



rare Kids

Australian Paediatric Surveillance Unit **NEWSLETTER** December 2017



Inside this issue:

Surveillance commencing in 2018 2

- Congenital Syphilis

AFP Reminder

Study updates

- JoRRP
- Vitamin K

• Microcephaly 3

2018 marks 25 Years of Surveillance for APSU

Congratulations 4

Fond farewells and a Welcome

In the News

Key publications 5

Upcoming Events 6

Christmas Wishes

Looking ahead

Surveillance Studies Commencing in 2018

- Congenital Syphilis

Surveillance Studies under development

- Hypophosphatasia (HPP)
- Trampoline Injuries

APSU Scientific Review Panel

2017 has been a busy year for the APSU Scientific Review Panel—we would like to extend a special thank you to Dr Fiona Mackie, Professor Bin Jalaludin, Professor Carol Bower, Dr David Burgner, Dr Mavis Duncanson, A/Professor Yvonne Zurynski and Professor Elizabeth Elliott for supporting the activities of the APSU.

Joining the SRP in September was Dr David Lester-Smith in the capacity of Medical Advisor—welcome David, we look forward to a very productive collaboration.



New Study Commencing in December 2017

Severe Injury Related to Disc Battery (SIRDB)

Poisons information centres and Emergency Departments (ED's) in Australia have seen increasing numbers of children presenting with possible battery exposures.

Although most ingested or inserted batteries can be easily removed or pass through the body uneventfully, severe injuries and deaths have been associated with ingestion or insertion of disc batteries, especially if they stay in situ for some time.

Battery-related injuries have increased with the increasing use of these batteries in common household products such as toys, torches, hearing aids, remote controls and ornaments.

This study is led by Dr Ruth Barker and aims to contribute to the review of manufacturing and packaging codes for products containing disc batteries.

Please start reporting Severe Injury Related to Disc Battery (SIRDB). It will appear on the APSU December 2017 report card.

Welcome to the December 2017 edition of the APSU's Rare Kids newsletter.

We hope that you find the newsletter interesting and informative.

We welcome your feedback by contacting us at SCHN-APSU@health.nsw.gov.au

The protocol, case report form and additional information for doctors and for the public have been uploaded to the APSU website: [Protocol](#), [Online Case Report Form](#), [Printable Case Report Form](#), [Information for Doctors](#), [Information for Parents](#)

Please complete the case details via the secure online case report form or simply download a PDF, complete and either email or fax back to the APSU.

Case Definition: Please report any child < 16 years of age with newly diagnosed injury related to disc or button battery ingestion or insertion that required procedural intervention either to remove the battery or to assess or repair damage related to the battery.

Exclusions: Please do not report cases where the battery has been ingested/inserted and it has passed/fallen out of the patient and the patient does not require a procedure to remove the battery or to assess or repair damage related to the battery.

Surveillance Study Commencing in 2018

Congenital Syphilis

Australia is currently experiencing a resurgence of infectious syphilis in its northern regions, and congenital syphilis may result in babies born to syphilis positive mothers. The World Health Organization (WHO) is committed to eliminating congenital syphilis globally with an overall goal of a 'reduction of incidence to 0.5 cases or less per 1000 live births.' The specific clinical management targets necessary to achieve this goal are: >90% of pregnant women to be screened for syphilis; and >90% of syphilis-seropositive pregnant women to receive appropriate treatment.

This study will enhance the current surveillance efforts through the Communicable Diseases Network Australia (CDNA) by enabling the linking of information on the mother and child to identify any gaps in health care provided to sero-positive women and their infants.

We are currently collaborating with the Kirby Institute and the New Zealand Paediatric Surveillance Unit to develop the APSU surveillance study. We anticipate that surveillance will commence in the first half of 2018 and the protocol, case definition and case report forms will be shared with all clinicians who report to the APSU.

Keeping Australia Polio Free — the Polio Expert Panel encourages all paediatricians to continue reporting all cases of AFP to the APSU

High quality acute flaccid paralysis (AFP) surveillance is essential to maintain polio-free status certification of Australia by the WHO. The WHO indicators of surveillance performance include a target annualised non-polio AFP rate of >1/100 000 children under 15 years of age.

The APSU protocol and case report form can be found here: [APSU Protocol](#), [Case Report Form](#)

Two stool samples for testing to exclude polio virus should be collected within 14 days of the onset of paralysis and sent to the National Polio Reference Laboratory.

More details at: www.vidrl.org.au/laboratories/poliovirus-reference/specimen-referral

Please do not hesitate to contact the APSU if you have any questions about this surveillance.

Surveillance Study Updates

JoRRP

Surveillance for Juvenile onset Respiratory Papillomatosis over the last 5 years (2012 – 2016) has shown declining rates of this severe outcome of human papilloma virus (HPV) infection with the introduction of the HPV vaccination program in Australia.

Estimated incidence rates have declined from 0.16/100,000 to 0.02/100,000 in 2016. Of the 15 cases reported, none of the mothers had been vaccinated for HPV and 86% of the children had a vaginal delivery.

JoRRP is characterised by repeated growth of benign exophytic papillomata in the larynx, often developing in early childhood. Affected children may require multiple surgical interventions and if the infection spreads to distal airways, tracheostomy may be required.

To read more about the APSU JoRRP study follow this link: [A Prospective Study of the Incidence of Juvenile-Onset Recurrent Respiratory Papillomatosis After Implementation of a National HPV Vaccination Program](#)

Vitamin K deficiency bleeding and haemorrhagic disease of the newborn: due to finish

Surveillance for Vitamin K deficiency has been on the APSU report card for the **last 24 years**. This study has been unfunded for many years and we have decided that we are no longer able to continue surveillance for this condition. We are in the process of analysing the data from the last 24 years.

Vitamin K Deficiency Bleeding will appear on the APSU card for the last time in January 2018 to enable any outstanding cases for 2017 to be reported. Please report any cases you may have seen but have not yet reported for 2017.

Microcephaly Update

Please report any child < 12 months of age with microcephaly when:

The Occipito-Frontal head Circumference (OFC) is more than **two standard deviations** or **<3rd percentile below the mean** for gestation*, age and gender according to standard growth charts (CDC Growth Charts— [CDC for Boys](#); [CDC for Girls](#)), presenting to you in the last month and whom you have not previously reported.

For Example: Any Male born at term with a OFC <32.1cm or Female born at term with a OFC <31.7cm would meet the criteria.

* use Fenton growth charts to adjust for gestational age/prematurity: [Fenton Chart for Boys](#); [Fenton Chart for Girls](#).

Please continue to report any new cases through the APSU reporting channel. Should you have any questions regarding this study, please feel free to write to us at: SCHN-APSU@health.nsw.gov.au



Since 1993 we have:

Sent 352,944 report cards - 321,179 cards have been returned

Maintained a response rate at >90%

Studied 68 conditions

Collected detailed data on ~8,330 children with rare disease

Published ~ 350 journal publications and reports

990 clinicians reported in 1993 by returning a report card in the post

1570 clinicians reported in 2017 – 97% report by e-mail

2018 will mark APSU's 25th Anniversary

25 years of surveillance is an enormous achievement for the APSU. We will be holding a session at the RACP Congress in May 2018 and we hope you will be able to join us to celebrate this important milestone – further details will be shared soon. Meanwhile, here are some key facts about the APSU:

- Established by Elizabeth Elliott in 1993
- 68 rare chronic and complex childhood diseases have been studied so far: infectious, genetic, mental health, rare injuries, rare immune disorders, etc.
- Up to 16 different conditions studied simultaneously
- Surveillance according to standardised case definitions and standardised datasets for each condition (demographics, diagnosis, treatment, complications, outcomes)
- Used by over 300 researchers (national groups of experts established for each condition)
- Often provides 1st national incidence estimates for rare childhood conditions
- Outcomes support policy, clinical practice and lead to further research

APSU staff have also attracted grants to enable activities other than surveillance:

- Systematic reviews / rapid reviews
- Surveys of Paediatricians (knowledge attitudes practice)
- Clinical audits
- Impacts on families and quality of life surveys
- Health services utility and costs studies (detailed activity and costing data)
- Establishing rare disease registries

Recent Events

Congratulations Professor Elizabeth Elliott

Professor Elizabeth Elliott, Director of the APSU, has been awarded the prestigious RACP **Howard Williams Medal** for 2017.

Each year at the RACP Congress the Paediatrics & Child Health Division officially acknowledges a person who has made an outstanding contribution to Paediatrics and Child Health in Australia or New Zealand.

Watch the RACP Howard Williams Medal 2017 presentation [here \(via RACP official YouTube channel\)](#)



Staff Changes in the APSU

We bid a fond farewell to **Dr Marie Deverell** who has returned to her home state of Western Australia after 5 years with the APSU. Marie was a key staff member who worked tirelessly to support the APSU system while developing new surveillance studies and supporting groups of investigators. We have missed her professionalism and her wide skill set which ranged from writing reports to complex data analysis and presenting findings to national and international audiences. We wish Marie every success in her new role in Perth.

We also said goodbye to **Amy Phu** who supported the APSU in a wide variety of ways ranging from developing databases and on-line data collection forms, to data analysis, literature reviews and clinical audits. Luckily Amy is not too far away at Westmead Hospital and we are keeping in touch. Of course Amy is flourishing in her new role and we wish her continued success.

We welcome **Dr Suzy Teutsch** (PhD) who comes with excellent experience and expertise which she most recently gained while working on the Australian Childhood Encephalitis (ACE) study and managing the Neonatal and Young Infant Herpes Simplex Virus Infection study in collaboration with Professor Cheryl Jones. Suzy started in early December and is already proving to be a valuable team member. We look forward to a successful working relationship with Suzy.

In the News

NEWCASTLE HERALD Wednesday December 06, 2017 theherald.com.au

NEWS

The curse of bottle babies

BY DONNA PAGE

WOMEN who drink should be using contraception to avoid having brain damaged babies, a Newcastle public health forum heard on Tuesday night.

Fetal alcohol spectrum disorder (FASD) expert Professor Elizabeth Elliott, of Sydney University, said it was a "false understanding" that disadvantaged women were at greater risk of having babies with the disorder that causes life-long brain damage.

Professor Elliott said the "standard understanding" was that FASD was more common among lower socio-economic families.

"In fact, the data suggests older women of high socio-economic status are most likely to drink during pregnancy," she said. "There are too many kids hidden or unrecognized living in high socio-economic families."

FASD, in which the unborn baby's brain is damaged by alcohol, is producing a wave of school children with learning difficulties.

The Australian Medical Association (AMA) is now instructing pregnant women to give up the gong.

The forum heard that up to 50 per cent of pregnancies are unplanned, creating a major risk for women who binge drink.

Professor Elliott said research found health professionals were reluctant to discuss FASD with parents, creating a hidden epidemic.

"Many are worried about asking about alcohol in pregnancy," she said. "The conversations are just not happening between women and GPs."

University of Newcastle's Professor John Boulton, a pioneer of FASD research, said doctors and nurses were "extremely uncomfortable" raising the issue of FASD with parents when diagnosing children because there was a "moral dimension" to the condition because it was entirely preventable.

"There is effectively not much redemption," he said.

"There is guilt, but no redemption."

About 150 people attended the forum at Newcastle City Hall organised by Newcastle Local Drug Action Team.

Other speakers included Dr James Fitzpatrick, head of alcohol and pregnancy and FASD research at Perth's Telethon Kids Institute, and Magistrate Andrew Eckhold, of Newcastle Local Court.

Newcastle has been selected by the federal government as one of two sites to lead the fight against FASD receiving more than \$500,000 over three years for a prevention program that focuses on increasing community awareness, prevention, skills development for workers and primary health diagnosis.

Dr Fitzpatrick said about one-third of juveniles in custody met the criteria for FASD and people with FASD were 19 times more likely to end up in jail.

Magistrate Eckhold said the hidden nature of the disorder was evidenced by the fact that he had "never" been presented with a report in the local court diagnosing a person with FASD.

EPIDEMIC: From left, Dr James Fitzpatrick, Magistrate Andrew Eckhold, Professor Elizabeth Elliott and Professor John Boulton at a public health forum looking at fetal alcohol spectrum disorder at Newcastle City Hall.

Professor Elizabeth Elliott, along with Professor John Boulton, Dr James Fitzpatrick and Magistrate Andrew Eckhold recently attended a public forum at Newcastle City Hall. The Forum was hosted by Newcastle Local Drug Action Team and focussed on FASD and it's impact in the local community.

Newcastle has been selected by the federal government to receive one of two \$500,000 grants over 3 years for a prevention program that will focus on increasing community awareness, prevention, skills development for workers and primary health workers.

To read the full article online, follow our [link here](#)

Key Publications for 2017

- Deverell M, Phu A, Zurynski Y, Elliott E. Australian Paediatric Surveillance Unit, Annual Report, 2015. *Commun Dis Intell Q Rep.* 2017; 41(2):E181-E185.
- Elliott E, Zurynski Y. Female genital mutilation is hurting Australian girls and we must work together to stamp it out. www.theconversation.com/female-genital-mutilation-is-hurting-australian-girls-and-we-must-work-together-to-stamp-it-out-71885
- Lacaze P, Millis N, Fookes M, Zurynski Y, Jaffe A, Bellgard M, et al. Rare disease registries: a call to action. *Internal Medicine Journal.* 2017; 47:1075–1079
- Mehr S, Frith K, Barnes EH, Campbell DE; FPIES Study Group. Food protein-induced enterocolitis syndrome in Australia: A population based study, 2012-2014. *J Allergy Clin Immunol* 2017; 140(5):1323-1330.
- Novakovic D, Cheng ATL, Zurynski Y, Booy R, Walker PJ, Berkowitz R, et al. A prospective study of the incidence of juvenile-onset recurrent respiratory papillomatosis after implementation of a national HPV vaccination program. *J Infect Dis* 2017; doi: 10.1093/infdis/jix498. [Epub ahead of print].
- Roberts J, Hobday L, Ibrahim A, Aitken T, Thorley B. Australian National Enterovirus Reference Laboratory annual report, 2014. [Commun Dis Intell Q Rep.](http://www.cdc.gov/mmwr/preview/mmwrhtml/mm6409a1.htm) 2017; 41(2):E161-180.
- Saddi V, Beggs S, Bennetts B, Harrison J, Hime N, Kapur N, et al. Childhood interstitial lung diseases in immunocompetent children in Australia and New Zealand: a decade's experience. *Orphanet J Rare Dis.* 2017; 12(1):133. doi: 10.1186/s13023-017-0637-x.
- Zurynski Y, Deverell M, Dalkeith T, Johnson S, Christodoulou J, Leonard H, et al. Australian children living with rare diseases: Experiences of diagnosis and perceived consequences of diagnostic delays *Orphanet J Rare Dis.* 2017;12(1):68. doi: 10.1186/s13023-017-0622-4.
- Zurynski Y, Elliott E. No compromise on female genital mutilation. *Journal of Paediatrics and Child Health.* 2017; 53:612–616. [Letter]
- Zurynski Y, Gonzales A, Deverell M, Phu A, Leonard H, Christodoulou, et al. Rare disease: A national survey of paediatricians' experiences and needs. *BMJ Paediatrics Open* 2017; 1:e000172. doi:10.1136/bmjpo-2017-000172.
- Zurynski Y, Phu A, Sureshkumar P, Cherian S, Deverell M, Elliott E. Female Genital Mutilation in children presenting to Australian Paediatricians. 2017;102(6):509-515.



Upcoming Events



Royal College of
Paediatrics and Child Health

Leading the way in Children's Health

Royal College of Paediatrics and Child Health Conference

"Children First—Ethics, Morality and Advocacy in Childhood"

15th—18th March, 2018, Glasgow UK

www.rcpch.ac.uk/annual-conference/rcpch-conference-2018

Royal Australian College of Physicians Congress

"Disruption for Healthy Futures"

14th—16th May, 2018, Sydney Australia

www.racpcongress.com.au



*We wish you and your family a happy and peaceful Christmas
and a prosperous New Year*

Thank you for your ongoing participation and support during 2017

The Team at the

Australian Paediatric Surveillance Unit



Australian Paediatric Surveillance Unit

Kids Research Institute
The Children's Hospital Westmead
Locked Bag 4001
WESTMEAD NSW 2145

p: (02) 9845 3005
f: (02) 9845 3082
e: SCHN-APSU@health.nsw.gov.au

We're on the Web

www.apsu.org.au



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RedCap Online Reporting Clinicians are now able to complete most case report forms online through our data capture management system (RedCap). Simply click on the link provided in the monthly report card email to complete the form. Links to the current APSU protocols and case report forms can be found in the monthly email report card and are also available for download on the [APSU website](http://www.apsu.org.au). If you have any difficulties accessing the links or would like more information about completing the forms online, please contact us.

Still returning the yellow card by post? If you are one of the remaining 95 clinicians out of 1600 still receiving the yellow report card we would like to make responding easier for you.

Not currently an APSU Contributor? We would very much like you to become a member of the APSU and to contribute data to support our unique national surveillance effort.

Changed your contact details? Please let us know your new details.

To start responding via email, to join the APSU or to notify us of your changed contact details please contact us on: SCHN-APSU@health.nsw.gov.au