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

was 13.2 µg/ml (mean control value 39.2 ± 8.3 µg/ml) (b) T4 values were always low: 1-6 µg/100ml (mean normal value 12.7 ± 3.06).(c) T3 values were between 37 and 145 ng/100ml, mean value was 102.75 (mean normal value: 229.19 ± 45.01).(d) Resin T3 uptake was always high: 53 to 74%, mean value 71% (normal: 36.3 ± 3.59.(e) Free T4 index (FTI) varied: normal in 5 cases, decreased (<2.1) in 7 cases.(f) TSH levels were normal (< 9 µU/ml)(g) T4/TBG ratio increased in 10 cases: 0.37-1.66 and was normal in 2 cases:0.34 and 0.25 (normal mean value 0.31 ± 0.05, normal range 0.21 - 0.37). In summary, 1) the incidence of the TBG deficiency was higher than reported by DUSSAULT (1/13,000).2) Severe TBG deficiency resulted in some disturbances in the measurements of thyroid hormone which might suggest hypothyroidism.3) The male predominance (11/12) and the reduced TBG level in mothers (2 families studied) were consistent with X-linked transmission.

93 I. DAB* and F. ALEXANDER * (Intr. by H. LOEB). Free University Brussels (V.U.B.) Dept. Ped., St.Pieters Ziekenhuis, Hoogstraat, 320, B-1000 Brussels(Belgium).

The mechanism of autogen drainage studied with flow volume curves.

At the annual meeting of ESPR in 1978 a new bronchial drainage technic has been presented. The method and efficiency were shown through a radiocinematographic study after a bronchography. At the present the autors studied many flow volume patterns (V̇/V) of tidal breathings, of forced vital capacity manoeuvres, of cough and compared them with V̇/V of autogen drainage to understand the mechanisms of the latter. For this purpose the cooperation was obtained from two C.F., two asthmatic and two chronic bronchorrheïc children. Systematically the pattern of autogen drainage was very comparable to a forced vital capacity manoeuvre except that it started at a lower volume, stopped at a higher volume and the highest flow of autogen drainage never reached the peak flow. It used however greatly the effort independant part of V̇/V. The V̇/V of a cough showed a peak flow even greater as in a forced vital capacity but the total volume expired was very small and many times interrupted by collapses of the airways. The authors conclude that cough is inefficient; a forced vital capacity can be efficient but is very exhausting and cannot be performed for a long period; autogen drainage is efficient and it uses the effort independant part of a forced vital manoeuvre; it can be performed even for half an hour without getting the child tired. Only the asthmatic children developped occasionnally a bronchospasm during the autogen drainage which became useless as shown by the occurence of very flat V̇/V curves.

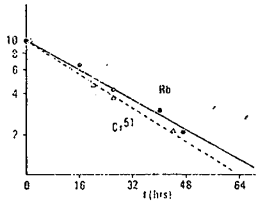
94 N. CONSTANTSAS*, and J. PALIS*(Intr. by C. Dacou-Voutetakis). 1st Department of Pediatrics of Athens University, Athens 617, Greece. A micromethod for the determination of globin synthetic ratios (GSR).

The GSR is an absolute criterion for diagnosis, investigation, and classification of thalassemias (thal). Its wide use is prevented by the tedious, time consuming processing of the numerous fractions obtained by conventional column chromatography. Electrophoresis is simpler, but gret dilution of the "hot" globin (newly synthesized by retics) in the "cold" hemoglobin of the mature red cells, results in unreliably low radioactivity of the separated globin bands. This can be overcome only by overloading the strips which compromises good resolution. Here, we report a procedure for the preparation of globin 10^4-fold "hotter", than currently obtained with established methods, by incubating 5 µl of packed 20-70% retics (isolated at 1,500xg on a preformed Percoll-Urografin density gradient) with 30 µC of ^{3}H-(leucine + lysine + proline + histidine +phenylalanine). Microzone electrophoresis of 2-5 µg of globin in urea-EDTA-thiol buffer, saturated with borate, gives sharp, well resolved α/β/γ bands. The GSR were determined in 75 individuals, in terms of cpm; 40 controls (β/α 1.01 ± .10); 20 β-thal traits (β/α .55 ± .10); 13 cases of β-thal major (β/α .17 ± .07, γ/α .35 ± .15); 2 α-thal traits (β/α 1.69, 1.41). In terms of specific activity, these values are not practically different, γ/β GSR, however, are up to 100-fold higher. This method allows easy handling by two persons, of 24-32 samples per week, at low cost, with repetitive assays. Hence, quality control is improved, and the scope of GSR determinations is expanded in post and antenatal thal, as well as in other hematological investigations.

95 D. DEL PRINCIPE, R. CESAREO°, B.M. TALLARIDA°, V.G. CIANCARELLI°, A. MENICHELLI°, M. RICCI° I Chair of Pediatrics, University of Rome, Italy.

Labeling blood platelets with stable tracers.

We investigated the possibility of labeling platelets by stable tracers. The concentration of the tracers was estimated by the Fluorescence X radioisotopic technique using a 3mCi Cd-109 source, a Xe-filled proportional detector and a single channel analyzer. In this paper we compared the in vitro survival of human platelets, anticoagulated with EDTA, stored at 22°C, labeled with stable Rb, with that of cells marked with ^{51}Cr. Three experiments were performed. A typical curve is shown. A similar investigation was performed in vivo in rabbits. The mean half-life deduced from ten animals was 22±3h (SE), for Rb labeled platelets; 18±2h for ^{51}Cr marked platelets. The elution rate of the stable tracer shows no difference in comparison with that of ^{51}Cr. Rb as a tracer offers several advantages (low toxicity, no exposure to radiation) and it can represent a valuable alternative to radioisotopes in the study of the platelet survival for the pediatric research.



96 L.Morlé*, F.Morlé*, R.Bouhass*, M.Aguercif*(Intr.by J.F. Desjeux).University Hospital,Oran,Algeria. Some aspects of beta-thalassemia in West-Algeria.

Control studies about patients suspected for thalassemia major revealed 8 cases homozygous for beta-thalassemia in 6 families:3 of them produce no beta A globin at all (beta° thalassemia type) and 5 others synthesize various amounts of beta chain (beta+ thalassemia type).Non alpha/alpha ratios were calculated after incubations of reticulocytes in vitro.They were very similar in the two types,changing from 0,13 to 0,34. However levels of HbF and alpha globin chain synthesis greatly differ in the beta+ thalassemia type (HbF percentage and alpha/non alpha ratios changing from 9 % to 67 % and 0,16 to 0,73 respectively.

7 cases from 4 families (association of beta-thalassemia and HbS) have been also studied: 4 cases produce only beta S globin chains (beta° thalassemia/beta S) and 3 others synthesize beta S and some beta A globin chains (beta+ thalassemia/beta S).

Here also non alpha/alpha ratios did not differ between the two types and vary from 0,37 to 0,43.However in the beta+ thalassemia/beta S cases one member of family more affected by anemia needed more transfusions.

Further clinical and biochemical characterization are now in progress in order to attempt some better classification of beta+ thalassemia in West-Algeria.