



#### Christmas Greetings

On behalf of the APSU Board, study investigators and our research collaborators, the APSU team wish you a happy and safe holiday period.

# APSU Update

Australian Paediatric Surveillance Unit  
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## December 2010

### Surveillance Studies

#### Concluding Study

##### Acute Rheumatic Fever

Thank you to all who reported cases for this study. Surveillance will conclude with the December 2010 card. To date, 141 cases have been confirmed. Please report any outstanding cases to the APSU and return all outstanding questionnaires to Sara Noonan, Menzies School of Health Research, C/- 8 Denham Drive, Valley View, SA, 5093. Ph: 08 8263 7801.

#### New studies to commence in 2011

##### Juvenile onset Recurrent Respiratory

**Papillomatosis (JoRRP)** – led by Dr Dan Novakovic and Dr Julia Brotherton. Juvenile onset Recurrent Respiratory Papillomatosis (JoRRP) is a condition in which benign papillomata develop and recur in the larynx in children. JoRRP usually develops in infancy or early childhood: the median age at diagnosis has been reported as four years.

The proposed APSU study will be the first prospective study of JoRRP worldwide. It will provide an estimate of incidence and demographics, age of onset at diagnosis and duration and nature of treatment as well as data on the mode of delivery, maternal history and HPV infection and vaccination status. HPV sequencing and genetic subtyping of lesions will inform future vaccine development. These data are likely to support further research into the biological, maternal and obstetric aspects of JoRRP.

##### Neonatal Herpes Simplex Virus Infection (HSV)

led by Prof Cheryl Jones. Completing 13 years of active surveillance through the APSU, has documented the spectrum of neonatal HSV disease in Australia from 1997 to 2009. The past surveillance study design did not provide opportunity to characterise morbidity in survivors or to document the frequency or management of recurrences of HSV infection. In addition, the international guidelines for treatment have changed and now recommend larger doses of antiviral (parenteral acyclovir) therapy for longer duration to limit progression of neonatal HSV disease in disseminated infection and to reduce the likelihood of early neurological recurrence. Thus, important trends and significant knowledge gaps in the epidemiology, management and outcome of HSV infection in the newborn period have emerged from the past surveillance study that require follow up.

The HSV protocol is being reviewed to enable collection of data on recurrent infections and outcomes through a follow-up questionnaire. This new knowledge will better inform clinical practice guidelines, and provide indirect evidence of efficacy of management and diagnostic changes.

#### One time survey

##### Croopyrin-Associated Periodic Syndromes (CAPS) -

led by Dr Sam Mehr. CAPS are extremely rare, potentially life threatening auto-inflammatory disorders. Three separate CAPS are recognised: Familial Cold-Auto-Inflammatory Syndrome (FCAS), Muckle Wells Syndrome (MWS) and Neonatal Onset Multi-Inflammatory Disorder (NOMID). The conditions represent a continuum of disease, with FCAS being the mildest and NOMID being the most severe.

The disorders are all associated with mutations of the NLRP3 gene (also known as CIAS1). NLRP3 encodes cryopyrin, a protein of the inter-interleukin-1 (IL-1) inflammasome involved in IL-1 $\beta$  production. NLRP3 mutations are thought to induce excessive IL-1 $\beta$  production and systemic inflammation follows. However in only 50-60% of patients a mutation is detected, suggesting genetic heterogeneity.

Delays in diagnosis may result in significant morbidity, including destructive arthritis, aseptic meningitis, deafness and neurological delay. Early recognition and treatment of CAPS is therefore paramount to prevent permanent joint, hearing and brain injury.

As CAPS are extremely rare, the study investigators aim to determine the current prevalence of CAPS in Australia. This will be a one-time survey and a **special one-off CAPS Report Card** will be sent separately to the usual APSU card. Clinicians will be asked to report any child with CAPS currently under their care, rather than newly diagnosed cases as is APSU routine.

#### INoPSU update

In October the APSU attended the 6<sup>th</sup> meeting of the International Network of Paediatric Surveillance Units (INoPSU) in Dublin. Fifteen representatives from eight different INoPSU units were present. Countries represented included; UK, Ireland, Canada, Australia, Netherlands, Switzerland and Portugal. Unfortunately,

**The APSU office will be closed from December 20 to January 3, 2011.**

Thanks to our Funders and Supporters. We wish to acknowledge the continued support of the APSU by:

Faculty of Medicine, University of Sydney; Australian Government Department of Health and Ageing; NHMRC Enabling Grant 402784; Division of Paediatrics and Child Health of the Royal Australasian College of Physicians; The Children's Hospital at Westmead

representatives from New Zealand, Wales, Greece, Germany and Latvia were unable to attend. The meeting provided an excellent opportunity for exchange of views on rare paediatric disease surveillance and to discuss issues that currently pose challenges to the units. Funding and the increasing need for confidentiality were a particular focus for discussion.

Yvonne Zurynski presented APSU data on severe complications of influenza.

Danielle Grenier from Canada and Yvonne Zurynski were elected as the new INoPSU co-chairs for the next three years. They will be replacing Daniel Virella from the Portuguese Unit.

For more details of the work of INoPSU visit [www.inopsu.com](http://www.inopsu.com) or email [bpsu@rcpch.ac.uk](mailto:bpsu@rcpch.ac.uk)



## Reporting Matters

### Keep a record of your cases

Please remember to fill out the yellow record sheet at the front of your 'blue folder' when notifying cases. This will be a useful prompt about which child you reported when you are filling out the questionnaire on the case. Misplaced your blue APSU folder? Let us know and we will send you a new one.

**We need timely data** - Please try to report cases for the month when they are seen. Don't forget that all questionnaires and protocols can be downloaded from the APSU website at [www.apsu.org.au](http://www.apsu.org.au). Please feel free to do so and complete the questionnaire as you see cases.

### Email or Snail Mail?

Did you know that 81% of our contributors are reporting by email? We like this as it saves our resources and makes it easier for you to report and keep in contact. Want to change from **reply paid mail to e-mail**? Yes? Then please contact us on 02 9845 3005 or email us at [apsu@chw.edu.au](mailto:apsu@chw.edu.au).

### \*\*\*\*New for APSU: On-line Reporting System\*\*\*

In the last evaluation of the APSU, paediatricians indicated that they would like to have an online reporting system. In response to this APSU has collaborated with the University of Sydney-ICT Services to develop an online reporting system. Many thanks to those who attended the

preliminary testing session held at CHW in November. Development of Phase 1 – automation of the monthly report card - is now completed. This will allow clinicians to record their monthly response of nothing to report or cases to report directly onto the web form. Additional features will allow direct update of contact details and viewing of reporting history. Phase 1 will be piloted starting with the January 2011 card.

A random group of 200 email contributors will be selected to facilitate this. Online questionnaires will be developed next year as part of phase 2.

Please contact our database manager, Nicole McKay on 02 9845 3007 or [nicolem8@chw.edu.au](mailto:nicolem8@chw.edu.au) for further information.

## APSU Biennial Research Report

The APSU Biennial Research Report (2007-2008) is available on the APSU website [www.apsu.org.au](http://www.apsu.org.au). We have a limited number of hard copies which can be requested by e-mailing [apsu@chw.edu.au](mailto:apsu@chw.edu.au) or please call 02 9845 3005.

## Recent Publications:

Blyth C, Best E, Jones C, Nourse C, Goldwater P, Daley A, Burgner D, Henry G, Palasanthiran P. Nontuberculous mycobacterial infection in children: a prospective national study. *Pediatric Infectious Disease Journal* 2009; 28(9):801-805.

He S, Zurynski Y, Elliott E. What do paediatricians think of the Australian Paediatric Surveillance Unit? *Journal of Paediatrics and Child Health* 2010; 46: 412-418.

Jaffe A, Zurynski Y, Beville L, Elliott E. Call for a national plan for rare diseases. *Journal of Paediatrics and Child Health* 2010; 46: 2-4.

Khandaker GM, Khandaker GM, Zurynski Y, Elliott E, Booy R. Mania in a patient with H1N1: Oseltamivir - the culprit or red herring? In reply: *American Journal of Psychiatry* 2010; 167(9):1129-1130.

Zurynski Y, Davey E, Elliott EJ. Australian Paediatric Surveillance Unit Annual Report, 2008 and 2009. *Communicable Diseases Intelligence* 2010; 34:285-289.

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