## 95 Assessment of bone mineralization by ultrasound in infants

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AIM - To develop a system to measure in bone, the velocity and attenuation coefficient of ultrasound as a function of frequency, in infants at the cotside and to determine its reproducibility.
METHOD - The infants wrist is placed between two transducers. They are at a fixed distance from each other in two containers each with a thin rubber membrane acoustic window. The radius is scanned and ten measurements taken for averaging.
Electrical pulses, controlled by a microcomputer, are produced at the transmitter output, which can excite both transducers. The received echoes are routed to an analogue-to-digital converter and the gated digital signals and their time of arrival are used to calculate the bone thickness and the velocity and attenuation coefficient of sound. This was repeated 5 times in one infant.
RESULTS - Bone thickness mean 5.46 mm SD 0.19, velocity mean 1630 m/s SD 22, attenuation coefficient mean 17.9 MHz/dB/cm SD 1.0.
CONCLUSION - Ultrasonic assessment of bone is a reproducible technique which we are using to study term and preterm neonates.

## 96 DIFFERENTIAL DIAGNOSIS OF HYPERPHENYLALANINAEMIA (HPA) AND FOLLOW UP OF SEVEN PATIENTS WITH TETRAHYDROBIOPTERIN (BH4) DEFICIENCY

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HPA is either due to phenylalanine hydroxylase deficiency or to BH4 deficiency. The latter should be excluded in all newborns with HPA using three methods: (1) oral BH4 loading test; (2) measurement of dihydropteridine reductase (DHPR) activity in erythrocytes; (3) investigation of the urinary pterin pattern. From 1979 until 1987 seven patients with BH4 deficiency have been detected in Munich. One of two girls with severe 6-pyruvoyl tetrahydropterin synthase (PTS) deficiency has been treated since the age of 7 weeks. Her psychomotor development is now, at the age of 11 years, normal whereas the other girl, 13 years of age, who has been treated since the age of 7 years, shows marked developmental delay. Three out of four patients with partial PTS deficiency (now 6-9 years of age) developed normally, the 4th patient (6 years old), however, presents with slight psychomotor disabilities. Early treatment of a patient with DHPR deficiency was highly unsatisfactory. This boy died at the age of 23 months.

## 97 Phosphorus metabolites in the human placenta estimated in-vivo by magnetic resonance spectroscopy.

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High energy phosphate metabolism of the human placenta has been studied in vivo using a localised 1D-chemical shift imaging magnetic resonance spectroscopy (MRS) technique. The position of the fetus and placenta were determined accurately by fast-scan magnetic resonance imaging (FS-MRI).

A General Electric "Signa" 1.5 Tesla system was used. Twelve third trimester pregnancies were studied. Seven of 12 normal pregnancies (gestation range: 33-36w) were suitable for analysis. In the remainder excess fat in the abdominal wall placed the placenta too far from the surface coil. Resonances due to inorganic phosphate (Pi) and nucleotide triphosphates (NTP, mostly ATP) were clearly distinguished. Large phosphomonoester (PME) and phosphodiester (PDE) signals were seen. Based on a total phosphorus content of 30 mmol/kg (Documenta Geigy 1: 220), a low estimate for ATP was 3.45 mmol/kg.

The size of each peak from 6 subjects was calculated as a fraction of the total phosphorus content and a normal range was established. The seventh subject had a twin pregnancy: one twin had died a few days previously and the PME and PDE signals fell outside the 95% confidence intervals.

In-vivo 31P spectra from the human placenta have not been previously reported. The ability to detect large resonances due to Pi and the products of phospholipid synthesis (PME) and catabolism (PDE) offers the opportunity of following placental maturation and its disorders.

## 98 PSYCHOLOGICAL AND SOCIAL FINDINGS IN ADOLESCENTS WITH PHENYLKETONURIA (PKU)

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In a retrospective study the psycho-social context of 34 early treated patients with PKU (mean age 14.6 y) and their parents were evaluated.
In the CFT 20 the patients reached a mean IQ of 93.6 which is significantly below the IQ of their mothers (98.2), that of their fathers (105.4) ($p<0.05$) and that of the normal population (100, $p<0.01$). In the MBI (personality inventory) the social context significantly differed from the normal population as the patients described "more support by parents, less autonomy, lower evaluation of their situation in school, less achievement motivation, less frustration tolerance, lower self-confidence and less health" ($p<0.05$). No significant differences regarding the scales "interpersonal behaviour, working discipline and creativity" were found.
The dietetic knowledge of most patients was poor and 55 % of them had poor dietary control despite regular instructions by doctors and dietitions. 93 % would prefer to discontinue the dietary regimen.
In view of these results and the demand to continue the diet during adolescence, systematic teaching programs must be established for a better coping with PKU.

## 99 EXOGENOUS SURFACTANT PHOSPHOLIPIDS STIMULATE ENDOGENOUS SATURATED PHOSPHATIDYLCHOLINE SYNTHESIS.

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Surfactant treatments result in increased synthesis and secretion of endogenous saturated phosphatidylcholine (Sat.PC) in adult rabbits. To elucidate the contribution to these effects of surfactant associated proteins and surfactant phospholipids in surfactant preparations we studied a mixture (MPCG) of dipalmitoylphosphatidyl choline and phosphatidyl glycerol (9:1), a lipid extract of natural surfactant (LENS), a phospholipid fraction of natural surfactant (PLNS) and a reconstituted natural surfactant (RNS). These surfactants were injected into the left mainstem bronchus after induction of oxygen resorption atelectasis of the left lower lobe of adult rabbits. Surfactant metabolism was studied by injecting $^{32}P$-orthophosphate 30 min and $^{3}H$ palmitic acid i.v. 6 h after the injections, followed by sacrifice at 10 h after the injections with subsequent alveolar wash and processing of the right lung and left lower lobe separately. For the $^{32}P$ incorporation into Sat.PC the ratios of the left lower lobe to right lung were: 0.87±0.15 (MPCG), 0.74±0.14 (LENS), 0.90±0.11 (PLNS), 0.86±0.14 (RNS) significantly higher than unmanipulated controls: 0.47±0.05 (mean±SD) ($P<0.05$). LENS tended to decrease the secretion of Sat.PC. These results suggest that Sat.PC synthesis is stimulated aspecificially by phospholipids from exogenous surfactant.

## 100 IMMUNE RESPONSE AFTER HETEROLOGOUS SURFACTANT TREATMENT OF NEWBORNS WITH RESPIRATORY DISTRESS SYNDROME (RDS).

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Curosurf, a surfactant preparation derived from minced pig lungs, used for the treatment of newborn infants with RDS, contains low molecular weight proteins. We studied the immune response after a single dose of Curosurf given intratracheally in 68 patients with RDS, 49 treated with Curosurf and 19 controls. Sera were taken before, 3 wks and 3 months after surfactant treatment. The sera were applied on frozen pig lung tissue, followed by an indirect peroxidase assay. Using this procedure any immune response to lung tissue components would be demonstrated as a brown-reddish deposit. In none of the controls nor in the sera obtained before Curosurf treatment we found any reaction. In 4 of the 49 Curosurf-treated patients we found a positive immune response, demonstrated by a clear brown staining of bronchial walls and brown granular deposits in alveolar cells, and in some lung tissue specimens collagen strands were also slightly brown coloured. Of these 4 patients, 2 showed a positive reaction 3 wks and 3 months after treatment, 1 only 3 wks after and 1 only 3 months after Curosurf treatment. We conclude that sera of some patients treated with Curosurf appear to contain antibodies against pig lung tissue components and surfactant proteins.