Australian Paediatric Surveillance Unit

PROTOCOL SHEET - CONGENITAL & IDIOPATHIC NEPHROTIC SYNDROME (from July 1998)

Background to study

Nephrotic syndrome occurs secondary to a number of glomerular diseases with variable prognoses. Despite being a common manifestation of glomerular disease in children, significant questions concerning the incidence, aetiology, associated morbidity and treatment of nephrotic syndrome remain unanswered. This study aims to determine the incidence of nephrotic syndrome in Australian children. Of the overseas incidence studies, only one performed in the USA in the 1950s was population based. This indicated an annual incidence in children aged below 16 years of 2 per 100,000. This and British studies suggested a higher incidence in African-American children, children from the Indian subcontinent and children from lower socio-economic backgrounds. Our study also aims to describe current management regimes, disease relapse rates and the spectrum of infectious and thrombotic complications in children with nephrotic syndrome. Information on the rates of adverse events will be used in future to determine the feasibility of randomised controlled trials of different treatment modalities and/or interventions to prevent complications of the disease.

Objectives:

Idiopathic nephrotic syndrome

1. To estimate the incidence of idiopathic nephrotic syndrome
2. To describe its distribution in relation to age, sex, socio-economic status, geography and ethnicity
3. To describe the steroid regimes and other treatments used in the first episode of idiopathic nephrotic syndrome
4. To describe disease relapse rates amongst steroid responsive children
5. To describe the frequency and type of infective and thrombotic complications

Congenital nephrotic syndrome

1. To estimate the incidence of congenital nephrotic syndrome in Australia
2. To describe its distribution in relation to age, sex, socio-economic status, geography and ethnicity
3. To determine the presentation, management and short-term outcome of children with congenital nephrotic syndrome

CASE DEFINITION AND REPORTING INSTRUCTIONS

Please report any new case seen in the previous month and fulfilling the following definition:

Idiopathic nephrotic syndrome

Any child aged >3 months and <15 years with oedema, proteinuria (•3+ on dipstick), hypoalbuminaemia (serum albumin <25g/L) and normal renal function (serum creatinine in normal range for age when not volume depleted) in the absence of persistent hypertension, systemic illness or macroscopic haematuria.

Congenital Nephrotic Syndrome

Any child aged •3 months with oedema, proteinuria (•3+ on dipstick) and hypoalbuminaemia (serum albumin <25g/L).

Follow up of notifications
A questionnaire requesting further details will be forwarded to practitioners who report a case of idiopathic or congenital nephrotic syndrome. **A copy of the questionnaire is enclosed for your information.**

A second questionnaire will be send one year after diagnosis of nephrotic syndrome requesting details of response to therapy, infectious and thrombotic complications and one year outcome.

Investigators
Dr Elisabeth Hodson (Chief), Dr Jonathan Craig and Mrs Narelle Willis at the Centre for Kidney Research, New Children's Hospital (Royal Alexandra for Children), Sydney in collaboration with the Australian Members of the Australia and New Zealand Paediatric Nephrology Association

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