but collectively they are significant and hence appropriate investigations should be considered. It is particularly important to consider IMD where more than one case of hydrops occurs in the same family, as this is a history of spontaneous abortion or unexplained deaths in close family members. We would like to describe our protocol for the investigation of fetal and neonatal hydrops for IMD as we have considered when the more common causes of hydrops have been excluded.

From the dead fetus a skin biopsy can be taken for HLA typing, if this should be requested as soon as possible. The cultured cells can be assayed for β-galactosidase (GM1 gangliosidosis, Morquio’s disease B, galactosialidosis), β-glucuronidase (Sly’s disease), β-galactosidase (Gaucher’s disease), α-neuraminidase (mucolipidoses I, II, III, and IVa), N-acetylgalactosamine-6-sulphate sulphatase (Morquio’s disease type A), sialic acid (sialic aciduria) and cholesterol esterification (Niemann-Pick C). DNA from the cultured cells can be investigated for expanded trinucleotide repeat sequence found in myotonic dystrophy.

In cases where hydrops has been diagnosed antenatally antenatal amniotic fluid can be tested for I-cell disease and cultured amniocytes may be examined. Other assays can be undertaken on cultured amniotic fluid cells or chorionic villus tissue (direct or cultured). In the live neonate investigations which can be performed on blood include enzymatic levels, plasma lyso- somal enzymes, plasma carnitine, iron, and ferritin (neonatal haemochromatosis). A skin biopsy may be preferable if a large number of lysosomal enzyme assays are to be assayed. A random urine sample may be examined for galactosialidosis and α-glucosidase deficiency. 

Do maternal β-sympathomimetics influence the development of retinopathy in the premature infant?

Editor.—Recent studies suggest that a number of factors other than the injudicious delivery of oxygen contribute to the development of retinopathy in the premature infant.1-2 One hundred and fifty infants on our neonatal unit who required oxygen treatment for more than 60 days were reviewed retrospectively in order to determine the range of factors contributing to this pathology.

All infants requiring oxygen for more than two weeks were examined at two weekly intervals by an ophthalmic surgeon (ES). Fourteen cases of retinopathy greater than stage 3 were identified. These patients were staged for gestation (±1 week), birth weight (±50 g), sex, and ethnic group with infants who had oxygen requirements for more than 60 days, but did not have retinopathy. The two sets of cases showed no significant differences in their Apgar scores, requirement for exchange transfusions, or the incidence of clinically significant patent ductus arteriosus.

Mothers of eight of the retinopathy group had received infusions of β-sympathomimetics (seven ritodrine, one salbutamol) to arrest prematurity labour: only one of the non-retinopathy group received ritodrine (p<0.008). This observation may be of physiological importance, as animal experiments have suggested that the retinal circulation is strongly influenced by these pharmacological agents acting jointly through the sympathetic nervous system and endothelial factors.3-4 β-Sympathomimetics are known to induce transient ischaemic changes in the myocardium of the newborn if given over long periods by intravenous infusion.4-5 Maternal ritodrine has been associated with reduced blood pressure in the newborn.5-6 In the premature infant in whom antenatal retinal perfusion is influenced by β-sympathomimetics, these drugs may predispose to retinopathy. This observation merits review in a larger series of infants. Further, it casts doubt as to the safety of β-sympathomimetics which are widely used in the therapy of prematurity labour with little evidence to support their efficacy.