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FAMILIAL' INFANTILE LIVER CIRRHOSIS AND HEMOLYTIC ANEMIA DUE TO CHRONIC COPPER INTOXICATION VIA TAP WATER FROM COPPER PIPES.

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We report a disease entity which occured in certain areas of West Germany and which was not diagnosed correctly in the past. At present, we are aware of 21 patients (pts) in 15 unrelated families with a total of 45 children. 11 pts died between 8 and 13 months of age, and 6 survived with liver cirrhosis. In the remaining 4 transaminases were initially elevated and turned to normal with correction of drinking water. All families lived in rural areas known for acid ground water. Drinking water was obtained from the own farm's wells with low pH (5.0 - 6.2) and was highly contaminated after passage through copper pipes. All pts were exposed to copper beginning shortly after birth; none of them was breast fed and all received milk formula prepared with the tap water. None of the 24 healthy siblings were exposed during the first three months of life. All pts developed a micronodular liver cirrhosis which resembled the "Indian Childhood Cirrhosis" and which was accompanied by significant copper accumulation in the liver. Common clinical symptoms included: enlargement of liver and spleen, anemia, bleeding disorders, muscular hypotonia. We believe that this disease might occur in other parts of the world if the environmental conditions are similar (acid drinking water and copper piping).

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EXTREMES OF MATERNAL AGE AND PARITY - A BIOLOGICAL OR ENVIRONMENTAL NEONATAL RISK FACTOR?

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We analyzed 15,000 consecutive births in an inner-city hospital. 51% of the mothers came from the nearby ultra-orthodox Jewish community of Mea-Shearim, unique in the stable socioeconomic status of its families, ensured by a family-size-linked social-support system. Two high risk groups, teenage primiparae (age $\leq$ 19 y) and grandmultiparae (parity $\geq$ 7), were studied. The incidence of low birth weight infants ($\leq$ 2500 g) was significantly lower ($p < 0.001$) for the teenage mothers from Mea-Shearim compared with teenage mothers from other areas of Jerusalem (12/190, 6.3% vs. 13/231, 14.7%). Similarly, low birth weight infants were significantly less common ($p<0.0002$) among grandmultiparae from Mea-Shearim compared with grandmultiparae from other areas of Jerusalem (56/1258, 4.5% vs. 55/616, 8.9%). The differences could not be explained by maternal age distribution, ethnicity, smoking and marital status. These results suggest that teenage and grandmultiparity, in a community with extensive social and economic support are not major biological risk factors.

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LONG-TERM FOLLOW-UP OF 203 CHILDREN WITH HODGKIN'S DISEASE /HD/ TREATED WITH MVPP AND INVOLVED-FIELD RADIOTHERAPY

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From 1971 to 1982, 203 children with HD /CS/PS I-IV/ were treated. Observation was ended June 30, 1988. The patients were under observation for at least six years. Complete remissions /CR/ were achieved in 95,6% children. Relapses occurred in 18,6% pts. Twenty pts died with the progression of HD and 9 due to complications. 150 pts remain the first CR / median, 114 months/ and without therapy/ median, 96 months/. The disease-free survival for all patients was 82,5% at 5 years and 80,1% at 10 years. Secondary malignancies / only solid tumors / developed in 3 pts and 2 of them died. Our experiences have prompted us to consider a new stage tailored protocol, probably generally less toxic and with increased efficacy especially in advanced HD.

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THE INFLUENCE OF LOW DOSES OF HEPARIN IN ARTERIAL CATHETERS ON THE COAGULATION OF PRETERM INFANTS.

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In preterm newborn infants little is known about the effect on the coagulation of low doses of heparin (2.5 IU/h) given through arterial catheters. In 13 infants with arterial catheters we studied venous values for activated partial thromboplastin time (APTT), thrombin time (TT), batroxobin time (BT), antithrombin III activity (ATIII) and factors I, II, V and VII, compared to those from 16 controls, on the third day of life. All infants were under 36 weeks of gestational age and weighed less than 2000 grams. Birthweight (BW), gestational age (GA) and coagulation parameters are shown (mean±SD).

	BW grams	GA wks	APTT sec	TT sec	BT sec
Study group	1390±300	30.2±1.3	54.0±19.5	20.8±8.2	23.4±10.6
Controls	1440±200	32.0±2.4	51.1±12.7	19.6±4.3	19.2± 2.7

	AT_{III} %	Factor I g/l	F_{II} %	F_V %	F_{VII} %
Study group	40.8±10.4	2.5±0.9	36± 9	77±21	49±18
Controls	46.6± 8.9	3.1±1.1	42±11	70±25	57±18

There was no significant difference in coagulation, weight or age. Conclusion: A dose of 2.5 IU/h of heparin does not affect coagulation of preterm infants.

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THROMBOEMBOLIC RISK IN CHILDREN WITH NEPHROTIC SYNDROME.

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We evaluated the "in vivo" activation of the hemostasis in 14 children (aged 4 to 13) with Nephrotic Syndrome (NS) at onset, determinating the blood platelet count (PLT, x 10^9/l), ß-thromboglobulin (ß-TG, ng/ml), platelet factor 4 (PF4, ng/ml), factor I (mg/dl), the inhibitors (AT-III -mg/dl- and PC:Ag -%-), and D-dimers (ng/ml).

	PLT	ß-TG	FP4	Fatt.I	AT-III/Ag	PC/Ag	D-dimers
NS (14)	375.8**	77.07***	31.4***	480.7***	22.6*	174.6***	80.8
	±11.1	±31.2	±8.8	±123	±6.8	±45.9	±45.6
C (10)	311.4	17.8	6.0	249.8	29.4	84.3	98.2
	±62.3	±6.4	±1.6	±64.9	±5.2	±15.5	±43.4

* $p < 0.02$ ** $p < 0.01$ *** $p < 0.001$ C = 10 healthy children of the same age

AT-III shows a positive correlation ($r = 0.80$; $p < 0.001$) with serum albumin; PC shows a negative correlation with serum albumin ($r = 0.40$; $p < 0.05$) and a positive one with urinary albumin ($r = 0.40$; $p < 0.05$). Thrombotic risk in Nephrotic Syndrome at onset does not seem to be caused by an inhibitor reduction for the increased PC levels; changes of platelet number and functions and of fibrinogen level assume more importance.

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ESR STUDY OF THE ERYTHROCYTE MEMBRANE SKELETAL PROTEIN NETWORK IN THE NEWBORN INFANTS

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Previously reported data obtained by electron spin resonance (ESR) assays demonstrated a lower availability of membrane sulfhydryl reacting groups in the erythrocytes of cord blood than in those of blood drawn after 4 days of life (Bracci et al. Pediatr Res 1988; 24:391). This finding was observed in red cells suspended in plasma. In the attempt to ascertain if the peculiarity of cord blood erythrocytes is due to a stable modification of the membrane structure or transitory interactions between membrane and plasma components, MAL-6 ESR assays were carried out in erythrocyte ghosts of cord blood and blood of 4 day old infants. The analysis of ESR spectra clearly demonstrate particular pattern at birth suggestive of significantly increased aggregation of spectrin in the cord blood red cells. After 4 days of life no significant differences between adult and infant spectra were observed. These important variations in the state of the erythrocyte membrane skeletal protein network may play a role on the changes of the rheological properties of newborn blood which have been reported to occur during the first days of life.