

AUSTRALIAN PAEDIATRIC SURVEILLANCE UNIT ANNUAL REPORT, 2013

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Introduction

This report provides an update on the surveillance conducted by the Australian Paediatric Surveillance Unit (APSU) during the period January to December 2013. The APSU facilitates national active surveillance of uncommon diseases of childhood including selected communicable diseases. This report includes data on the following conditions: acute flaccid paralysis (AFP), congenital cytomegalovirus (cCMV), congenital rubella, perinatal exposure to HIV and paediatric HIV infection, neonatal herpes simplex virus (HSV), congenital varicella, neonatal varicella, severe complications of varicella and juvenile onset recurrent respiratory papillomatosis (JoRRP). Surveillance of severe complications of influenza was undertaken during the influenza season (July to September 2013).

Methods

Australian Paediatric Surveillance Unit

The APSU study protocols and case definitions are developed with collaborating study investigators who provide specialised clinical and research expertise for each condition studied and listed in the Table. Conditions under surveillance are listed on the APSU report card, which is sent to approximately 1,400 practising paediatricians and child health specialists every month. Response rates to the APSU monthly report card have remained over 90% for the last 20 years, and were 90% in 2013.

Over 85% of contributors report via email. Contributors respond each month whether or not they have a case to report for any of the conditions listed on the report card. The APSU collects de-identified clinical and/or laboratory data via a case report form completed by the doctor looking after the child. Completed case report forms are forwarded to study investigators. All study protocols and case report forms are available on the APSU website (www.apsu.org.au).

Paediatric Active Enhanced Disease Surveillance

The Paediatric Active Enhanced Disease Surveillance (PAEDS) system was initiated in 2007 by the APSU and the National Centre for Immunisation Research and Surveillance of

Vaccine Preventable Diseases.¹ PAEDS (www.paeds.edu.au) is a hospital-based surveillance system reliant on active case ascertainment by specialist surveillance nurses. PAEDS operates in 5 tertiary hospitals in the capital cities of 5 states; New South Wales, Victoria, South Australia, Western Australia and Queensland. PAEDS complements surveillance conducted by the APSU for AFP and varicella complications.

Results

Acute flaccid paralysis

Pooled data from the APSU and PAEDS systems are submitted regularly to the Polio Expert Panel. In 2013, there were 62 confirmed cases of AFP: 28 ascertained by PAEDS, 12 by APSU and 22 by both systems. The target of a non-polio AFP rate of ≥ 1 per 100,000 children under 15 years of age has been consistently reached for the last 6 years (2008–2013). PAEDS nurses ascertain the majority of AFP cases from the 5 tertiary paediatric hospitals where PAEDS operates. The APSU mainly contributes cases from non-PAEDS hospitals such as Sydney Children's Hospital, John Hunter Children's Hospital, Geelong Hospital, Toowoomba Hospital etc. These data contribute towards Australia fulfilling its requirements as stipulated by the World Health Organization (WHO) required AFP surveillance as part of the Global Polio Elimination Strategy and maintenance of the Polio-Free Certification by WHO. All cases are reviewed by the Polio Expert Panel. In 2013, 61 cases of AFP were classified as non-polio AFP and 1 case was classified as polio compatible. The main diagnoses associated with reported cases of AFP were Guillain-Barré syndrome, transverse myelitis and acute disseminated encephalomyelitis.

Congenital cytomegalovirus

The total number of reports of cCMV continue at approximately 16–17 cases per year. In 2013, a total of 16 confirmed cases and 3 probable cases were reported to the APSU. There was a total of 247 confirmed cases reported during the study period 1999–2013. The number of reports suggest a continuing under-recognition and underreporting of cCMV, as the investigators reported in 2011,³ and as continues today. Testing for cCMV after detection of sensorineural hearing loss (SNHL) on neonatal hearing screening is not routine in all

Table: Confirmed cases identified to December 2013 and for the total study period, and reported rates per 100,000 of the relevant child population

Condition	Date surveillance commenced	Questionnaire returned (%)	Number of confirmed cases 2013	Reported rate for 2013 (per 100,000)	Number of confirmed cases for total study period	Reported rate for total study period (per 100,000 per annum)
Acute flaccid paralysis	Mar 1995	100	62*	1.42†	779	1.01†
Congenital cytomegalovirus	Jan 1999	91	16	5.17‡	247	6.48‡
Congenital rubella (with defects) [§]	May 1993	100	3	0.06	54	0.07
Perinatal exposure to HIV	May 1993	90	43	13.89‡	587	10.99‡
HIV Infection	May 1993	100	1	0.02	84	0.10
Neonatal – herpes simplex virus infection	Jan 1997	91	14	4.52‡	154	3.57‡
Infant – herpes simplex virus infection	Jan 2012	91	3	0.96	3	0.50
Congenital varicella	May 2006	No notifications	Nil	Nil	2	0.10‡
Neonatal varicella	May 2006	75	3	0.97‡	22	1.07‡
Severe complications of varicella	May 2006	50	1	0.02†	50	0.15†
Juvenile onset recurrent respiratory papillomatosis**	Data for 2012	91	7	0.16†	7	0.16†
Severe complications of influenza††	Data for 2013	100	3	0.07†	10	0.12†
	Influenza season each year since 2008	93	13	0.30†	289	1.13†

* Includes all cases of acute flaccid paralysis reported via the Australian Paediatric Surveillance Unit or Paediatric Active Enhanced Disease Surveillance. All cases have been classified by the Polio Expert Panel as 'non-polio AFP' according to World Health Organization criteria.

† Notification was received by the Australian Paediatric Surveillance Unit, clinical data had not been returned at the time of submission.

‡ Influenza surveillance was conducted each year since 2008 during the influenza season, July to September except in the pandemic year (2009) when surveillance occurred from June to October.

§ Based on population of children aged less than 15 years.

|| Based on number of births.

†† Based on population of children aged less than 16 years.

** Based on population aged less than 12 months.

Confirmed cases and probable cases are reported; a probable case is defined as a papilloma visualised by endoscopy but the histology results are pending.

All reported rates are based on child population estimates published by the Australian Bureau of Statistics.²

All of the figures were correct at the time of submission and agreed by the chief investigators for each condition.

jurisdictions despite cCMV being an important cause of SNHL. cCMV is found in 5%–10% of neonates with SNHL of an otherwise unknown cause,⁴ including in Sydney (Rawlinson 2014 personal communication),⁵ although screening of children failing universal hearing screening is often practically difficult.⁵ Potential treatment with antiviral therapy to prevent long-term neurodevelopmental sequelae is limited by the lack of routine screening for CMV in pregnant women and in neonates,³ although this may change with recent efficacy analyses.⁶

Congenital rubella

There were four notifications of congenital rubella to the APSU during 2013. One was a duplicate notification. Three cases met the case definition criteria for congenital rubella with defects (microcephaly, hearing and vision impairments) and were consistent with congenital rubella syndrome. Two of the confirmed cases were children born in Australia (1 child born in 2013 in Victoria and 1 child born in 2012 in the Northern Territory) to mothers who had been born overseas (Thailand and Indonesia). Another child reported in 2013 was born overseas (in 2012) and diagnosed after arrival in South Australia at about 12 months of age. Prior to this, the last confirmed case of congenital rubella was reported in 2008. These recent cases highlight the need to remain vigilant by continuing surveillance efforts, continuing prevention of congenital rubella by screening all women (especially immigrant women) of childbearing age for rubella antibodies, and by maintaining high coverage of rubella vaccination in children.

Perinatal exposure to HIV and HIV infection

There was a total of 43 confirmed cases of perinatal exposure to HIV reported to the APSU in 2013. In addition, there was 1 case of HIV infection reported to the APSU in 2013. Over the total study period (1993–2013) 556 cases of perinatal exposure to HIV and 84 cases of neonatal HIV infection have been reported. The number of HIV perinatal exposures has remained relatively steady, but fewer infants acquire HIV infection as women take up interventions including avoiding breastfeeding, antiretroviral therapies and caesarean section.

Neonatal herpes simplex virus

In 2012, the case definition was amended to include disease in the new-born as well as in infants aged 1 month to up to 1 year of age. In 2013, a total of 17 cases were reported to the APSU; 14 in neonates and 3 in infants aged over 1 month of age. Since 1997, a total of 154 cases of HSV were reported by the end of 2013.

Congenital, neonatal and severe complications of varicella

No cases of congenital varicella were reported to the APSU during 2013. The last reported case of congenital varicella was in 2007. There were 4 cases of neonatal varicella reported to the APSU; completed case reports were received for three of these and all three met the case definition criteria. All 3 infants were exposed to varicella after birth; the details of the infective contact were unknown for 2 infants; the other one was exposed to a sibling who had chickenpox. All infants required hospitalisation due to the varicella infection (3–6 days), and all were treated with Aciclovir. Of the 2 notifications of children hospitalised with severe complications of varicella, 1 completed case report was received. The child was 10 years of age and spent 15 days in hospital with encephalitis and ataxia. This child had mild ataxia at discharge from hospital. A history of contact with a person known to have varicella was not provided.

Juvenile onset recurrent respiratory papillomatosis

JoRRP is a rare condition that develops in childhood and is typically found in children aged less than 12 years. It is the most common cause of benign neoplasms of the larynx in children and is caused by persistent infection of the upper airways with human papillomavirus (HPV) genotypes HPV 6 or HPV 11 acquired via vertical transmission before or during birth. HPV6 and HPV11 are targeted by the prophylactic quadrivalent HPV vaccine, meaning that JoRRP is now potentially a vaccine preventable disease, with high vaccination coverage achieved among Australian women aged 12–26 years by 2009.^{7,8} There were 15 notifications during the total study period (2012–2013) with completed case reports received for 14 notifications (93%). Of these 14 completed case reports there were 3 duplicate notifications and 1 error, leaving 10 cases. Of these 10, there were 6 confirmed cases and 1 probable case in 2012 (total 7) and one confirmed case and 2 probable cases in 2013 (total 3). There has been a decrease in the number of cases of JoRRP reported to the APSU in 2013.

Severe complications of influenza

The data for 2013 showed a marked reduction in the number of cases of influenza with severe complications reported to the APSU. There was a total of 13 confirmed cases of influenza with severe complications in children less than 15 years of age reported to the APSU in 2013, compared with 56 confirmed case reports during 2012. Eight cases had influenza B, and 5 cases had influenza A. The median age was 4.3 years (range from 10 days

to 14.4 years). Complications included the following: pneumonia with oxygen requirement, need for mechanical ventilation, seizures, myocarditis, pericarditis, shock, acute encephalopathy and rhabdomyolysis. Similar complications have previously been reported in children with pre-existing chronic conditions as well as in previously healthy children.⁹ Eight cases had not been vaccinated for influenza in the previous 12 months and the vaccination status of a further 5 cases was unknown.

Of the 13 cases, five were admitted to the Paediatric Intensive Care Unit. Of these 5 cases, four had not been vaccinated for influenza and the vaccination status was not known for 1 case. Underlying chronic conditions of these patients included: neuromuscular disorder with intellectual disability, lung disease (with and without intellectual disability) and asthma. There were 2 deaths reported during 2013.

Conclusions and future directions

The APSU has conducted national surveillance of rare diseases for 20 years and continues to provide a valuable data on a number of serious rare childhood diseases. The APSU 20 year annual report is available for download on the APSU web site (www.apsu.org.au).

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Chief Investigators of APSU surveillance studies:

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References

1. Zurynski Y, McIntyre P, Booy R, Elliott EJ, Paeds Investigators Group. Paediatric active enhanced disease surveillance: a new surveillance system for Australia. *J Paediatr Child Health* 2013;49(7):588–594.
2. Australian Bureau of Statistics. Australian Demographic Statistics. September 2013. ABS Cat no: 3101.0 Canberra; Australian Bureau of Statistics: 2013
3. McMullan B, Palasanthiran P, Jones C, Hall B, Robertson P, Howard J, et al. Congenital cytomegalovirus—time to diagnosis, management and clinical sequelae in Australia: opportunities for earlier identification. *Med J Aust* 2011;194(12):625–629.

4. Korver AM, de Vries JJ, Konings S, de Jong JW, Dekker FW, Vossen AC, et al. DECIBEL study: Congenital cytomegalovirus infection in young children with permanent bilateral hearing impairment in the Netherlands. *Journal of clinical virology : the official publication of the Pan American Society for Clinical Virology*. 2009;46 Suppl 4:S27–S31.
5. Williams EJ, Kadambari S, Berrington JE, Luck S, Atkinson C, Walter S, et al. Feasibility and acceptability of targeted screening for congenital CMV-related hearing loss. *Arch Dis Child Fetal Neonatal Ed* 2014;99(3):F230–F236.
6. Cannon MJ, Griffiths PD, Aston V, Rawlinson WD. Universal newborn screening for congenital CMV infection: what is the evidence of potential benefit? *Rev Med Virol* 2014;24(5)291–307.
7. Brotherton JML, Murray SL, Hall MA, Andrewartha LK, Banks CA, Meijer D, et al. Human papillomavirus vaccine coverage among female Australian adolescents: success of the school-based approach. *Med J Aust* 2013;199(9):614–617.
8. Brotherton J, Gertig D, Chappell G, Rowlands L, Saville M. Catching up with the catch-up: HPV vaccination coverage data for Australian women aged 18–26 years from the National HPV Vaccination Program Register. *Commun Dis Intell* 2011;35(2):197–201.
9. Elliott EJ, Zurynski YA, Walls T, Whitehead B, Gilmour R, Booy R. Novel inpatient surveillance in tertiary paediatric hospitals in New South Wales illustrates impact of first-wave pandemic influenza A H1N1 (2009) and informs future health service planning. *J Paediatr Child Health* 2012;48(3):235–241.