In the Name of GOD

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Research Affairs

Disease Registry Unit

REGISTRY PROPOSAL: (1-2 PAGES excluding references)

A. Brief Intro to Registry –Literature Review (1-2 paragraphs)

Idiopathic nephrotic syndrome is the most common etiology for this disease in children and is characterized by massive proteinuria, hypoalbuminemia, hyperlipidemia and edema. The response to steroid is of a great value from prognosis point of view in comparison with pathologic pattern. The incidence of idiopathic nephrotic syndrome is varied by age, gender and geographic region. In Europe and America, the incidence of nephrotic syndrome is 1-3 in 100000, and its cumulative prevalence is 16 in 100000 in children under 16 years of age. Geographic and racial differences are understood and in Asians the incidence is 6 times more than average which indicates genetic and environmental factors in pathogenesis of the disease. Familial cases, especially among steroid-resistant cases are well-known and are usually occurred between the age of 2 to 7 and is more frequent in boys. Complications of this disease are acute kidney injury, infections, thrombosis, growth disorders and chronic kidney disease.

B. Registry Objectives (what is it you are specifically looking at, trying to reach?)

Considering the importance of this disease, there are a lot of different registries around the world that collect the demographic data, etiology and prognosis of patients with nephrotic syndrome especially the congenital and steroid-resistant cases. Podnote registry collects data of 98pediatric nephrology centers, the data considering renal and extra renal manifestations, histopathologic, genetic and familial history are collected and are followed in a prospective manner. European reference network for rare kidney diseases is a concersium consisting of 38 pediatric and adult nephrology centers in European countries that in addition to a lot of other kidney diseases, SRNS(steroide-resistant nephrotic syndrome) is also registered. In Iran, there aren't any registry about this disease but there are different centers working on nephrotic syndrome that articles from Dr. Safaee and Dr. Hooman can be pointed out as their outcome.

C. Registry Design (participants, data collection, statistical analysis plan, etc.)

Therefor pediatric nephrology research center decided to run the registry for nephrotic syndrome. In this registry, all the patients are diagnosed by nephrotic syndrome from neonatal to 18 years of age that refer to mofid hospital are collected using hospital files and HIS system. A questionnaire which is designed beforehand is filled and patients are followed by having remission, relapse, complications and kidney injury. In the case of no referral to hospital after registration, patients are monitored by calls every six months and are pointed out in case of relapse, non-responsiveness to steroids or complications of treatment. This data is entered in a software and ultimately, data analysis by an epidemiologist will be done.

D. Timeline for your Registry project (project deadlines set by you and your mentor)

The duration of registry for evaluation of early complications is about two year

E. Who will provide support and feedback and how often will this occur?

In the future this project will be installed in other children hospitals of Tehran and will spread by cooperation of Iranian society of pediatric nephrology. Right now, this registry is structured as two GP thesises and after preparing preliminary data, will be continued as a prospective research every six months.