlike Legg-Calvé-Perthes disease, AVN in children with chronic renal failure is frequently asymptomatic and has no sex predilection.

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Congenitally short trachea with left mainstem bronchus obstruction by the ductal remnant: a syndrome of airway obstruction on an extrinsic vascular basis

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Congenital short trachea (CST) has diminished tracheal rings, a high carina, and has been primarily reported as a pathologic finding in syndromes and in spina bifida patients. Its main clinical association has been with inadvertent intubation of a bronchus, usually the right, during attempted endotracheal intubation. We report an infant with CST and left lung hyperinflation. Inability to image the thoracic trachea and airway led to bronchography, which revealed the carina to be at the thoracic inlet; the distal left mainstem bronchus was extrinsically narrowed. MRI and coronal CT showed the major vessels to be at their usual level. At operation a tight band effect on the left mainstem bronchus was found, due to the overlying ligamentous ductal remnant. Division and ligation of the band relieved the obstruction. This appears to be a rare and possibly new cause of vascular-related airway disease. Similar findings have been noted in the past with post-right pneumonectomy patients as well as in agenesis of the right lung. In these two entities the rotation of the heart into the right chest produced extrinsic compression of the bronchus by the ductal remnant and the adjacent pulmonary artery and aorta.

Poster Presentations

Iatrogenic disorders

Iatrogenic disorders of percutaneous renal biopsy

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More than 300 articles have appeared in the world literature describing techniques, results and complications of percutaneous renal biopsy during the past 40 years, but there were only a few pediatric data among them. Over a period of 6 years ultrasound-guided percutaneous renal biopsies were performed 45 times in 41 children aged 1–16 years. Each patient underwent ultrasonography on the 1st and 3rd day following renal biopsy, and one child had computed tomography. The blood pressure, bleeding time, clotting time and prothrombin time were normal in all cases. Two children had decreased creatinine clearance. Four patients had a total of eight complications, including gross hematuria lasting for more than 12 h (one case), pain lasting for more than 12 h (one case), subcapsular hematomas (three cases), perirenal hematoma (one case) and intrarenal hemorrhage (two cases). We did not find the complication of intra-renal arteriovenous fistulas. Palpable hematoma, infections, and death did not occur. According to the literature the complication rate seemed greater where the creatinine clearance was decreased. When kidney tissue was not obtained, there was a higher incidence of complications, but this was not the case in our group. There was no relation between patient's age, size of biopsy needle and rate of complications. All iatrogenic disorders were improved by conservative treatment. They did not require blood transfusion or invasive treatment. Kidney needle biopsy by ultrasound guidance is a relatively safe procedure. The complications should be detected by early ultrasound examination.

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Conventional radiography vs US, vs CT, and vs MRI

Renal osteodystrophy: evaluation by MRI

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Renal osteodystrophy is an important finding in children with chronic renal failure (CRF). The term renal osteodystrophy is used to include skeletal disorders of patients with CRF: osteitis fibrosa, osteomalacia and/or "rickets," osteosclerosis and the rarer associated extraskeletal calcifications. Current research is attempting to unravel the pathogenetic events and underlying mechanism of uremic osteodystrophy. These include an early occurrence of secondary hyperparathyroidism which has been attributed to phosphate retention, hypocalcemia, the impaired production of 1,25(OH)₂D, the altered "set point" for PTH secretion, the skeletal resistance to PTH and the diminished degradation of PTH. Moreover, aluminum intoxication and metabolic acidosis may have a pathogenetic role in renal osteodystrophy. In the past few decades, the astonishing progress in life prolongation of patients with CRF has greatly enlarged the number of subjects with these metabolic problems and has brought to light severe and novel forms of the disease previously not frequently encountered. The aim of this study was to evaluate the role of magnetic resonance imaging (MRI) in the detection and evaluation of osseous damage in children with CRF. We studied 14 children (11 male, three female; mean age 13 ± 3 years range 2–16 years). Eight out of 14 patients were treated conservatively (GFR 8–40 ml/min per 1.73 m²; mean duration of CRF 6 ± 3 years) and six patients were undergoing hemodialysis (mean duration of HD 7 ± 3 years). In all patients, the femora and the pelvis were examined with 0.5-T MRI, using T1-weighted spin echo (TR 500, TE 25 ms) and inversion recovery (STIR; TR 1200, TE 13, TI 120 ms) sequences in the coronal and/or in the sagittal planes. MRI showed the following patterns of bone involvement: (a) diffuse thickening of cortical bone with various degrees of medullary channel restriction; (b) structural change of bone marrow component, with abnormal predominance

of yellow bone marrow in comparison with reports in the literature for healthy children of the same age; (c) evidence of intramedullary pathological tissue areas ("brown tumors"). We observed a significant correlation between the MRI findings and the severity of CRF. In conclusion, our study suggested that MRI may give valuable information about the progression of bone marrow in children with CRF. Moreover, MRI can be useful in the follow-up of the extent of bone damage.

Asymptomatic deep neurofibromas in neurofibromatosis type 1: correlation of US, CT, MR and pathologic findings

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Purpose: Neurofibromas involving deep visceral and somatic nerves in the course of neurofibromatosis type 1 (NF-1) are reported with a prevalence of 4–11%. Among these, 4–13% are diagnosed as malignant tumors. The aim of our study was to compare the role of various imaging methods in this group of pediatric patients.

Materials and methods: We evaluated 35 children with NF-1 but without symptoms of abdominal and thoracic neurofibromas. US, CT and MR imaging were performed.

Results: We detected chest and abdominal neurofibromas in five patients (four male, one female). In all cases, the radiologic diagnosis of neoplasm was confirmed by surgical and pathologic findings.

Conclusion: The results of our study demonstrate the usefulness of diagnostic work-up in NF-1. Continuous monitoring of the patients with US, CT and MR imaging will decrease the risk of unoperable and malignant neoplasms, improving surgical results.

High-resolution computed tomography and color Doppler sonography in the imaging of children with Lemierre syndrome

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Purpose: Lemierre syndrome is characterized by an acute oropharyngeal infection due to *Fusobacter*, suppurative thrombophlebitis of the internal jugular vein and systemic dissemination. The aim of this study was to evaluate the role of high-resolution computed tomography (HRCT) and color Doppler sonography (CDS) in the diagnosis of Lemierre syndrome in children.

Patients and method: Two adolescent girls aged 15 and 17 years suffering from Lemierre syndrome were studied with CDS and CT in the search for thrombophlebitis of the internal jugular vein and with HRCT for pulmonary abscesses.

Results: In each patient, CDS showed extensive thrombosis of the internal jugular vein which was not clearly demonstrated by CT. HRCT showed smaller peripheral nodules than conventional CT. HRCT yielded good depiction of the multiple cavitated nodules of various sizes typical of this entity.

Conclusions: The early use of CDS and HRCT in the case of sepsis following anaerobic angina allows prompt and non-invasive recognition of Lemierre syndrome, which may result in severe complications and death if the diagnosis is delayed or unsuspected.

Cystic fibrosis: chest radiography or computed tomography?

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Introduction: Cystic fibrosis (CF) is the most common life-shortening inherited disease among caucasians. The upper respiratory tract is involved in most CF patients. The lower respiratory involvement in CF patients accounts for well 90% of morbidity and mortality. A variety of scoring systems have been developed in the attempt to quantitative the severity of clinical and radiographic disease in individual patients.

Purpose: The purpose of this study was to adapt the scoring system described by Bhalla et al. to computed tomography (CT) and compare it with the chest X-ray (CXR) scoring system described by Brasfield.

Materials and methods: Twenty-seven patients (aged from 4 to 34 years) with CF were examined using CT and CXR. All CXR and CR scans were independently scored in a blinded fashion by two radiologists, using the system described by Brasfield (CXR) and a system adapted from Bhalla et al. (CT).

Results: The results were analyzed statistically. There was good correlation between the two observers. The scoring systems showed statistically significant correlation with percentage ratios CT/CT (0.928 and CXR/CXR (0.89). There was also a good correlation between the CT score and the CXR score (0.927).

Conclusions: There is good correlation between the two modalities (CXR and CT); however the radiographic scoring system involves the use of subjective terms. The CT scoring system emphasizes the importance of some changes in CF, such as bronchiectasis, peribronchial thickening and mucus plugging and incorporates the grading of some CF complications: collapse, consolidation and bullous disease.

Chest imaging

Congenital lung lesions: prenatal diagnosis and postnatal follow-up. Can we rely on the postnatal chest X-ray?

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A number of difficult management decisions arise following the prenatal diagnosis of congenital lung lesions. In many cases the lesions are relatively small and are of uncertain clinical significance. We report on the outcome of 21 lung lesions diagnosed on routine prenatal ultrasound scanning. The relative contributions that chest radiography, ultrasound and CT scanning make to both diagnosis and postnatal management will be discussed. In 14 cases, a complex "mass" was noted prenatally containing both cystic and solid echogenic components. One case has not yet reached term. Two pregnancies were terminated; postmortem examination confirmed extensive cystic adenomatoid malformation (CAM) in both. Of the 11 live-born infants all were asymptomatic, apart from mild transient respiratory distress in one. Only 10 were investigated postnatally. The other infant was asymptomatic and discharged without follow-up! Chest radiography was abnormal in nine, but was diagnostic only in two. Although diagnostic in one infant, demonstrating a solitary cyst, ultrasound was generally unhelpful. CT was helpful in the five infants scanned. Three children have undergone surgery, with CAM lung being confirmed in two and a combined CAM/sequestered segment in one. The presumed diagnoses in the other seven included CAM/sequestrations, a bronchogenic cyst and pulmonary arteriovenous malformation. All remain well and asymptomatic at a mean age of 26 months. Four prenatal scans showed solid but small echogenic masses thought to represent sequestrations. Birth is awaited in one. The others were asymptomatic at birth and remain well at a mean age of 18 months. Chest radiography showed mild abnormality in two. Hypoplastic left lung was diagnosed prenatally in one case and confirmed postnatally by chest radiography. The infant is well at 16 months. Three intra-abdominal pulmonary sequestrations were detected prenatally. Ul-

trasound showed small complex masses related to but separate from the left adrenal in all three cases. Surgery was performed in two with confirmation of the diagnosis. The other infant, aged 2 years, remains well. Thus the prognosis for children with prenatally diagnosed lung lesions is generally good. Initial chest radiography was abnormal in all infants who have so far required surgery.

Neonatal *Pneumocystis carinii* pneumonia: radiographic appearance

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Background: *Pneumocystis carinii* pneumonia (PCP) is the most common and serious opportunistic infection in children with AIDS and the other immunodepressive conditions. The disease was also observed in premature debilitated infants.

Purpose: To evaluate the pulmonary appearance preceding PCP in newborns, the chest radiographic patterns during the infection and the sequelae of the neonatal pneumocystosis.

Patients and methods: From 1991 to 1994 at our institution 15 premature neonates with gestational age 32 ± 4 weeks, Apgar score 4.6 ± 2 and body weight 1470 ± 560 g (mean \pm SD), and with respiratory support for longer than 1 week, were treated because of PCP. Parasites were detected in bronchoalveolar lavage fluid by a monoclonal antibody technique (immunohistochemical staining method). We reviewed retrospectively chest radiographs obtained in the first 2 days after birth, in the period of parasite detection and during clinical recovery or the late period of the disease.

Results: Of 11 neonates investigated after birth, eight had radiological symptoms of RDS and three had pneumonia acquired in utero. *Pneumocystis carinii* parasites were detected at 28 ± 14 day of life (mean \pm SD). The chest X-ray pattern observed at the time of parasite detection had started 14 days (median) earlier and showed ground glass opacities in all patients. In 12 of 13 newborns the air bronchograms were obtained and the opacities were seen to occupy the whole lung fields with peripheral lesions, although the changes were more pronounced in perihilar regions in five patients. Two children had additional patchy and linear consolidations in the medial lung regions. Five children had air leakage symptoms: two pneumothorax and five interstitial emphysema. Early after the radiological pneumocystosis symptoms became evident, chest X-ray in five of 13 patients showed more evident infiltrations in the perihilar regions, or lesions only there, from the late one in the developed disorder [occurring 4 days (median) later]. Seven of 14 children died, at 72 (median) days of life: one died of PCP at 20 days of life, and six developed BPD. BPD was also radiologically evident in two survivors. Radiographic features of the next five patients during the next 2–6 weeks of treatment were unchanged and similar to the pattern at the time of parasite detection.

Conclusions: (1) PCP occurs in preterm neonates born with respiratory distress syndrome or pneumonia. (2) Radiographs demonstrate pulmonary compromise in PCP early, compared with evident clinical symptoms and laboratory detection. (3) BPD is the common sequelae (57%) of neonatal PCP and mechanical ventilation. (4) Mortality was 50% in this patient group during the first 4 months of life.

The pulmonary manifestations of vertically acquired HIV

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Purpose: To identify the spectrum of respiratory disease in a large cohort of vertically HIV infected children, the natural history and the response to therapy, compared with the known spectrum of adult HIV-related respiratory disease.

Methods: This was a retrospective analysis of the serial chest radiographs performed on 30 vertically HIV infected children (age range 6 weeks to 12 years, mean 36 months) over a 4-year period at a national referral centre. The radiographs were assessed by two observers and a consensus obtained for the presence of ground glass opacification, lobar/multifocal consolidation, reticulonodular shadowing, peribronchial thickening and bronchial dilatation. The presence of pneumothorax, pneumatoceles or mediastinal lymphadenopathy was documented and serial change assessed. Where possible, findings were correlated with clinical and laboratory data.

Results: Ground glass shadowing was present in 8/30 patients, with a widespread distribution due to PCP in three patients, related to cytomegalovirus pneumonitis following ventilation for *Pneumocystis carinii* pneumonia (PCP) in two patients. Three patients with lobar ground glass shadowing responded to empirical antimicrobial therapy. Consolidation was present in 14/40 patients, the presenting feature of PCP in five, related to pneumococcus in three patients, cryptococcus in one patient and atypical tuberculosis in one further patient. Parainfluenza III was isolated in two patients with multifocal consolidation. Repeated multifocal consolidation was related to aspiration pneumonitis and chronic reflux in one patient and to terminal pulmonary haemorrhage in a patient with lymphoma. Reticulonodular shadowing occurred in 6/30 cases, due to LIP in five patients and following cryptococcal pneumonia in one. Peribronchial thickening/dilatation was seen in 9/30 patients: following repeated infections in five cases, chronic aspiration pneumonitis in one, chronic LIP with traction bronchiectasis in two and PCP in one. Mediastinal/hilar lymphadenopathy was seen in 9/30 patients: associated with repeated infections in four cases, *Mycobacterium avium-intracellulare* in two and due to LIP in three. Cardiomegaly was present in one of two patients with proven clinical cardiomyopathy.

Conclusions: Abnormal chest radiographs were seen in 26/30 symptomatic patients, with LIP occurring in 20% (6/30); only one developed lymphoma. In contrast to adult disease, clinical and radiological response occurred in two patients ventilated for PCP and two patients treated for LIP.

High-resolution CT in children suffering from gastroesophageal reflux and recurrent aspiration

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Recurrent aspiration has been believed to be the most likely cause of pulmonary disease. The aim of the study was to evaluate and compare the usefulness of HRCT, chest radiography and the common diagnostic methods in children with recurrent aspiration. HRCT, chest radiography, barium oesophagogram, intraoesophageal pH probe monitoring, bronchoscopy and pulmonary function testing were done in 26 patients (aged from 7 months to 17 years, mean 8 years) with gastroesophageal reflux. Surgical antireflux procedures (gastropexy, narrowing of the hiatus oesophageus) have been performed in 19 patients so far. All patients have improved with the surgical management, and many are asymptomatic concerning bronchopulmonary disease. HRCT showed bronchiectasis and peribronchial thickening in 13 children, consolidation in 15 children and emphysema in 1 child. Only six patients did not have any pulmonary abnormality. Eleven per cent with normal chest radiographs and 35% with questionable findings on chest radiography had pulmonary abnormalities on HRCT. HRCT is a valuable tool in disclosing and diagnosing pulmonary abnormalities caused by recurrent aspiration, even in younger children without the possibility of breathhold and cooperation.

High-resolution CT in children with pneumonia resistant to therapy

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The diagnosis of invasive *Aspergillus* infection is difficult to establish in patients with seriously impaired host defence mechanisms. The aim of the study was to evaluate HRCT in such children with pneumonia resistant to therapy. Chest radiography and HRCT were performed in 26 children aged from 10 months to 20 years. Nineteen patients had leukaemia, one lymphoma, one cystic fibrosis, one HIV infection (AIDS) and haemophilia, one bone marrow transplantation, two lung infiltrates of unknown cause and one Epstein-Barr virus-associated haemophagocytosis syndrome. We found 10 cases of invasive aspergillosis, four cases of lung abscess (one oesophagobronchial fistula), two interstitial edema, one graft versus host reaction, five alveolar infiltrates and four cases without pathological pulmonary findings. HRCT is a valuable method for early detection of specific infection, especially *Aspergillus* pneumonia, in children with leukaemia and other severe disorders.

Role of conventional radiographic methods in the diagnosis of hydatid cysts

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Echinococcus pulmonum is a relatively frequent tumor of childhood caused by parasitic infection. Macedonia is an endemic area due to poor social and hygienic conditions. The nonspecific symptoms often lead to confusion with pneumonia, tuberculosis or pleural effusion. In this study we investigated 19 patients (age range 5–8 years) hospitalized in 1993 and 1994. Besides laboratory methods, diagnosis is based on plain chest film and conventional tomography, but in some difficult cases ultrasound and CT investigations were performed. Antibodies to antigen blood test was performed in 16 patients, and 14 positive results were confirmed. Twelve patients were examined by ultrasound. The cyst was seen in the lung only when it was localized near the chest wall of diaphragm. CT was performed only three times. Of the 19 patients, 11 were male and eight female. Nine patients had cysts on the right side, eight on the left lung, while three had liver cysts diagnosed by ultrasound. In one case cysts were found on both sides. One child who was hospitalized because of pleural effusion had a subdiaphragmatic liver hydatid. The cyst had ruptured into the right lung. It was confirmed by CT and treated surgically. Based on the results obtained by each method we came to the conclusion that conventional radiographic and laboratory investigations are in many cases sufficient for the correct diagnosis to be made.

Percutaneous balloon dilatation in congenital pulmonary and aortic stenosis

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The purpose of this study is to announce our first results of balloon dilatation of stenotic pulmonary valves, stenotic aortic valves and recoarctation. From May 1993 we performed 27 balloon valvuloplasty procedures and one balloon dilatation in a child with recoarctation of aorta. Twenty-one procedures were performed in infants and children with isolated pulmonary valve stenosis, five in neonates with critical stenosis and two in infants with tetralogy of Fallot. In all patients except one the results were satisfactory: de-

creased right ventricular pressure, decreased gradient between right ventricle and pulmonary artery. Percutaneous balloon valvuloplasty was attempted in four newborns (2000–4000 g) with critical aortic valve stenosis and congestive heart failure. In all of them the procedure was fully effective. Stenosis was relieved as assessed by a significant decrease in transvalvular pressure gradient. Moderate aortic insufficiency after the procedure occurred in one infant (grade II–III regurgitation). All patients are well, but the follow-up period in this group of patients is not very many months. In conclusion, in our opinion percutaneous valvuloplasty is a safe and efficient procedure.

Good clinical and radiological response to treatment in a child with familial primary pulmonary hypertension

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Prognosis is poor in children and adults with primary pulmonary hypertension. Cardiac strain is manifested in clinical findings as well as in X-ray. The degree of pulmonary hypertension can now be accurately measured with Doppler from the tricuspidal regurgitation. A 6-year-old girl was admitted to our hospital in June 1992 due to epistaxis. Microcytic and microangiopathic anemia was detected. The primary chest X-ray revealed very prominent pulmonary arteries, and peripheral vascularity was poor due to pulmonary hypertension. The patient's sister had died at 3 years of age due to right-sided heart failure during infection. At autopsy the cardiac and pulmonary findings were found to be typical of pulmonary hypertension. Despite digoxin, diuretic and anticoagulation therapy and night-time oxygen administration, also at home, the patients' clinical condition deteriorated. She could walk only a few metres and had a poor appetite, and consequently lost weight. The patient was entered on the heart-lung transplantation list. Extensive tests of the blood coagulation system yielded normal results. She failed to respond in the pulmonary vasodilator test (nifedipine), and thus vasodilator treatment was not started. The treatment was further intensified. Hyperalimentation via nasogastric tube was commenced. Gradually the patient improved, as was also evident on X-ray and echocardiography examinations. In July 1993 peripheral vascularity was increased in the chest X-ray control, while the prominence of the pulmonary arteries was decreased. The patient's condition was better, and she is now attending school. In ECG and ultrasound the situation is also better. The situation worsened temporarily during infections. The patient has been removed from the transplantation waiting list. One third of adults with primary pulmonary hypertension show a response to vasodilators, and their prognosis is better than in those who do not respond. In children this disease is rare and the epidemiological knowledge of the disease is scanty. Anticoagulant therapy is always indicated. In conclusion, pronounced pulmonary hypertension, usually irreversible, can decrease during therapy and is detectable on chest X-ray.

Urological imaging

Isolated renal vein injury in childhood

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Purpose: To describe the imaging findings in isolated renal vein injury.

Methods and results: A boy fell from a height of 7 m and presented with blunt abdominal trauma. CT showed a great amount of contrast

material in a large perirenal hematoma with clot formation without massive renal shattering. The proximal part of the right renal artery was identified but the distal part was not visible. On angiography the right renal artery was not visible, although otherwise normal at surgery, because of compression by the huge hematoma. Therefore the diagnosis was not suspected preoperatively. On CT, isolated renal vein injury often causes minimal and delayed nephrographic visualization of the enlarged kidney. The capsule appears thicker than in the case of arterial injury, and there may be a rim, which is usually thicker than in renal artery thrombosis. In some cases, there is a large surrounding hematoma with normal kidney on CT.

Conclusion: The findings on diagnostic imaging of isolated renal vein injury are difficult to assess but may be useful in avoiding nephrectomy. In one case, successful autotransplantation after "bench surgery" was performed.

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DMSA scan in rats with pyelonephritis: comparison between a gamma-camera method and histology

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Purpose: To test a newly developed method for analysing DMSA scintigraphy from kidney examinations, a study was performed where the gamma-camera results could be compared to histology.

Method: DMSA scintigraphy was used to examine 36 rats, using a large filled of view camera equipped with a pin-hole collimator. Twenty-eight of the rats were examined 5 and 28 days after infection to evaluate uptake defects in the infected areas. Eight of the rats were not infected and were used as reference. The images were recorded and stored for later computer analysis. The kidneys were removed and prepared for light microscopy. Comparison was made between scintigraphic findings and histology with regard to inflammatory changes.

Result: The findings from the computer analysis of the DMSA scintigraphic correlated well with histology. The new method is more sensitive to changes in the side distribution of DMSA uptake than the conventional right/left quotient.

Conclusion: The method we use to analyse DMSA scintigraphy from gamma-camera examinations is more sensitive than the conventional right/left distribution. The method also enables us to study regional defects.

Bilateral cystic renal involvement in Turner's syndrome

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Turner's syndrome is one of the most common human chromosomal abnormalities. Renal diseases are frequently recognized in

this syndrome and occur in up to 70 % of patients. Renal unilateral cystic lesions have been previously reported in Turner's syndrome (4/364 or 1.09 % of patients, considering 11 published papers). Urographic features of renal cysts in Turner's syndrome are quite difficult to detect. Recently, high-resolution sonography has replaced urography as the imaging method to screen for renal anomalies, and a high incidence of renal cysts in patients with Turner's syndrome has been reported (16 %). A 22-year-old woman with Turner's syndrome who presented normal urographic findings during childhood and adolescence was recently referred to our Institution for further evaluation because of moderate renal failure. On renal sonography both kidneys were found to be of normal size and presented increased echogenicity with numerous small cysts (maximum size 18 mm). Bilaterally some small nodular echogenic areas without acoustic shadow were also found. To our knowledge this is the first report of bilateral cystic lesions in Turner's syndrome. The finding of echogenic nodular areas is of uncertain significance and has not been previously reported. This feature will be further investigated by CT and MR. Ultrasound examination is now considered the first-line method for detection of renal abnormalities, also in Turner's syndrome patients. The presence of numerous renal cysts in our patient suggests the desirability of earlier echographic evaluation in all pediatric patients with Turner's syndrome.

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Bilateral renal oncocytoma in a child

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The authors report a case of bilateral renal oncocytoma in a 13-year-old child presenting with a long-lasting abdominal tumor. Ultrasound showed a large left renal tumor and a smaller one in the right kidney. CT showed that the left tumor was heterogeneously enhancing and revealed two other lesions. Surgery was performed, and histology showed bilateral renal oncocytoma.

Extrarenal Wilms' tumor

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We report a case of an extrarenal retroperitoneal Wilms' tumor in the right adrenal gland with metastasis. CT, MRI and ultrasonographic appearances of this extrarenal Wilms' tumor were those of a relatively homogeneous and solid mass. We discuss the need to consider this diagnosis when confronted with a retroperitoneal juxtarenal mass and the advantages of performing chemotherapy before surgery, and evaluate CT and ultrasonography in this case. This tumor is very rare. The median age of the patients is 3.6 years. The retroperitoneum is the most common site. It has been postulated that extrarenal Wilms' tumor arises from extrarenal displacement of metanephric or mesonephric remnants.

Ureteric cyst: a variant of ureterocele disproportion

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Background: The so-called ureterocele disproportion [1] is a rare type of double collecting system defined by the association of an ectopic ureterocele with a dysplastic renal upper pole. The upper pole ureter decreases in size upwardly and loses patency in its proximal portion.

Case report: We report a variant of this condition found in a female child evaluated for urinary infection. The upper pole ureter had involuted at its two extremities but remained as a dilated segment (the ureteric cyst) in its mid-portion.

Discussion: The pathophysiology of this anomaly can be compared to that of incomplete allantois obliteration leading to urachal cyst. This case, proven by surgery and pathology, illustrates the complementary nature of ultrasound, voiding cysto-urethrography and intravenous urography in the evaluation of complex double systems. Each of those techniques alone would have missed the diagnosis. In accordance with previous reports [2], surgical excision rather than percutaneous drainage was performed to prevent infection and/or recurrence.

Conclusion: Although it is rare, ureteric cyst should be considered in the differential diagnosis of a lateral cystic retroperitoneal mass in a child.

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Prolapsus mucosae ureteris

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Introduction: Prolapse of the ureter's mucous membrane is a very rare anatomical condition. Prolapse is simply a protrusion of the ureter into the bladder. In contrast to ureterocele, which is covered by bladder mucosa, the ureteral prolapse is covered by ureteral mucosa.

Case report: A 10-year-old boy was admitted with dorsal pain in the right kidney region and macroscopical haematuria. Ultrasonography and CT revealed no anomaly of the upper urinary tract. In the bladder, on the right side, an ureterocele-like formation was seen by both procedures. Intravenous urography showed a filling defect in the bladder, and some contrast filling was seen inside this mass. Cystography showed a filling defect in the bladder. There was no reflux. By cystoscopy a polyp-like mass was identified in the ostium of the right ureter with a necrotic surface. These examinations suggested a right "atypical ureterocele" without dilatation of the upper urinary tract. At surgery the intracystic mass was removed. Histology showed a prolapse of the right ureter's mucous membrane.

Discussion: This anomaly may exist for a long time without any complaint, but may be the source of any infection and haemorrhage. In spite of several different diagnostic procedures the diagnosis and differential diagnosis is difficult. While ureterocele causes dilatation in the upper urinary tract, there is no dilatation in the case of prolapse because its ostium is not narrowed.

Conclusion: If we see what looks like an atypical ureterocele, we should think of this anomaly.

Double cloacal malformation in xiphoomphaloischio pagus tripus

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We present a rare case of xiphoomphaloischio pagus twins, each with a cloacal malformation and a large obstructed vagina. The twins, initially diagnosed prenatally by ultrasound, were further imaged after delivery as part of their preoperative evaluation for separation. Initial evaluation by MRI demonstrated two kidneys in each twin; however, on nuclear renal scan only one functioning kidney and a single ureter to a common bladder was seen in each twin. A second bladder without ureteral openings was found during endoscopy. In addition, the twins shared a common atretic distal colon that communicated with the two cloacal malformations on contrast studies. We emphasize the value of contrast studies with fluoroscopy over other imaging modalities in the evaluation of this complex deformity.

Testicular tumors in patients with adrenogenital syndrome: sonography versus MRI

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Uni- or bilateral benign testicular tumors are known in patients with adrenogenital syndrome. Differentiation from testicular malignancies by ultrasonography is sometimes difficult. The aim of the present study was to compare the ultrasonographic appearance of these lesions with the presentation on MRI. Twenty patients with adrenogenital syndrome between 1 and 24 years of age underwent high-resolution scrotal ultrasound. Six patients with testicular lesions detected by ultrasound were examined by MRI. Ultrasonographically, the lesions were hypoechoic with well-defined margins. Distribution was solitary or multifocal, uni- or bilateral. On MRI the lesions appeared hypointense in the otherwise hyperintense normal testicular tissue. Further criteria for differentiation from testicular malignancies could not be obtained by MRI. Therefore ultrasonographic monitoring is the method of choice for detection of testicular lesions in patients with adrenogenital syndrome, and for follow-up examination to document decrease of tumor size during hormonal therapy.

Musculoskeletal imaging

Brachydactyly type A 4 in four families: association with microcephaly and other malformations

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In brachydactyly A 4 the middle phalanges of the second and fifth fingers are short and the middle phalanx of the lateral toes are missing. Type A 4 brachydactyly is exceedingly rare and the four families that have been reported suggest autosomal dominant transmission. We report on four families with A 4 brachydactyly. Previously unreported findings include: microcephaly, patent ductus arteriosus, hallux valgus, duodenal atresia, restricted elbow movement and syndactyly of toes. Longitudinal follow-up suggested that hypoplasia or absence of the diaphyseal ossification centers is the primary phenomenon. Epiphyses, although somewhat delayed, were generally normal. Our observations suggest that absence of the middle phalanges of the toes may be caused by fusion.

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Ultrasound assessment of the effectiveness of splints in developmental delay of the infant hip

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The management of developmental delay of the hip (DDH) varies widely, with some clinicians relying on clinical examination alone and others using ultrasound assessment to guide management. There is uncertainty regarding the optimum age to perform ultrasound examination and apply splintage, the duration of splintage necessary and the best type of splint to use. We reviewed the ultrasound reports of 339 children referred for clinical instability of the hip, and the examinations and records of all children who were reported as having dysplasia of Graf type II C or worse, to assess the effect of splintage. All the ultrasound examinations were performed by radiologists. Four treatment groups were identified: those who received no treatment, and those who were treated within the first 6 weeks of life with the Von Rosen splint, the Craig splint or the Pavlic harness. A normal outcome was defined as Graf type I by the age of 12 weeks, abnormal as Graf type II or III, and dislocated as Graf Type IV.

The results were as follows (number of hips):

	No splintage	Von Rosen	Craig	Pavlic
Normal	11	12	11	11
Abnormal	11	2	8	11
Dislocated	1	0	0	2
	23	14	19	24

Use of the Von Rosen splint results in fewer abnormal hips than use of the Pavlic harness ($P < 0.05$) or no treatment ($P < 0.05$). Use of the Craig splint and the Pavlic harness showed no statistical benefit compared with no treatment. These early results suggest that ultrasound-guided management with early splintage of dysplasia of type II C or worse, using the Von Rosen splint, is the most effective treatment for DDH. A prospective trial continues.

Cystic tuberculosis of bone: mimicking radiographic appearance of Langerhans cell histiocytosis

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The incidence of tuberculosis of bones and joints has decreased markedly in the past three decades. Cystic tuberculosis is a rare variety of tuberculosis associated with disseminated lesions of the axial and the extra-axial skeleton. Multiple cystic bone lesions in tuberculosis are much more commonly encountered in children than in adults. The purpose of this report is to present a patient with cystic tuberculosis involving multiple bones and to discuss the differential diagnosis briefly. An 8-year-old girl complained of pain and swelling of the left knee. Initial radiography of the left knee showed a well-defined small osteolytic lesion at the distal metaphysis of the left femur. Then she felt masses in the skull and right clavicle. As the masses had increased in size, biopsy was carried out. Langerhans cell granuloma was diagnosed pathologically,

suggesting either tuberculosis or Langerhans cell histiocytosis. The patient was then referred to our hospital. Radiographs at the time of admission demonstrated multiple osteolytic lesions involving the skull, right clavicle, right scapula, ribs, left iliac bone, distal metaphysis of the left femur, and epiphysis of the left tibia with progression. No surrounding osteosclerosis could be identified. These findings appeared to represent Langerhans cell histiocytosis. No lesions could be seen in the spine. A chest radiograph was clear. A second biopsy from the right clavicle finally proved the diagnosis of *Mycobacterium tuberculosis*. The patient had had BCG vaccination 3 years previously. She had no family history of tuberculosis. Following antituberculous therapy, the multiple osteolytic lesions improved slowly. A number of diseases may demonstrate multiple osteolytic lesions. In acid-fast bacilli infection of bone, *Mycobacterium tuberculosis*, *Mycobacterium bovis* (BCG vaccination-induced infection), or atypical mycobacteria may show a radiographic appearance similar to that in this case. Langerhans cell histiocytosis, metastasis from neuroblastoma, leukemia, lymphoma, and osteomyelitis are the most common causes of multiple osteolytic lesions in infants and children. Sarcoidosis, generalized fibromatosis, and lymphangioma, which are rare, also may cause multiple osteolytic lesions. Metastases from neuroblastoma and leukemia mainly involve the metaphysis, with somewhat irregular margins. Langerhans cell histiocytosis usually occurs, in the shafts of the long bones, surrounded by a sclerotic margin. Although the diagnosis of tuberculosis may be difficult, as in this case, tuberculosis should be included in the differential diagnosis. The early diagnosis of tuberculosis is important, as prompt and adequate treatment will preclude excessive damage to bones and joints.

Effect of US-guided fluid aspiration in the Ilizarov limb lengthening procedure

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Background: The Ilizarov limb lengthening procedure can be followed by means of X-ray and US. The fluid collection in the distraction gap can be detected by US.

Case reports: Comparison of two cases illustrates the therapeutic effect of US-guided fluid aspiration. *Case 1:* The 6-cm lengthening of the left femur of a 13-year-old girl was necessitated by a trauma. In the distraction gap, an accumulation of fluid 13 mm in diameter was detected. This was not aspirated. X-ray examination revealed a lack of the ossification process. The Ilizarov apparatus could be removed only after 9 months. The ossification process was complete after 12 months. *Case 2:* The 4-cm lengthening of the right tibia and fibula of a 10-year-old girl was performed in correct a congenital shortening. In the distraction gap, an accumulation of fluid 15 mm in diameter was detected. This was aspirated under US guidance. After 1 week, there was no longer any sign of fluid. The Ilizarov apparatus was removed after 14 weeks, and the ossification process was complete after 6 months.

Discussion: US examinations permit determination of the corticotomy gap length, assessment of the development of new bone, and detection of certain complications. US-guided fluid aspiration can reduce the time of the procedure and promote the completion of the ossification process. In order to co-ordinate the rate of distraction and new bone formation, it is necessary to use X-ray and US examinations together. Early detection and therapy facilitate the avoidance of a prolonged ossification process.

Pathologic fractures in children

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Purpose: The purpose of this exhibit is to display illustrative cases of the wide spectrum of pathologic fractures in children.

Methods: Representative examples of pediatric pathologic fractures are selected from clinical material encountered at two affiliated institutions over the past 5 years. The pictorial essay is organized by large categories and includes key clinical and imaging features.

Results: In children the presence of unusual fractures raises the question of underlying congenital or acquired conditions. Osteogenesis imperfecta, congenital pseudarthrosis of the tibia, and osteopetrosis represent the first group. Pre-existing bone conditions may be tumor-like diseases or benign bone tumors (bone cyst, aneurysmal bone cyst, non-ossifying fibroma, enchondroma) or malignant bone neoplasms (Ewing's, osteosarcoma, metastatic neuroblastoma). The process of growth is at the basis of the third category, including apophysitis, stress fractures, fractures due to metabolic disease (rickets, storage disease) or to endocrine disorders like hyperparathyroidism, osteoporosis related to hypercorticism (often iatrogenic). The fourth group combines fractures secondary to neurologic or neuromuscular disease and concerns primarily the limbs. The last category relates to circumstances following orthopedic or oncologic treatment.

Conclusion: Children may have congenital as well as acquired bone weakness. Fractures may be the initial symptom of an underlying disease. Some entities bear some site specificity of the fractures (osteoporosis, fibrous dysplasia). Knowledge of the primary underlying process may lead to adjustment of the management plan and may alter the ultimate prognosis. The most frequently encountered problem in differential diagnosis is the issue of child abuse. Plain radiography is still the first modality used, before technetium scan, CT and quantitative bone densitometry.

MRI appearance of epiphysis, growth cartilage and metaphysis in osteochondrodysplasias

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Purpose: To assess the magnetic resonance imaging (MRI) features of epiphysis, growth plate and metaphysis in patients with osteochondrodysplasias.

Material and methods: Eleven patients aged from 5 to 17 (mean 9) years were selected for the study: four patients affected by achondroplasia, four affected by hypochondroplasia and three affected by pseudoachondroplasia. On X-ray all patients had epiphyseal, growth plate and metaphyseal abnormalities in relationship with osteochondrodysplasias. All patients were studied with MRI (0.5 T). T1-weighted (500/20) and T2-weighted (1800/25–80) spin echo sequences were obtained on knees in the sagittal or coronal plane. Slice thickness was 5 mm of 10 mm. Morphology and signal intensity of epiphysis, growth plate and metaphysis were evaluated on MRI.

Results: MRI clearly detected epiphyseal, growth plate and metaphyseal alterations due to defects of growth, especially for cartilaginous structures. Signal intensities of epiphysis, cartilaginous epiphysis and growth plate and metaphysis were normal.

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Bladder exstrophy in the maturing skeleton: radiographic appearance in 44 patients

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Exstrophy of the bladder is an uncommon congenital abnormality of the urinary tract occurring in approximately 1 in 10,000–50,000 live births. The bladder abnormality may be part of a spectrum of visceral malformations that have been well documented and which include epispadias and cloacal exstrophy. Widening of the pubic symphysis is invariably present and is the hallmark plain radiographic feature. Little information can be found in the literature concerning other bone and joint abnormalities that may be present in these patients. We have reviewed the pelvic radiographs of 44 patients with bladder exstrophy, 35 male and nine female (current age range 25–48 years). Premature degenerative changes in the hip joint were seen in 27 cases (61%). In addition, other bony abnormalities were seen in 29 patients (66%). These included transitional vertebrae, spina bifida occulta and unilateral or bilateral crescentic defects in the medial iliac crest which may be related to previous surgical procedures. The latter abnormality has not, to our knowledge, been previously described. The bone and joint abnormalities in bladder exstrophy are described. We discuss the clinical relevance of these findings in addition to possible mechanisms that may account for their occurrence.

Pseudoachondroplasia: conventional radiology versus MRI

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Clinical data: A boy aged 5 years, 7 months with short-limbed dwarfism, extreme hyperlordosis of the lumbar spine and valgus deformity of the legs. Delivery uneventful, birth weight 2900 g, body length 52 cm, head circumference 34 cm. Growth rate within the 1st year of life normal. Decreasing growth rate and development of disproportional dwarfism in the 2nd year of life.

Radiologic findings: Shortness of all tubular bones. Platyspondyly. Mushroomed metaphyseal irregularities in the long tubular bones. Short tubular bones with crown-like deformities. Biconcave platyspondyly of the lumbar vertebra bodies. Acetabular roofs irregular confined, femoral heads small for age, femoral necks very short.

MRI findings: Cartilage layer of the distal femur and proximal tibia epiphyses very thick. Patella completely cartilaginous. Lumbar vertebra bodies with extremely thick cartilaginous epiphyses.

Conclusion: In pseudoachondroplasia, MRI provides important additional information about the cartilage structure of the growth plates in the spine and in the long bones. MRI findings demonstrate clearly that the platyspondyly is only simulated because of the invisibility of the cartilage in the conventional X-ray films. Delay of ossification, as seen from the epiphyses of long bones, contrasts with the evidence of cartilage hypertrophy as proven by MRI.

The ball and socket ankle joint deformity presentation of a new case and review of literature

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Introduction: The ankle is a hinge joint. We want to draw attention to a type of ankle with a spherical head of the talus which lies in a socket-formed distal end of the tibia and the fibula. This condition first described in 1958 [1], is named ball-and-socket ankle (BSA).

Presentation of case: A 15-year-old male patient presented with shortage of the right lower limb (5.5 cm, 5.6%; upper leg 3%, low-

er leg 10%) for surgical correction. In addition there was a missing fifth ray at the right foot. Range of motion was the same for both feet at physical examination. Radiographs first showed a BSA on the right side. There was a fusion of talus, navicular, and calcaneum into one bone, and fusion of the second and third cuneiform. There were no further deformities. The mother denied use of drugs during pregnancy, and no similar case with absent toe or shortage of a leg is known in the family.

Review of literature: Since the first description, 206 instances of the BSA have been reported in 175 patients. In 80% of cases the deformity is single-sided with a preference for the right side (right : left = 2 : 1, $P < 0.001$). Male sex predominates (male : female = 2 : 1). In only one case was familial occurrence supposed. In common, symptoms related to the ankle are rare; only 2% of the patients presented with an instability of the ankle and 3% with pain. Few degenerative changes occur in the BSA. Mean age at diagnosis was 10 years (range 2–50 years). In most cases associated deformities were the chief complaint.

Associated abnormalities:

- Eighty-eight percent shortage of the leg at the same side, the tibia is more affected than the femur. In 80% of cases of femoral shortage the lateral part of the femur is more affected than the medial, resulting in a slight valgus alignment of the knee.
- Hypoplasia of the fibula or elevation of the lateral malleolus is common.
- Deformities of the foot: 11 club feet, 14 congenital flat feet, nine metatarsus adductus varus (4 of 5 patients bilateral).
- Eighty percent tarsal synostoses, thereof 72% talo-calcaneal, sometimes including the navicular and the cuboid, 18% talo-navicular without inclusion of the os calcis. Cases without synostoses include some deformities of the subtalar joint with reduced mobility. Often there are fusions between cuneiforms or bases of metatarsals.
- Fifty-five percent oligodactyly; in most cases the fifth ray is missing, never the first ray. Aplasia of the second or the third ray is often combined with aplasia of the second or third cuneiform. Hypoplasia or fusion of two rays is rarely found.

Conclusion: Our patient was quite a typical case of BSA, as he showed unilateral appearance, shortage of the leg, missing lateral ray, a large fusion in the hindfoot and fusion of two cuneiforms. Callus distraction was performed to correct the length discrepancy, and an instability in the ankle was treated with suitable shoes.

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Neurological imaging

Persistent ventriculus terminalis in the newborn: sonographic findings

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Spinal ultrasonography of 200 newborns revealed a small ovaloid lesion of variable size (length 10 mm, diameter 2–5 mm) between the conus medullaris and the cauda equina in 20 patients. Spontaneous regression of the cyst could be observed in follow-up examinations during the first weeks of life. We suppose that this cyst corresponds to the ventriculus terminalis, the most distal dilated portion of the central lumen of the spinal cord. During embryogenesis the ectoderm forms the brain and the spinal cord by developing the neural plate, the neural folds and the neural tube. The most caudal portion of the spinal cord evolves from a caudal cell mass by canalisation and retrogressive differentiation. Multiple microcysts and clumps of cells appear in the cell mass, devel-

oping an ependyma-lined tubular structure. This most caudal segment forms the conus medullaris, the filum terminale and a focal dilatation of the central canal, the ventriculus terminalis. From the 38th day of gestation the cell mass and the central lumen decrease in size. We assume that the regression of the ventriculus terminalis takes a varying period of time and that it may therefore still be visible during the first weeks of life, seen most easily by spinal ultrasonography.

Brain ultrasound for prediction of outcome at 5 years in extremely low birth weight infants (ELBWI)

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Improved prenatal and neonatal care have contributed to the increased survival of ELBWI. These tiny infants, however, have an increased risk of sequelae such as cerebral palsy, retinopathy, school difficulties and chronic lung disease. These sequelae are of great concern both to the parents and to society. The purpose of our study was to analyse the neonatal risk factors for these problems in ELBW (<1000 g) infants. During the period 1982–1987, 96 (56%) ELBWI survived at the neonatal intensive care unit (NICU) of the Children's Hospital, University of Helsinki. At 5 years of age all children underwent neuropsychological testing and a neurological examination. The data from the neonatal period were collected retrospectively. They included the social background of the family, duration of the pregnancy, birthweight, and several parameters from the first day of life and from the NICU treatment. The clinical risk index for babies (CRIB) score was calculated from these data for all infants. Cranial ultrasound was performed several times on all infants. Of the infants, 11% had neurological abnormalities, 16% had retinopathy, 19% had emotional disturbances, 17% had delayed verbal development, 32% had visuo-motor abnormalities, and 20% had slight to severe intellectual impairment. In 53% of the infants, all these parameters were normal. Intellectual impairment was associated with severe intra-ventricular haemorrhage (IVH), low gestational age, low birthweight, and long respirator care. Logistic regression analysis indicated that third grade IVH and sepsis were the most important of these. Other outcome variables showed similar associations. However, neurological abnormalities were associated only with severe, fourth grade IVH.

Schizencephaly: clinical and radiological evaluation in 15 patients

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Background: Schizencephaly is a developmental disorder of cell migration, characterized by cortical surface clefts which may be uni- or bilateral and are most commonly located near the pre- and postcentral gyri.

Material and methods: We studied the main clinical and radiological features of 15 patients with schizencephaly between 1982 and 1994. Eight of the affected patients were female and seven male. The presenting symptoms were nonspecific, including seizures, hemiparesis, tetraparesis and developmental delay. Seizures occurred in six patients. No infantile spasms or neonatal convulsions were observed. Seizures began between 1 and 12 years: these six patients had motor abnormalities and retardation. All six had partial and generalized seizures with predominant EEG epileptic activity localized on the side of the lesion: unilateral (3) and bilateral

(3). All patients (15/15) had computed tomography (CT), and 11 also had magnetic resonance imaging (MRI).

Results and discussion: CT and MRI revealed evidence of schizencephaly in all cases. The cleft is usually oriented vertically from the midsylvian fissure area: unilateral (8), right (5), left (3), bilateral asymmetric (7). Five patients with epilepsy had diffuse heterotopia. Schizencephaly is an extreme variant of cortical dysplasia. The clefts are bordered by dysplastic gray matter. The clinical outcome of patients with schizencephaly is very variable. Mental retardation may be mild or severe. Seizures occur in only 40 % of patients. Motor dysfunction correlates with the locations of the clefts.

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Cerebral MR findings in Williams syndrome (WS)

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WS is an uncommon disease characterized by facial dysmorphism, aortic stenosis, hypercalcemia and renal, dental and osseous abnormalities. The patients present mental retardation with poor visuospatial and visuomotor capacities and relatively spared linguistic abilities. It is well known that patients with WS and those with Down syndrome (DS) may share some neurological abnormalities. The aims of this study were: (1) to evaluate the anatomical aspects of WS by MR; (2) to compare features observed in WS to those of patients with DS. Ten patients with WS and 10 with DS were examined: age range was 9 months to 14 years in both groups. All patients with WS were found to have a poor cerebral development corresponding to microcrania, whereas the cerebellum was normal. By contrast, those with DS showed both structures equally affected. Other findings in our patients are summarized below:

	WS	DS
Frontal cortex	Normal	Hypoplastic
Limbic system	Normal	Asymmetrically reduced
Corpus callosum	Elongated	Rounded

In conclusion, MR clearly shows the characteristic findings of WS, so it is helpful in diagnosing this disease. Furthermore, it is very useful in differentiating WS from the other syndromes presenting mental retardation.

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An unusual arteriovenous malformation of the spinal cord in a young child

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Vascular malformations of the spinal cord are considered uncommon lesions. In children symptoms start prior to the age of 16 in 50–60 % of cases, but the correct diagnosis is made in only 20 %. We present the case of a 10.5-month-old boy admitted with a sudden onset of hypotonia and hyporeflexia of the lower limbs. Lumbar puncture was performed and showed an elevated protein level and a high white and red blood cell count in the CSF. MRI of the spinal cord demonstrated the presence of a medullopomyelia, a hemorrhagic lesion located in the conus and tortuous vessels posterior to the spinal cord. The diagnosis of arteriovenous fistula was proposed and confirmed by phase-contrast MRA. These findings were in agreement with the selective spinal angiogram: a direct arteriovenous fistula fed by the anterior spinal artery was visualized. At a distance from the fistula, the draining vein carried an aneurysm complicated by a pseudoaneurysm. The lesion was treated by surgical ligation of the supplying artery at the level of the fistula. MRI, MRA and angiography were repeated after surgery and demonstrated that treatment had been successful. Spinal cord arteriovenous fistulas are high-flow lesions located in the subpial space. The presentation of these lesions is most frequent in children and characterized by chronic venous congestion and progressive neurologic deficit. Prognosis is poor without treatment. The presence of a ruptured aneurysm on the venous outflow site causing intramedullary hemorrhage is a rare feature in this pathology. MRI is the imaging technique to be used first whenever a spinal cord arteriovenous malformation is suspected. Angiography depicts the exact vascular components and determines surgical or interventional therapy.

Gastrointestinal imaging

Plummer-Vinson syndrome in children

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We report the case of two children (14 and 10 years old) suffering from sideropenic dysphagia. Radiological investigations revealed thoracic esophageal stenosis, confirmed by endoscopy. After iron replacement therapy both children completely recovered. The esophageal stricture disappeared. We will discuss the etiology, pathophysiology and treatment, in children of this rare syndrome which is well known in adults under the name of Plummer-Vinson syndrome.

Jejunal duplication cyst displaying five-layer appearance of the wall: a preoperative US diagnosis

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Duplications of the GI tract are congenital anomalies that can occur in any segment of the tract. A double-layered wall appearance of echogenic mucosa and anechoic muscularis has been described to be pathognomonic of an alimentary duplication [1]. The author reports on a case of preoperatively diagnosed jejunal duplication cyst in a 7-year-old girl. Ultrasonography revealed a cystic mass which was noted to change in shape during the course of examination. Using a high-resolution (7.5 MHz) probe, the characteristic five-layer structure of the GI tract was revealed in most parts of

the wall; to my knowledge an observation not previously reported. One should recognize the alimentary duplications as a rare but curable cause of abdominal symptoms on the basis of their typical, either double or, probably more characteristic, five-layer wall appearance.

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Stricture of the ascending colon in a child with cystic fibrosis treated with conventional-dose pancreatic enzymes

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In recent reports a possible association between strictures of the ascending colon in cystic fibrosis and high-dose pancreatic enzyme treatment was suggested [1, 2, 3]. We describe an infant with cystic fibrosis who presented with intestinal obstruction and a right lower quadrant mass. The sonographic and contrast enema findings were similar to those described previously. We also describe the CT findings. The patient was treated conservatively and feeding was resumed after 4 days. In contrast to previous cases described, our patient was always treated with conventional doses of pancreatic enzymes. This case further demonstrates that a cause and effect relationship still remains to be established before withdrawing what has so far proven to be a beneficial treatment.

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Ileocolic intussusception: another cause of inversion of the superior mesenteric vessels

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Inversion of superior mesenteric (SM) vessels has been reported in malrotation, intraabdominal masses and, rarely, in normal children. There is no reported association of inversion of the SM vessels and intussusception. We report two infants (8 and 10 months old) with distal ileocolic intussusception and abnormal relationship of the two mesenteric vessels. Since the sonographic findings were atypical for intussusception, this sign was considered indicative of midgut malrotation with volvulus. At surgery a distal ileocolic intussusception was found (the intussusceptum was advanced into the sigmoid colon) and reduced. No midgut malrotation was found. Follow-up sonographic examination 4 months after surgery demonstrated a normal anatomic relationship of the SM vessels in both cases. It is concluded that an abnormal mesenteric vascular arrangement can arise in – among other circumstances – cases of distal ileocolic intussusception.

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MRI of the pelvis in the imperforate anus complex

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Fifteen infants (nine male, six female) with imperforate anus had pelvic MRI examinations over a 14-month period. The age range was from 1 day to 9 months, with the mode being 2 days. No sedation was used in 12 neonates. Nine newborns were imaged preoperatively in the first 3 days of life. Oral sedation was used in two older infants, and general anaesthesia in one. The following protocol was used: sagittal, coronal, axial T1 sequences with sagittal T2 (FSE) through the pelvis, employing 4-mm slice thickness and 1-mm intersection gap. Normal spinal cord and conus were visible in 13 patients and not adequately visualised in two studies. Three hydronephrotic kidneys were seen in two infants and unilateral renal agenesis in one. One hypoplastic undescended testis was not evident on MRI, though later found at surgery. Three known rectourethral fistulas were not visible on MRI. Only four sacral segments were evident in two patients. Rectosigmoid meconium was of high T1 and T2 signal in all nine neonates examined in the first 3 days of life, but after this age rectal contents were generally isointense to muscle on all pulse sequences. “High” rectal anomalies, with the rectum ending above the levator sling, were found in four patients, “intermediate” lesions in two and “low” anomalies in nine patients. Pelvic MRI can reliably assess most of the anomalies associated with the imperforate anus complex but may have low sensitivity to recto-urethral fistulas in newborns. MRI also provides valuable anatomic information in babies with imperforate anus, can help predict the likelihood of future continence and occasionally alters surgical management.

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Spontaneous resolution of ceftriaxone-associated biliary pseudolithiasis

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Ceftriaxone, a third-generation cephalosporin, can induce biliary pseudolithiasis which mimics true cholelithiasis both clinically and radiologically. This complication resolves spontaneously after discontinuation of the drug. We report a case of an 8-year-old boy who developed gallbladder pseudolithiasis and wall thickening 4 days after the initiation of ceftriaxone therapy. Serial ultrasound examinations and a plain X-ray of the abdomen verified the association of ceftriaxone intake with the development of increasing amount of pseudolithiasis (increasing calcium precipitation) and the gradual disappearance of lithiasis after discontinuation of ceftriaxone.

Imaging of tyrosinemia

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Tyrosinemia type I is a rare hereditary metabolic disease caused by a deficiency of fumarylacetoacetase (FAH), the last enzyme in ty-

osine degradation. The disease manifests as progressive liver damage and renal tubular dysfunction. Hepatocellular carcinoma may occur in the chronic form. Hepatic imaging is essential to assess and follow the progress of liver disease and to detect malignant changes at the earliest stage. We present the spectrum of ultrasound and CT hepatic changes in 5 patients with tyrosinemia type I.

Imaging of cystic mesenchymal hamartomas of the liver: review of 11 patients

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To evaluate the impact of the available imaging modalities, we retrospectively reviewed the files of 11 patients with the final diagnosis of cystic mesenchymal hamartoma of the liver (CMHL). They were collected over a 13-year period extending from April 1981 to December 1994. There were three girls and eight boys. Three times, antenatal US disclosed the hepatic cystic mass. In the other eight patients an abdominal mass led to the postnatal diagnostic work-up. Mean age at diagnosis in these patients was 15 months (SD 17 months, range 7 days to 57 months). Final diagnosis was supported eight times by surgical and pathological findings. In three patients, diagnosis relied only on clinical findings and imaging modalities. In two of these patients US survey showed disappearance of the mass respectively at 12 months and 3 years 10 months. The last patient was lost to follow-up. US was performed in every patient, CT in seven, angiography in six and MR in one. The hepatic origin of the mass was ascertained by US and CT. The mean CHML diameter was 8 cm (SD 6 cm, range 2.5–25 cm). In nine patients the mass was formed by the juxtaposition of fluid-filled micro- and macrocysts with septations. In two patients, there was a macrocystic unilocular lesion. In two patients there were several foci of CHML. No calcifications were found. Hepatic angiogram showed one or several avascular masses, with enhancement of the compressed surrounding normal hepatic parenchyma. No portal or hepatic vein thrombosis was found. CMHL is a rare condition. In the literature, biliary cyst, biliary cystadenoma and hepatic lymphangioma are other names for CMHL, which seems to be more a tissular dysplasia than a true tumoral condition. Differential diagnosis should include cavernomatous angioma, hepatoblastoma or hepatic mesenchymal sarcoma with central necrosis and nodular focal hyperplasia. Regarding evolution, in our patients two kinds of CMHL seemed to exist: on one hand, large CMHL revealed in young infants by abdominal mass which led to surgery, and on the other hand, small CMHL disclosed on prenatal or neonatal US which may regress. In this selected population, US follow-up may be an alternative to surgery. US is the most helpful imaging modality. It shows the lesion and strongly suggests the diagnosis of CMHL from the macrocystic pattern. However, the microcystic component may be falsely regarded as solid because of the high number of interfaces. CT and MR help in depicting the microcystic component of the CMHL and in evaluating the total extent of the lesion; they support US in the evaluation of the vascular relationships of the mass, which determine resectability. Angiography may still be useful in the case of a huge tumor closely related to the hepatic vein confluence or to the portal bifurcation.

A new sign on the plain roentgenogram of Williams syndrome: supernumerary ribs and supernumerary vertebrae

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Supernumerary ribs were seen in 15 (55.6%) of 27 cases of Williams syndrome (WS). Supernumerary vertebrae were also noted in seven

(50%) of 14 plain supine films available. Most of the pairs of 13th ribs were symmetrical and long compared with the 12th ribs. In 12 cases, the tip of the 13th rib reached the mid-clavicular line. In the other three cases, the extra ribs were short and asymmetrical. Of seven patients with 13 pairs of ribs, five had five lumbar vertebrae and two had six lumbar vertebrae. Of seven with 12 pairs of ribs, two had six lumbar vertebrae, with no relation to age, gender or other combined anomalies. The supernumerary rib from the cervical vertebra is seen in many normal individuals and in some cases of Turner syndrome. Chromosome 8 trisomy syndrome and incontinentia pigmenti are also listed [1] as featuring "supernumerary ribs" but WS is not. It is well known that congenital cardiovascular anomalies are frequently combined with vertebral or rib anomalies. During the past year we have paid special attention to the number of ribs in ordinary chest films. In total, 7087 films were read and 13 pairs of ribs were found in five cases of tetralogy of Fallot, two cases of asplenia syndrome, two cases of pulmonary arterial atresia with ventricular septal defect, and one case of ventricular septal defect, dextrocardia, DeLange syndrome and 4p-syndrome. In most cases, however, the 13th ribs were asymmetrical or very short. The incidence of 13 pairs of ribs in these diseases was not as high as in WS. The first ribs of WS are normal in size and shape, and the extra ribs arise from the 13th thoracic or the first lumbar vertebral body. WS has been identified by hemizygosity at the elastin locus; 7q 11, 23 [2], but the mechanism of the supernumerary ribs is still unknown. To the author's knowledge, these findings have not been described in the standard textbooks and can be considered as a new, simple and effective sign of WS.

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Cushing syndrome, bilateral adrenal hyperplasia and hemangiomatosis of the liver in a newborn

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Cushing syndrome is uncommon in neonates. A 3-week-old baby girl was investigated by ultrasound because of palpable kidneys. Extensive, bilateral adrenal masses were found, 4–5 cm in diameter, with mostly solid, partly mixed echostucture. The hormonal tests, with cushingoid features and clitoral hypertrophy, showed moderate cortisol and excessive androgen production, not suppressible by dexamethasone. At 4 months of age bilateral adrenalectomy was performed. Unexpectedly, multiple, small focal liver masses were found and biopsied. A multicenter study of histological samples showed bilateral adrenal hyperplasia and liver hemangiomatosis. Since operation, continuous hormone substitution has been administered, and the cushingoid features and clitoral hypertrophy have disappeared. During the follow-up, ultrasound and CT scan showed a moderate increase in number of the liver lesions. The diagnostic and therapy of this extremely rare combination, hormonal active adrenal hyperplasia and multiple liver hemangiomatosis, will be discussed.

Sonography of salivary glands in children

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The aim of the study was to evaluate the usefulness of ultrasonography in parotid and submandibular pathology in children. During 1993 and 1994, 60 children underwent ultrasound examination of parotid and submandibular glands, 45 because of symptoms of major salivary disease and 1 with other pathology of the neck. Salivary glands were evaluated using a Sonoline SL-2 device with a 5-MHz linear array transducer. The age of the children ranged from 1 day to 16 years. Results of ultrasound examination in a group of 45 children with salivary disorders are presented. In 15 of them sialography was also performed. The results of diagnostic procedures are compared.

The final diagnoses were as follows:

Acute inflammation	10
Chronic inflammation	15
Sjogren syndrome	2
Sialolithiasis	7
Tumors	11

Our results indicate that sonography is a useful procedure for diagnosis of salivary gland disorders.

Atresia of the femoral vein and spinal cord malformation: report of a case

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Purpose: Report of a case of Klippel-Trenaunay syndrome associated with a spinal cord arteriovenous malformation.

Methods and results: A 19-month-old girl had an enlarged right lower limb with cutaneous angioma. Phlebography revealed an atresia of the right superficial femoral vein. Surgical banding of the ipsilateral femoral artery was performed to decrease the length of the limb, but without success. Seven years later, the child presented with severe back pain. Diagnostic angiography of the spine revealed an intramedullary vascular malformation. Post-mortem examination demonstrated a posterior extramedullary intraspinal arteriovenous fistula fed by the anterior spinal artery.

Conclusion: The association of Klippel-Trenaunay syndrome with an extramedullary arteriovenous fistula has already been described in several cases and can be partially explained on a developmental basis.

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Magnetic resonance imaging of the shoulder in children with brachial plexus birth palsy

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Purpose: To study the value of magnetic resonance imaging (MRI) in the diagnosis of abnormalities associated with obstetric brachial plexus palsy.

Patients and method: Six patients (three boys, three girls) aged 8 months to 10 years (mean 4 years, 11 months) suffering from

Erb-Duchenne brachial plexus birth palsy were prospectively studied with MRI. A group of 11 healthy children was used as control. **Results:** MRI anatomy of the normal growing glenohumeral joint is presented. A hypoplastic and flattened posterior part of the glenoid fossa and a blunted posterior labrum were found in all patients. Four patients had a blunted anterior labrum and a flattened humeral head. Four patients presented with a posterior subluxation of the humeral head.

Conclusions: These results suggest that MR imaging provided a nonionising and noninvasive method to demonstrate the early abnormalities of the shoulder associated with obstetric brachial plexus palsy, which may prompt orthopedic correction.

Craniofacial fasciitis of childhood

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Craniofacial fasciitis is a tumor-like fibroblastic lesion. Only nine cases of cranial fasciitis and one case of facial fasciitis have been reported in children. The ages of these reported cases ranged from newborn to 6 years of age, mean 18 months. We present a 15-month-old white male with facial fasciitis. He was presented with a 2-month history of gradually increasing right infraorbital mass. The preoperative CT of the face showed a soft tissue mass with dense curvilinear calcification in the outer rim in the right infraorbital region anterior to the maxillary sinus. No bony destruction or inflammatory changes in the sinuses or orbit were present. The lesion was completely resected surgically. The pathology showed that the tumor consisted of a loose, haphazard proliferation of spindle- to stellate-shaped fibroblasts set in a myxoid matrix. The immunohistochemical studies showed positive for vimentin and negative for desmin, HNF-35, actin and S-100, which made the diagnosis of rhabdomyosarcoma or a neurogenic tumor unlikely. The electron microscopic study showed spindle cells with a feature of fibroblastic differentiation. The postoperative MR showed no residual tumor. The patient did well without recurrence of tumor.

Reversible change in the direction of the portal flow: a case of a leukemic child with hepatosplenic candidiasis

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Portal hypertension and a change in the direction of the portal flow are known to occur associated with irreversible damage of the liver parenchyma. The portal flow can be reliably examined by colour Doppler ultrasound. Although the change of the direction of the portal flow is usually thought to be permanent, we present a case in which the portal flow direction returned to normal after 2 years. We present Doppler curves with the arterial flow visible in the background, verifying the direction of the portal flow. Acute myeloid leukemia was diagnosed in a 10-year-old boy in March 1991. The patient was in remission after two courses of chemotherapy. He had several infection episodes during neutropenia, and in April ultrasound examination revealed a large, slightly hyperechoic liver, enlarged kidneys and splenomegaly. Later focal lesions developed in liver, spleen, kidneys and pancreas. Hepatosplenic candidiasis was confirmed by liver biopsy. Despite long term amphotericin-B therapy, the liver contracted and showed an irregular echo pattern as well as focal lesions, which later became more echogenic while several new lesion appeared in the spleen. In August 1992 ascites was seen on ultrasound, and afterwards the amount increased rapidly. Three months later the portal flow was noted to have changed its direction away from the liver. A month later esophageal varices were diagnosed and were later treated several times. The spleen was removed because of persistent infection in it, and thereafter the patient slowly improved. The liver still remained grossly

abnormal in the repeated ultrasound examinations. More than 3 years after the onset of the infection the portal flow was seen to have changed towards the liver again. The amount of the ascites was very small and no new varices emerged. Although the infection caused irreversible liver damage, the change of the direction of portal flow is not necessarily irreversible.

Testicular adrenal rests in an 11-year-old patient with congenital adrenal hyperplasia – 21 OH deficiency: US findings

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Introduction: Patients with congenital adrenal hyperplasia (CAH) are known to have testicular lesions due to ectopic adrenal rests. Sonography permits the identification of such lesions [1, 2]. To our knowledge, only patients older than 12 years are presented in the literature [3]. We report the case of an 11-year-old patient with CAH (21 OHD). Sonography performed because of poor response to therapy, showed adrenal rests in both testis.

Case report: A 7-year-old patient with CAH (21 OHD), treated since the age of 1 month, was referred to our clinicians for evaluation of therapeutic response. US examination of testis was negative. When the patient was 11 years old, due to therapeutic response worsening, a sonographic examination was performed with a mechanical, annular multifrequency probe, 7/13 MHz (Toshiba). Near the mediastinum testis some rounded, hypoechoic, well-margined areas were detected (diameters 4–8 mm on the right, 3–6 mm on the left). These findings were considered adrenal rest tumors and therapy was modified; a new follow-up sonographic examination of testis was scheduled.

Conclusions: The present case report confirms that testicular lesions caused by ectopic adrenal rest can be present in CAH patients; these lesions can be detected with ultrasound. We describe the case of a CAH patient 11 years of age; only CAH patients older than 12 years are described in the literature. When therapy response worsens, testicular sonographic is mandatory. Very high frequency (13 MHz) probes should be used to better advantage.

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Ovarian adrenal rests in patient with congenital adrenal hyperplasia – 21 OH deficiency: US findings

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Introduction: Ectopic adrenal rests (EAR) are known to be a possible finding in testes of male patients with congenital adrenal hyperplasia (CAH). Some authors have proved their existence in CAH males by ultrasound investigations and were able to detect microscopic lesions in apparently normal testes [1, 2]. At present, to our knowledge, adrenal rests have been never detected by ultrasound (US) in ovaries of female CAH patients, although their existence has been hypothesized [3].

Case report: A 16-year-old girl with CAH due to 21-hydroxylase deficiency (21 OHD), treated with glucocorticoids since the age of

1 month, was referred because of poor control of the disease. Among the performed investigations, US of the pelvis (5 MHz sector probe) showed in both the ovaries some nodular hypoechogenic areas, of about 1 cm, surrounded by a thin, hyperechogenic “halo”. The suspicion of the presence of EAR in the ovaries was confirmed by histologic studies. A US examination after a period of adequate treatment showed a reduction in size of the ovarian nodular areas with an enhance of the hyperechogenic “halo”.

Conclusions: US examination of adrenal rests has not been described in the ovary, although some authors were able to detect them in CAH males’ testes. In this case, the US findings suggesting the presence of adrenal tissue were confirmed by histologic examination. In girls with CAH showing a poor hormonal control, we suggest performing a US scan of the pelvis in order to detect any ectopic adrenal tissue. As in CAH males, the presence of EAR probably causes a poor response to treatment and required a period of therapy with high glucocorticoid doses.

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Malignant pheochromocytoma in neurofibromatosis type 1: a case report

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Pheochromocytomas are rare tumors in children, and most are benign. We report a 14-year-old boy with hypertensive crises because of diffuse metastatic pheochromocytoma of the left adrenal gland. At presentation a diagnosis of neurofibromatosis type 1 was made because of typical cutaneous signs. The primary tumor, pulmonary and skeletal metastases were evident on a single conventional X-ray film of the chest, as well as the underlying genetic disease itself, and were confirmed by helical CT scanning and at operation. Six months after diagnosis and operation of the primary tumor followed by chemotherapy, the boy was still in clinical stable condition and therapy with MIBG was started.

Hyrtil’s anastomosis: occurrence, morphology and relation to umbilical artery blood flow waveforms and fetal growth

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Introduction: The blood flow waveform is almost always identical in the two umbilical arteries. This is true also in cases with diastolic zero flow and when the placenta is remarkably asymmetrical as a consequence of either malformation or extensive infarction. This can be explained by the existence of an anastomosis between the two umbilical arteries. This anastomosis was first mentioned by B.S. Albin in 1748 and described in more detail in 1870 by the Austrian anatomist J.Hyrtil, after whom it is named. Risk pregnancies are today monitored by Doppler examinations of umbilical artery blood flow velocity waveforms. The aim of this study has been to investigate the occurrence and appearance of Hyrtil’s anastomosis and to relate it to blood flow waveforms and fetal growth.

Method: Sixty-nine pregnancies at risk for small growth for age (SGA) were monitored by Doppler registration of the umbilical ar-

tery blood flow waveforms, which were classified as normal (class 0) or pathologic (class 1–3) according to Laurin et al. (1987). Post partum the arterial system of the placentas was infused with BaSO₄ suspension for 30 min under constant pressure of 100 mm Hg. Repeated X-ray images were taken with the umbilical cord in different positions. After fixation the placentas were sectioned and perpendicular pictures were made. Anastomoses that could not be clearly classified by X-ray images only were dissected under a stereo microscope.

Results: An anastomosis could be identified in 65 of 69 placentas. In 53 instances the anastomosis was represented by a true vessel, whereas in five it was represented by a fenestration between the two arteries. In another three the umbilical arteries coalesced. In four cases anastomosis could not be classified. The anastomosis was truly absent in three cases. Another case had a single umbilical artery. In 25 of 61 classified cases, one of the umbilical arteries branched before the anastomosis, and in one case both did so, whereas in the remaining 35 instances the anastomosis was between the two main arteries. The occurrence and type of anastomosis were not found to be related to blood flow waveforms or fetal growth. The anastomosis was localized to within the umbilical cord at a maximum of 20 mm from the insertion in six cases, to the cord insertion in 48 cases and in the placental surface in 11 cases. The length of the anastomosis varied between 0 and 17 mm and the diameter between 0.5 and 6 mm. Neither the length nor the diameter was related to blood flow or fetal growth.

Conclusions: An anastomosis between the umbilical arteries was found in more than 90% of all placentas. In no case was more than one anastomosis found. The type and size was highly variable but showed no relation to blood flow or fetal outcome. Hyrtl's anastomosis explains the similarity in blood flow waveforms between the two umbilical arteries.

A sonographic method of assessing bone age

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Introduction: Ultrasound (US) has proved excellent in diagnosing many hip disorders of childhood and adolescence. Although increasing evidence exists that the anterior aspect of the cartilage of the femoral head (FHC) can be accurately imaged by US, little information is available concerning the extent of and changes in thickness of the FHC during the developmental age. This study was designed (1) to analyze the relationship between the thickness of the FHC and the degree of skeletal maturation in a pediatric population, and (2) to verify whether the measurement of the thickness of FHC may be used for bone age assessment.

Materials and methods: US examination of the hips was performed in 213 subjects (99 boys and 114 girls), aged 1.9–14 years (mean 8.1 years, standard deviation 3.5 years). Seventy-four were scanned after they had undergone hand and wrist X-ray examination for osseous maturation by the Greulich and Pyle method. Chronological age, standing height and body weight were recorded for all subjects. Images were obtained using a real-time scanner equipped with a 7.5-MHz linear transducer. To avoid interobserver errors, all US examinations were carried out by the same person, without any comparison between the two sets of photographs. A single scan of each hip in every subject was obtained, using an anterior approach with the probe placed along the axis of the femoral neck. The FHC was defined as the width of the anechoic space between the ossified nucleus and cartilage surface, close to the epiphyseal side of the physal plate. Correlation of the sonographic measurements with the independent variables chronological and

skeletal age, height and body weight were obtained. For statistical analysis the paired *t*-test and the Pearson's correlation were used. The significance level was set at 90% of confidence limits.

Results: Close correlations were found between FHC and chronological age ($r = 0.88$), skeletal age ($r = 0.88$), standing height ($r = 0.89$) and body weight ($r = 0.80$). Based on chronological age, the study population was divided into 13 groups. For each group, the mean value and standard deviation of FHC was registered. Since skeletal development at all ages is more advanced in girls than in boys, separate standards were established.

Discussion: The results of this study indicate that hyaline FHC gradually decreases in thickness during growth, as the deposition of new epiphyseal bone proceeds with age. This phenomenon is substantiated by the very strong correlation between the thickness of FHC and the chronological and skeletal ages. Although it is not definitely proven whether the FHC can be used as index of biological maturity, it is reasonable to assume that changes in the thickness of the articular cartilage are closely related to skeletal maturation. Further investigations are needed to expand the present series, assess the precision and accuracy of the method, determine racial and ethnic differences and confirm whether the depth of hyaline FHC may provide additional information on skeletal maturation. It seems that this new method can be used as a primary screening tool for developmental derangements in childhood or as an ancillary tool in cases in which skeletal age determination is difficult on conventional X-ray due to a discrepancy in maturity between the different bones of the left hand and wrist.

Coexistence of adrenocortical carcinoma with abdominal ganglioneuroma in a child

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We present the case of a boy aged 3 years, 5 months with a, to our knowledge, never previously listed association of an adrenocortical carcinoma and an abdominal ganglioneuroma. The patient was examined because of general seizures and hypertension. US examination of the kidneys, intravenous pyelogram and abdominal CT showed the presence of a right suprarenal mass (4.7 × 4.2 cm). A coexisting small soft tissue mass in the retrocrural and paracaval region was considered to be a lymphadenopathy. After exploratory laparotomy, the pathologic diagnosis of a right adrenocortical carcinoma and a prevertebral ganglioneuroma was made. Adrenocortical carcinomas are very rare and highly malignant tumors in childhood. Only 300 cases have been identified, 75% of them in children under the age of 5 years. These neoplasms mostly have an active hormone secretion, tend to be locally aggressive, and metastasize to the lung, liver, lymph nodes and bone. In our case there was a misleading second retroperitoneal neoplasm which turned out to be a ganglioneuroma (a rare, benign neurogenic tumor arising from the sympathetic ganglia). We present this unusual case because of the difficulty in radiological diagnosis. The imaging investigations and their findings are discussed and correlated with literature reports.

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