nemes. These defects were more numerous in the patients without bronchiectases. Our observations suggest that anomalies of the bronchial ciliary microtubular system may not only be congenital but also acquired, and could be an important factor explaining some cases of repeated airway infection and bronchiectases.

16 K. SCHOPFER*, B.J.WILKINSON* and K. BAERLOCHER. Children's Hospital, St. Gallen Switzerland and Department of Biological Sciences, Illinois State University, Illinois 61761 USA. Characterisation of antistaphylococcal IgE antibodies in patients with recurrent S. aureus infection, eczema and hyperimmunoglobulinemia E.

Antistaphylococcal IgE antibodies have recently been demonstrated in patients with recurrent staphylococcal infections, eczema and hyperimmunoglobulinemia E. The formation of these IgE antibodies may be a manifestation of an aberrant immunological response to S. aureus infection and may be related to the undue susceptibility of staphylococcal infections in these patients. In an attempt to further investigate this immunological disorder the IgE antibody binding site within the staphylococcal cell wall was studied. Cell walls from various staphyloccal strains were purified and IgE binding was measured using a solid phase radioimmunoassay. Binding occurred only to purified cell walls (PCW) from S. aureus. Since the glycan backbone and the peptide subunit of the peptidoglycan (PG) of the PCW tested are believed to be identical it is concluded that the IgE antibodies are directed to the interpeptide bridge or an unknown antigenic site within the PG of S. aureus cell walls. This hypothesis is further studied using synthetic analogues of PG interpeptide bridges.

17 R. SEGER*, P. JOLLER*, J. WUEST*, D. GARZONI*, A. RAPP* and G. DUC. Department of Paediatrics and Institute of Medical Microbiology, University of Zurich. Unmasking of the Thomsen-cryptantigen in necrotising enterocolitis (NEC): Bacteriological and immunological findings.

Clostridial infections have been associated with neonatal NEC. Neuraminidase, produced by Cl. perfringens and other clostridial species, cleaves neuraminic acid from the red blood cell glycoprotein and thus exposes a hidden receptor, the so-called Thomsen cryptantigen (T-Ag). Clostridial infection in NEC can thus be recognised by demonstration of the T-Ag on the patient's red blood cells. Among 22 cases with clinical signs of NEC 7 revealed T-antigen-positive erythrocytes. In one of those circulating neuraminidase was detectable for a short period. In 2 cases clostridial species were isolated from blood or peritoneal fluid cultures. The filtrates of these cultures were able to expose the T-Ag on normal red blood cells. Commercially available purified neuraminidase from Cl. perfringens unmasked the T-Ag on normal neutrophils and impaired function as measured by their killing of staphylococcus aureus. Our data indicate that screening of patients with NEC for the presence of T-Ag allows to delineate a subgroup of patients infected with neuraminidase-producing microorganisms. They also suggest that in this subgroups non-specific defense may be impaired.

18 F.LAURENTI*, R.FERRO*, G.ISACCHI*, F. MANDELLI*, G. BUCCI. Depts. of Pediatrics and Hematology-Univ. of Rome, Italy. Granulocyte transfusion in very premature infants with sepsis.

In premature infants with sepsis several defects of PMN function have been documented, suggesting that these infants can be equated to neutropenic patients. We, therefore, evaluated the effectiveness of PMN transfusion in 11 very premature newborn infants with sepsis (b.w.820-1200 g; g.a. 25-29 wks). The diagnosis of sepsis was established according to clinical signs and to blood picture, and was always confirmed by blood cultures. Transfusion of packed PMNs (0.5×10^{10} cells in 20 ml, obtained by leukafiltration of ABO/R h compatible blood) was given after sepsis was ascertained and repeated in the subsequent days. Group A (6 cases) received a mean of 1 unit per 2.2 symptomatic days, whereas Group B (5 cases) received a mean of 1 unit per 6 symptomatic days. The mortality rate was significantly lower in Group A than in Group B (17% vs. 100%; $p < 0.02$). No side effect was observed. The present results suggest that PMN transfusion may represent a substantial improvement in the treatment of these infants. Adverse effect of the treatment needs further evaluation, but at present possible risks seem to be reasonably low.
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19 L. CORBEEL, R. EECKELS*, G. VAN DEN BERGHE*, H. DEVLIEGER*, J. JAEKEN*, P. BRACKE* (Dept. of Paediatrics, University of Leuven), and L. HUE*, B. LEDERER*, T. de BARSY* (Dept. of Biochemistry, University of Louvain and I.C.P., Brussels, Belgium).
Clinical and biochemical findings before and after portacaval shunt (PCS) in type Ib glycogen disease (GSD).

An 11-year-old girl with statural age of 5 yrs. and hepatomegaly (15cm) had fasting hypoglycaemia, acidosis, increased serum cholesterol, triglycerides and uric acid, and increased liver glycogen (7.5%). There was no rise in blood glucose after i.v.galactose or fructose but glucagon gave a delayed response. Type Ib GSD was suggested by the normal activity of glucose-6-phosphatase (G-6-Pase) and of other glycogenolytic enzymes in frozen liver. After PCS, at 12 6/12 yrs., height increased by 29cm in 3 yrs. Serum cholesterol decreased from 620 to 230mg/dl and triglycerides from 2400 to 200mg/dl. At oral GTT peak values for glucose (mg/dl) and insulin (U/l) were respectively 210 and 50 before, 290 and 90 after PCS. Higher than normal utilisation of (2-^{3}H,U-^{14}C)-glucose was shown by a $^3H/^{14}C$ ratio of 24% after 60 min.(mean ± S.E.M.: 59% ± 7 in normals and 92% ± 3 in type Ia GSD) and can be explained by peripheral hyperinsulinism. Assay of G-6-Pase in a fresh liver homogenate prepared in 0.25 M-sucrose revealed only 29% (96% ± 11 in 3 controls) of the enzyme activity as compared with an homogenate in H_2O. The latter finding may be in favour of the hypothesis of Narisawa et al. (Biochem. Biophys. Res.Comm. 83: 1360, 1978) postulating a defect of the microsomal G-6-transport system in type Ib GSD, but is difficult to reconcile with the isotopic data.

20 M. ODIEVRE, H. BISMUTH*, P. DOUILLET*, N. MOATTI*, M. BRIVET* and C. BAUSSAN*. Unité de Recherche d'Hépatologie infantile, INSERM U 56. Clinique de Pédiatrie, Université Paris-Sud. Unité de Chirurgie Hépato-biliaire, Villejuif & Laboratoire Central de Biochimie, Hôpital de Bicêtre, 94270 Bicêtre, France. Portal diversion in glycogen storage disease (GSD): studies in 11 cases.

Successful treatment of GSD by portal diversion was reported with subsequent improvement in growth and return to normal dietary habits. An end-to-side portacaval shunt has been performed in 11 patients aged 7 months to 11 1/2 years (6 with type I and 5 with type III GSD). In 10 patients, patency of the anastomosis was confirmed. In 9 of them, a dramatic improvement of physical activity was observed in spite of persistent hypoglycemia and hyperlactacidemia. Liver size decreased in 7. A threefold decrease in blood cholesterol level ($\bar{m}$ 164 mg/dl vs. 415 peroperatively) was observed in 8 patients and a tenfold decrease in blood cholesterol level ($\bar{m}$ 170 mg/dl vs. 1548) in 6 out of 7 tested patients. Uricemia, increased in 4 cases, returned to normal. An increased linear growth rate ($\bar{m}$ 0.66 cm/month vs. 0.27) was noted in only 5 patients. Two type I patients died, 2 years (thrombosis of the anastomosis) and 6 months (intercurrent infection) after surgery. These results show that the most important benefits of surgery are improvement of general condition and normalization of lipids and uric acid metabolism.

21 FARRIAUX JP.*, RIBET M.*, DHONDT JL.*, ARDOUIN P.*,(Intr. by Corbeel L.)- Service de Génétique et Maladies Héréditaires du Métabolisme-LILLE (F) - *Portacaval shunt (PCS) in type IIa hypercholesterolemia; results from three cases.*
3 cases of type IIa hypercholesterolemia (homozygous LDL receptor-negative type) were treated with PCS. Follow-up is 46,23 and 3 months respectively (cases 1, 2, 3).
1) clinical and biological tolerances were good (no hepatic nor cerebral abnormality); cutaneous xanthomas slowly decreased.
2) cholesterolemia decreased (pre-operative levels: respectively 20.6 - 23.2 and 25.8 mmol/l; post-operative levels: 10.3, 15.5 and 12.9 mmol/l.
3) insulin and glucagon levels progressively increased (present basal insulin level: 20 µU/ml, glucagon: 500 pg/ml in case 1);results were normal in the 3rd case 2 months after shunt (insulin: 6 µU/ml, glucagon: 34 pg/ml).
4)in case 2, PCS thrombosed during the post-operative period; 18 months later a mesoiliac shunt was as efficient as PCS.
5) in cases 1 and 2 coronarography showed no change of coronary lesions, however; stabilization might be considered as favourable.
6) a diet low in lipids and cholesterol, inefficient before PCS became indispensable in the post-operative period. An increase of 30% of blood cholesterol occurred when the patient returned on a free diet. Continuous noctural gastric drip feeding (with a follow-up of 12 months in case 1) did not give better results than a well controlled diet.
7) a decrease of blood cholesterol during total parenteral feeding in the pre-operative period was a good index of the success of the PCS in the 3 cases.

22 Th.ANGERPOINTNER*,O.LINDERKAMP,H.STALLINGER*,J.STRUCK*, K.RIEGEL;Pediatric Surgical Hospital and Dept.of Neonatology of the Children's University Hospital,Munich
The effect of hypoxia on cardiac performance in newborn piglets-evaluation by the electromagnetic method.*
Tolerance of hypoxia differs among species and with age. Within age groups there is considerable individual variation which is as yet poorly understood.In response to hypoxemia cardio-circulatory adaptation plays a central role. We studied the changes of cardiac performance in artificially ventilated piglets aging 8 to 42 hours (n=1o) under fluothane-N_2O-O_2-anaesthesia. After a stable control period arterial Po_2 was lowered from 6o-9o mmHg to 3o-4o mmHg (hemoglobine-oxygen-saturation 7o-9o%) by lowering FiO_2 at otherwise constant conditions.
Mean heart rate rose continuously by + 38% at 111± 85 min hypoxia time (p o,oo1).However,cardiac output, stroke volume,cardiac power, mean aortic blood pressure, aortic peak flow reached their maxima (+11 to +32%;p o,o1),and vascular resistance its minimum (-17%; p o,o1) already during the first 3o min. They subsequently fell below (-6o to -9o%) or surmounted (+45%) control levels.These changes were accompanied by increasing metabolic acidosis.Average survival time was 188± 98 min. Survival time depended exclusively on the animal's ability to increase its heart rate (K=o,83).It was shorter in piglets who started with heart rates above the average control level (133± 61 min)than those with heart rates below it (227± 1o4 min).
It is to be concluded that newborn piglets are unable to compensate sustained hypoxemia of such degree by means of cardiac performance longer than 3o min.