

PS155 BACTERIOPHAGE ASSAY FOR GALACTOSE AND GALACTOSE-1-PHOSPHATE IN NEONATAL SCREENING FOR GALACTOSEMIA IN TAIWAN

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The most frequently used screening test for galactosemia is Beutler's fluorescent spot test for galactose-1-phosphate (Gal-1-P) uridyl transferase and glucose-6-phosphate dehydrogenase (G6PD) activity. Owing to the high incidence of G6PD deficiency and the warm climate, Beutler's test is not suitable for neonatal screening in Taiwan. We have adopted, for the purpose of screening for galactosemia, the Paigen bacteriophage assay as modified by Yoshida for measuring the total amount of galactose (Gal) and Gal-1-P in dried blood spots. From January 1984 to June 1987, 90,062 neonatal blood spot samples collected from all over Taiwan were screened. There were 356 (0.3%) positive cases (>6mg/dL). One of them (>20mg/dL) was confirmed as galactosemia. Using enzymatic methods, the Gal and Gal-1-P in the blood spot sample were found at levels of 58.6 and 67.8 mg/dL, respectively. The case had been exchange-transfused twice due to hyperbilirubinemia and was in the neonatal ICU. The infant was 10 days old when we reported the result by telephone. After the milk was changed to galactose-free formula, the infant recovered dramatically. Subsequently the erythrocyte transferase activity was found to be 0.04 mU/gHb (reference: 290-470 mU/gHb). The case is 2.5 years old at the present time and is developing normally.

PS156 INCREASED METHAEMOGLOBIN LEVELS IN NEONATAL HYPERBILIRUBINAEMIA (NHB)

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Neonatal hyperbilirubinaemia is reported to be more common in prematurely born infants. It is considered to be due to an immature hepatoexcretory function, decreased UDP-glucuronyl transferase activity or due to increased beta-glucuronidase activity. In such babies the development of lung antioxidant enzyme system is reported to be deficient, and the babies may also have vitamin E deficiency. In 56 cases of NHB we observed increased levels of methaemoglobin ($4.0 \pm 0.5\%$ compared to $1.5 \pm 0.25\%$ in normal babies). The levels of vitamin E, superoxide dismutase and glutathione peroxidase activities were comparatively lower than the normal in the cases studied. A decrease in superoxide dismutase may result in methaemoglobinemia leading to NHB.