The Sky's the Limit

29 May - 1 June 2008, Sun City

Allergy Society of South Africa (ALLSA)

PRIMARY IMMUNODEFICIENCY IN SOUTH AFRICA – LESSONS FROM A REGISTRY

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Primary immunodeficiencies (PIDs) are rare diseases caused by inherited defects. Established prevalence figures for European, American and Australasian countries are available on electronic databases. No such figures exist for South Africa or for the majority of African countries. With the clinical burden of secondary immunodeficiencies due to HIV and malnutrition these potentially treatable deficiencies are easily missed. General awareness is poor with suboptimal outcome for many. Routine molecular diagnosis is not available in SA, but previous studies have highlighted novel mutations of BTK deletions in agammaglobulinaemia and clustering of complement C6 deficiencies.

Data of over sixty live consenting patients were recorded at the PID Registry, Tygerberg Hospital since November 2006. They reflect a wide spectrum of immunodeficiencies from all over South Africa. Age at diagnosis varies from 3 to 727 months, with delayed diagnosis for common variable immunodeficiency especially. Male patients predominate as expected with X-linked inheritance pattern of the more common antibody deficiency states. Only one black patient is recorded and the majority of patients reside in the Western Cape with sample bias to an area with PID clinic services. Interesting subgroups of hereditary fevers, disseminated warts and ectodermal dysplasias were included. A separate group of 45 complement C6 deficiency patients is known to the Allergy Institute of the University of Cape Town where a cohort with hereditary angioedema is also followed up.

Over sixty percent of patients are on immunoglobulin replacement therapy, which is frequently haphazard and insuffiently monitored. Resultant infectious morbidity such as bronchiectasis, deafness, growth failure and developmental delay are frequently observed.

Parental advocacy and level of informed involvement in childhood PID are crucial predictors to outcome in countries with low level of awareness. With data collection the registry aims at networking between doctors, healthcare workers and patients for earlier diagnosis and improved outcome.

PENICILLIN ALLERGY IN CHILDREN – OFTEN MISDIAGNOSED?

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Background: Penicillin antibiotics are commonly implicated in allergic reactions in children without tests being performed to confirm or refute allergy. Many children are inappropriately labelled penicillin allergic and are treated with alternative antibiotics that may be less effective and more expensive.

Objective: The aim of this retrospective study was to determine the prevalence of true penicillin allergy in children with self-reported allergy.

Methods: Clinical and laboratory data of children referred to the Allergy Clinic at Red Cross Hospital for evaluation of suspected penicillin allergy between July 2002 and June 2007 were analyzed. *Clinical data* included sex, age at first reaction, co-existing atopy and nature of adverse reaction. *Tests* included CAP-RAST® for penicillin V, penicillin G, ampicillin, and amoxil, skin prick tests (SPT) as well as penicillin challenge test. The time interval between adverse drug reaction and evaluation was recorded.

Results: Data of twenty subjects were analyzed. Penicillin allergy was confirmed in 5/20 (25%) subjects. Four were SPT +ve and 2 CAP-RAST +ve. The median age at reaction was 2 years; all were atopic, all presented with urticaria ± angioedema. The median time interval from reaction to evaluation was 2 months.

Penicillin allergy was excluded in 15/20 (75%) subjects. CAP-RAST, SPT and challenge tests were negative in all patients. Median age at reaction was 2,5 years. 3/15 were atopic; 6/15 presented with a maculopular rash, 6/15 with urticaria ± angioedema and 3/15 with an unidentified rash. The median time interval from reaction to testing was 20 months. All non-allergic patients subsequently received penicillin without adverse events.

Conclusions: Penicillin hypersensitivity is relatively uncommon in children. SPT and challenge testing is required to confirm or refute the diagnosis. Accurate diagnosis avoids the morbidity, mortality and economic cost associated with unnecessary withholding of penicillin therapy.

EFFECT OF DESLORATADINE ON EXERCISE-INDUCED BRONCHOCONSTRICTION

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Background: Exercise-induced bronchoconstriction (EIB) is a significant problem in asthmatic patients. The link between allergic rhinitis and asthma is now well established. Patients with allergic rhinitis may have EIB.

Objective: This study compared the effects of desloratadine and placebo on EIB in a group of patients with allergic rhinitis and EIB.

Methods: This was a double blind placebo controlled, randomized, crossover study. Exercise challenge tests were performed before and after 7 days of treatment with either 5 mg desloratadine or placebo. Patients then underwent a washout period for 7 days and were crossed over to receive either 5 mg desloratadine or placebo. The exercise challenge tests were repeated.

Results: Desloratadine had no effect on the reduction in percentage fall in FEV_1, the AUC (0-60min) and the time to recovery.

Conclusions: Desloratadine had a no effect in attenuating the bronchoconstriction caused by exercise in patients with allergic rhinitis and exercise induced bronchoconstriction.

ATOPY IN HIV-INFECTED AND NON-INFECTED CHILDREN IN PRETORIA, SOUTH AFRICA

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Introduction: The relationship between the development or aggravation of a pre-existing atopic state and HIV has not been thoroughly investigated in the South African context. HIV-positive patients have been shown to have a higher prevalence of atopy in some international studies in the early stage of their disease but this has not been documented in children.

Methodology: A prospective convenience sample study of children aged 3 months to 12 years attending the HIV clinic were recruited into the study. Information regarding the child's personal and family history of atopy was recorded. The WHO HIV clinical staging, CD4 counts were recorded. An age and sex-matched control group of healthy children attending routine follow up at the cardiology and the neurology clinic were included. Spin prick tests (SPT) for common aeroallergens were conducted in all patients.

Results: A total of 100 patients were included in the study with 50 in each arm. 10% of the HIV-infected patients in comparison to 16% of controls had positive SPT for aeroallergens. Of the HIV-infected patients a high number of patients had allergic rhinitis and eczema (60% and 68% respectively). There is a lack of correlation between CD4 count and any SPT positivity (r=0.011), CD4 count and presence of reported asthma (r=-0.020), and CD4 count and reported presence of dermatitis (r=-0.06). CD4 count was not statistically different between children with and without family history of atopy p=0.68.

Only the oral presentations at 'The Sky's the Limit' appear in SAJCH, *owing to space constraints.*