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PHOSPHOMONOESTERS, PHOSPHODIESTERS AND PHOSPHOLIPID BILAYERS STUDIED BY MAGNETIC RESONANCE SPECTROSCOPY (MRS) OF INFANT BRAINS AND LIVERS. J Moorcraft, RM Dixon, NK Ives, PL Hope, NM Bolas, B Rajagopalan, TAD Cadoux-Hudson, GK Radda. Dept of Paediatrics, John Radcliffe Hospital and MRC Clinical MR Unit, University of Oxford, England.

Phosphomonoesters (PME-mainly phosphoethanolamine), and phosphodiester + phospholipid bilayers (PDE + PB) can be quantified non-invasively using phosphorus MRS. Spectra from the livers of 7 normal infants (gest'n 27-40w, age 3-76d), and the brains of 12 other normal infants (gest'n 37-42w, age 1-14d) were obtained by MRS, using phase modulated rotating frame imaging (PMRFI) for spatial localisation. Global metabolite ratios relative to adenosine triphosphate (ATP) are shown with comparable data from adults. Liver and brain have more PME ($p < 0.001$), and liver has less PDE + PB ($p < 0.02$), in the infant than in the adult.

	Brain		Liver	
	Adult(n=7)	Infant(n=12)	Adult(n=17)	Infant(n=7)
PME/ATP	0.70±0.34	1.91±0.52	0.32±0.08	0.98±0.30
PDE+PB/ATP	3.32±1.32	2.57±0.76	1.44±0.34	1.12±0.22

±1SD

Depth resolved PMRFI data show high PME/ATP at all depths in infant brain and liver. PDE+PB does not increase with depth in the infant brain, unlike the fully myelinated adult brain. The sum of PDE(mainly phospholipid breakdown products)and PB is reduced compared to adults in localised spectra of white matter ($p < 0.01$)

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LONGTERM INTELLECTUAL DEVELOPMENT IN 86 CHILDREN WITH PHENYLKETONURIA (PKU).

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Dietary treatment in phenylketonuria (PKU) prevents severe impairment of psychomotor development. However, there are unequivocal recommendations in regards to diet liberalization. Aim of this retrospective study was to examine intellectual development in eighty-six individuals with "classical" PKU, who were treated early in life.

Intellectual development was recorded using the Kramer, Hawik & Hawle test at the age of 4, 6, 8, and 12 years.

Intellectual development was not influenced by the time dietary treatment was started, probably due to the fact that treatment in all patients was introduced within the first two months (mean±SD: 23±8 days, range: 6 to 61 days).

age (n)	6 years (86)	8 years (75)	12 years (44)
IQ (mean±SD)	105±14	100±14	101±15

Compared with their non-PKU brothers and sisters PKU children showed significantly lower IQ at the age of 6 years (105±14 vs 114±14, $P < 0.001$). IQ of the parents did not differ from IQ of the PKU children at the age of 12 years.

A significant association was observed between the quality of diet control (measured by Guthrie-test) and intellectual outcome: IQ at 6,8, and 12 years significantly correlated with quality of diet control during all observation periods ($r = 0.388$, $P<0.001$). Children with "strict" dietary control - already at the age of 2 years- showed significantly higher IQ than those with "poor" dietary control. These observations underline the importance of a strict adherance of PKU patients to the diet to acchieve adequate intellectual development.

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Topographical Localization of Peroxisomal Acyl-CoA Ligases: Differential Localization of Palmitoyl-CoA and Lignoceroyl-CoA Ligases.

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We found that peroxisomal lignoceroyl-CoA ligase, like palmitoyl-CoA ligase, is present in the peroxisomal membrane. To further define the role of peroxisomal acyl-CoA ligases we examined the transverse topographical localization of enzymatic sites of palmitoyl-CoA and lignoceroyl-CoA ligases in the peroxisomal membranes. Proteolytic enzyme treatment inhibited palmitoyl-CoA ligase activity in intact peroxisomes and lignoceroyl-CoA ligase activity was inhibited only if peroxisomes were disrupted prior to trypsin treatment. Antibodies to palmitoyl-CoA ligase and to peroxisomal membrane proteins (PMP) inhibited palmitoyl-CoA ligase in intact peroxisomes, and no pool of "latent" activity appeared when antibody-treated peroxisomes were disrupted. On the other hand, disruption of PMP antibody-treated peroxisomes with detergent resulted in the appearance of a "latent" pool of lignoceroyl-CoA ligase activity. These results demonstrate that the enzymatic site of palmitoyl-CoA ligase is on the cytoplasmic surface whereas that for lignoceroyl-CoA ligase is on the luminal surface of peroxisomal membranes. The implication of these findings to X-adrenoleukodystrophy will be discussed.

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EFFECT OF POLYETHYLENE GLYCOL-SUPEROXIDE DISMUTASE AND CATALASE ON BLOOD-BRAIN BARRIER PERMEABILITY AFTER ISCHEMIA-REPERFUSION IN THE NEWBORN PIG. Olivier P. Thélin, Robert Mirro, William M. Armstead, Sam Zuckerman, David W. Busija, and Charles W. Leffler. The University of Tennessee, Memphis, Memphis, TN.

Introduction: Transport of urea through the blood-brain barrier (BBB) and superoxide anion radical production on the brain's surface are both increased during post-ischemic reperfusion. **Study objective:** To see if the alteration of the BBB penetration to urea could be linked to superoxide anion generation, 20 minutes of total cerebral ischemia followed by 2 hours reperfusion was produced by increasing intracranial pressure with a hollow parietal bolt. **Interventions:** Newborn pigs were pretreated with systemic polyethylene glycol-superoxide dismutase (PEG-SOD), 1000 U/kg, and polyethylene glycol-catalase (PEG-CAT), 10,000 U/kg. ^{14}C-Urea transfer into the brain was measured in pretreated (n=5) and in control animals (n=5). **Main results:** Transfer of ^{14}C-Urea into the brain is not significantly altered by pretreatment with PEG-SOD and PEG-CAT; K_{in} transfer constant = 37.0 ± 1.6 in treated group vs 34.0 ± 2.4 $cm^3 \cdot g^{-1} \cdot s^{-1} \cdot 10^6$ in control group (mean ± SEM). **Conclusion:** The results of the present study suggest that superoxide anion generation does not directly contribute to the alteration of urea transfer through the BBB during ischemia-reperfusion in the newborn pig.

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Rhizomelic Chondrodysplasia Punctata: Metabolic Studies in Isolated Peroxisomes.

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The Rhizomelic form of chondrodysplasia puntata (RCDP) is a fatal autosomal recessive peroxisomal disorders. Clinically, it is characterized by abnormal calcification of extremities, dwarfism, cataracts, skin changes and severe mental retardation. The biochemical findings were abnormal activities dihydroxy-acetonephosphate acyltransferase (DHAP-AT) oxidation of phytanic acid whereas oxidation of lignoceric acid was normal.

Peroxisomes isolated from two cell lines of RCDP and control each were compared for biochemical studies. The RCDP peroxisomes had the same density (1.178 gm/ml) as control peroxisomes. The residual activity (0.5% of control) of DHAP-AT was observed only in the peroxisomes from RCDP and no such activity was observed in any other region of the gradient. The rates of activation and oxidation of lignoceric acid was normal in peroxisomes from RCDP. The peroxisomes from RCDP contained 3-ketoacyl-CoA thiolase in the unprocessed form (44 KDa) whereas peroxisomes from normal peroxisomes contained both unprocessed (44KDa) and mature (41 KDa) forms. These results suggest that processing of 3-ketoacyl-CoA thiolase takes place in peroxisomes and recognition signals for its transport into peroxisomes were normal in RCDP.

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EARLY SUPPLEMENTARY FEEDING AND BREASTFEEDING DURATION IN NORTHERN THAILAND. DA Jackson, SM Imong, L Wongsawasdii, K Amatayakul & JD Baum. Inst. of Child Health, Univ. of Bristol, UK & Res. Inst. for Health Sciences, Chiang Mai University, Thailand.

Infant feeding patterns were studied prospectively from birth to 2 years of age in 60 infants, randomly selected from 36 northern Thai villages. Early introduction of supplementary foods was typical with 68% of infants receiving regular supplements by 6 weeks of age. Infants began supplements significantly earlier if their father was a farmer ($p=.006$), they were born during rice planting or rice harvesting period ($p=.011$) into households with many adults ($p=.05$). Early supplementary feeding may permit Thai mothers to devolve infant feeding to other adults in the household at times when extra farm labour is required. Breastfeeding duration was prolonged despite early supplementation. Median duration was 12 months and 7% of infants were still breastfed at 24 months. Early supplementary feeding per se was not a significant predictor of duration of lactation. However, short lactation was significantly associated with infant formula given as the first supplement ($p=.009$), younger mothers ($p=.011$) and more children in the household ($p=.028$). Weight or length gain from 0-3 months was not associated with early supplementation or feeding of infant formula, after adjusting for nutrient intake, sex and birthsize.