but no O antibody response. Three doses of PTAP produced a normal Schick conversion and an antibody rise to between 0.01 and 0.1 A.U. per ml. Salk vaccine produced poliovirus neutralizing antibody titres to 1/16 or more, but with evidence of an impaired response.

In vitro lymphocyte transformation with PHA showed 20–25% blast cells at 3 days. No blast cell transformation occurred when the patient's lymphocytes were cultured in the presence of heat-killed pneumococci isolated from the CSF during an attack of meningitis two weeks previously: a control showed 74%. Chromosome analysis showed a normal female karyotype.

The tendency to recurrent infections associated with eczema, low serum IgM, low circulating lymphocytes, impaired delayed hypersensitivity reactions, and impaired response to polysaccharide antigens recalls the Wiskott-Aldrich syndrome (Cooper *et al.*, 1968; Blaese *et al.*, 1968). The immunological defect is identical, but the patient is a girl with low isohaemagglutinins and normal platelets.

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A Possible Case of Niemann-Pick Disease.

H. J. DEGENHART, H. E. ZOETHOUT, W. C. DE BRUIJN and G. J. M. HOOGHWINKEL (Rotterdam). A 5-month-old boy was admitted to hospital with mild cyanosis, early finger clubbing, dyspnoea, and tachypnoea. A chest x-ray showed increased hilar shadows and reticulation extending into the periphery of the lung fields. There was no calcification of the adrenals. During hospitalization liver and spleen gradually increased in size. Cherry-red spots were not present in the fundi. He showed moderate general mental retardation, with poor motor development. He died at the age of 18 months. Bone-marrow biopsies showed numerous large, foamy cells with a vacuolated cytoplasm. These vacuoles by electron microscopy appeared to consist of concentric lamellae, around an electron-lucent centre. In the lamellae the unit-membrane structure could be observed. Storage of sphingomyelins at these places is likely. Liver sphingomyelin was only twice normal and amounted to 19% of the phospholipids. Liver cholesterol was increased 2–3 times. Plasma and erythrocyte lipids were normal. In the bones a highly abnormal amount of liquid fat was found: 96% triglyceride, principally triolein, 3% cholesterol.

The clinical and electron microscope features were similar to those in Niemann-Pick disease. The chemical data suggest a new syndrome.

Generalized GM₁ Gangliosidosis: An Electron-microscopic Study of the Brain. H. ROELS (University of Ghent). Generalized gangliosidosis (Landing-

Norman disease) is an inborn error of metabolism characterized by the absence of the lysosomal enzyme β -galactosidase and storage of GM₁ ganglioside.

The electron microscopic study of a biopsy of the frontal lobe of a patient suffering from this disease shows characteristic lesions in the different cell types: cyto-membranous bodies in the nerve cells, membranous vesicular bodies in the oligodendroglia and astrocytes and pericytes, empty large vacuoles in the endothelium of the capillaries.

The intracellular localization of the ganglioside and the lysosomal origin of the different inclusions are discussed.

Neonatal Hepatitis. J. F. SALTET (Rotterdam). 5 cases of neonatal giant cell transformation are described. The first 4 are 2 pairs of sibs, one of the cases being anencephalic. The last case is the sixth child in a family, the other children being normal.

There is a morbid entity in the newborn with the following characteristic features: (1) familial appearance in sibs; (2) survival for a few days only; (3) prominent dyspnoea and cyanosis, with low blood sugar level; (4) haemolysis may be present; (5) jaundice is absent; (6) the placenta is enlarged, probably as a result of interference with blood flow in the cirrhotic liver.

At necropsy the following features are found. The liver is smaller than normal and has a rough surface. There are large cells in the liver (containing 20 to 40 nuclei) derived from the normal liver cells. There is striking iron storage in these giant cells and in other organs such as the pancreas, thyroid, renal tubule cells, and heart muscle cells, but not in the spleen. In most cases there is an increase of islet cell tissue in the pancreas.

Only in the proved cases of virus infection is it justified to speak of giant cell hepatitis. In other cases it is better to use the term giant cell transformation.

These changes have been referred to as neonatal haemochromatosis, but in view of recent work indicating a specific defect of gastroferrin in this condition, the term is better avoided when referring to giant cell transformation in the newborn, where the lesion may well be an enzyme defect in the liver.

Recent Progress in the Knowledge of Histiocytosis X. E. BASKET and C. NEZELOF (Paris). Tissues from 39 cases of histiocytosis X have been studied since 1965. They include 6 specimens of lung tissue, 4 skin biopsies, and 20 fragments of bone.

In each case, on electron microscopic study, characteristic granules were observed within the cytoplasm of the pathological cells. The granules appeared as rod-shaped profiles, 420 Å thick, composed of a central osmiophilic core and a double outer sheath. A transverse striation of 100 Å periodicity was seen in the central core.

After 6 days, tissue cultures of eosinophilic granuloma material showed the presence of large polykaryocytes. Rod-like profiles were observed as long as 3 months after the explantation.

Identical structures were seen within the cytoplasm of the dendritic clear cells of the epidermis; they were thought to be characteristic normal organelles of these cells.

Additional studies will have to be carried out to try to reconcile the presence of these structures in the Langerhans cells of normal epidermis and in the pathological cells of histiocytosis X.

Renal Changes in Henoch-Schönlein Purpura.

E. F. GLASGOW (The Children's Hospital, Birmingham). The light microscopical changes in Henoch-Schönlein purpura are variable, depending on the severity of the disease and the time of biopsy. Percutaneous biopsies from 9 children between the ages of 3 and 13 years were examined by both light and electron microscopy and the latter appearances were similarly variable.

In capillary loops which appeared normal with light microscopy there was some thickening of the basement membrane, with fusion of foot processes if proteinuria was present. In adjacent areas of focal sclerosis there was marked accumulation around the mesangial cells of basement membrane-type material which was continuous with the basement membrane of the loop. Scattered through this material were discrete stellate islands of mesangial cytoplasm which was indistinguish-

able from endothelial cytoplasm where they were contiguous.

In more severely affected glomeruli, the endothelial cells additionally demonstrated hyperplasia with increased amounts of cytoplasm and absence of fenestrations. Frequently endothelial cells situated at the mesangial pole showed an appearance suggestive of phagocytosis within their cytoplasm.

In addition to thickening, the basement membrane presented a scalloped appearance on its endothelial surface and it was not observed to be ruptured. Podocytes occasionally showed hyperplasia in addition to fusion of foot processes. In glomeruli showing more complete sclerosis, characteristic collagen fibrils were recognized in the urinary space and within the obliterated capillary loops. Such fibrils were not observed in isolated focal areas of sclerosis.

Demonstrations

Demonstrations included the following: chromosomal abnormalities, lipoidoses of bone-marrow, the histology of the thymus in stress and infection, autoimmune thyroiditis, hemilateral degeneration of the liver, interstitial emphysema in the early perinatal period, morbus Bournville, congenital toxoplasmosis, and normal and abnormal fetuses.