PRPP, and inorganic phosphate (P_i) levels are somewhat higher in mutant cells, and MB elevates PRPP. Addition of AN plus MB depletes P_i, PRPP and adenine nucleotides in normal cells, but in the mutant R5P and PRPP rise and nucleotides remain unaltered. We conclude that oxidative PS is not essential for R5P generation: hence G6PD-deficient cells have no defect in PRPP or nucleotide synthesis. They are also protected from the combined effect of AN and MB, which is based on P_i depletion in normal fibroblasts. P_i is the prime modulator of PRPP synthetase in vivo.

Supported by the Academy of Finland, NIH (18197), the Kroc Foundation, and V.A. Medical Research Service.

79 G.KOSZTOLÁNYI*, K.JOBST*, M.KELLERMAYER* and A.LUDÁNY*/Intr.by J.MESTYÁN/. Departments of Paediatrics and Clinical Chemistry, University Medical School,Pécs,Hungary. Comparison of surface charge and ADP induced electrokinetic behaviour of fetal and adult platelets.

The electrophoretic mobility of washed platelets as well as of platelets suspended in diluted plasma obtained from adults and newborns was practically the same. No significant difference could be observed in the pH-mobility relationship of the two types of platelets. These comparative studies indicate that the actual charge density, i.e. the number and sign of the charges groups at the fetal and adult platelet surface are essentially identical.

Significant difference between the two platelet population was found, however, in the mobility changes induced by ADP. On the basis of "cross over" experiments between the platelets and plasma of adults and newborns it seems likely that the different behaviour of fetal platelets arises from a dissimilarity between adult and fetal plasma. The adult plasma might have a factor which is not present in the fetal plasma. Preliminary results indicate that this factor is a plasma component with mol.wt. about 1o ooo.

80 M.OBLADEN*and I. KLATT*(Intr.by H. BICKEL) Universitäts-Kinderklinik,69 Heidelberg,FRG. A Synthetic Surfactant Substitute.

A crystalline mixture of 90 % Dipalmitoylphosphatidylcholine (DPPC) and 10% Dipalmitoylphosphatidylglycerol (DPPG) was analyzed for its suitability as a surfactant replacement using a specifically designed modified Wilhelmy balance.A suspension prepared by vigorous shaking in 0.9% NaCl at 20° and 37° did not adsorb to the air-water interface (γmax 72.6 dyn/cm, γmin 69.8 dyn/cm,S.I. 0.04). When prepared in multilayered liposomes after drying,the material was adsorbed to the surface, spread rapidly to a film,and was highly surface-active (γmax 70.4 dyn/cm, γmin 3.7 dyn/cm,S.I. 1.83). After solubilization with ultrasound,a clear solution resulted which was not surface-active (γmax 72.2 dyn/cm, γmin 56.1 dyn/cm,S.I. 0.39) due to the formation of stable vesicles unable to form a film at the surface. Compared to DPPC alone which adsorbs to the surface in more than 90 minutes at 37°C,the material investigated adsorbed to a surface-active film in less than 10 minutes. The minimal film concentration of DPPC-DPPG displaying maximal surface-tension lowering ability was 2.55 $\mu g/cm^2$ in the liposomal preparation.
No local or general toxicity was found in rabbits after tracheal instillation of the surfactant substitute during mechanical ventilation. Autohistoradiography showed the 3-H-labeled material at the alveolar wall 30 minutes after instillation into the tracheal tube.

81 **J.J.PIETRZYK.Clinical Genetics Department, Institute of Pediatrics,Kraków,Poland.**

Genetic analysis of HLA and spina bifida association.

The HLA typing and routine segregation analysis of HLA haplotypes were performed in the group of 68 families with single and multiple cases of spina bifida /SB/.A significant associations of SB with HLA-B27 allele /Chi2=78.073 p<0.0145/ and HLA-A3,B27 haplotype /Chi2=7.371 p<0.01/ were found.The observed distribution of B27 among the affected children fits the distribution expected on the assumption that this antigen makes the zygote more susceptible to the abnormal neural tube development /Chi2=0.161 p>0.5/.The significant relative risk of SB development given B27 allele and HLA-A3,B27 haplotype was 3.4/p<0.0005/ and 4.6 /p<0.005/,respectively.The analysis of parental HLA phenotypes revealed significantly higher frequency of common HLA antigens shared by both members of the couples as compared to the expected values /Chi2=314.040 p<0.0005/.The couples which shared two or three HLA antigens yield the highest realitive risk of SB for their children /RR=17.8 p<0.0005/.The results raise the possibility that HLA antigens may interact with other developmental factors during the ontogenesis. Non-random association of HLA antigen and HLA haplotype with SB,as well as the very high frequency of common HLA antigens among the parents of the affected children might be used in identification of risk families.

82 J.C. ROUGE*, L. TISSOT* and G.C. LACOURT* (Intr. by P.C. Sizonenko). Dpt of Anesthesia, Pediatrics and Genetics, University of Geneva, Geneva, Switzerland. Effects of continuous positive airway pressure breathing (CPAP) after pediatric open heart surgery.

CPAP is an advance in the treatment of pulmonary dysfunction after cardiac surgery. The effects of different levels of CPAP on lung functions were determined immediately after weaning from the respirator in 14 children. The following parameters were measured: - tidal volume (V_T), compliance (C_L), resistance (R_{TL}) and blood gases ; the work of breathing (W_T) was calculated.

CPAP (cmH_2O)	0	5	10	15	0
	Mean initial value (±SEM)	Change from mean initial value (in percent)			
C_L (ml/cmH_2O)	21 ± 4,9	+ 47	+ 57,6*	+ 86,2*	+ 84,3
V_T (ml)	122 ± 20	+ 15*	+ 8,6	+ 25,8*	+ 7,9
PaO_2 (kPa)	12,6 ± 1	+ 19	+ 23*	+ 32,5*	+ 30,9
$PaCO_2$ (kPa)	4,4 ± 0,2	+ 1,8	- 7*	- 6,1*	- 12,3*

* Significant at the 5 % level.

Simultaneous beneficial effects on C_L, R_{TL}, W_T and PaO_2 were obtained with increasing values of CPAP up to 15 cmH_2O, the prefixed maximum value in this study. No deleterious effects on hemodynamics were found.

83 P. SCHWARTZE* (intr. by L. Corbeel). Department of Pathophysiology, School of Medicine, Karl-Marx-University, Leipzig GDR.
Does rotatory stimulation or handling influence the development of vestibular system?

Rabbits were used to test whether repeated vestibular stimulation or handling during the first 1o postnatal days accelerates the development of vestibulo-oculomotor reactions. The animal material was devided into two experimental groups, and each of these into three subgroups: stimulated rabbits, handled rabbits and controls. Meanwhile the handling procedure was the same, two different rotatory stimulation programs were used in the respective subgroups from the 1st to 12th postnatal day. At this day chronic electro-oculographic electrodes were implanted to all of the stimulated and handled animals and the controls. Nystagmic eye movements (NEM) were recorded daily during a standard rotation stimulus between the 12th and 2oth day. No systematic differences were observed between number and latency of NEMs of stimulated, handled and control animals. Further, no correlation was found between the speed of body weight increase and of nystagmic parameters in the subgroups.

84 M. GARCIA-FUENTES*, A. RUBIO*, J.L. ARCE*, E. BUREO*, V. MADRIGAL* and M. LOPEZ-COLLADO* (Intr. by J. Rodríguez-Soriano). Dept. of Pediatrics, National Med. Center "M. de Valdecilla", School of Medicine. Santander, Spain. Alterations of the complement and coagulation systems in meningococcal infections.

Serum levels of complement components (C1q, C4, C3, C5, C9, C3PA and C1I), platelets, prothrombin time (PT), fibrinogen concentration and fibrin degradation products were measured at admission in 93 children (mean age 3.1±2.0 y)with meningococcal infections, 86% type B. Results were compared with an age matched normal group. Patients were classified in three groups: 21 with meningitis without systemic manifestations; 39 with uncomplicated septicemia and 33 with septicemia and shock.
C1q was decreased ($p<.001$) in the three groups; C3 was also low but only in the last two groups was significantly diminished ($p<.005$). Forty-seven patients, regardless of the groups, showed a prolonged PT and 7 out of these 47 showed a disseminated intravascular coagulation. These 47 patients had lower levels of C1q ($p<.02$), C3 ($p<.05$), C5 ($p<.05$), C3PA ($p<.005$) when compared with the remaining patients with normal PT. Values of PT in all patients correlated well with the levels of C1q ($p<.05$), C4 ($p<.01$), C3 ($p<.001$), C5 ($p<.001$), C9 ($p<.001$) and C1I ($p<.05$). These results suggest that activation of the classical pathway of complement occurs in all patients with meningococcal infections, even in benign cases, and that such activation may be related to the alteration of the coagulation system.

85 F.LAURENTI^,R.BALDUCCI^,P.CRISPINO^, F.MALAGNINO^, and D.PALERMO^(Intr. by Bucci). Depts of Pediatrics and Hematology, CNR Centre for Respiratory Viruses, Univ. of Rome, Italy. Functional activity of packed polymorphonuclear leukocytes (PMN) obtained by leukafiltration.

We recently obtained a striking increase of the survival rate in very small pre-term infants with sepsis through daily transfusions of packed PMN (2Oml/Kg equal to 0.5 x 10^{10} cells). In order to increase the availability of PMN concentrates and to reduce the risk of sentization, it would be useful to transfuse repeatedly, in the same patient, PMN collected from the same donor. We, therefore, evaluated the rate of functional decay of packed PMN obtained by leukafiltration and

kept at +4C° for 48 hrs, for phagocytosis, candidacidal activity, chemotaxis and spont. & stim. NBT test (for methods see : Ital. J.Pediat. 4:571,1978). Phagocytosis, candidacidal activity and NBT test remained unchanged at 24 and 48 hrs (104% and 100%, 94% and 93%, 94% and 92% respectively of initial values). Chemotaxis only showed a slight decrease (98% and 73%). The present results show a satisfactory functional stability of PMN collected by leukafiltration, suggesting that the same concentrate may be effectively transfused for three consecutive days.

86 L.Businco,A.M.Menghi°,P.Rossi°,P.Delle Femmine°,E.Galli °
Dpt.of Pediatrics(I),Centre Respiratory Viruses CNR, University of Rome. Zinc therapy in an infant with Acrodermatitis Enteropathica:clinical and immunological result.

Acrodermatitis Enteropathica(AE), a rare autosomal recessive disorder is characterized by onset before age 1,failure to thrive diarrhea,alopecia,a vesiculobullous eruption around the mouth, anus hands and feet,high susceptibility to infection.The disorder is related to zinc metabolism and low level of plasma zinc have been reported in these patients.A cellular chemotaxis defect zinc dependent has been demonstrated by Weston et al.(Arch.Dermatol. 1977).We have investigated a 5 month old infant who developed the clinical features of AE when weaned from breast milk to cow's milk. Serum zinc concentration were 50µg%.The evaluation of the immune system revealed a normal T and B lymphocyte functions but a severe defect of neutrophil chemotaxis was present.The therapy with zinc sulphate 40mg/os/daily induced a dramatical clinical improvement, the restoration of plasma zinc level and the correction of neutrophil cellular chemotaxis.When the zinc therapy was discontinued cellular chemotaxis decreased(Tab.).

		Zn mg. 40/os			
Serum Zn (103 ± 9)	50	101	97	92	48
Cell.Chemo. M.I.(522 ± 225)	0	600	<100	<100	130
Hum. " " (522 ± 225)	300	880	240	200	300
Weight Kg.	4.5	6.1	7.3	7.5	8.4
Weeks	0	2	12	14	16

87 A. LUCAS*, T.E. ADRIAN*, S.R. BLOOM* and A. AYNSLEY-GREEN. University Department of Paediatrics, Oxford and Department of Medicine, Hammersmith Hospital, London. DIFFERENCES IN ALIMENTARY HORMONE RESPONSES BETWEEN BREAST AND BOTTLE-FED INFANTS.

Comparative aspects of breast and bottle (formula) feeding have received considerable attention, yet there is little information on gastrointestinal differences between infants fed in these two ways. We compared plasma levels of 10 alimentary hormones in 33 6-day old normal term infants who were breast fed, with 39 such infants fed on a modified cow's milk formula (Cow and Gate Premium). Each infant contributed with ethical approval only one plasma sample taken at the time of a routine blood test. These samples were taken either basally or at 25, 60 or 120 min. after a feed. Formula-fed infants have significantly higher basal levels of motilin ($p < 0.05$), neurotensin ($p < 0.05$), vasoactive intestinal peptide ($p < 0.02$) and gastric inhibitory peptide ($p < 0.02$), and had larger insulin ($p < 0.05$) and neuro tensin ($p < 0.01$) responses to feeding. The formula-fed infants showed postprandial elevations of entero-glucagon ($p < 0.02$) and pancreatic polypeptide ($p < 0.02$) and depression of motilin ($p < 0.02$) while none of these three responses were seen in breast-fed infants. Plasma concentrations of glucagon, gastrin and secretin were, however, identical in the two groups. These differences may reflect feed composition, or lower total milk intake in breast-fed infants prior to the study (feed volumes were, however, similar during the study itself). A strong relationship between dietary experience and hormone release has been demonstrated.

88 LUCAS, A*, Lucas, P.J.* Baum, J.D. University Department of Paediatrics, John Radcliffe Hospital, Oxford. FLOW AND COMPOSITION DETERMINATION OF SUCKLED BREAST MILK.

Although human milk has been studied extensively, this milk has usually been collected unphysiologically by mechanical expression of the breast. Expressed breast milk (EBM) may differ from sucked breast milk (SBM), the milk that the infants obtain during breast feeding. The calculation of SBM content requires simultaneous measurement of milk flow and composition throughout suckling since these two variables change continuously during a feed. We have weighed 132 six day old infants at different times during feeding on each breast (each infant contributing only one weighing datum) and using cross sectional data analysis we have demonstrated that the breast has an exponential emptying pattern with a mean of 50% and 85% of the feed from each breast flowing from mother to infant after two and four minutes respectively. Preliminary data indicate that this same flow pattern occurs during a feed at one month after delivery. We describe how a nipple shield, equipped to sample milk continuously during feeding can be used to calculate the nutrient content of SBM by matching flow with composition data. We also describe a micro-miniaturised ultrasonic flowmeter incorporated into a nipple shield that measures milk flow throughout feeding. Our studies are of relevance to the accurate determination of infants' nutritional requirements in this country and in the developing world.

89 F.DONZELLI^,C.BOCCHESE^,L.VERGANI^,P.A.BATTISTELLA^, F.F.RUBALTELLI, C.ANGELINI^. Department of Pediatrics and Neurology, University of Padova Hospital, Padova, Italy. Free and acyl-carnitines in human milk and colostrum.

We observed that newborns fed soy based formulas essentially free of carnitine demonstrated lower levels of free and acyl-carnitines in plasma than newborns that were breastfed or were fed with other types of milk. We, therefore, inferred an essential role of carnitine in nutrition during the first weeks of life and studied the concentration of free carnitine, acyl-carnitines, triglycerides and proteins in colostrum and human milk. We analyzed 93 samples of human milk obtained from 14 mothers. Of these, 9 were obtained during the first 5 days of lactation while 5 were specimen obtained during the second month of nursing. The milk was collected at the onset and at the end of each feeding from each breast once in the morning and evening. Colostrum when compared with mature milk showed increased concentration of free carnitine (45±12.56 versus 32.41±9.63 SD nmol/ml) and proteins (15.95±3.96 versus 7.88±1.83 SD mg/ml) but reduced content of triglycerides (6.00±3.38 SD versus 11.89±6.64 SD nmol/ml). All these changes were significant ($p < .001$). Single samples in each mother showed significant correlation both in colostrum and mature milk in the levels of free carnitine and proteins. Colostrum is, therefore, an important nutrient in the metabolic transition between fetal and postnatal life: it contains essential aminoacids in the form of proteins, and is low in triglycerides. The elevated level of carnitine seems to promote the newborn adaptation in utilizing long chain fatty acid. During fetal life glucose and amino acids are prevalently used as a metabolic fuel and fatty acid oxidation is low.

90 J. JAEKEN*, M. VANDERSCHUEREN-LODEWEYCKX*, P. CASAER*, L. SNOECK*, L. CORBEEL, E. EGGERMONT*, R. EECKELS*
(Dept. of Paediatrics, University of Leuven (B)
Familial psychomotor retardation with markedly fluctuating serum prolactin, FSH and GH levels, partial TBG-deficiency, increased serum arylsulphatase A and increased CSF protein: a new syndrome?

Identical twin-sisters (born at 36 wks; birthweight 2.2 and 3.0 kg) presented at 2 years of age with marked psychomotor retardation and bone-age of 1 year. Physical growth and phenotype were normal. Repeated investigations revealed: markedly fluctuating basal serum prolactin (778-5652 µU/ml; nl < 800), FSH (17-55 mIU/ml; nl< 10) and GH (2-144 ng/ml; nl < 10), but normal LH; low TBG (1.1 and 1.2 mg/dl; nl 1.6-2.4) also present in the father, with otherwise normal thyroid function including TRH test, arylsulphatase A moderately increased in serum (mean 293 and 272 nmol/ml; nl 30-130) but not in leukocytes, without increase of other lysosomal enzymes, and increasing CSF protein. Normal results were found for GH response to i.m. glucagon, urinary excretion of 17-keto and 17-hydroxysteroids, at funduscopy and for lymphocyte karyotype (Giemsa banding), buffy coat of blood leukocytes and electronmicroscopy of conjunctiva. Sella tursica was normal on x-ray. Cortical and cerebellar hypotrophy was evident on CAT-scan. Electromyography was normal but nerve conduction velocity was delayed (30-31 m/sec; nl 50 ± 1). A nerve and muscle biopsy is planned. At this stage we have no satisfactory explanation for these unusual findings.

91 J.NAFFAH*,C.AKATCHERIAN* (Intr. by V.M. Der Kaloustian). Department of Human Genetics Faculté de Médecine, USJ, and the Hôpital Hotel Dieu de France, Beirut, Lebanon. A new syndrome of osteopathic dwarfism.

A probably new syndrome of osteopathic dwarfism was discovered in three siblings - a boy and his two sisters born of a first cousin marriage. At birth, the appearance was normal except for a significant micrognathia. At the end of the first year, a high fever of unknown etiology lasted over many months before the onset of swelling and pain of the big joints. There was no lymphadenopathy, no splenomegaly, no biological signs of rheumatoid arthritis. The prolonged corticosteroid therapy failed to prevent ankylosis of the big joints and of the interphalangeal joints. Mesomelic dwarfism became obvious at the end of the third year. On X-ray, the epiphyses were fragmented, the metaphyses were mostly irregular and flared, the diaphyses were thin and curved at places. The carpal bones were fragmented. The spine was normal and the facial bones were dysplastic. All the biological values were normal, especially the urinary mucopolysaccharides, aminoacids, the thyroid and parathyroid function tests. The I.Q. and the pubertal development were also within normal limits.A biopsy revealed dystrophic endochondral ossification.

92 FARRIAUX JP*, DHONDT JL.*, CARTIGNY B.*, LOEUILLE GA.*, GUILLEMYN R.*, (Intr. by Corbeel L.) - Service de Génétique et Maladies Héréditaires du Métabolisme de l'Enfant - LILLE (F) - *Congenital Thyroid Binding Globulin (TBG)deficiency. Its incidence on a screening program for neonatal hypothyroidism.* - During 13 months,76650 newborn infants have been screened for congenital hypothyroidism by blood spot T4 and TSH measurements at 6 days of life.Twelve cases of TBG deficiency were detected,indicating prevalence of 1/6,400 births. Follow-up data on these children are reported: (1)patients: 11 males and 1 female without clinical symptoms obvious of hypothyroidism.(2) at 30 days of life: (a) TBG RIA concentrations (TBGK^R), CEA)were between 2.4 and 13.2 µg/ml; in 9 males TBG was between 2.4 and 7.5 µg/ml and in the 2 remaining males 10.9 and 13 µg/ml; in the girl TBG level