New Zealand Paediatric Surveillance Unit
PROLONGED INFANTILE CHOLESTASIS

Aim: **To determine the incidence and underlying causes of prolonged infantile cholestasis**

Backgound:
Infantile cholestasis is an uncommon condition resulting from many causes. There is currently no information on the incidence and aetiology in New Zealand. A British study found the incidence of biliary atresia to be 1/16,700 births. A French study (that included French Polynesia) suggested a higher incidence in French Polynesia. A retrospective review of biliary atresia presenting to Princess Mary Hospital in Auckland between 1978 and 1986, demonstrated a 10 fold higher incidence in Polynesians compared to Caucasians.

International studies performed in Australia, Sweden and United Kingdom found biliary atresia the most common cause of infantile cholestasis, and was present in 20-35% of reported cases. Other well known causes are infections, Alagille Syndrome, Trisomy 21 and Total Parenteral Nutrition. There are some data suggesting the frequency of metabolic diseases as a cause of infantile cholestasis has increased.

The prognosis and outcome of infants with cholestatic jaundice varies with the underlying diagnosis. A proportion of infants will progress to have chronic liver disease irrespective of intervention while others will have resolved disease with no long-term complications. Paediatric orthotopic liver transplantation has been available in New Zealand for the last year and it is very likely that infants with chronic liver disease will eventually require liver transplantation as a life saving procedure. The timing for transplantation itself varies significantly, between the first year of life to adulthood dependent on the underlying cause and the tempo of deterioration in liver synthetic function. Follow-up data will assist in predicting the number of children requiring transplantation in New Zealand.

There has been neither previous attempt to document the incidence of infantile cholestasis nor any of its other underlying causes as a prospective national study in New Zealand.

Case Definition and Reporting Instructions

Jaundice in infants with liver disease aged 2 weeks post term to 6 months (inclusive) and present for at least 2 weeks, with the conjugated bilirubin fraction at least 20% or higher of total bilirubin. This may be accompanied by pale stools and dark urine.

Follow-up of positive returns:

A short questionnaire requesting demographic and clinical details will be forwarded to respondents reporting a case. Further information will be requested at 1, 2 & 5 years to determine long-term outcome.

Investigators:

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