Burkitt's lymphoma and with infectious mononucleosis, was mentioned as a possible candidate virus.

Action of Vincristine Sulphate on Gliomas. G. Pearse and L. P. Lassman (General Hospital, Newcastle upon Tyne). Film.

Cell Culture Studies in Cystic Fibrosis. G. B. Reed, A. D. Bain, and W. M. McCrae (Royal Hospital for Sick Children, Edinburgh). Fibroblasts cultured from skin biopsies of children with cystic fibrosis and their parents were studied, using a variety of methods including a simple histochemical stain for metachromasia, toluidine blue O. With Eagle's medium supplemented with pooled human serum, the degree of cellular metachromasia (per 1000 cells surveyed) found in parents and controls was similar, but in affected children there was a greater degree of metachromasia. The degree of metachromasia in the cultured cell was compared with the chemical content of acid mucopolysaccharide and with the in vitro incorporation of $^{35}S$.

Up to the present it has not been possible to relate the degree of metachromasia to cell chemical content nor to the isotope uptake.

Preliminary studies of cultured histiocytes derived from the buffy coat of peripheral blood specimens of controls, parents, and affected patients have been carried out. Despite in vitro conditions similar to those employed for fibroblast cultures, it has not been possible to distinguish at present, by metachromatic staining, any significant difference between controls, parents, or affected patients.

Further studies are necessary in order to understand the mechanisms leading to cytoplasmic metachromasia in cultured cells and in order to investigate the relation of this metachromasia to a particular disease, in this case cystic fibrosis of the pancreas.

Wolman's Disease. A. J. Barson (Department of Pathology, University of Manchester). A case of Wolman's disease was described in a female infant who died at 7 weeks of age in the Hospital for Sick Children, Toronto. This is a rare familial disorder of lipid metabolism in which there is an accumulation of esterified cholesterol in many abdominal organs, especially the small intestine. Characteristically both adrenal glands are calcified.

This infant presented at 1 week of age with persistent vomiting and abdominal distension. A laparotomy performed at 2 weeks of age failed to show a mechanical obstruction, but the diagnosis was made from tissue biopsied at this time and confirmed later at necropsy.

For the first time in this disease large quantities of ceroid were demonstrated in the liver, spleen, adrenals, lymph nodes, and particularly within the lamina propria of the small intestine. It is postulated that the ceroid resulted from the oxidation of the accumulated cholesterol esters. Because ceroid is so inert, it acts as an impermeable seal progressively impairing intestinal absorption.

Accessory Lungs with Foregut Connexion. A. A. M. Gibson (Yorkhill Children's Hospital, Glasgow). A case was reported of a male baby with bilateral accessory lungs who died at the age of 7 weeks from infection. The accessory lungs were situated symmetrically in the posterior thorax on either side of the midline just above the diaphragm. Each lung was single-lobed and lay in a separate pleural cavity. A fistula resembling a normal main bronchus connected the lungs with the posterior wall of the stomach. The blood supply to the lungs was from an accessory artery arising from the abdominal aorta, and the venous return was to the portal vein. There was also a congenital diaphragmatic hernia on the left side.

Osmiophilic Inclusions in the Human Lung in the Perinatal Period. G. Gandy and W. Jacobson (Addenbrooke's Hospital, Cambridge). Published in Archives of Disease in Childhood, under the title 'Hyaline Membrane Disease' (1970, 45, 289).

Oxygen-induced Bronchiolar Changes in Infancy.* R. C. Rosan, T. C. Durbridge, M. M. Bieber, and M. C. Cogan (Department of Pathology, Stanford Medical School, California, U.S.A.). 80–100% oxygen at atmospheric pressure causes similar sequential changes in the bronchioles of infant humans, guinea-pigs, mice, and young adult rats. The earliest cellular changes are reactive. Within a few days, there is necrosis. Metaplasia appears in 1–2 weeks, and later there is hyperplasia. This chemical bronchiolitis results in irregular aeration, lobular distortion, and finally lobular emphysema in survivors of more than 2 weeks. In such survivors, histological and clinical evidence of pulmonary hypertension is the rule. It is feasible to follow the progress of the disease in humans by exfoliative cytology, in which case increasing abnormality of cells is the rule. Increasing difficulty with secretions is also common, due both to loss of an effective sheet of respiratory mucosa and to increased secretion of viscous mucus. The latter can be documented by electronephoresis.

The development of a metaplastic bronchiolar mucosa is a prominent part of late adaptation to chronic oxygen intoxication. These cells are characteristically rich in ribosomes. Even as early as 4 days it can be shown histologically, autoradiographically, and by electron microscopy that there are many ribosomes in surviving mucosal cells. Our data show conclusively that early oxygen toxicity does not decrease net ribosome synthesis in homogenates of whole lung examined on sucrose gradients. On the contrary, it appears that a period of ribosomal synthesis precedes the development of metaplasia by several days. When these 'early' ribosomes are examined by molecular techniques, we find that their protein structure is anomalous and that their response to stimulation by polyuridine and polyadenine is erratically defective. The data are the same for infant humans and guinea-pigs. These features suggest that ribosomes may play a hitherto unsuspected role in the regulation of

*Supported by Grant 646 of the Council for Tobacco Research, U.S.A.
Cell culture studies in cystic fibrosis.

G B Reed, A D Bain and W M McCrae

Arch Dis Child 1970 45: 710
doi: 10.1136/adc.45.243.710-a

Updated information and services can be found at:
http://adc.bmj.com/content/45/243/710.1.citation

Email alerting service

These include:
Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

Notes

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/