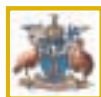


Australian Paediatric Surveillance Unit

■ **Surveillance Report 2002-2003**



The Royal Australasian  
College of Physicians  
Paediatrics & Child Health Division



Australian Government  
Department of Health and Ageing

**Australian Paediatric Surveillance Unit  
Surveillance Report 2002-2003**

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We acknowledge the input of Ms Karen Pattinson and  
Ms Ingrid Charters in the preparation of this document.

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ISSN: 1443-3524



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The Royal Australasian  
College of Physicians

**Paediatrics & Child Health Division**



**Australian Government**

**Department of Health and Ageing**

The APSU acknowledges The Royal Australasian College of Physicians,  
Paediatrics and Child Health Division for funding this report

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## ■ Minister for Health and Ageing The Honourable Senator Tony Abbott



The Australian Paediatric Surveillance Unit has made an important contribution to the national surveillance of rare childhood diseases since it was first established in 1993.

As the only available method of national data collection for most of the conditions studied, the Australian Paediatric Surveillance Unit has become a resource in the development of evidence-based programs that protect and promote the health of our children.

The data collected by the Unit include: the surveillance of acute flaccid paralysis to identify cases of poliomyelitis; the monitoring of disease activities for measles, mumps, rubella; and the surveillance of hepatitis C infections.

The data collected by the Unit has assisted my Department in providing evidence to inform and evaluate immunisation guidelines and other health programs. For example, the Unit plays an ongoing role in monitoring the success of the young adult measles, mumps and rubella immunisation campaign by documenting cases of congenital rubella in Australia.

I thank and congratulate the paediatricians and child health clinicians who support the activities of the Unit by providing information on a wide range of communicable and non-communicable diseases as well as the Unit's staff.

A handwritten signature in black ink, appearing to read 'Tony Abbott', with a long horizontal line above the name.

### ■ President, Paediatrics and Child Health Division Royal Australasian College of Physicians Professor Don Robertson



It is a great pleasure to be able to write to introduce the Progress Report of the Australian Paediatric Surveillance Unit. The Unit is familiar to all paediatricians in Australia in two major ways. One is for the work that it does, and the evidence that it provides which assists paediatricians in their day to day paediatric practice. The other is the emails, (or yellow cards for some),

which arrive like clockwork every month requesting notifications of patients that fit the particular criteria for studies conducted through the APSU at that particular time.

The output of the Unit has grown significantly over the years. It has a research, and research publication record, that all

paediatricians can be proud of. It has an international reputation for the data it provides, and also for the high report return rates each month. The Paediatrics and Child Health Division, with the Royal Australasian College of Physicians is proud of its association and linkages with the APSU. We expect to develop these further and to share in APSU successes in future years.

A handwritten signature in black ink that reads "Don Robertson".

#### Professor Don Robertson

Professor and Head, Department of Paediatrics,  
University of Adelaide.

Head, Department of Clinical Immunology/Rheumatology  
Women's and Childrens Hospital, Adelaide.

### ■ Chief Executive Officer, Royal Australasian College of Physicians Mr Craig Glenroy Patterson



As a unit of the Paediatrics & Child Health Division, The Royal Australasian College of Physicians is appreciative of the valuable work undertaken by the Australian Paediatric Surveillance Unit (APSU).

The College also acknowledges the high level of voluntary co-operation of Fellows providing information through the Unit.

The College, through its Paediatrics & Child Health Division, looks forward to a continued and rewarding association with the APSU.

A handwritten signature in black ink that reads "Craig Patterson".

#### Mr Craig Glenroy Patterson

Chief Executive Officer

The Royal Australasian College of Physicians

## ■ Chair, APSU Board Professor Carol Bower



By almost any measure, the Australian Paediatric Surveillance Unit (APSU) has been highly successful over its 12 years of operation. It is well supported by enthusiastic paediatricians (who consistently return notification cards each month in high numbers) and by researchers (who use the APSU system to conduct research). The output from studies conducted using the APSU has

contributed importantly to clinical and public health practice and policy (summarised in table 1 of this report).

It is perhaps surprising, therefore, that the APSU Board and Director have found it difficult to secure adequate ongoing funding for the Unit. In raising this issue, I hasten to acknowledge the enormous contributions to the operation of the Unit over the years from several research funding bodies, the Commonwealth Department of Health and Ageing, The Clive and Vera Ramaciotti Foundation; the Financial Markets Foundation for Children, The University of Sydney and The Children's Hospital at Westmead, amongst others. The difficulty, rather, is in obtaining adequate recurrent funding for the infrastructure of the Unit and such difficulty is not unique to APSU – it is also found with other ongoing surveillance systems and databases. I thus note, with great pleasure (and relief), that the medium-term future of the Unit now seems secure, thanks to the continued commitment of the Commonwealth and the Faculty of Medicine at The University of Sydney.

One of the many advantages of this relative security of funding, is the time it will free up for the APSU staff to invest in expanding

the functions of the APSU, rather than seeking funding. These may include exploring opportunities to enhance case ascertainment through collaboration with other research groups; developing international research collaborations to study diseases of public health significance in several countries simultaneously; and increasing post-graduate training and research opportunities in the APSU. Like the staff of the APSU, the Board will also have more time for other considerations. One of the items high on our agenda will be ways of maximising opportunities for APSU-based research to address national health priorities.

I congratulate Associate Professor Elliott and her staff, paediatricians, and APSU researchers and supporters on the achievements documented in this Report. I look forward to the successes continuing to contribute to important and valuable research in child health in Australia.

**Professor Carol Bower**

Principle Research Fellow, Division of Population Health Sciences  
Clinical Professor, Centre for Child Health Research, Telethon  
Institute for Child Health Research

Clinical Professor, School of Population Health, University of  
Western Australia

## ■ Associate Professor Elizabeth Elliott



The information included in the APSU Progress Report for 2002-3 reflects the ongoing commitment of Australian Paediatricians to improving and updating our knowledge about uncommon but high impact diseases in childhood. Thanks to the participation of clinicians from around the country, we have access to meaningful national data on a range of these diseases, as opposed to the local, non-representative data that would otherwise be available from individual clinicians and paediatric centres.

Internationally there has been a recent surge in interest in uncommon disorders. In the USA (where an estimated 25 million people are affected by ~6,000 rare diseases) the NIH has established an Office of Rare Diseases. The NIH emphasises the need for collaborative research "Because of the small number of affected patients in any one location, rare disease research requires the collaboration of scientists from multiple disciplines and the capacity to share access to geographically distributed national research resources and patient populations." Also highlighted is the fact that the study of rare disease may provide insights into the diagnosis or management of more common diseases: "Knowledge about rare diseases may offer leads for scientific advancement in other rare diseases and in more common diseases." ([www.nih.gov/news/pr/nov2003](http://www.nih.gov/news/pr/nov2003)).

In Europe, Orphanet, a database for rare diseases funded by the European Union, aims "to improve the diagnosis, management and treatment of rare diseases", which affect an estimated 25 million Europeans ([www.orpha.net](http://www.orpha.net)). In Australia and elsewhere, the establishment of paediatric surveillance units has gone some way to providing otherwise unavailable national information on rare childhood diseases. We estimate that in Australia approximately 1.6 million Australians have a rare disease, half of them children. These diseases have a huge impact on children and their families and many are chronic and demand considerable health resources.

However, research funding is largely directed into common diseases with immediate public health impact and it has been difficult, in Australia and elsewhere, to obtain adequate funding for research into uncommon diseases. We hope that as awareness

of rare diseases increases, so will the need for and availability of research resources.

One of the highlights in 2002-3 was the APSU 10th year celebratory meeting held in Hobart in association with the Annual Scientific Meeting of the Royal Australasian College of Physicians. Prof Fiona Stanley opened the meeting and agreed to be Patron of the APSU. Dr Christopher Verity, our guest speaker from the British Paediatric Surveillance Unit presented results of surveillance for Progressive Intellectual Deterioration, including new-variant Crutzfeld Jacob Disease, in the UK. The meeting was well attended and showcased the numerous studies performed through APSU since 1993.

I wish to acknowledge the hard work of Ms Donna Rose, APSU Scientific Co-ordinator and Ms Karen Pattinson, Administrator and thank them for their contribution to the unit. Thanks also to the APSU Board Chair Prof Carol Bower, members of the APSU Board and Scientific Program Committee and Prof Don Robertson, President of the Division of Paediatrics and Child Health. Once again I'd like to thank all paediatricians and child health clinicians who contribute data to our surveillance activities.

We are grateful for the support received for APSU activities from the Department of Health and Ageing, the Royal Australasian College of Physicians (especially the Division of Paediatrics and Child Health), the Faculty of Medicine of the University of Sydney, and The Children's Hospital at Westmead. However, increased resources are required to ensure that the data collected from clinicians through the APSU continues to be of the highest quality, can be supplemented where appropriate by data from other sources, and is disseminated widely and in a timely fashion to inform clinical and public health practice.

A handwritten signature in black ink, appearing to read 'Elizabeth Elliott'.

**Associate Professor Elizabeth Elliott**

Associate Professor, Discipline of Paediatrics and Child Health,  
University of Sydney

Consultant Paediatrician, The Children's Hospital at Westmead  
Director, The Australian Paediatric Surveillance Unit  
Convenor, International Network of Paediatric Surveillance Units



## Patron

### Professor Fiona Stanley AC

Director Telethon Institute for Child Health Research  
Professor, School of Paediatrics and Child Health  
The University of Western Australia

## Board

### Carol Bower\* (Board Chair from Jan 2003)

Principle Research Fellow, Division of Population Sciences and  
Clinical Professor, Centre for Child Health Research and School of  
Population Health, The University of Western Australia and the  
Telethon Institute for Child Health Research.

### Elizabeth Elliott\*

Associate Professor, Discipline of Paediatrics and Child Health,  
University of Sydney and Consultant Paediatrician, The Children's  
Hospital at Westmead. Director, The Australian Paediatric  
Surveillance Unit and Convenor, International Network of  
Paediatric Surveillance Units.

### Nigel Dickson\*

Senior Lecturer, Department of Preventive and Social Medicine  
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Paediatric Surveillance Unit.

### Elizabeth Hallam (to June 2004)

General Paediatrician, Hobart, Tasmania and Member,  
Divisional Committee for Paediatrics and Child Health, RACP.

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### Paul Lancaster

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Health, University of New South Wales.

### Helen Longbottom\* (to Dec 2002)

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### Elisabeth Murphy

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### Michael Nissen

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### Lesley Podesta

Assistant Secretary, Communicable Diseases Branch,  
Dept of Health and Ageing.

### Don Robertson (Board Chair to Dec 2002)

Professor and Head, Department of Paediatrics, University of  
Adelaide and Head, Department of Clinical Immunology/  
Rheumatology Women's and Children's Hospital Adelaide.

### Donna Rose\*

Scientific Coordinator, APSU

### Greg Sam (to Nov 2003)

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### Susan Skull\*

Head, Clinical Epidemiology and Biostatistics Unit,  
Royal Children's Hospital, Melbourne.

### Jenean Spencer (from December 2003)

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Department of Health and Ageing

### Barry Taylor

Professor and Head of Paediatric Section, Department of  
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Co-Director New Zealand Paediatric Surveillance Unit.

### Neil Wigg\*

Executive Director, Community Child Health Service, Royal  
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Professor, Department of Paediatrics and Child Health University  
of Queensland.

### John Ziegler

Clinical Immunologist and Head, Department of Immunology  
and Infectious Diseases, Sydney Children's Hospital, Randwick.  
Associate Professor, School of Women's and Children's Health,  
University of New South Wales.

\* Board and Scientific Review panel members

## ■ INSTITUTIONS COLLABORATING WITH THE APSU 1993-2003

### National Organisations

- Australia and New Zealand Paediatric Nephrology Association
- Australian CHARGE Association
- Australian Enteric Pathogens Surveillance Scheme
- Australian Polio Expert Committee
- Australian Institute of Health and Welfare
- Australian Society of Clinical Immunology and Allergy
- Commonwealth Department of Health and Ageing
- National Centre in HIV Epidemiology and Clinical Research
- National Centre for Immunisation Research and Surveillance of Vaccine Preventable Diseases
- National Notifiable Diseases Surveillance System
- National Perinatal Statistics Unit
- OzFoodNet: Australian Enhanced Foodborne Disease Surveillance
- Rett Syndrome Association
- Rett Syndrome Research Program

### New South Wales

- CAMSHNET
- Centre for Kidney Research
- Centre for Mental Health, NSW Health
- Children's Hospital at Westmead
- Gastroenterology & Liver Unit, Prince of Wales Hospital
- Institute for Neuromuscular Research
- Hunter Genetics
- Westmead Hospital
- Macleay Hastings Health Service
- Millennium Institute of Health Research
- NSW Birth Defects Register
- NSW Centre for Perinatal Health Services Research
- NSW Health
- Paediatric HIV Services Unit, Sydney Children's Hospital
- Royal Prince Alfred Hospital
- Royal North Shore Hospital
- Sydney Children's Hospital
- University of NSW
- University of Sydney
- South Eastern Sydney Area Health Service
- South Eastern Area Laboratory Services
- South Western Sydney Area Health Service

### Victoria

- Australian Mycobacterium Reference Laboratory Network
- Centre for Adolescent Health
- Victorian Infectious Diseases Reference Lab
- Monash Medical Centre
- Murdoch Children's Research Institute
- Public Health Group, Dept Human Services, Royal Women's Hospital, Melbourne
- Royal Children's Hospital, Melbourne
- University of Melbourne

### Queensland

- Mater Children's Hospital
- Princess Alexandra Hospital
- Queensland University of Technology
- Royal Children's Hospital, Brisbane
- Tropical Public Health Unit
- University of Queensland

### South Australia

- Flinders Medical Centre
- Institute of Medical Veterinary Science
- Mycobacterium Reference Laboratory, Adelaide
- South Australian Health Commission
- Women's and Children's Hospital, Adelaide

### Western Australia

- Curtin University
- Disability Services Commission
- King Edward Hospital, Perth
- Pathcentre Queen Elizabeth II Medical Centre
- Princess Margaret Hospital for Children, Perth
- Royal Perth Hospital
- Telethon Institute for Child Health Research

### Tasmania

- Royal Hobart Hospital

### Northern Territory

- Alice Springs Hospital
- Royal Darwin Hospital

### International Organisations

- Great Ormond St Hospital, London UK
- Hospital for Sick Children, Toronto Canada
- Oakland Childrens Hospital, USA
- Westkids, Auckland NZ

### INOPSU

- British Paediatric Surveillance Unit
- Canadian Paediatric Surveillance Programme
- Cyprus, Greece Paediatric Surveillance Unit
- German Paediatric Surveillance Unit
- Latvian Paediatric Association
- Malaysian Paediatric Surveillance Unit
- Netherlands Surveillance Unit
- New Zealand Paediatric Surveillance Unit
- Papua New Guinea Paediatric Surveillance Unit
- Portuguese Paediatric Surveillance Unit
- Swiss Paediatric Surveillance Unit
- Republic of Ireland Paediatric Surveillance Unit
- Welsh Paediatric Surveillance Unit

## FUNDING AND SPONSORSHIP



### Australian Government

#### Department of Health and Ageing

The Commonwealth Department of Health and Ageing is currently the APSU's principal funding agent. Funding provides infrastructure support for APSU studies that relate to communicable and vaccine-preventable conditions.



### The Royal Australasian College of Physicians

#### Paediatrics & Child Health Division

The APSU is a division of the Division of Paediatrics & Child Health, of the RACP. The APSU acknowledges the RACP in its generous funding of the 2002-03 Surveillance Report



The APSU Director is supported by the Discipline of Paediatrics and Child Health, Faculty of Medicine, University of Sydney.



The Children's Hospital, Westmead, generously provides office space and infrastructure support.

### Healthway, WA

A Healthway health promotion research grant contributes towards the surveillance of Fetal Alcohol Syndrome, which was initiated in 2001.



Roche Products Pty Limited sponsors the ongoing surveillance of Vitamin K deficiency bleeding with an annual grant.



The Financial Markets Foundation for Children (FMFC) is the financial and marketing community's charity. Between 1995 and 1999, the FMFC was the major sponsor of the APSU.



Majura Wines generously sponsors the APSU wine draw prize in 2002-2003



Orlando wines generously sponsored the APSU wine draw prize from 1999-2002.

### Financial contributions have been received from the following study groups using the APSU to conduct surveillance

- Acute flaccid paralysis: Department of Health & Ageing
- Adverse effects from complementary and alternative medicine: Women's and Children's Health, The Royal Women's Hospital and The Royal Children's Hospital, Melbourne
- CHARGE association: Australian CHARGE Association
- Congenital cytomegalovirus infection: Virology Division, Dept of Microbiology, SEALS, Sydney Children's Hospital
- Fetal alcohol syndrome: Telethon Institute for Child Health Research, Healthway, WA
- HIV/AIDS, perinatal exposure to HIV: National Centre in HIV Epidemiology and Clinical Research
- Munchausen by proxy syndrome: Child Protection Unit, The Children's Hospital at Westmead
- Neonatal herpes simplex virus infection: Dept of Immunology and Infectious Diseases, The Children's Hospital Westmead Herpes Simplex Virus Research Lab
- Rett syndrome: Telethon Institute for Child Health Research, USA National Institutes of Health, Rett Syndrome Association of Australia
- Vitamin K deficiency bleeding: NSW Health, Roche.

### Previous sources of funding are gratefully acknowledged

- Allen & Hanburys
- AMP
- ANZ
- Centre for Prevention of Psychological Problems in Children, Children's Hospital at Westmead
- Glaxo Smith Kline
- Clive and Vera Ramaciotti Foundation
- CSL
- Davies Collison Cave Attorneys
- NSW Department of Health
- Nutricia Australia
- Paediatric Research Society of Australia and New Zealand.

## ■ The APSU

The APSU was established in 1992 and surveillance commenced in May 1993. The APSU conducts national active surveillance of rare diseases of childhood, including infectious and vaccine preventable diseases, genetic disorders, childhood injuries and mental health conditions, with reporting by Australian child health specialists. Surveillance through the APSU provides the only available method of national data collection for most of the childhood conditions studied.

The primary aim of the APSU is to document the epidemiology of the conditions under surveillance, their clinical features, current management and short-term outcomes. The APSU's secondary aims are to provide a mechanism for national collaborative research and to disseminate data acquired by the Unit to inform best practice, appropriate prevention strategies and optimal health resource allocation.

The APSU is a unit of the Division of Paediatrics and Child Health, Royal Australasian College of Physicians. It is based at The Children's Hospital at Westmead and supported by the Faculty of Medicine, University of Sydney. The APSU Board, comprising prominent paediatricians, epidemiologists and public health personnel, oversees the management of the Unit. The APSU Scientific Review Panel evaluates applications to conduct studies through the Unit for suitability and scientific merit. The activities of the APSU are funded in part by the Federal Department of Health and Ageing through their communicable diseases program.

### Primary Aims:

- To accurately document the number of Australian children presenting with specific uncommon diseases (or complications of diseases) to child health specialists;
- To record the geographic distribution, clinical features, current management and short-term outcome of rare diseases.

### Secondary Aims:

- To provide a mechanism for national collaborative research in child health;
- To issue updated clinical and diagnostic information to clinicians caring for children with the conditions being studied;
- To disseminate information acquired by the Unit which will guide best practice, appropriate prevention strategies and optimal health resource allocation.

## Contributors to the APSU

Contributors to the APSU are clinicians known to be working in paediatrics and child health in Australia. These are predominantly Fellows of the Division of Paediatrics and Child Health, Royal Australasian College of Physicians, however other child health specialists including paediatric surgeons and child psychiatrists also participate in surveillance. Clinicians are identified through the Division of Paediatrics and Child Health (RACP), the Australasian Association of Paediatric Surgeons and subspecialty interest groups (Figure 1). In 2003 an average of 1050 clinicians participated in monthly surveillance.

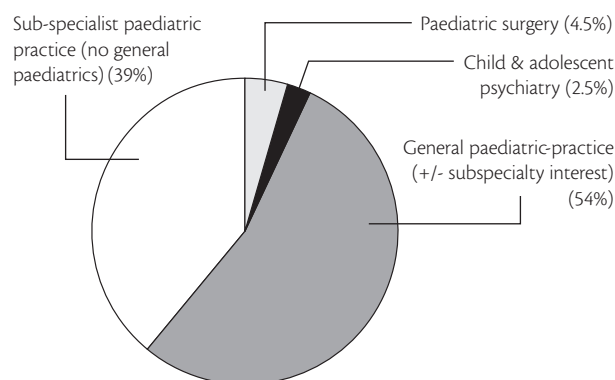


Figure 1. Practice characteristics of contributors to the APSU in 2003

## Operation of the APSU

Each month all clinicians are sent either a reply-paid report card or an e-mail 'card' listing conditions currently being studied through the APSU. Clinicians are asked to report children newly diagnosed with any of the conditions listed. Investigators conducting a study are informed weekly of new cases reported by APSU contributors. The Investigator then sends a brief questionnaire to the clinician requesting further de-identified information. Investigators are responsible for collation, analysis and publication of this data (Figure 2), and report study findings annually to the APSU.

## Selection of conditions for study

Individuals or organisations may apply to study a condition through the APSU and applications undergo a process of peer review and revision before being listed on the monthly report card.

To satisfy the criteria for study a condition:

1. must be sufficiently uncommon so that the system is not over-burdened;
2. must usually result in referral to a paediatrician or related specialist;
3. must provide information that satisfies the study aims and that is not available from other sources.

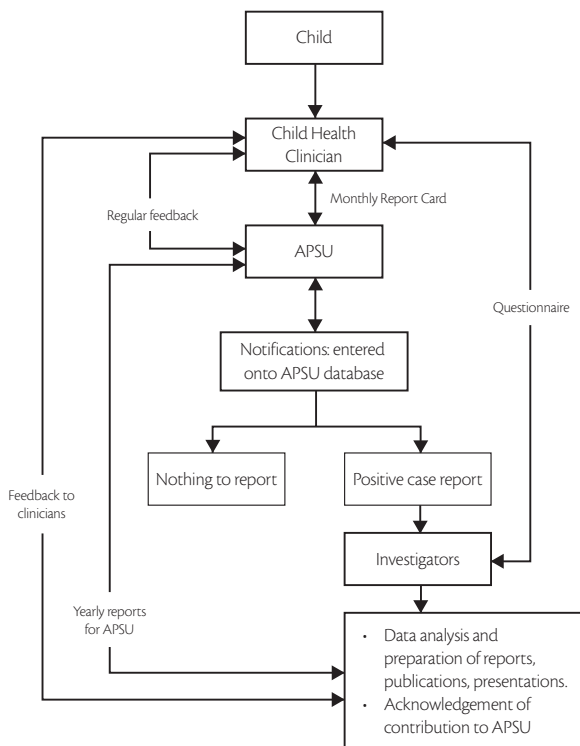


Figure 2. Operation of the APSU

Conditions are usually studied for one to three years, although provision for on-going study may be granted for diseases of public health significance or with very low incidence.

## Conditions Studied

Between 1993 and 2003, the APSU monitored 32 uncommon childhood conditions. Some of the major findings of studies conducted through the APSU are documented in Table 1. Between 2002 and 2003 studies of Childhood conversion disorder, Anaphylaxis following food ingestion, Early onset eating disorder, and Hepatitis C virus infection commenced. Surveillance of CHARGE association was completed in 2002. Studies on Childhood Conversion disorder, Munchausen by proxy syndrome and surveillance of Anaphylaxis following food ingestion was completed at the end of 2003.

## Case Classification

The APSU aims to estimate the incidence of selected conditions and to provide information which is representative of the Australian population. Maximal and unbiased case ascertainment is a high priority. Over-reporting rather than under-reporting of cases will help achieve this and duplicate reports are encouraged. Accurate classification of reported cases is facilitated by use of a unique identification code that is provided by clinicians on the study questionnaires. This enables investigators to identify duplicate reports.

To confirm a case, the clinical information reported in the questionnaire is used. The method of classifying cases is shown in Figure 3.

### Valid

- A **confirmed case** is one confirmed by the Investigator to satisfy the diagnostic criteria.
- A **probable case** is one that does not completely meet the diagnostic criteria but is highly probable on the basis of information available.

### Invalid

- A **duplicate case** is one that has already been reported
- An **error** is a case that has been reported but which:
  - 1) does not fulfil the diagnostic criteria, or
  - 2) has had diagnosis revised by referring clinician, or
  - 3) APSU report card was ticked by mistake.

### Unknown

Insufficient follow-up information is available to the Investigator or information is not received by APSU from the investigator.

Figure 3. Classification of reported cases

The APSU encourages Investigators to use multiple sources of case ascertainment where possible. This is particularly important in remote areas, where children have limited access to paediatricians and are often seen by general clinicians. Reported rates for conditions ascertained through the APSU therefore represent a minimum estimate of these conditions in the relevant Australian populations.

# Surveillance Overview

**Table 1 Key findings of national surveillance conducted through the APSU 1993-2003**

Conditions Under Surveillance	Commencement date	Key findings, implications and publications
<b>Infectious / vaccine preventable</b>		
Acute flaccid paralysis	Mar 1995	APSU reports via DoHA Polio Expert Committee to WHO and data contributed to 'polio-free' certification by WHO. (3,14,19,25,26,30*)
Congenital cytomegalovirus (CMV) infection	Jan 1999	Identified predominate CMV isolate in congenital infection and used isolates in placental model of transmission. (27,43*)
Congenital varicella	Mar 1995-Dec 1997	Identified that birth defects also occur with 3rd trimester infection; pregnancies should be monitored and infants' eyes examined for visual impairment. (1)
Neonatal varicella	Mar 1995-Dec 1997	Early identification, treatment (acyclovir, Ig) recommended. (1-3)
Congenital rubella	May 1993	Identified burden of imported cases, gaps in the vaccination program and need for immigrant screening. (10,11*)
Haemolytic uraemic syndrome	Jul 1994-Dec 2001	Identified Shiga-toxin producing E.coli prevalent in Australia; provided national data during HUS outbreak and informed code of production for fermented meats. (4;5) (32,33*)
Hepatitis C virus infection	Jan 2003	Monitoring an emerging disease of national significance; identified need for screening/management guidelines.
HIV/AIDS, Perinatal exposure to HIV	May 1993	Enhances mandatory reporting, identifies perinatal exposure and maternal risks to inform perinatal health. Identified adults as main infection source. (50,51,52*)
Hospitalised pertussis in infancy	Jan 2001-Dec 2001	Informed revision of immunisation schedule in 2003 to recommend vaccination of teenagers. (6) (41,48*)
Invasive Haemophilus Influenzae infection	Jan 1998-Dec 2000	Confirmed success of Haemophilus influenzae Type B vaccination policy. (7)
Kawasaki disease	May 1993-Jun 1995	Identified that young children may not fulfil international diagnostic criteria. (8)
Neonatal herpes simplex virus infection	Jan 1997	Identified HSV Type 1 as predominant type in neonatal infection in Australia and high frequency of eye disease. Informs perinatal management and vaccine development. (4,8,31,34,35,54,55*)
Subacute sclerosing panencephalitis	Jan 1995-Dec 1998	Very rare, reflecting high uptake of measles vaccination. (9)
<b>Congenital / genetic disorders</b>		
Arthrogyposis multiplex congenita	Jan 1996-Dec 1998	Documented risk factors, informed development of new disease classification and informed causal pathways. (9)
CHARGE association	Jan 2000-Dec 2002	Increased awareness of diagnostic criteria for CHARGE, diagnosis of 87% of cases in first year of life. (10)
Congenital adrenal hyperplasia	Aug 1995-Dec 1997	Enabled cross validation of potential neonatal screening program. (3)
Congenital & idiopathic nephrotic syndrome	Jul 1998-Jun 2001	Identified non-adherence to evidence-based management. (10,11)
Extrahepatic biliary atresia	May 1993-Dec 1996	Identified late diagnosis and need for education. Quantified transplantation needs. (12)
Fetal alcohol syndrome	Jan 2001	Indigenous children over-represented; siblings often affected. Informed causal pathways and educational strategies. (53*)
Hirschsprung disease	Jan 1997-Dec 2000	Primary surgical procedure most used is Soave operation. (7) (23*)
Prader-Willi syndrome	Jan 1998-Dec 2000	First DNA confirmed estimate of birth prevalence. PWS often missed clinically in infants – education needed. (24*)
Primary immunodeficiency disorders (PID)	Jan 1997-Dec 1999	Documented numbers affected, need for immunotherapy, bone marrow transplant. (13)
Rett syndrome	May 1993-Apr 1995; Jan 2000	Enabled molecular epidemiological study of national cohort, phenotype/genotype correlation, international collaboration genotyping. (1,2,9,12,15,16,18,28,29,37,49*)
Severe combined immunodeficiency	May 1995-Dec 2001	Confirmed bone marrow transplant after early diagnosis has good outcome. (10) (44*)

**Table 1 Key findings of national surveillance conducted through the APSU 1993-2003**

Conditions Under Surveillance	Commencement date	Key findings, implications and publications
<b>Mental health issue</b>		
Childhood dementia	May 1993-Jun 1995	First national study worldwide. Clarified diagnostic criteria, identified large group with no identified cause. (21*)
Childhood conversion disorder	Jan 2002-Dec 2003	First study to document the burden of illness in Australian children and to clarify psychosocial risk factors. (10) (42*)
Munchausen by proxy syndrome	Jan 2000-Dec 2003	First study to document impact of the diagnosis on clinicians. Data informing development of management policy. (13*)
Early onset eating disorder	Jul 2002	First epidemiological study <13yrs. Contributing to debate on relevance of adult diagnostic (DSM) criteria to children. Simultaneous Canadian study.
<b>Other injury / illness</b>		
Anaphylaxis following food ingestion	Jul 2002-Dec 2003	Peanut most common cause. Also other; nuts, soy, shellfish implicated. (36*)
Complementary and alternative medicine	Jan 2001-Dec 2003	Sentinel adverse effects documented in infants and children. (17,38,39,40*)
Near drowning	May 1993-Dec 1996	Neurological outcomes poor; age determines near drowning site. Commonly home pool. (22*)
Vitamin K deficiency bleeding	May 1993	Monitoring disease during policy changes to vitamin K prophylaxis and universal use of new vitamin K preparation. (14)

\*These publications are located in publications and presentations 2002-3 pg. 59.

- Forrest JM, Mego S, Burgess MA. Congenital and neonatal varicella in Australia. *Journal of Paediatrics and Child Health* 2000; 36(2):108-13.
- Burgess MA, Forrest JM. Congenital Varicella. *Journal of Paediatrics and Child Health* 1995; 31(6):564.
- Elliott EJ, Williams K, Peat J, Mildenhall S, Redmond D (Eds.). *Australian Paediatric Surveillance Unit Fifth Annual Report, 1997*. Sydney: Australian Paediatric Surveillance Unit 1998.
- Elliott EJ, Robins-Browne RM, O'Loughlin EV, Bennett-Wood V, Bourke J, Henning P et al. Nationwide study of haemolytic uraemic syndrome: clinical, microbiological and epidemiological features. *Archives of Diseases of Childhood* 2001; 85:125-131.
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- Elliott EJ, Ridley G, Morris A, Redmond D, Williams G, (Eds.). *Australian Paediatric Surveillance Unit Eighth Annual Report, 2000*. Australian Paediatric Surveillance Unit. 2001 Sydney, ISSN: 1443-3524.
- Royle J, Williams K, Elliott EJ, Sholler G, Allen R, Isaacs D, Kawasaki disease in Australia 1993-1995. *Archives of Diseases in Childhood* 1998; 78:33-39.
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- Elliott EJ, Williams K, Ridley G, Morris A, Redmond D, Williams G, (Eds.). *Australian Paediatric Surveillance Unit Seventh Annual Report, 1999*. Australian Paediatric Surveillance Unit. 2000 Sydney, ISSN: 1443-3524.
- National Health and Medical Research Council, Paediatric Division of of the Royal Australasian College of Physicians, et al. *Joint statement and recommendations on vitamin K administration to newborn infants to prevent vitamin K deficiency bleeding in infancy*, 2001. Canberra: NH&MRC.

## Response Rates

Figure 4 shows the average number of clinicians sent a monthly APSU report card and the average annual response rate. In 2003, 1050 clinicians participated in the monthly surveillance of 14 conditions, with an overall response rate of 96%. This maintains the excellent participation level of contributing clinicians since APSU's inception in 1993. Reporting by e-mail was introduced in

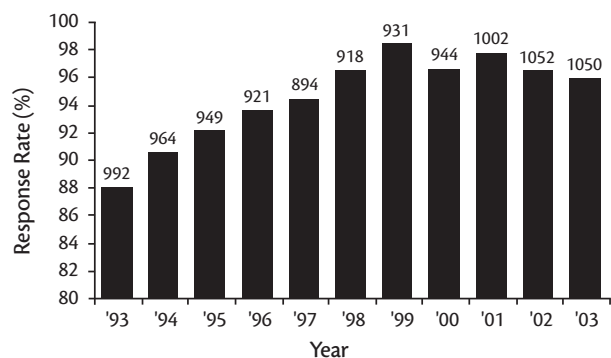


Figure 4. APSU annual response rate (%) and average number of contributing clinicians 1993-2003

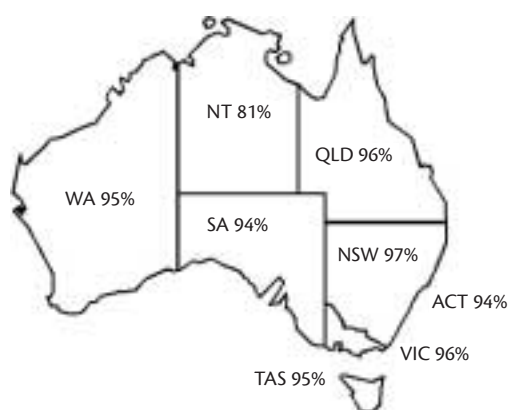


Figure 5. APSU Response rates by state and territory for 2003

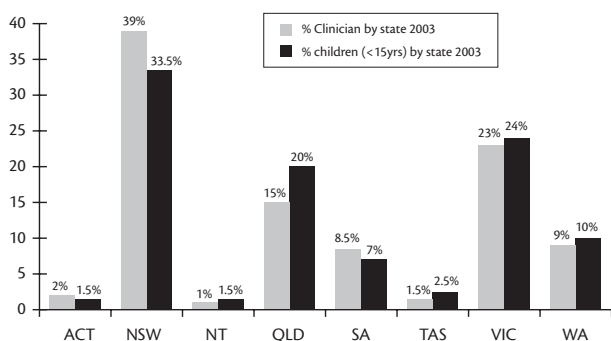


Figure 6. Population distribution by state and territory for children aged <15 yrs and clinicians reporting to APSU

<sup>1</sup> Austats: Population by age and sex, Australian states and territories. Australian Bureau of Statistics 2004; 3201.0

February 2001. In 2003, 52% of clinicians reported by e-mail with an average response rate of 96%. Response rate by state for 2003 is shown in figure 5.

Figure 6 shows the population distribution by state for children < 15 years in 2003 according to the ABS Statistics.<sup>1</sup> NSW has the greatest proportion of children with 33.5%, Victoria 24% and Queensland 20%. Correspondingly, NSW has the greatest number of participating clinicians (408; 39%), Victoria (246; 23%) and Queensland (161; 15%). The distribution of clinicians across Australia is indicated in figure 6 below.

## Respondent workload

Sixty four percent of clinicians had no cases to report and ticked the "nothing to report" box each month. Seventeen percent of clinicians reported one case, 8% reported two and 5% three cases. Six percent (63) of clinicians reported four or more cases during 2002-3 (Figure 7).

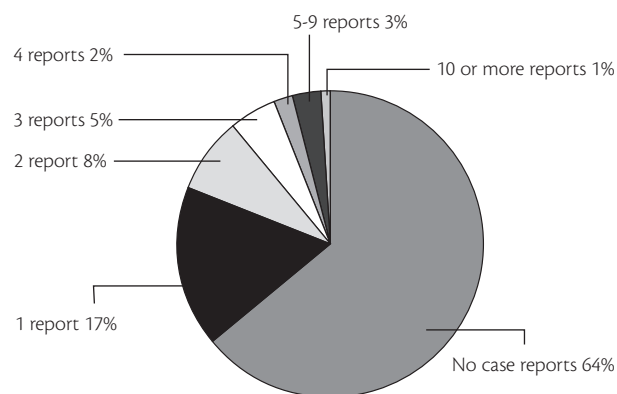


Figure 7. Respondent workload in 2002-3: Distribution of clinicians making zero to more than ten reports to the APSU

## Summary of surveillance study results 2002-2003

Incidence can be estimated on the basis of the number of reports of newly diagnosed cases in a defined population in a defined period of time. Population figures for the denominator are obtained from the Australian Bureau of Statistics. Incidence is expressed either as the number of new cases per 100,000 live births per annum (for conditions diagnosed before 12 months of age), or per 100,000 children under five or under 15 years per annum. Table 2 shows reported rates, with 95% confidence intervals, for conditions studied through the APSU. As 100% case ascertainment is unlikely to be achieved by any one surveillance scheme, reported rates represent estimates of minimum incidence. For conditions where case ascertainment has also occurred through complementary sources, (including Perinatal exposure to HIV, Acute flaccid paralysis, Haemolytic uraemic syndrome and Rett syndrome) cases from more than one source have been included to estimate incidence.



**Table 2 Summary of results of studies conducted to December 2003**

Conditions Under Surveillance	Commencement date	Duration of study (years)	Total confirmed cases	Reported Rate (95% CI) <sup>a b c d</sup>
<b>Infectious / vaccine preventable</b>				
Acute flaccid paralysis	Mar 1995	8.75	287	0.82 (0.73-0.93) <sup>b</sup>
Congenital cytomegalovirus (CMV) infection	Jan 1999	5	31	1.78 (1.4-2.52) <sup>b</sup>
Congenital rubella (with defects)	May 1993	10.5	49	0.117(0.08-0.15) <sup>b</sup>
Hepatitis C virus infection	Jan 2003	1	12	0.3 (0.16-0.53) <sup>b</sup>
Perinatal exposure to HIV (Birth Prev)	May 1993	10.5	222	8.00 (6.98-9.12) <sup>e</sup>
Perinatal infection after exposure to HIV (Birth Prev)			39	1.4 (0.99-1.92) <sup>e</sup>
Neonatal herpes simplex virus infection	Jan 1997	6	59	3.4 (2.6-4.4) <sup>a</sup>
<b>Congenital / genetic disorders</b>				
CHARGE association	Jan 2000-Dec 2002	3	23	2.8 (1.8-4.3) <sup>a</sup>
Fetal alcohol syndrome	Jan 2001	3	53	0.443 (0.332-0.58) <sup>d</sup>
Rett syndrome	Jan 2000	4	158	8.5 (7.9-9.1) <sup>c</sup>
<b>Mental health issues</b>				
Childhood conversion disorder	Jan 2002-Dec 2003	2	194	2.44 (2.1-2.8) <sup>b</sup>
Munchausen by proxy syndrome	Jan 2000-Dec 2003	4	58	0.4 (0.3-0.5) <sup>b</sup>
Early onset eating disorder	Jul 2002	1.5	45	*
<b>Other injury / illness</b>				
Anaphylaxis following food ingestion	Jul 2002-Dec 2003	1.5	110	1.84 (1.51-2.22) <sup>b</sup>
Complementary and alternative medicine	Jan 2001	3	171	1.43 (1.22-1.66) <sup>b</sup>
Vitamin K deficiency bleeding	May 1993	10.5	28	1.01 (0.67-1.46) <sup>a</sup>

\* Rate for early onset eating disorder is not calculated. Refer to report page 30.

a Reported incidence (95% CI) per 100,000 live births – includes confirmed and probable cases

b Reported incidence (95% CI) per 100,000 children <15 years

c Reported prevalence (95% CI) per 100,000 @ 2003 in Aust born females aged 5-18yrs.

d Reported incidence (95% CI) per 100,000 children < 5yrs confirmed + probable cases FAS

e Reported birth prevalence (95% CI) per 100,000 live births

## ACUTE FLACCID PARALYSIS (AFP)

- After reaching the World Health Organization (WHO) AFP case notification target in 2000 and 2001 (1 case/100,000), Australia did not reach the target in 2002 and 2003 due to low reporting from Victoria, SA and WA.
- Adequate faecal specimens were received from only 26% of all eligible cases in 2002 and 24% in 2003, well below the WHO target of 80%.
- Extensive investigation excluded VAPP as the cause of acute flaccid paralysis (AFP) in a child from whose faeces a poliovirus was isolated in 2002.
- A novel enterovirus, EV75, was isolated from one child with AFP in 2003.
- A study of AFP notification rates from each state of Australia concluded that the engagement of state based Departments of Health to promote and monitor AFP as a public health issue may assist in increasing Australia's AFP notification rate.

### Background

AFP surveillance was initiated in 1995 to comply with the WHO standards for certification of poliomyelitis eradication. Paediatricians notify AFP cases to the APSU using the monthly report card or to the Victorian Infectious Diseases Reference Laboratory (VIDRL) directly by telephone. APSU notifications are sent to VIDRL for follow-up. The WHO annual surveillance target for non-polio AFP cases is 1/100,000 children aged <15 years. This is equivalent to 40 cases per year in Australia. The National Polio Expert Committee (PEC) reviews all notified cases of AFP and classifies them as confirmed poliomyelitis; polio-compatible; or non-polio AFP. Faecal specimens collected from AFP cases are tested at the WHO accredited national polio reference laboratory situated at VIDRL.

### Objectives

- To determine the notification rate of AFP in children aged <15 years of age in Australia;
- To determine whether AFP is caused by poliovirus infection and, if so, whether it is a wild, vaccine or vaccine-derived strain of poliovirus;
- To determine other causes, and the clinical picture, of AFP in Australia.

### Case Definition

Any child resident in Australia and aged <15 years with acute onset of flaccid paralysis in one or more limbs or acute onset of bulbar paralysis.

### Results

During 2002-3, the National Polio Expert Committee (PEC) reviewed 96 notifications. These are classified in figure 8. Fifty-seven cases were classified as non-polio AFP (Figure 8). The notification rate for confirmed cases for 2002-3 was 0.71/100,000, 29% below

the WHO target. However, Queensland, New South Wales, Tasmania, the Australian Capital Territory and the Northern Territory reached the accepted target during the reporting period.

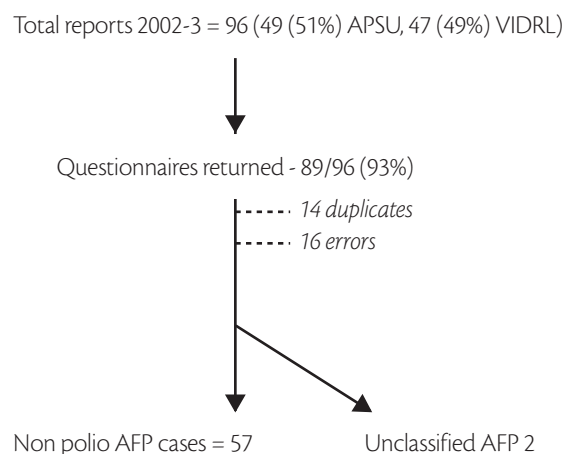


Figure 8. AFP surveillance data 2002-3



Figure 9. Geographic distribution AFP cases 2002-3 (includes seven notifications for whom no clinical information is available)

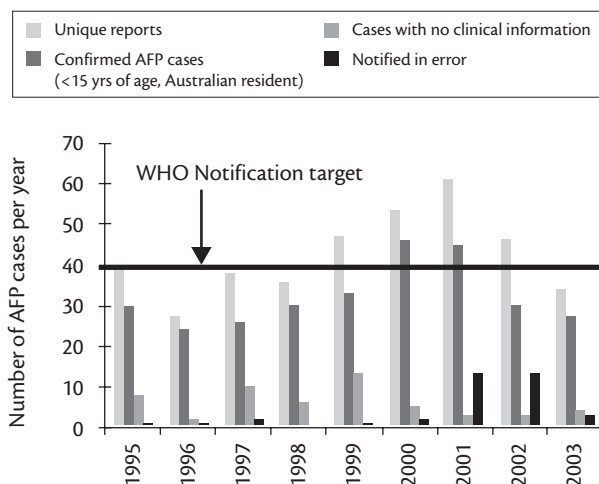


Figure 10. AFP Surveillance 1995-2003.

As described previously, Guillain-Barre syndrome was the most common single diagnosis (35% of confirmed cases), followed by transverse myelitis (18%) in 2002-3. Only 26% of cases in 2002 and 24% of cases in 2003, had adequate faecal specimens, well below the WHO target of 80%. Adequate sampling is defined as two faecal specimens collected 24 hours apart and within 14 days of onset of paralysis. No faecal specimens were available for cases first notified to the APSU in 2002 but this improved in 2003 with seven cases first notified to the APSU having at least one faecal specimen collected.

One case of AFP was reported in 2002 was initially considered a potential case of vaccine associated paralytic poliomyelitis (VAPP) in a contact. A poliovirus type 3 was isolated from a patient whose sibling had been immunised eight weeks earlier with oral polio vaccine (OPV). The patient had been immunised at 2, 4 and 6 months of age with no pre-school booster. No significant rise in titre between acute and convalescent sera was detected to poliovirus type 3 in the patient. The PEC reviewed the available clinical and laboratory data and classified the case as non-polio AFP. The final diagnosis was acute focal mononeuropathy (with anterior horn cell involvement) and the isolation of the poliovirus type 3 was considered incidental.

During 2003 enterovirus type 71 was isolated from the faeces of one child with AFP. This virus was genetically related to viruses of genogroup C1 detected in the Malaysian peninsular during 1997-2000. A novel enterovirus type 75 was isolated from another case of AFP in 2003.

## Conclusion

Australia's notification rate of confirmed AFP cases in 2002 and 2003 declined to levels reported between 1995-1999, after having reached the WHO target in both 2000 and 2001 (Figure 10). The decline in notifications is unlikely to be due to a decrease in cases of AFP. Queensland has continued to exceed the WHO notification rate (Figure 9) and incomplete ascertainment in Victoria has been documented through capture recapture studies in 1995-7 (National Documentation for Certification of

Poliomyelitis Eradication in Australia, Commonwealth of Australia 2000 pp127-131, and 1998-2000 (Bull WHO 2002; 80:846-51).

Faecal specimens were more likely to be collected in cases first notified to VIDRL. This may be because direct notification to VIDRL allows for arrangements to be made for faecal collection and transportation to the national polio reference laboratory at VIDRL. All paediatricians are encouraged to report cases of AFP to both VIDRL and APSU.

A Sabin-like poliovirus was isolated from a child with AFP in 2002 and was not classified as VAPP. This emphasizes the need for the laboratory investigation of all AFP cases. In 2003 enterovirus types 71 and 75 were isolated. This demonstrates the need for the specific identification of enteroviruses, especially from cases of AFP.

## Report authors / study investigators

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Associate Professor Elizabeth Elliott; Discipline of Paediatrics and Child Health, University of Sydney, Locked Bag 4001, Westmead, NSW 2145.

## Publications arising from this study:

### Original Articles

- Kelly H, Brussen KA. Apparent improvement in AFP surveillance in Australia. *Australian and New Zealand Journal of Public Health* 2002; 26:281-282.
- Morris AM, Elliott EJ, D'Souza RM, Antony J, Kennett M, Longbottom H. Acute flaccid paralysis in Australian children. *Journal of Paediatric Child Health*. 2003 Jan-Feb;39 (1):22-6.
- Thorley BR, Brussen KA., Stambos V, Yuen LK, Kelly HA. Annual report of the Australian National Poliovirus Reference Laboratory and summary of acute flaccid paralysis surveillance, 2001. *Communicable Disease Intelligence* 2002; 26:419-27.
- Thorley BR, Brussen KA, Stambos V, Kelly H. Annual report of the Australian National Poliovirus Reference Laboratory, 2002. *Communicable Disease Intelligence*. 2003; 27(3):352-6.
- Whitfield K, Kelly H. Estimating the incidence of acute flaccid paralysis in Victoria, Australia using the two-source capture-recapture method. *Bulletin World Health Organisation*. 2002; 80:846-851.
- Wood N, Thorley B. Towards global poliomyelitis eradication: The successes and challenges for a developed country. *Journal of Paediatrics and Child Health*. 2003; 39:647-50.

### Presentations

- Thorley B. *Poliomyelitis due to vaccine-derived polioviruses in the Philippines*. 8th National Public Health Association of Australia Immunisation Conference, Melbourne, 16-17 May 2002.
- Thorley B, Paladin F, Shimizu H. *Poliomyelitis due to vaccine-derived polioviruses in the Philippines*. XIIIth International Congress of Virology, Paris, 27 July - 1 August 2002.
- Whitfield K, Kelly H. *Sensitive surveillance? Estimating non-poliomyelitis acute flaccid paralysis in Victoria (1998 - 2000)*, 34th PHAA Annual Conference, Adelaide, 30th Sept - 2nd Oct, 2002.

### ADVERSE EFFECTS FROM COMPLEMENTARY ALTERNATIVE MEDICINES (CAM)

- Like any other treatment, Complementary Alternative Medicines (CAM) has the potential for causing adverse events.
- Several areas of particular concern have been identified through this study: significant dangers associated with dietary restriction in children; potential risks of CAM use in pregnancy; potential risks from overdose of CAM and the need for safe storage/child resistant packaging; dangers associated with substituting conventional medication with CAM.
- Clinicians need to ask about CAM use and be prepared to discuss potential benefits and potential risks.

#### Background

Paediatricians are aware that CAM is being used extensively in Australia, both in children with common childhood conditions (eg. Echinacea for the common cold) and in those with serious or chronic conditions (eg. a wide range of CAM in children with cancer).<sup>1</sup>

Conventional drugs must pass through a very comprehensive regulatory system before they can be licensed for sale or prescription. In contrast, compounds used as a part of CAM are not subject to similar regulations. It is likely that CAM use in children includes the full spectrum of therapeutic benefit: from good therapies with important potential roles in health care through to ineffective but harmless therapies and dangerous therapies. There are currently limited data about adverse events associated with the use of CAM. Apart from the APSU there is no other systematic national means of collecting national information about CAM-related adverse events in children.

#### Objectives

- To provide data on the incidence of major adverse events associated with the use of CAM in children in Australia;
- To provide information for paediatricians and other health practitioners about specific adverse events associated with the use of particular forms of CAM.

#### Case Definition

The occurrence of any adverse event, suspected or confirmed to be associated with the use of CAM, occurring in children up to 15 years of age, where:

- An “**adverse event**” is any unfavourable and unintended sign (including an abnormal laboratory finding), symptom or disease associated with the use of CAM, whether or not it is confirmed to be related to the therapy.
- “**Complementary or Alternative Medicine (CAM)**” includes any health care practice other than one intrinsic to the current conventional system.

#### Severity of adverse events are classified as follows:

- **Mild** – an adverse experience which is easily tolerated by the patient, causing minimal discomfort and not interfering with everyday activities (eg. a minor rash).
- **Moderate** – an adverse experience, which is sufficiently discomforting to interfere with normal everyday activities (eg. nausea and vomiting requiring time away from school).
- **Severe** – an adverse experience which is incapacitating and prevents normal everyday activities and/or requires therapeutic intervention such as the use of a prescription drug or hospitalisation.
- **Life threatening** – the patient is perceived to be at risk of death from the event as it occurred (eg. an anaphylactic reaction).
- **Fatal** – the patient died.

#### Examples of CAM

Detailed definitions of these and other CAM therapies can be found at our web-site, <[www.CAM-study.org](http://www.CAM-study.org)>:

Naturopathy; Homeopathy; Herbal therapy; Traditional Chinese Medicine; Kinesiology; Colour Therapy; Chiropractic; Hypnotherapy; Therapeutic Massage; Alexander Technique; Osteopathy; Reiki; Moxibustion; Acupuncture; Acupressure; Reflexology; Ayurveda; Aromatherapy; Therapeutic Touch; Yoga.

#### Results and discussion

The classification of the 29 reported cases of adverse events associated with CAM in 2002-2003 are shown in figure 11. Of the 29 reports, 25 were confirmed cases. The distribution of these is indicated in figure 12 below.

#### Of the 25 cases reported these are broadly classified as:

- Adverse events associated with a failure to use conventional therapy (Table 3) OR
- Adverse events associated with the use of medicinal CAM (Table 4).

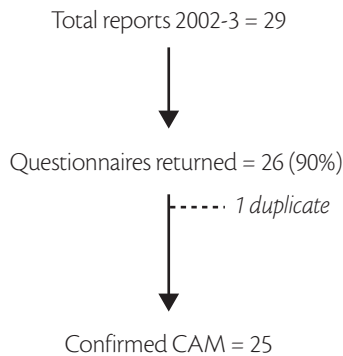


Figure 11. CAM surveillance data summary 2002-3



Figure 12. CAM distribution of confirmed cases 2002-3

**Table 3: Adverse events associated with a failure to use conventional therapies.**

CAM	ADVERSE EVENT
CAM used instead of anticonvulsants	Life-threatening seizures
Multiple CAM treatments	Delayed management of cerebral palsy
Chiropractic treatment irritable infant	Delayed diagnosis of UTI
Naturopathy for diabetes and change to insulin dose	Symptomatic hyperglycaemia
Failure to immunise in favour of CAM	Hib pneumonia
Failure to use anticoagulant in favour of CAM	Lung infarction

The severity of adverse events ranged from mild to fatal. Of particular concern were reports of adverse events (including dehydration or malnutrition) associated with significant dietary restriction in children, which is potentially extremely dangerous and resulted in one death. Other areas for concern included reports of adverse effects of CAM on the fetus when used during pregnancy. Risks associated with accidental or excessive ingestion of CAM were also identified. In one child, ingestion of a herbal remedy for vomiting resulted in acute hepatic toxicity requiring liver transplantation. Problems associated with changing from conventional therapy to CAM were also reported. In one child, substitution of a CAM for an anti-epileptic agent resulted in life threatening seizures.

The number of reports to APSU is likely to be an underestimation of the actual number of events occurring. This may result from paediatricians failing to report events or not recognising them. Alternatively, events may be detected by health care workers such as general practitioners, or families may not disclose CAM use or present for review. The higher number of reports from Victoria compared with other states is likely to reflect a higher awareness and reporting rate, rather than a greater number of adverse events.

## Conclusion

These sentinel cases highlight the fact that adverse events associated with the use of CAM do occur in Australian children. The perception that CAM is natural and therefore harmless needs to be changed. Safety monitoring of any therapy is an important part of optimal care.

In the interest of public safety, a good case can be made for improving the regulatory framework within which CAM is practised. This is particularly relevant to the use of CAM in children, given that they do not seek these forms of therapy for themselves but rely on the quality of decisions made by others. Clinicians need to be aware of the high rates of CAM use, should include questions about use in their patient history, and should be prepared to discuss potential benefits and adverse effects with families.

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**Table 4: Adverse events associated with use of medicinal CAM**

CAM	ADVERSE EVENTS
Homeopathy and dietary restriction	Malnutrition, sepsis and death
Homeopathic medicines*	Seizure and apnoea
Herbal remedy for vomiting*	Liver failure requiring liver transplant
Herbal medicines*	Intra-operative bleeding
Diet and fluid restriction	Dehydration, encephalopathy and refeeding syndrome
Alternative medical practitioner prescribed triiodothyronine for obesity	Admission with thyrotoxicosis
IM injection of vitamins daily	Left sciatic neuropraxia
Oral and IV calcium supplements from alternative practitioner	Severe hypercalcaemia
Gingko and brahmi overdose	Admitted for observation
Food supplement with taurine and inositol	Vomiting requiring IV rehydration
Homeopathy (pulsatilla) for cough	Vomiting, drowsiness and fever
Homeopathy and dietary restriction	Malnutrition with oedema
High dose vitamin B	Carotenaemia
Rice milk for constipation	Failure to thrive
Vitamin mixture	Allergic reaction
Chamomile tea in excessive amounts	Worsening constipation
Herbal medicine adulterated with steroid	Steroid excess

\*Mechanism unknown

## Publications arising from this study:

### Original Articles

1. South M, Lim A. Use of complementary and alternative medicine in children – too important to ignore. *J Paediatr. Child Health* 2003; 39; 8:573.

### Abstracts

2. Lim A, Cranswick N, Skull S, South M. Adverse Events associated with the use of Complementary and Alternative Medicine in Australian children. *Clinical Pharmacology and Therapeutics*. 2002; 71(2): P76. (Presented American Society for Clinical Pharmacology and Therapeutics Meeting, Atlanta USA March 2002. Poster Presentation and invited trainee oral presentation).
3. Lim A, Cranswick N, Skull S, South M. Surveillance of Adverse Events Associated with the Use of Complementary and Alternative Medicine. *Journal of Paediatrics and Child Health* 2002;38(5) A3. (Presented Royal Australasian College of Physicians Annual Scientific Meeting, Brisbane May 2002: Finalist Rue Wright Memorial Prize).

### Presentations

4. Lim A, Cranswick N, Skull S, South M. The Australian Health and Medical Research Congress.. Oral Presentation: *Adverse events associated with the use of complementary and alternative medicine in children*, Melbourne Australia November 24-29, 2002.

## ■ CHARGE association

- The complex diagnostic issues and multi-system involvement in CHARGE association require genetic review and a multidisciplinary team approach.

### Background

CHARGE association (Syndrome) is a non-random pattern of congenital abnormalities including coloboma, choanal atresia and cardiac, renal, genital and gastrointestinal abnormalities. The incidence of CHARGE association in Australia is not known as no systematic study has been performed. International incidence estimates range from 0.1-1.2 per 10<sup>5</sup> live births. Diagnosis of this condition has been hindered by inconsistencies in diagnostic criteria. Diagnosis may also be delayed, as major features of the

**Table 5: CHARGE association criteria used in the APSU study**

Major criterion	Includes
Coloboma	Iris, retina or choroid
Choanal atresia	Unilateral or bilateral; membranous or bony; stenosis or atresia
Characteristic ear abnormalities	External ear (lop or cup shaped); Middle ear (ossicular) malformations, Chronic serous otitis; Mixed deafness; Cochlear defects
Cranial nerve dysfunction	Anosmia; Facial palsy (unilateral or bilateral); Sensori-neural deafness, Vestibular problems and swallowing difficulties
Minor criterion	Includes
Genital hypoplasia	Males: micropenis, cryptorchidism; Females: hypoplastic labia Both: delayed and incomplete pubertal development
Developmental delay	Delayed motor milestones, hypotonia, mental retardation
Cardiovascular malformations	All types: especially conotruncal defects (eg. Tetralogy of Fallot / Aortic arch anomalies) and Atrioventricular canal defects
Orofacial clefts	Cleft lip and / or palate
Tracheo-oesophageal fistula	Tracheo-oesophageal defects of all types
Distinctive face	Unilateral facial weakness / asymmetry, broad forehead, flat malar region and small mid-face, and thin lips

disorder may not be readily identifiable, particularly in young children. Adding to the confusion, the CHARGE association shares many of its features with other genetic and birth defect syndromes, such as Velocardiofacial syndrome.

### Objectives

- To estimate the incidence of CHARGE association seen by child care specialists in Australia;
- To describe the epidemiology of CHARGE association;
- To describe the clinical features of CHARGE association;
- To document the morbidity and mortality of CHARGE association.

### Case Definition

This study uses the revised consensus criteria Blake (et al, 1998)<sup>1</sup> recommended to enhance clinical diagnosis and facilitate research. Clinicians were asked to report **any child under 16 years of age seen during the previous month with a newly diagnosed CHARGE Association** – that is live-born children with:

- three or more major criteria alone or
- one or more major criteria with at least two minor criteria (see Table 5).

### Results

The classification and distribution of the 61 reports of CHARGE association made to the APSU between January 2000 and December 2002 are shown in figures 13 and 14. The estimated incidence of CHARGE association in Australia is 2.8 per 10<sup>5</sup> live births (95% CI 1.8-4.3 per 10<sup>5</sup>).

### Sample Characteristics

Of the 23 children with CHARGE association, 12 (52%) were female. The median age of children when given the provisional diagnosis of CHARGE association was 12 days (range 1-75 days), 15 (65%) children were given a provisional diagnosis of CHARGE association before age one month. None of the children were known to have a parent or sibling with CHARGE. Karyotype was available for 20 (87%) children, all were normal. However, the use of FISH deletion 22q11.2 to detect and exclude velocardiofacial syndrome was known to have been done for two children only.

## Surveillance Overview

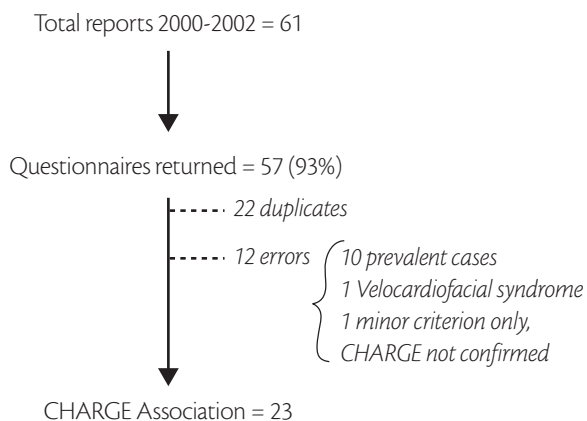


Figure 13. CHARGE surveillance data summary 2000-2

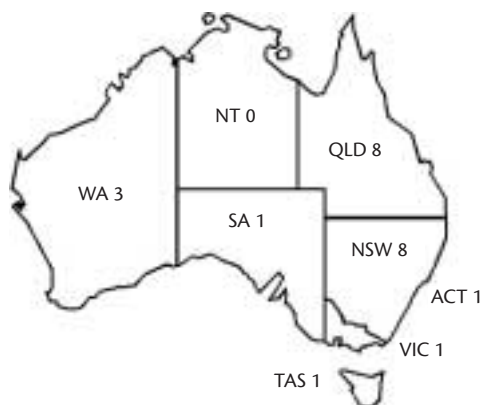


Figure 14. Distribution of children with CHARGE 2000-2

### Clinical picture

Only four children experienced a perinatal period unaffected by any of the following complications: poly- or oligohydramnios (n=7, 30%); preterm birth (n=13, 57%) or resuscitation required at delivery (n=16, 70%). Six of the reported children are known to have died in the first year of life, three of those in the first month of life. Three children died from sepsis, one from Epstein's anomaly, one child died unexpectedly from suspected aspiration and life support was withdrawn from one severely affected child with multiple anomalies.

All children presented with a complex of major and minor CHARGE criteria (Table 6). A range of other problems were reported in 20 (87%) of the children, the most common of these were abnormal kidneys (n=6, 26%), bowel problems (n=5, 25%), seizures (n=3, 13%), and immune deficiency (n=2, 9%).

Most of the children reported required lengthy hospitalisation around the time of birth for the investigation and management of multiple problems. At the time of diagnosis, a developmental

Table 6: Distribution of characteristics reported for the 23 children reported with CHARGE Association

MAJOR CRITERION	n	%
Coloboma	21	91
Choanal atresia	14	61
Characteristic ear abnormalities	22	96
Cranial nerve dysfunction	17	74
MINOR CRITERION	n	%
Genital hypoplasia	12	52
Developmental delay*	10	44
Cardiovascular malformations	19	83
Orofacial clefts	6	26
Tracheo-oesophageal fistula	3	13
Distinctive face	12	52

\* does not include data on the 6 children who died

\*\* Canadian study criteria include 4 major criteria as above and 7 minor criteria (including 6 criteria above plus growth deficiencies)

specialist was also known to be involved in the care of four of the surviving children. The study investigators do not have ongoing access to information on the health service needs of children with CHARGE and this is an important issue for future research.

### Conclusion

Although CHARGE association is the most likely diagnosis for children meeting the stated criteria, other diagnoses still need to be considered. Velocardiofacial (Sphrintzen) and DiGeorge syndromes should be distinguished on clinical grounds. FISH analysis for 22q11.2 deletion will confirm these diagnoses in most instances.

All the children will have their DNA analysed using Comparative Genomic Hybridization (CGH) for the newly discovered CHD 7<sup>2</sup> (Chromodomain Helicase DNA binding protein 7 gene deletion). The microdeletion is located on the long arm of chromosome 8 and analyses will be performed by Dr Conny vanRavenswaaij in Nijmegen University, Holland in 2005.

Blake et al in the Canadian Paediatric Surveillance Unit used four major (same as our study) and seven minor criteria<sup>3</sup>. These criteria are indicated in table 6 above. For a firm diagnosis the authors accepted all four or three major criteria and three minor criteria. The addition of seventh criteria (growth deficiency) is not believed to improve diagnostic accuracy as many factors impact on growth.

The complex diagnostic issues and multi-system involvement in CHARGE Association require genetic review and a multidisciplinary team management approach. The paediatrician has a pivotal role



to play in the coordination of care and advocacy for children with CHARGE association and their families. Most parents join CHARGE parent association and continue to collaborate in research on this condition. A further long-term study assessing the neurodevelopment and education of this cohort is being planned.

## Acknowledgements

- Child health specialists contributing to the Australian Paediatric Surveillance Unit.
- The Australasian CHARGE Parent's Association for their financial support.

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## Publications arising from this study:

### Presentations

4. Williams G. *The APSU CHARGE study*. 5th International CHARGE Meeting in Cleveland, USA. July 2003.
5. Williams G. *Sleep disorders in Children with CHARGE*. 5th International CHARGE Meeting in Cleveland, USA. July 2003.
6. Williams G. *The CHARGE Brain*. 5th International CHARGE Meeting in Cleveland, USA. July 2003.

## CONGENITAL CYTOMEGALOVIRUS INFECTION (cCMV)

- Flu-like illness during pregnancy was a common feature in mothers of children with confirmed cCMV infection.
- Women who present with a febrile illness during pregnancy should be considered for serological testing for CMV in order to identify children who may need supportive treatment or early intervention.

### Background

Congenital Cytomegalovirus Infection (cCMV) remains an important and serious cause of congenital viral infection in Australia. It is estimated that the number of children with cCMV infection in Australia is between 200 and 800 cases per year. However, 90% of infected children are asymptomatic.

In the 10% of symptomatic children, clinical features range from moderate enlargement of the liver and spleen (with jaundice) to fatal illness (cytomegalic inclusion disease). Of symptomatic children, 80% to 90% will develop complications within the first few years of life, including hearing loss, visual impairment, developmental delay and neuromotor disabilities. Between 5% and 10% of infected infants who are without symptoms at birth will subsequently have varying disabilities recognised later in life, the most common being sensorineural hearing loss.

### Objectives

The study aims:

- To determine the incidence of cCMV and suspected cCMV in Australian children;
- To determine the presenting features and clinical spectrum of disease due to cCMV;
- To determine the current therapy in use for cCMV infection;
- To determine the epidemiology of cCMV including prevalent cCMV subtypes prior to trials of vaccines and antivirals.

### Case Definition

#### Definite congenital CMV infection

- Any child from whom CMV is isolated in the first three weeks of life, from urine, blood, saliva, or any tissue taken at biopsy.

#### Suspected congenital CMV infection

- Any child up to 12 months of age, in whom CMV is isolated from urine, blood, saliva or any tissue taken at biopsy AND/OR
- a positive serum IgM is found AND
- in whom clinical features exist that may be due to intrauterine CMV infection.

### Results

The classification and distribution of the 42 reports of CMV made during 2002 and 2003 are reported in Figures 15 and 16 below. Forty clinicians made a report of cCMV to the APSU during the study period and 2 reports came through other sources. Clinical data was provided for only 18(40%) of notified cases. Of the 42 notifications 5 were confirmed cases and 10 were a probable cases. The geographic distribution of these cases is indicated in Figure 16.

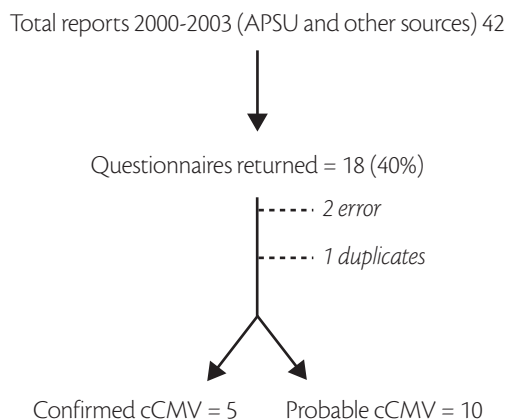


Figure 15. CMV surveillance data 2002-3



Figure 16. CMV distribution of confirmed and probable cases 2002-3

**Table 7. Clinical signs of CMV infection in children\*.**

Clinical sign	Number of affected children	Clinical sign	Number of affected children
Microcephaly	1	Petechia, purpura	2
Hepatitis	3	Pneumonitis	1
Hepatomegaly	3	Small for gestational age	1
Intracranial calcifications	3	Splenomegaly	2
Jaundice	2	Thrombocytopenia	2
		Maternal illness during pregnancy	4

\* Some children have more than 1 sign. No child presented with developmental delay or encephalitis

## Clinical Features

Petechiae, purpura, thrombocytopenia, jaundice, hepatomegaly, splenomegaly and hepatitis are the most consistent features of cCMV infection. The most common feature in cases of cCMV infection was maternal flu- like illness present during pregnancy in four of the five confirmed cases (Table 7).

## Conclusion

The current incidence of CMV is not known. Reported rates of cCMV infection to APSU are low and may reflect the low questionnaire return rate from clinicians. This return rate is lower than all other APSU studies and may reflect the complexity of the questionnaire which has been redesigned for 2005 with the aim of increasing participation rates.

Women who present with a febrile illness during pregnancy should be considered for serological testing for CMV in order to identify children who may need supportive treatment or early intervention.

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1. Revello, M. G. & Gerna, G. Diagnosis and management of human cytomegalovirus infection in the mother, fetus, and newborn infant. *Clinical Microbiology Reviews* 2002; 15:680-715.

## Publications arising from this study:

### Original Articles

2. Trincado DE, Rawlinson WD. Congenital and perinatal infections with cytomegalovirus. *Journal of Paediatric Child Health* 2001; 37:187-192.

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### Presentations

5. Munro S. C., Trincado D. E., Maine, G. and Rawlinson W. D. *A Study of cytomegalovirus infection in Pregnancy*. Presented at the 9th International Cytomegalovirus Workshop, Maastricht, The Netherlands, 2003.
6. Trincado D. E., Munro S. C. and Rawlinson W. D. *Human cytomegalovirus infection of the placenta and congenital disease*. Presented at the 27th International Herpes Virus Workshop, Cairns, Australia, 2002.

## CONGENITAL RUBELLA

- Most cases of congenital rubella in Australia are 'imported', that is they occur in children born to unvaccinated women who are born outside Australia.
- Two cases were notified after a recent resurgence of rubella in Queensland. Both were children of Australian born mothers who may have missed the school vaccination programme.
- Broad immunisation coverage in childhood and detection and vaccination of susceptible women of child-bearing age before they become pregnant or immediately post-natally is necessary to prevent further cases. Screening and vaccination of immigrant women is also important.

### Background

Rubella vaccination in Australia commenced in 1971 for schoolgirls and susceptible ante-partum women. Since 1989, both boys and girls have received routine measles-mumps-rubella (MMR) vaccine at twelve months of age. In 1994, vaccination of teenage boys was introduced and in 1998 all primary school aged children received a second dose of MMR vaccine. Since 1999, all children receive two doses of MMR vaccine before they start primary school.

### Objectives

- To document the incidence of congenital rubella infection;
- To determine the vaccination status of mothers of affected infants;
- To monitor the effectiveness of the current vaccination program.

### Case Definition

Any newly diagnosed child or adolescent <16 years of age who, in the opinion of the notifying paediatrician, has definite or suspected congenital rubella, with or without defects, based on history, clinical and laboratory findings.

### Results

There have been 103 notifications of congenital rubella since 1993. (Figure 17) and 11 notifications of congenital rubella infection in 2002-2003. Of the 7 newly confirmed cases, five were born to women whose country of birth was outside Australia. These included one severely affected infant from Western Australia who died on day four of life. This infant's mother was infected with rubella while pregnant in Indonesia. Three infants were notified from NSW. One child was born in Indonesia and the Indonesian mother was infected there. The other two cases from NSW were born in Australia to mothers who were born overseas (Goa and the Philippines). Both mothers of these infants were not vaccinated prior to pregnancy. A fifth infant was born in Victoria in 2001, but was not notified to the APSU until 2002. The parents were Fijian and it is not known where the mother acquired her infection.

The confirmed cases from Queensland were born to Australian-born mothers. Both women reportedly missed rubella school girl vaccination due to illness and had no follow up vaccination arranged. Both infants had severe bilateral deafness. The distribution of the 7 confirmed cases are indicated in Figure 18.

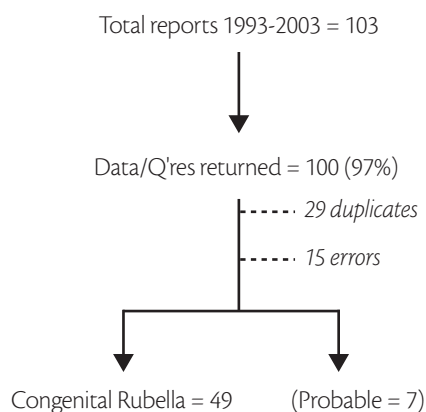


Figure 17. Congenital Rubella Surveillance data summary 1993-2003

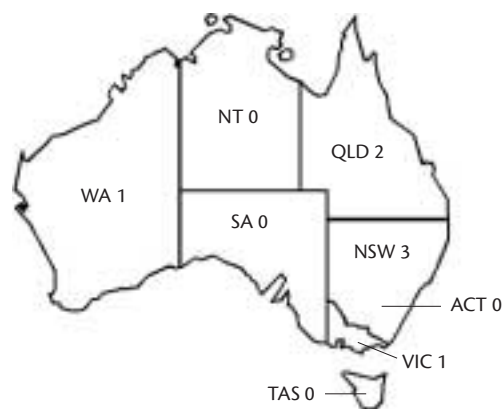


Figure 18. Congenital Rubella – distribution of cases 2002-3

## Conclusion

Two cases of congenital rubella were reported from Queensland in 2003, after an increase in rubella notifications in that State in 2001-2002<sup>1</sup>. The national Measles Control Campaign in 1998 aimed to give measles-mumps-rubella (MMR) vaccine to all unvaccinated preschoolers and a second dose to primary schoolchildren. Following the campaign no children with congenital rubella defects were born to Australian residents during the 5 years 1998 to 2002, according to reports to the Australian Paediatric Surveillance Unit. However, two cases were reported in Australian born mothers in 2003. Five imported cases also occurred. Broad immunisation coverage and detection and vaccination of susceptible women of child-bearing age before they become pregnant are necessary to prevent further cases.

## References referred to in text:

1. Forrest JM, Burgess M, Donovan T. A resurgence of congenital rubella in Australia. *Communicable Disease Intelligence*. 2003; 27(4):533-535.

## Report authors/study investigators

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## Publications arising from this study:

### Original Articles

2. Forrest JM, Burgess M, Donovan T. A resurgence of congenital rubella in Australia. *Communicable Disease Intelligence*. 2003; 27(4):533-535.
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## ■ CONVERSION DISORDER

- Conversion disorder is not “rare” in paediatric practice.
- Children with conversion disorder may often have complex family-social concerns that need to be managed.
- A multidisciplinary approach (including medical and mental health providers) to the diagnosis and management of medically unexplained symptoms in childhood is recommended. This will minimise diagnostic delay and may reduce the risk of subsequent episodes involving medical and mental health providers.

### Background

Conversion disorder refers to a disturbance of body function characterised by neurological, sensory or motor symptoms that have no organic explanation and for which an underlying psychological cause is proposed. In this study we aimed to describe the clinical features in Australian children, presenting to child health specialists with conversion disorder.

### Objectives

- To estimate the incidence of conversion disorder seen by child health specialists in Australia;
- To describe the clinical and other associated features of conversion disorder;
- To describe the investigation and management of children with conversion disorder in Australian children.

### Case Definition

Any child aged <16yrs presenting with a newly diagnosed episode of Conversion disorder according to the following criteria:

- One or more symptoms or signs affecting voluntary motor or sensory function. (Children with pain or fatigue only are excluded from this study);
- Symptoms / signs cannot be adequately explained by a medical condition after full investigation;
- No evidence that symptoms are intentionally produced;

Consistent with DSM-IV clinical criteria, the diagnosis of conversion disorder should only be made if symptoms cause significant distress or impairment

### Results

The classification and distribution of the 310 reports of Conversion disorder made to the APSU during 2002 and 2003 (Figures 19 and 20). One hundred and nineteen clinicians (11% of the APSU contributor list) made a report of conversion disorder to the APSU during the study period. Although child and adolescent psychiatrists made the majority of reports (46%), 30% of reports were made by clinicians in general paediatric practice.

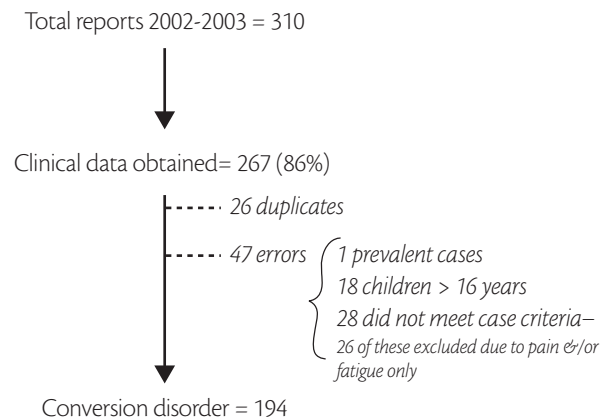


Figure 19. Conversion surveillance data summary 2002-3

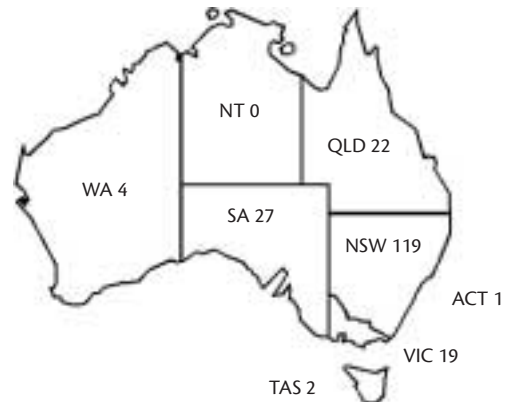


Figure 20. National distribution of confirmed Conversion disorder cases 2002-3

### Sample Characteristics

Consistent with the adult literature, more girls than boys (n=142, 71%) were diagnosed with conversion disorder. The age range was 3.06 to 15.98 years (mean 11.77 years; SD 2.70). Conversion disorder is an episodic condition and for 22% of the 194 cases, the presentation notified was not the initial episode of conversion disorder. Three children re-presented with a new episode of conversion during the two-year study period.

## Clinical picture

The clinical picture of children presenting with conversion disorder was often complex. One third of the sample presented with three or more symptoms or signs of conversion disorder. The most common symptom clusters were pseudoseizure (23%); motor concerns (including weakness, abnormal movement, or paralysis) in 64% of cases; sensory symptoms (including parasthesias or lack of sensation, sight or hearing difficulties) in 24%; and respiratory concerns (including psychogenic cough or stridor) in 19% of children. Pain (56%) and/or fatigue (34%) were commonly reported in addition to conversion symptoms.

## Duration of symptoms prior to diagnosis

Although just under 50% of cases were diagnosed within one month of symptom onset, 23% of children reported symptoms for more than six months prior to diagnosis. The average time to diagnosis was not observed to be significantly different between children presenting with initial or subsequent episode of conversion disorder.

## Antecedents

Reported rates of a history of mental illness was high for both the children (47%) and family of origin (30%). Anxiety disorder was the most frequently reported additional mental health diagnosis in the children (n=45, 23% of cases).

The association of conversion disorder with adverse life events has been noted since the nineteenth century. Reporting clinicians identified at least one significant antecedent life event or stressor for 63% of cases (Figure 21).

Two or more significant events or stressors were recorded for 34% of children. Family separation or loss (32% of cases) was the most frequently reported stressor (Figure 21).

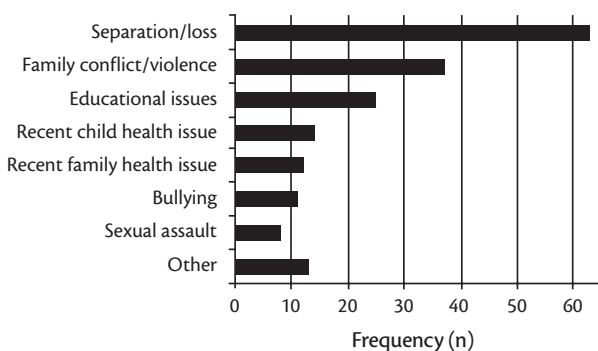


Figure 21. Antecedent stressors reported by clinicians

## Investigations and management

The reported rate of health service usage was high. Almost all children underwent multiple investigations and had contact with

multiple clinicians. Seventy three percent of children were seen by a psychiatrist or psychologist and 14% received a psychotropic medication for a co-morbid psychiatric disorder.

Admission to hospital was required in 70% of cases, for a mean of 10.2 days (range=1-90 days). There was a trend for shorter periods of hospitalisation for children with initial episodes of conversion (mean 9.1 days) than for children diagnosed with subsequent episodes of conversion disorder (mean 13.6 days).

## Conclusion

The excellent contribution by APSU clinicians to this study has enabled description of the diverse range of presentations in children diagnosed with conversion disorder. This study has also demonstrated that conversion disorder in children is not uncommon and is associated with a significant burden for the child, family and the health system.

This national "snapshot" of the condition provides invaluable evidence which will inform future longitudinal research studies to monitor the burden of disease, health services usage and relapse rates in Australian children with conversion disorder. This study emphasises the need for a multidisciplinary approach (involving medical and mental health providers) to the diagnosis and management of medically unexplained symptoms in childhood.

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## Publications arising from this study:

### Abstracts

- Rose D, Kozłowska K, Nunn K, Morris A, Et al. A picture of conversion disorder in Australian children in 2002. *Journal of Paediatrics and Child Health* 2003; 39(suppl), A8.

### Presentations

- Rose D, Kozłowska K, Nunn K, Morris A. A picture of conversion disorder in Australian children in 2002. APSU Young Investigators Award Finalist, 2002. RACP Annual Meeting, 2003.
- Rose D, Kozłowska K, Nunn K, Morris A. A picture of conversion disorder in Australian children in 2002. Invited presentation APSU Annual Scientific Meeting, Hobart, May 28-29, 2003.

## EARLY ONSET EATING DISORDER (EOED)

- Children as young as five may display the cognitive symptoms of eating disorder and concerted food avoidance.
- In addition to weight loss or failure to gain weight, many children with early onset eating disorder have significant medical co-morbid and/or mental health problems.
- International comparisons between Australia, Canada and the UK data will be possible because of research on EOED that is currently being conducted through the Canadian Paediatric Surveillance Unit (2003) and the British Paediatric Surveillance Unit since 2004.

### Background

Epidemiological studies suggest that the prevalence of eating disorders, including anorexia nervosa, has been increasing in adolescents over the last 50 years. However, there is wide variation in the few available estimates of rates of eating disorders in children under 13 years of age.

There is ongoing debate in the literature about how to apply the current diagnostic criteria for eating disorders to children and younger adolescents. Current DSM-IV criteria for anorexia nervosa require evidence of patient concerns about body weight, disturbed body image and fear of weight gain. However, these criteria may not accurately reflect the clinical features in young children for several reasons:

- Young children may not report fear of weight gain while at a low weight but may do so only when weight has been restored to a more healthy level.
- Children may be unable to express distress in terms of body shape and self-perception but may instead describe somatic symptoms such as abdominal pain or discomfort once re-feeding commences.
- While the presence of secondary amenorrhoea is an important diagnostic feature for anorexia nervosa in post-menarchal girls it is a developmentally inappropriate criterion in young girls, in whom a history of delay in onset of puberty may be important.
- The DSM-IV criteria specifies that weight should be <85% of expected weight for height, however, this may lead to an underestimate of the severity of low weight in younger children in whom linear growth has also been affected.

We do know that significant medical and psychological complications arise from starvation, weight loss or lack of appropriate weight gain during childhood and adolescence, making this group of conditions important to recognise and treat appropriately.

### Objectives

- To estimate the incidence of early onset eating disorders seen by child health specialists in Australia;
- To describe the epidemiology of early onset eating disorders;
- To describe the range of clinical features at presentation, including other psychiatric illness;

- To compare the features at presentation in this population with current DSM IV criteria;
- To describe the acute medical complications experienced by children with early onset eating disorders;
- To describe the therapeutic interventions used in management;
- To contribute data to an international comparison of the diagnosis of management of early onset eating disorders.

### Case Definition

Children aged 5-13 years inclusive, newly diagnosed with an early onset eating disorder, defined as:

- Determined food avoidance AND
- weight loss or failure to gain weight during a period of growth, not due to any identifiable organic cause AND
- Child is admitted to hospital OR managed within the community.

The first 12 months of the study excluded children managed within the community. However, variation in national management strategies for this condition resulted in a change to the case definition from August 2003 to include children managed in the community.

### Results

The classification and distribution of the 86 reports of early onset eating disorder made to the APSU between July 2002 and December 2003 are reported in figures 22 and 23 below. Of the reported cases, 71% are female and 38% are younger than 12 years of age.

### Clinical picture

The clinical features of children reported with EOED are shown in table 8. The median duration of symptoms of eating disorders was six months (Range: 1-24 months). All children exhibited food avoidance behaviour and over 91% were preoccupied with food. In the six months prior to diagnosis four children (9%) failed to gain any weight. Decreased weight was observed in 89% of cases. Median weight loss was 6 kg (Range: 2 to 16 kg). Eight of the nine females who had reached menarche had secondary amenorrhoea.



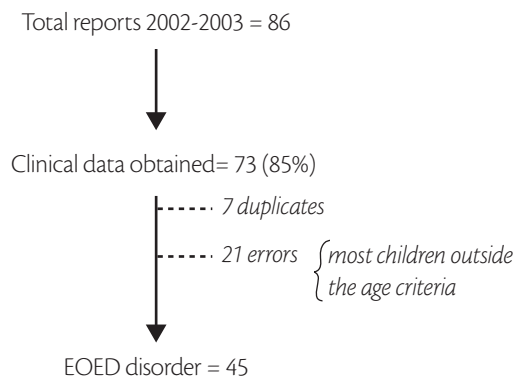


Figure 22. EOED surveillance data summary 2002-3

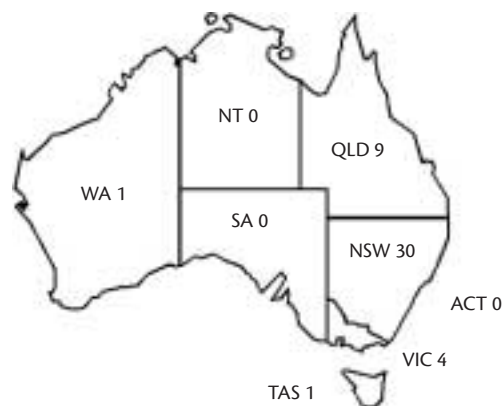


Figure 23. National distribution of confirmed EOED cases 2002-3

Sixty nine percent of Australian children were reported to have a concurrent mental health concern, most commonly depression (18/45, 40%) and/or anxiety (17/45, 38%). A family history of mental illness was reported for 16 children (36%), six of whom had family member with a history of anorexia or bulimia.

Surveillance of early onset eating disorder in Canadian children (5-12 years of age) commenced in January 2003. Clinical features of the 63 children (83% female) with early onset eating disorder identified in the first 12 months of the Canadian study are included in Table 8.

### Management

Hospitalisation was a reporting criterion for the first 12 months of the APSU study, therefore a clear picture of national management strategies for young children with eating disorder is not yet available. Of the 45 Australian children diagnosed with an eating disorder 91% were hospitalised. Duration of admission ranged from four to more than 70 days. Almost all children received multi-disciplinary care and over 60% were managed in a specialist eating disorder unit. Two-thirds of children hospitalised (n=27/41) received naso-gastric tube feeding. Just under half the children were prescribed psychotropic medication for concurrent mental health problems. All children were alive at the time of reporting.

Table 8: Clinical features of Australian and Canadian<sup>#</sup> children with early onset eating disorder

SYMPTOMS at PRESENTATION	Australian %	Canadian %
Food Avoidance	100	100
Preoccupation with food	91	88
Fear of weight gain	76	86
Preoccupation with weight	69	84
Misperception body shape	67	72
Excessive exercise	47	58
Self-induce vomiting	18	11
Denial of severity	87	65
Diuretic or laxative abuse	0	0
Weight loss	89	89

### OTHER CLINICAL FEATURES\*

Bradycardia (<36 beats/min)	53	28
Temperature (<35.5°C)	40	13
Hypotension (systolic BP<80)	18	29

\* 1st 12 months of APSU surveillance hospitalisation was reporting criteria  
 # Data courtesy of Dr Anne Morris, Co-investigator Canadian Team

### Conclusion

These preliminary findings indicate that while not all children reported with early onset eating disorder meet full DSM-IV criteria for anorexia or bulimia nervosa, they are generally at significant biological and psychological risk. The profile of clinical features at the time of diagnosis is generally consistent with those being reported in the Canadian cohort, although hypothermia and bradycardia were reported in a higher proportion of Australian children. This may be an indication of severity associated with the requirement that only children who were hospitalised be reported during the first year of the APSU study.

The surveillance of early onset eating disorder in Australia, Canada and the UK will provide much needed epidemiological data on the burden of this disease in young children and enable the documentation of variation in national and international management practices.

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## FETAL ALCOHOL SYNDROME (FAS)

- Since 2001, there have been 53 children with confirmed Fetal Alcohol Syndrome (FAS) reported to the APSU. These children are frequently in care and/or born to mothers with multiple substance use and are demanding of health and education resources.
- During the study period 22 of the children reported failed to meet the case definition of FAS used in this study.<sup>1</sup> For some of these notifications there was insufficient clinical information, making it difficult to apply our stringent case definition criterion. The majority of these children had alcohol related effects.
- We wish to thank clinicians who notified cases of FAS and provided us with clinical and other information.

### Background

FAS was first identified in the 1970s and has been described as a preventable tragedy.<sup>2</sup> FAS is caused by maternal alcohol consumption during pregnancy and represents the most severe effects of exposure to alcohol *in utero*. Children with FAS display a wide range of physical defects and disabilities, however the cardinal features are:

- Minor cranio-facial abnormalities;
- Prenatal and/or postnatal growth deficiency;
- Evidence of damage or dysfunction of the central nervous system.

A definite diagnosis of FAS requires identification of all three of these features as well as confirmation of prenatal alcohol exposure.

Most epidemiological studies on FAS have been performed in the USA and Canada where birth prevalence estimates range from 0.26-7.2 per 1,000 live births<sup>3,4</sup>. In The Western Cape Province in South Africa birth prevalence is 39 per 1,000 births<sup>5</sup>. However, data from the Western Australian Birth Defects Registry (1987-1997) suggest a birth prevalence of only 0.18 per 1,000 live births (0.02 per 1,000 non-Indigenous live births; 2.76 per 1,000 Indigenous live births)<sup>6</sup>. A study of children born in the Northern Territory (NT) (1990-2000) estimated a birth prevalence of 0.68 per 1,000 live births (1.9 per 1,000 Indigenous live births)<sup>7</sup>.

### Objectives

- To estimate the incidence of newly diagnosed FAS seen by child health specialists in Australia;
- To describe the epidemiology of FAS (gender, age, geography, ethnicity, SES);
- To describe the clinical features of FAS including developmental and physical co-morbidity;
- To describe use of health services by children with FAS;
- To document the use of other harmful substances eg drugs, by mothers of children with FAS;
- To increase clinicians' awareness of FAS by provision of information on clinical features and diagnostic criteria.

### Case Definition – consistent with the Institute of Medicine<sup>1</sup>

Clinicians were asked to report any child aged <15 years with newly diagnosed:

#### 1. Fetal Alcohol Syndrome – alcohol exposure confirmed, defined as:

- Evidence of prenatal alcohol exposure AND
- All characteristic cranio-facial abnormalities AND
- Pre-natal or post-natal growth deficiency AND
- Structural abnormalities or dysfunction of the CNS

#### 2. Partial Fetal Alcohol Syndrome – alcohol exposure confirmed, defined as:

- Evidence of prenatal alcohol exposure AND
- All characteristic cranio-facial abnormalities AND
- Pre/post natal growth deficiency OR
- Some characteristic cranio-facial abnormalities AND
- Structural abnormalities or dysfunction of the CNS

#### 3. Suspected Fetal Alcohol Syndrome – exposure not confirmed, defined as:

- All characteristic cranio-facial abnormalities AND
- Pre/post natal growth deficiency AND
- Structural abnormalities or dysfunction of the CNS

### Results

Of the 53 children who met the Institute of Medicine's<sup>1</sup> case definition for FAS used in this study, 38% of children had FAS with alcohol exposure confirmed, 58% children had Partial FAS with alcohol exposure confirmed and 4% of children had Suspected Fetal Alcohol Syndrome with alcohol exposure not confirmed (Figure 24).

The reported birth prevalence for FAS for 2001-2003 in this study is 2.41 per 100,000 live births (95% CI 2.30-2.52). Eighteen children with confirmed FAS were under 12 months old. The median age at the time of diagnosis was 2.8 years (newborn to 12 years), 55% were male and 53% were identified as Indigenous. Information on Indigenous status was not available for five children. Cases were reported from all states except Tasmania (Figure 25).

The family history was incomplete for some children in this study. Of the 53 children, 15% of mothers only had an educational attainment of primary school, 32% completed secondary school and the education of the remaining 51% was unknown. Twenty six percent of children had siblings with FAS, however, 40% of cases had no recorded information about siblings.

Only 42% of the cases were living with their biological parent(s), 13% lived with their grandparents or other relatives, and 43% were adopted or fostered.

Seventy two percent of children were exposed to other substances in utero. These included nicotine (66%), cocaine (1%), heroin (3%), glue or solvents (2%), marijuana (26%), and other drugs including carbamazepine, benzodiazepines, naltrexone, sodium valproate, morphine, pethidine, amphetamine and chlorpromazine.

All children had been referred to one or more health related agencies. These included specialty paediatric services (76%), child development or disability services (38%), respite services (11%), psychological medicine services (13%), the department of community services (68%) and remedial education services (30%).

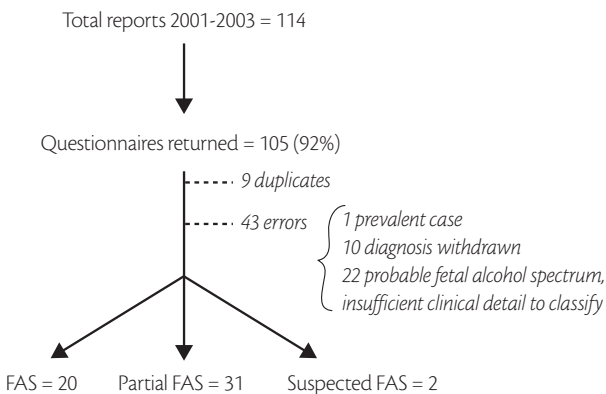


Figure 24. FAS surveillance data summary 2002-3

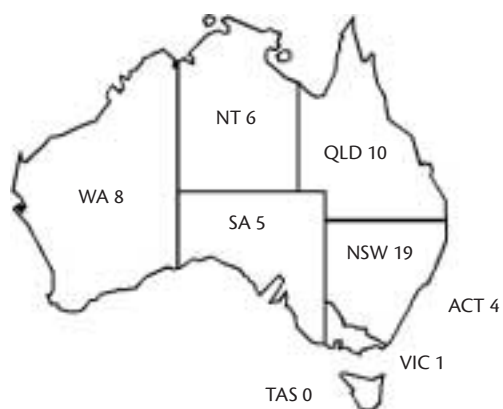


Figure 25. Distribution of FAS Cases 2002-3 (including confirmed FAS, partial FAS and suspected FAS)

## Conclusion

Based on the first three years of the APSU study, the reported birth prevalence was much lower than rates reported in North America<sup>(3,4)</sup>, South Africa<sup>5</sup>, WA<sup>6</sup> and the NT<sup>7</sup>. This may reflect under diagnosis or under-reporting and should be regarded as a minimal estimate. Other difficulties include the obtaining of infant data in children in foster care and ensuring the validity of retrospective data on alcohol consumption during pregnancy. Furthermore the average age of diagnosis was 3 years, suggesting the diagnosis is frequently missed at birth. A study from WA investigating the knowledge and practices of health professionals suggests that health professionals lack knowledge on diagnosis and management of FAS.

Aboriginal and Torres Strait Islander children living in remote communities may be under-diagnosed and under-reported to the APSU due to lack of access to paediatric services. Despite this Aboriginal and Torres Strait Islander children with FAS or suspected FAS are over-represented in this study. Many are not living with their parents and for many children complete data is not available.

FAS contributes significant social, medical and educational burdens to affected children, their families and the community. In order to address these burdens, estimates of the size of the problem need to be addressed. Information through the APSU provides an estimate of the numbers of children with FAS seen by paediatricians and provides invaluable descriptive data about both clinical features and health service usage.

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## Publications arising from this study:

### Presentations

1. Bower C, Elliott E, Payne J, Morris A, Ridley G, Haan E. Fetal Alcohol Syndrome, Australian Paediatric Surveillance Unit Scientific Meeting, Hobart, 29 May 2003.
2. Elliott E, Payne J, Bower C, Morris A, Ridley G, Haan E. FAS in Australia: the role of the APSU, Birth Defects Society Scientific Meeting, Sydney, 21 February 2003.
3. Elliott E, Bower C, Payne J, Morris A, Ridley G, Haan E. Fetal Alcohol Syndrome in Australia: Our Place, Our Dreaming, Queensland Council of Social Services, Indigenous Children's Services Unit, Townsville, 23-24 September 2003.
4. Payne J, Bower C, Elliott E, Morris A, Ridley G, Haan E. Fetal Alcohol Syndrome in Australia, Population Sciences Meeting, Telethon Institute for Child Health Research, Perth, 25 September 2002.
5. Payne J, Bower C, Elliott E, Morris A, Ridley G, Haan E. Surveillance of Fetal Alcohol Syndrome in Australia, Away Days, Telethon Institute for Child Health Research, Perth, 26-27 November 2002.
6. Payne J, Bower C, Elliott E, Morris A, Ridley G, Haan E. Fetal Alcohol Syndrome, Pilbara Education Support Conference: Working with Aboriginal Children with Special Needs, 13-14 June 2003.
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## ANAPHYLAXIS FOLLOWING FOOD INGESTION

- There were 110 confirmed cases of food anaphylaxis in children reported to the APSU in 2002-03.
- Peanut was the leading cause of anaphylaxis and was involved in 30% of reported cases.
- Seventy five percent of reported cases were first episodes of food anaphylaxis and occurred mainly in infants and preschool children at home.
- Four children were admitted to intensive care units. These children were all aged over 8yrs.
- One death occurred in a 15yr old female with a history of asthma, who ingested peanut in salad dressing and subsequently participated in physical activity.

### Background

Anaphylaxis is a life threatening, acute allergic reaction requiring prompt medical treatment. In children the most common triggers of anaphylaxis are foods, insect bites and drugs.<sup>1</sup> Food allergy is common in childhood. An estimated 6-8% of children aged under three years suffer from food allergies and there is evidence that rates are increasing.<sup>2</sup> However, the incidence and mortality from anaphylaxis associated with food is not known. The Australian Paediatric Surveillance Unit study represents the first national study of food anaphylaxis in Australian children.

### Objectives

- To estimate the incidence of anaphylactic reactions to food in Australian children and their outcome;
- To describe clinical features of anaphylactic reactions to food and their management;
- To identify probable causative allergens;
- To identify steps which might be taken to assist carers and families regarding acute management.

### Case Definition

Anaphylaxis is defined as an immediate life-threatening, multisystem hypersensitivity response involving at least two of the following four systems:

- skin: rash (angioedema, urticaria);
- respiratory: airway obstruction (stridor, wheeze);
- gastrointestinal: vomiting, abdominal pain, diarrhoea;
- cardiovascular compromise: (eg. cyanosis, hypotension).

Clinicians were asked to notify children aged <15 years who had presented to a hospital or medical practitioner and required medical treatment (adrenaline being the appropriate treatment).

Children were excluded if their symptoms manifested as asthma or wheeze alone, or if the allergic reaction was mild and required no adrenaline treatment.

### Results

The classification of the 304 reports of anaphylaxis made to the APSU during 2002 and 2003 are indicated in figure 26.

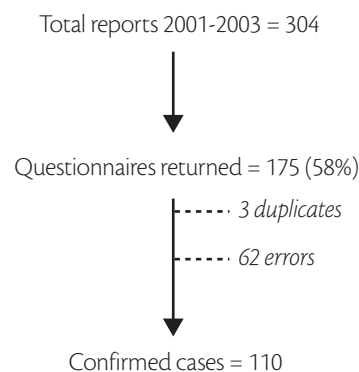


Figure 26. Anaphylaxis surveillance data for 2002-3

Questionnaires were returned for only 175 (58%) of APSU notifications in 2002-3 due to the complexity of the questionnaire. The female: male ratio was 6:1. The age range was 3 months to 15 years (mean 4.5 years). The most common allergens reported were peanut (30% of cases) and tree nuts (20% of cases) and cashews were the most common tree nut implicated.

Seventy five percent of anaphylactic reactions occurred in the child's home. The most prevalent symptom cluster was skin rash and/or angioedema with respiratory compromise.

Four children were admitted to intensive care units and there was one reported death. This occurred in a 15-year-old girl, who ingested peanut in salad dressing and the girl then developed symptoms after participating in physical activity.

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2. Sampson H, Mendelson L, Rosen J. Fatal and near- fatal anaphylactic reactions to food in children and adolescents. *New England Journal of Medicine* 1992;327(6): 380-4

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### Publications arising from this study:

#### Abstracts

1. Dosen A, Ziegler JB. National surveillance of anaphylaxis in children following food ingestion. *Journal of Paediatrics and Child Health* 2003; 39(suppl): A3-A4.

## HEPATITIS C VIRUS INFECTION (HCV)

- The diagnosis of Hepatitis C virus infection in Australian children is rare and is usually due to vertical transmission from the mother or intravenous drug use in the child.
- Most children with HCV infection are asymptomatic at diagnosis. Although routine screening for HCV infection in pregnancy is performed in many centres, postnatal screening of HCV-exposed infants and of older children at risk of HCV infection is not uniform across the country.
- We suspect that HCV may be under-diagnosed in Australia and need to explore reasons for this. Screening and diagnostic guidelines are urgently needed for paediatricians.

### Background

HCV infection poses a significant public health problem in Australia, with an estimated 1.1% adults infected. However, the epidemiology and natural history of HCV infection in Australian children is unknown. Annual laboratory surveillance describing the number of Australian children with HCV antibodies suggests that the prevalence of HCV in children is relatively low. However, epidemiological information obtained from laboratory surveillance is limited. Therefore, a prospective, national, active surveillance study was commenced in 2003 to estimate, the incidence and describe the epidemiologic factors, natural history and clinical management of HCV in children presenting to paediatric services.

### Objectives

- To determine the reported incidence of newly diagnosed HCV infection in Australian children;
- To describe the clinical presentation, investigation and management of newly diagnosed HCV infection;
- To document the presence of known risk factors for HCV infection;
- To determine the prevalence of co-infection with Hepatitis B Virus (HBV) and/or Human Immunodeficiency Virus (HIV) in Australian children with newly diagnosed HCV infection.

### Case Definition

Any child <15 years of age with newly diagnosed hepatitis C virus infection, defined as:

- at least one confirmed positive anti-HCV antibody test performed at age greater than or equal to 18 months OR
- a positive anti-HCV antibody test on a single occasion AND a positive test for HCV RNA (PCR or RT-PCR) on single occasion at any age > 1 month of age OR
- a positive HCV RNA test (PCR or RT-PCR) on two separate occasions.

### Results

Of the 12 confirmed cases of childhood HCV infection notified in 2003, all were born in Australia and the ratio of males and females was 1:1. No child was reported to be co-infected with HIV or hepatitis B virus. Ten children were asymptomatic at diagnosis, one child had hepatomegaly and one child had lethargy. The median age at diagnosis was 4.25 years (age range 6 weeks to 13 years).

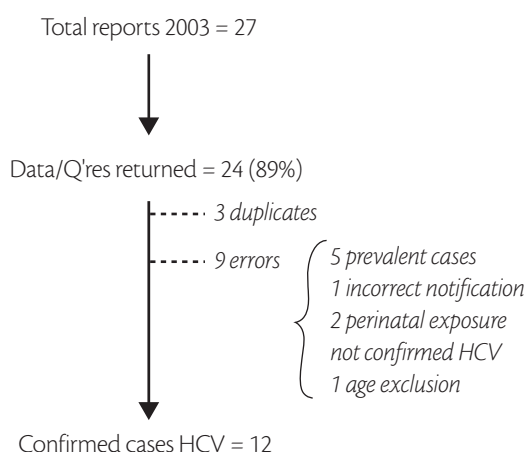


Figure 27: HCV surveillance data summary 2002-3

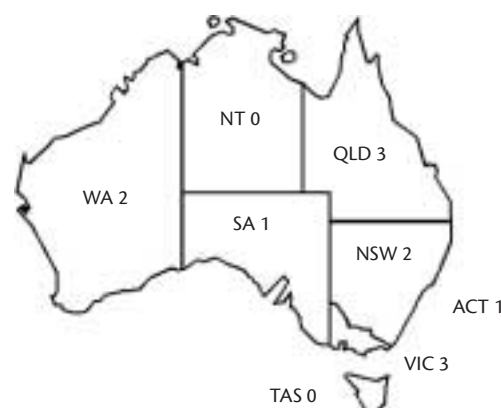


Figure 28: Distribution of confirmed HCV cases 2002-3

Two children were diagnosed at less than 18 months of age (by positive HCV RNA tests). The principal risk factor for childhood HCV infection was being born to an HCV positive mother (n=7/7 where maternal HCV antibody status known). Mothers acquired HCV infection through IV drug use (>80%), tattooing, piercing or needle sharing. Three children had a personal history of injecting drug use. The data summary and distribution of the 12 confirmed cases are indicated in figure 27 and 28 above.

### Conclusion

The rate of HCV infection in Australian children reported by Australian Paediatricians is very low (0.3 per 10<sup>5</sup> children < 15 years). HCV infection in Australian children is usually due to vertical transmission or childhood intravenous drug use and most children are asymptomatic at diagnosis.

Although the study findings are consistent with previous global studies, the small number of HCV cases identified nationally raises the possibility of under diagnosis or under-reporting. We are in the process of cross matching cases reported to the APSU with cases identified through alternate data sources to check for under-ascertainment.

Education is needed to raise awareness of HCV infection in children amongst health professionals and Australian guidelines for the diagnosis and management need to be developed. Ongoing surveillance and long term follow up studies are also needed to further define the natural history HCV infection in Australian children.

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## HIV INFECTION, AIDS AND PERINATAL EXPOSURE TO HIV

- Mother-to-child transmission rates are significantly lower if maternal HIV infection is diagnosed before the birth and interventions are used. These include antiretroviral treatment in pregnancy, elective caesarean delivery if appropriate, and avoidance of breastfeeding.
- Only 0.9% of children born in Australia to women diagnosed in the antenatal period acquired HIV, compared with 64.3% of children born to women diagnosed at, or after the birth of the child.
- The APSU makes a major contribution to surveillance for perinatal exposure to HIV in Australia and was the only notifying source for 78.1% of perinatally exposed children reported in 1999-2003.

### Background

National surveillance for perinatal exposure to HIV, paediatric HIV infection and AIDS is carried out through the APSU in collaboration with the National Centre in HIV Epidemiology and Clinical Research (NCHECR). The study aims to provide more complete ascertainment of perinatal exposure to HIV among Australian children than is available through existing surveillance mechanisms for notifying diagnosed cases of HIV infection and AIDS.

Perinatal exposure to HIV is now the most frequently reported source of HIV infection in Australian children, following the virtual elimination of the risk of exposure to HIV through the receipt of contaminated blood or blood products. Approximately 75% of children born to women with HIV infection do not acquire HIV infection perinatally, potentially resulting in an underestimate of the rate of perinatal exposure especially among women who were unaware of their HIV infection during pregnancy.

The risk of mother-to-child HIV transmission may be reduced from 25% to less than 2% among women whose HIV infection is diagnosed before delivery, by interventions such as antiretroviral therapy, elective caesarean delivery and avoidance of breastfeeding.

### Objectives

- To identify new cases of perinatal exposure to HIV, paediatric HIV infection and AIDS;
- To describe the pattern of perinatal exposure to HIV in Australia;
- To monitor the perinatal HIV transmission rate, and use of interventions for reducing the risk of mother-to-child transmission;
- To describe the natural history of paediatric HIV infection.

### Case Definition

Any child born to a woman with HIV infection, whether or not the child has HIV infection, and any child under 16 years of age with diagnosed HIV infection.

### Sources of ascertainment

Cases of perinatal HIV exposure were notified to the National Centre in HIV Epidemiology and Clinical Research (NCHECR) through the Sydney Children's Hospital from 1982, the APSU from 1993 and through assessment of perinatal exposure among children born to women whose HIV infection was newly diagnosed from 1995. Cases of newly diagnosed HIV infection in mothers and

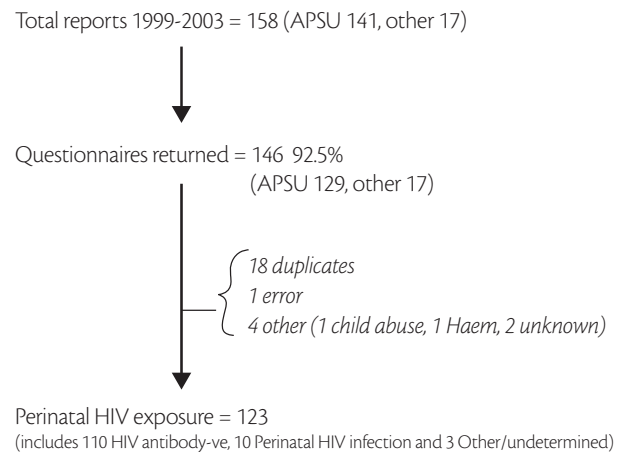


Figure 29: HIV surveillance data summary 1999-2003



Figure 30: Cases of perinatal HIV exposure 1999-2003

children were notified by State/Territory health authorities. For the purpose of this report, we have included all cases notified through the APSU and through State/Territory health authorities for the period 1999-2003.

## Results

A total of 123 children with perinatal exposure to HIV were reported, including 96 cases reported through the APSU only, 16 reported through national HIV/AIDS surveillance activities only and 11 reported through both APSU and national HIV/AIDS surveillance activities. Based on these 123 cases, the reported incidence for perinatal exposure to HIV was 9.87/100,000 (95%CI 8.2-11.7) live births. The data summary of the 123 confirmed perinatal exposure and the geographic distribution of cases is shown in Figure 29 and 30 respectively.

By 31 March 2004, 110/123 (89.4%) perinatally exposed children were confirmed as HIV antibody negative and 10/123 (8.1%) had HIV infection. HIV status remained undetermined at 31 March 2004 for 3 cases. The reported incidence of perinatal infection was 1.4 (95%CI 0.99-1.92).

The timing of the mother's diagnosis of HIV has been shown to affect the rate of transmission of HIV between mother and child. Women whose HIV infection was diagnosed antenatally made use of interventions for reducing the risk of mother-to-child HIV

**Table 9: Reported cases of perinatal exposure to HIV in children born in 1999-2003 (by timing of the woman's HIV diagnosis and interventions to reduce risk of vertical transmission)**

Intervention	Woman's HIV diagnosed before child's birth (antenatal)	Woman's HIV diagnosed at or after child's birth (postnatal)
	No. infected/No. exposed	No. infected/No. exposed
<b>Mode of delivery</b>		
Elective Caesarean	1/46	0/0
Other delivery	0/62	9/14
<b>Mode of feeding</b>		
Breast fed	0/0	4/7
Not breast fed	1/92	1/2
Not reported	0/16	4/5
<b>Antiretroviral Therapy</b>		
Treated	1/101	0/0
Not treated	0/5	9/14
Not reported	0/2	0/0
<b>Total<sup>1</sup></b>	1/108	9/14

<sup>1</sup> Does not include 1 woman whose date of HIV diagnosis was not reported.

transmission (Table 9). Only one of 108 children (0.9%) born to women whose HIV infection was diagnosed antenatally acquired HIV infection whereas 9 of 14 (64.3%) children whose mother was diagnosed postnatally were infected. The date of HIV diagnosis for a mother of 1 exposed child was not reported.

## Conclusion

National surveillance indicates that perinatal exposure to HIV and paediatric HIV infection remain rare among Australian children. The APSU makes a major contribution to surveillance for perinatal exposure to HIV in Australia and was the only notifying source for 78.1% of perinatally exposed children reported in 1999-2003. Women whose HIV infection was diagnosed antenatally have made use of interventions to minimise mother-to-child HIV transmission rate. However, perinatal HIV infection in children continues to occur, predominantly among children whose mothers' HIV infection remains undiagnosed in pregnancy.

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## Publications arising from this study:

### Reports

1. National Centre in HIV Epidemiology and Clinical Research. HIV/AIDS, viral hepatitis and sexually transmissible infections in Australia Annual Surveillance Report 2003. National Centre in HIV Epidemiology and Clinical Research, The University of New South Wales, Sydney, NSW. 2003 ISSN 1442-8784.

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## NEONATAL HERPES SIMPLEX VIRUS INFECTION (HSV)

- Neonatal herpes simplex virus infection is uncommon in Australia, with 59 confirmed cases in the period from 1997 to 2003, giving a reported incidence of 3.4/100,000 live births.
- APSU data suggests that there are considerable delays in the initiation of treatment of HSV once diagnosed. Twenty five percent of HSV-infected neonates in our study had delays of >7 days before therapy commenced. This is consistent with US literature, which suggests that little inroads have been made in the last 20 years in reducing the interval between onset of symptoms of neonatal HSV disease and initiation of life-saving intravenous therapy. According to a recent US review the reasons for the delay include non-specific clinical signs at presentation and the rarity of the condition.<sup>1</sup>
- Strategies to reduce delay in diagnosis and initiation of treatment are needed to further reduce mortality and morbidity from this condition.

### Background

Neonatal HSV infection is a potentially lethal complication of genital, (or occasionally, orolabial) HSV infection in a care-giver. It may manifest as disease localised to the skin, eye or mouth, as encephalitis, or as a disseminated visceral infection with or without central nervous system involvement. The latter two presentations are associated with high mortality and long term morbidity even with effective antiviral treatment. Early recognition and treatment of localised disease reduces the risk of progression to disseminated infection.

Definition of the epidemiology of neonatal HSV infection in Australia will enable the rational application of new or improved therapeutic and preventative strategies for this condition in our local population.

### Objectives

- To estimate the incidence of neonatal HSV infection in Australia;
- To determine the proportion of babies with disseminated HSV infection, localised disease or encephalitis;
- To determine the mode of acquisition of HSV infection.

### Case Definition

Any baby <= 28 days old with clinical evidence suggestive of HSV infection:

- HSV isolated from any site OR
- HSV detected in CSF by PCR (in the presence of CSF pleocytosis or other evidence of HSV encephalitis) OR
- Specific HSV-IgM detected in baby's serum OR
- Mother sero-converted or IgM positive and baby has typical clinical manifestations OR
- HSV isolated from mother around the time of delivery, and the baby has typical clinical manifestations.

### Results

One hundred and eighteen reports of HSV were made to the APSU from 1997 to 2003. Clinical data was provided for 115 (97%) of notifications and a total of 59 cases of neonatal HSV infection

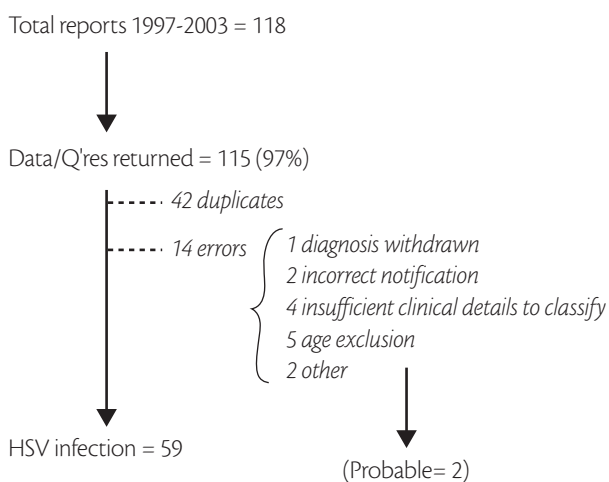


Figure 31: HSV surveillance data summary 1997-2003



Figure 32: Distribution of confirmed HSV cases 1997-2003

**Table 10. Confirmed neonatal HSV infection 1997-2003**

Presentation	Number of cases 2002	Number of cases 2003	Number of cases 1997-2003
Generalised excluding CNS	1	0	4
Generalised, including CNS	1	1	9
CNS alone	3	1	8
Skin, eye, mouth	5	4	29
Pneumonitis	0	0	6
Intrauterine	1	0	1
Unclassified	0	0	2
<b>TOTAL</b>	<b>10</b>	<b>6</b>	<b>59</b>

were confirmed (Figure 31). The geographic distribution of cases is shown in Figure 32. Of the 59 confirmed cases of HSV, 10 were identified in 2002 and 6 in 2003 (Table 10).

In the 7 years of the study the most prevalent presentation was with skin, eye or mouth involvement (29 cases). In 9 children there was generalised infection, including CNS and in 8 children there was CNS involvement only. A summary of all clinical presentations is found in Table 10 above.

## Conclusion

HSV is uncommon in Australia, with 59 confirmed cases in the period from 1997 to 2003, giving a reported incidence of 3.9/100,000 live births. This is in marked contrast to the USA, where there is up to a ten fold greater incidence.<sup>1</sup> The differences in incidence rates may be due to a greater risk of acquiring genital HSV disease in child bearing women in the US, and/or a possible protective effect of prior immunity from HSV-1, which is greater in Australia, against genital HSV-2 infection.

Because HSV is not notifiable in Australia or overseas, the collection of prospective national data through the APSU has provided some invaluable information about this uncommon but important infection. Data and treatment are often delayed. APSU data suggest that there are considerable delays in the initiation of treatment of HSV once diagnosed. Twenty five percent of HSV-infected neonates in our study had delays of >7 days before therapy commenced. This is consistent with US literature, which suggests that little inroads have been made in the last 20 years in reducing the interval between onset of symptoms of neonatal HSV disease and initiation of life-saving intravenous therapy. According to a recent US review the reasons for the delay include non-specific clinical signs at presentation and the rarity of the condition.<sup>1</sup>

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**Associate Professor Suzanne Garland**; Director, Dept. Microbiology & Infectious Diseases, Royal Women's Hospital, Carlton, VIC 3053

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## MUNCHAUSEN BY PROXY SYNDROME (MBPS)

- Symptom profiles described by the caregiver were complex – over 40% presented with reports of multiple health concerns/symptoms.
- The fabrication, induction or exaggeration of illness in a child by a caregiver is associated with a range of risks for the child and has a significant impact on the family, the health service and diagnosing clinicians.
- Fabrication or exaggeration of symptoms was the key feature of > 90% of presentations and 43% of cases and/or their siblings were known to statutory child protection authorities.

### Background

Munchausen by Proxy Syndrome (MBPS) was first described in 1977. The use of this term today is controversial, however in this study it describes the fabrication, induction or exaggeration of illness in a child by a caregiver. Although not mutually exclusive, the three main ways in which caregivers may induce or fabricate illness in a child include:

1. Fabrication or exaggeration of signs or symptoms, including the fabrication of medical history;
2. Falsification of or tampering with documents (eg. results), medical records or specimens;
3. Induction of illness or symptoms in the child by a variety of means including withholding or giving excessive amounts of prescribed medications.

Diagnosis of the condition is made difficult by the many different possible clinical presentations resulting from these patterns of caregiver behaviour. The different types of behaviour by the caregiver can create a wide spectrum of risk for the child. For some children the detrimental impact is relatively minor (over-presentation to health care-providers), for others it is intermediate (unnecessary investigations and hospitalisation) and for some the risk is severe (physical injury or death). A study on MBPS conducted through the British Paediatric Surveillance Unit estimated an annual incidence of 0.5 per 10<sup>5</sup> children under 16 years of age and 2.8 per 10<sup>5</sup> under the age of one year. This study represents the first attempt to estimate the incidence of MBPS in Australian paediatric practice.

### Objectives

- To estimate the incidence of MBPS seen by child health specialists in Australia;
- To describe the range of presenting symptoms, the interval between first consultation and diagnosis and the clinical suspicion that led to diagnosis;

- To describe current practice in relation to diagnosis and management of MBPS;
- To describe health service use as a result of MBPS;
- This study adopted an inclusive approach to case ascertainment to develop a better understanding of the spectrum of this syndrome and to gather information about the problems faced by clinicians in making this diagnosis.

### Case Definition

Any child under the age of 15 years, in whom it is suspected that physical or psychological symptoms or signs have been exaggerated, fabricated, or induced by a caregiver, with the deliberate intent of misleading the doctor.<sup>a</sup>

### Results

The classification and distribution of the 98 reports of confirmed or suspected MBPS made to the APSU between January 2000 and December 2003 are shown in Figures 33 and 34. Of the 58 confirmed or suspected cases with MBPS, 64% were reported to

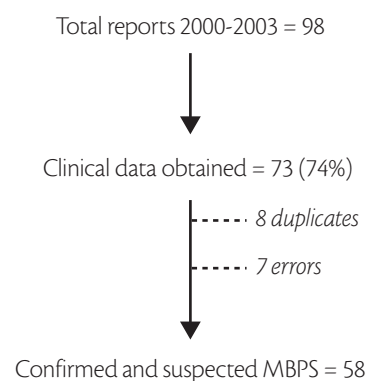


Figure 33: MBPS surveillance data summary 2000-3

<sup>a</sup> Requirements for notification of MBPS to statutory agencies vary from state to state in Australia. Clinicians were encouraged to notify the relevant statutory agencies when clinically appropriate and/or as required in their state.

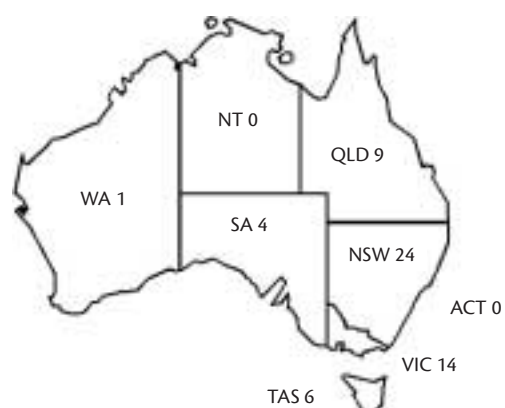


Figure 34: MBPS distribution of confirmed and suspected cases 2000-3

APSU by general paediatricians, 24% by medical/developmental sub/specialists and 12% by child protection or mental health specialists.

The reported rate of MBPS in Australian children aged 0-15 years in this study is 0.4 per 10<sup>5</sup> (95% CI 0.3 -0.5 per 10<sup>5</sup>) and 0.6 per 10<sup>5</sup> (95% CI 0.2-1.3 per 10<sup>5</sup>) in children under one year of age.

### Clinical Picture

The mean age of children at the time of report was 4.9 years (range= 0-14 years). The age distribution of reported children at diagnosis is presented in Figure 35. Fifty three percent of the children reported were male.

In 86% of cases the caregiver seeking treatment for the child was the mother. Twenty five (43%) of the cases and/or their siblings were known to the statutory child protection authority prior to the diagnosis of MBPS.

Fabrication or exaggeration of symptoms were caregiver behaviours associated with most of the presentations (Figure 36). It is noteworthy that in over 20% of the cases, more than one mechanism for the induction or fabrication of symptoms was reported (eg. poisoning and fabrication of a symptom for which the child had a history).

In 43% of cases the symptoms fabricated were associated with a condition for which the child had a previous history. Seizures were described as a principal reason for presentation for 26% of cases. However consistent with previous research, symptom profiles described caregivers were often complex, with over 40% of cases presenting with reports of multiple health concerns/ symptoms.

Clinicians reported suspecting MBPS within one month of the first consultation with symptoms ultimately attributed to MBPS in 52% of cases. In fact, MBPS was suspected within one day of

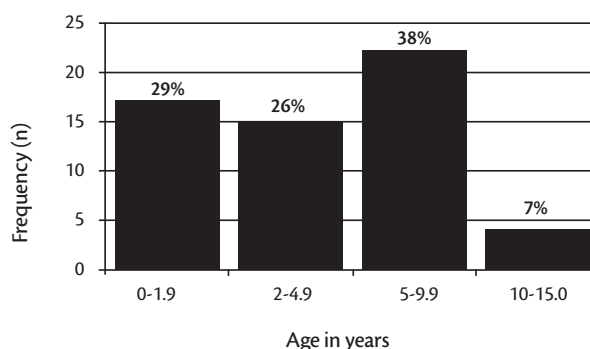


Figure 35: Age distribution of children diagnosed with MBPS

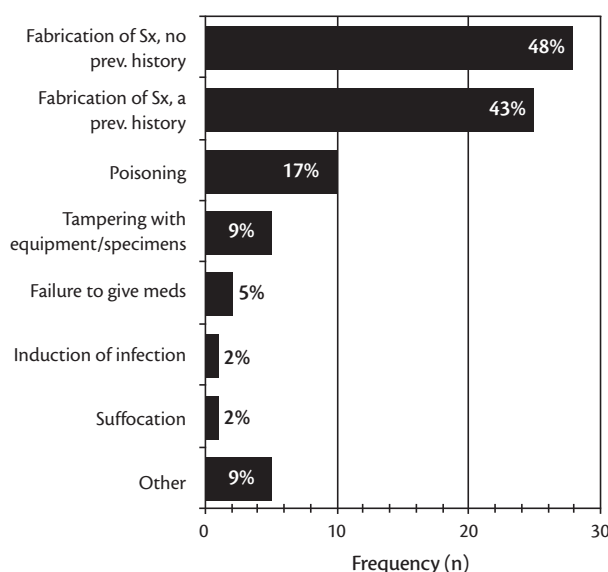


Figure 36: Caregiver behaviours associated with the induction or fabrication of symptoms\*

\* 31% of cases >1 behaviour

presentation in 40% of cases. However, in 10% of cases, clinicians first suspected MBPS at between 12 and 100 months following presentation with symptoms ultimately attributed to MBPS. Most clinicians describe multiple sources of suspicion regarding the case presentations. The following were the most frequently reported reasons for suspecting MBPS: the clinical findings being inconsistent with care-giver report, history anomalies (from bizarre disease combinations to inconsistent reporting), frequent health care seeking behaviour, symptoms not being observed in other care settings and suspicious care behaviour/presentation. Consistent with the complexity of many of these cases, there is not a clear relationship between time to suspecting MBPS and the nature of caregiver behaviours associated with the presentation.

## Short-term Outcomes

**Child:** A spectrum of immediate effects on the children were reported by clinicians. One death was reported, associated with poisoning with methadone. Some clinicians noted the loss of educational or “normal life” opportunities, others commented on the psychological effects of the abusive behaviour. Clinicians reported that most children suffered numerous unnecessary investigations and interventions.

**Family:** Most children had been notified to the statutory child protection authority as at risk of harm at the time of reporting to APSU. The outcomes of these notifications for families are generally unknown, however we are aware that specialist child protection services were involved with 62% of cases and several children were removed from the care of their families around the time of diagnosis.

**Health service:** High health service usage was associated with these children. The median number of doctors in addition to the clinicians reporting the cases, was five (range=0-50). Sixty seven percent of children were hospitalised around the time of diagnosis and the median length of admission was 10 (range=1-200) days.

**Clinicians:** Ninety percent of the reporting clinicians indicated that these cases had a significant impact on their practice, with almost one third of clinicians indicating that their involvement with these families had caused them significant personal distress. Three clinicians described caregiver behaviour towards them as aggressive.

## Conclusion

This study suggests that Munchausen by proxy syndrome is uncommon in Australia, affecting approximately 0.4 per 10<sup>5</sup> children under 15 years of age. This is a slightly lower incidence rate than that reported by the British study.<sup>1</sup> Considering our study questionnaire response rate (70%) is lower than most APSU studies, and the disproportionately strong response from the state of Tasmania, it is probable that our data represents an underestimate or *minimum estimate* of the national incidence of MBPS.

The British study reported a much higher rate of MBPS in babies; this pattern was not observed in this APSU study. Despite the likely underreporting of cases, this APSU study is interesting in that it confirms that older children can also be affected by fabrication or induction of illness by a caregiver. Another interesting outcome of this work has been the collection of information from clinicians about their experiences of caring for families in which the fabrication or induction of illness by a caregiver is suspected.

It is acknowledged that the implications of the suspicion of MBPS for children and their families can be enormous. However the consequences of MBPS for the clinicians, and health services, are rarely acknowledged. The substantial resource costs associated with caring for these families was noted by many reporting clinicians, as were frustrations at disappointment at perceived shortcomings in interagency cooperation. A significant minority of clinicians also described their personal distress associated with cases they reported.

MBPS, or the fabrication or induction of illness by a caregiver, may place a child at significant risk of harm. The controversy surrounding the terminology used to describe the fabrication or induction of illness in a child by a caregiver shifts the focus from the welfare of the child. Although rare, this diagnosis has an impact on outcomes for the child, their family and the responsible clinician.

MBPS is a complex problem that requires a multidisciplinary and dedicated team approach both in terms of diagnosis and management. There needs to be careful consideration as to how these cases are managed in the future in this country. A coordinated network for support and evaluation of suspected cases should be considered as an option to improve the care of families, as well as providing much needed support to health providers involved in the care of these families. The development of national and state guidelines for the assessment and diagnosis of this condition is recommended.

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## ■ RETT SYNDROME

- During 2002-03 there were 11 confirmed cases and 6 probable cases of Rett syndrome reported in Australia through the APSU.
- Phenotype and genotype analyses are currently underway in identified children and their families.
- There is an association between Rett syndrome and mutations in the MECP2 gene and genetic testing is now used in conjunction with clinical diagnosis to identify people with the disorder.

### Background

Rett syndrome is a neurological disorder almost exclusively affecting girls. Development appears to be normal until approximately 18 months of age, when development stalls and then regresses. People with Rett syndrome often develop seizures, scoliosis, gastrointestinal disorders and autonomic abnormalities. Ultimately, people with Rett syndrome are usually severely intellectually and physically disabled for the remainder of their lives. In 1999, the association between Rett syndrome and mutations in the MECP2 gene was identified. Molecular genetic testing is now used in conjunction with a clinical diagnosis to identify people with the disorder.

### Objectives

- To identify every person with Rett syndrome in Australia, born during or after January 1976;
- To describe the epidemiology (including survival analysis of a range of endpoints) of Rett syndrome;
- To obtain information about the progression of the disorder;
- To investigate the association between genotype and phenotype;
- To evaluate Rett syndrome management strategies;
- To identify factors in the family and community that promote optimal functioning and health for the child/young person with Rett syndrome and her family as a whole.

### Case Definition

Any child born during or after January 1976, with newly diagnosed or possible Rett syndrome, measured against clinical criteria or genetic testing. The clinical criteria include:

- Normal head circumference at birth;
- Deceleration of head growth between five months and four years;
- Loss of acquired purposeful hand skills between ages six and 30 months temporarily associated with communication dysfunction and social withdrawal;
- Development of severely impaired expressive and receptive language and presence of apparent severe psychomotor retardation;
- Stereotypic hand movements such as hand wringing/squeezing, clapping/tapping, mouthing and

“washing/rubbing” automatisms appearing after purposeful hand skills are lost;

- Appearance of gait apraxia and truncal apraxia/ataxia between one and four years.

### Method

Genetic studies had been performed on 83.4% of girls and young women in the study by the end of 2003. Phenotype-genotype correlation with the molecular analysis is being undertaken in laboratories in Sydney and Perth. During 2002, the second phase of the follow-up study was completed. One hundred and ninety six families completed a comprehensive study and data have been used in the phenotype-genotype analyses described above. Analysis of healthcare and morbidity data collected in the 2000 follow-up study was carried out in 2002 and was compared to data collected from controls.

During 2002 the pilot phase of InterRett, an online searchable Rett syndrome phenotype database (<http://www.ichr.uwa.edu.au/rett/irsa>), was focussing on international participants and funded by the International Rett Syndrome Association. The study was launched in October 2003 following a pilot phase carried out between January and September, 2003 and involving 118 families from 12 countries and their managing clinicians.

Analysis of clinical aspects of scoliosis in Rett syndrome using Australian data was carried out in 2003. These analyses were complemented by an online international study focussing on parental perceptions and knowledge of scoliosis in Rett syndrome. Data from the 2000 and 2002 follow-up questionnaires were also used in 2003 to analyse Rett syndrome behaviour profiles by age and mutation.

### Results

At the end of 2003 there were a total of 259 cases of Rett syndrome aged 27 and under identified through the epidemiological study of Rett syndrome in Australia. During 2002-03 there were 28 reports of Rett Syndrome. These are given as Figures 37 and 38 below. Of these reported cases, 11 were confirmed cases and 6 probable cases.

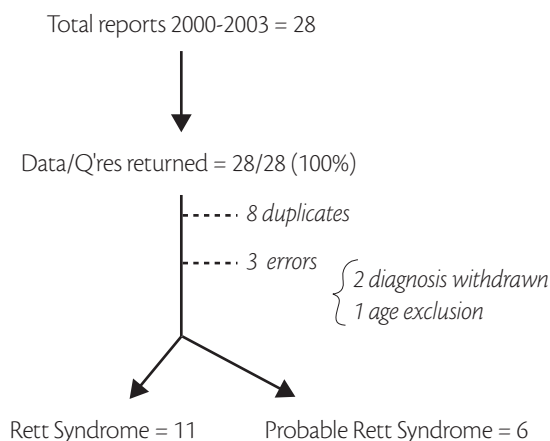


Figure 37: Rett surveillance data summary 2002-3

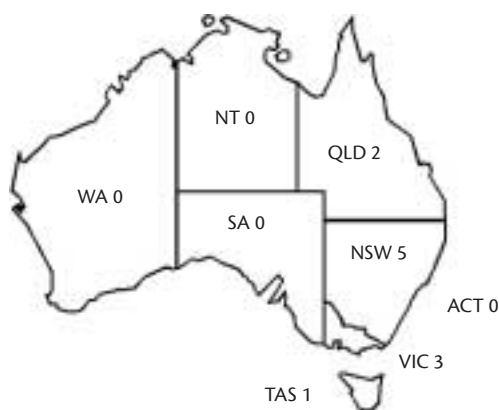


Figure 38: Distribution of confirmed Rett cases 2002-3

## Conclusion

We appreciate the ongoing commitment of paediatricians to the APSU Rett syndrome study. Their early referral and liaison with families has facilitated participation from parents. We welcome contact from clinicians who feel that a diagnosis is possible.

Successful funding has been obtained from the USA National Institutes of Health to continue the longitudinal study for five years. Participating centres include the Royal Children's Hospital Melbourne, The Children's Hospital at Westmead and Princess Margaret Hospital, Perth. The funding will provide further expansion of the study, to include: identification of clinical features; molecular genetic testing, clinical assessments, EEG and ECG studies, bone densitometry and autonomic nervous system evaluation in children with Rett.

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Dr Martin Delatycki; Victorian Clinical Genetics Services, Royal Children's Hospital, Parkville, VIC.

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# VITAMIN K DEFICIENCY BLEEDING (VKDB) INCLUDING HAEMORRHAGIC DISEASE OF THE NEWBORN

- The reported incidence for confirmed and probable cases was 1.01/100,000 live births.
- Despite NHMRC guidelines, Vitamin K was not given at birth to 10 infants.
- Parents should be informed of the significant morbidity and mortality associated with VKDB.

## Background

Vitamin K prophylaxis given by intra-muscular injection at birth was introduced in the 1950s in order to prevent haemorrhagic disease of the newborn in neonates considered at high risk of intracranial haemorrhage. In the 1970s, the recommendation was changed to include all newborn infants.

The epidemiology of vitamin K deficiency bleeding (VKDB) came into focus in early 1993 following the publication of a study in August 1992 by Golding et al<sup>1</sup> which reported an association between the use of intramuscular vitamin K and childhood malignancy.

In response to the study in January 1993, the NHMRC, the Australian College of Paediatrics and the Royal Australian College of Obstetricians and Gynaecologists issued a joint statement on vitamin K prophylaxis indicating that vitamin K could be given orally to healthy full term infants in a 3 dose regimen at birth, 3 to 5 days and in the 4th week<sup>2</sup>. Further studies<sup>3,4</sup> did not substantiate this association and revised recommendations were released in March 1994. In December 1999 the Australian Drug Evaluation Committee registered Konakion MM Paediatric, a new formulation of vitamin K (phytomenadione) containing 2mg in 0.2ml for intramuscular (IM) and oral use. This is currently the only preparation available in Australia.

The current NH & MRC recommendations<sup>5</sup> for prophylaxis with Konakion MM are as follows:

- *For all healthy neonates:* 1 mg by intramuscular injection at birth. Alternatively, 2 mg orally at birth, at the time of newborn screening (three to five days of age) and at four weeks.
- *For neonates with special risk factors\*:* 1 mg by intramuscular injection at birth. If the neonate has special risk factors and weighs less than 1.5 kg, then 0.5 mg is recommended.  
\* (infants who are pre-term, unwell or unable to tolerate or absorb vitamin K)

Vitamin K deficiency bleeding, including haemorrhagic disease of the newborn was listed for reporting through the APSU in May 1993 and clinicians were asked to retrospectively notify cases diagnosed from January 1993.

It is imperative that surveillance for this condition continues following the introduction of Konakion MM. This report summarises the data collected through the APSU during the period 1993 to 2003.

## Objectives

- To describe the epidemiology of VKDB in Australia;
- To estimate the morbidity and mortality associated with VKDB;
- To evaluate the efficacy of various regimes of vitamin K prophylaxis.

## Case definition

Any infant less than six months of age with spontaneous bruising/bleeding or intracranial haemorrhage associated with prolonged clotting time, (not due to an inherited coagulopathy or disseminated intravascular coagulation) and in whom the bleeding disorder corrects with vitamin K.

## Case Classification

A broad reporting definition was used to ensure complete case ascertainment. Notified cases were reviewed by study investigators and classified according to the following criteria:

**Confirmed cases** – Infants with coagulation studies as follows: INR greater than 4, prothrombin time greater than 4 times control value, platelet count normal or elevated; and in whom coagulation abnormalities corrected within 24 hours of vitamin K administration.

**Probable cases** – Infants with a clinical history and findings that strongly suggest a diagnosis of VKDB, but in whom coagulation studies were either cited as abnormal but results were not available to the investigators or were not performed.

**Unknown cases** – Infants with a clinical history and findings that made a diagnosis of VKDB possible, but unlikely and in whom there were no laboratory data to confirm or refute the diagnosis.

**Errors** – cases for which laboratory investigations and response to vitamin K therapy excluded the diagnosis of VKDB.

Table 11. Cases of Vitamin K deficiency bleeding, Jan 1993-Dec 2003

Year	No of notifications	Confirmed	Probable	Duplicate	Unknown	Error
1993*	47	6	4	14	15	8
1994	10	2	0	2	4	2
1995	4	1	0	0	1	2
1996	6	3	0	1	0	2
1997	2	1	0	0	0	1
1998	12	1	2	6	0	3
1999	3	0	0	0	1	2
2000	9	6	0	2	1	0
2001	5	0	0	1	0	4
2002	8	1	0	3	1	3
2003	3	1	0	0	1	1
<b>Total</b>	<b>109</b>	<b>22</b>	<b>6</b>	<b>29</b>	<b>24</b>	<b>28</b>

\* Surveillance commenced May 1993 with initial retrospective reporting to January 1993.

Table 12. Characteristics of definite &amp; probable cases of Vitamin K Deficiency bleeding, 1993-2003

Australia (n=28)		
	n	%
Case Classification		
Confirmed case	22	79
Probable	6	21
Disease profile		
Classical	4	14
Early onset	1	4
Late onset	23	82
Vitamin K given at birth		
Yes #	18	64
Not given	10	36
Type of Vitamin K at birth		
IMI	8	44.5
Oral	8	44.5
IVI	2	11
Breast fed	26	93
Term gestation (37-42 weeks)	25	89
Liver disease	11	39
Main site of bleeding*		
Skin	13	46
Gastrointestinal tract	8	29
Intracranial	6	21
Umbilical	5	18
Nose	2	7
Circumcision	1	4
Median age of onset in days (range)	42 (0 to 180)	

<sup>1</sup>Both were preterm infants who received the first two doses of vitamin K intravenously and the third dose as an oral preparation

\*Site of bleeding may be from multiple sites

# Some infants received inadequate dose IM

## Results and discussion

In the period January 1993 to December 2003, the APSU received 109 notifications of VKDB (Table 11). Twenty-two cases were classified as confirmed, six as probable cases and 24 as unknown. There were 29 duplicate cases and 28 errors.

Based on the confirmed cases, the reported incidence for the study period was 0.79/100,000 (95%CI 0.50-1.20) live births. The reported incidence for confirmed and probable cases was 1.01/100,000 (95%CI 0.67-1.46) live births. Clinical characteristics of the confirmed cases for 1993 to 2003 are shown in table 12.

## Conclusion

Vitamin K should be given to all infants in accordance with the current National Health & Medical Research Council guidelines. When consent is sought, parents should be informed of the significant morbidity and mortality. If oral vitamin K is given, the need for 3 doses should be stressed. We thank clinicians for continuing to notify cases.

## Acknowledgement

This surveillance study is generously sponsored by Roche Pty Ltd.

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## ■ STUDY PROTOCOL

### Haemoglobinopathies (excluding carrier states)

#### Background

Haemoglobinopathies are recessively inherited blood disorders for which there is no cure except, under certain circumstances, bone marrow transplantation. While haemoglobinopathies are rare, gene carriage occurs with high frequency in certain populations. For example, the frequency of the carrier state for  $\beta$ -thalassaemia is as high as 1 in 5 in Greece and Italy, the frequency of the carrier state for sickle cell anaemia is as high as 1 in 5 in equatorial Africa and alpha thalassaemia and haemoglobin E carrier status are common amongst Asian populations.

The number of children in Australia affected with a haemoglobinopathy is unknown. However the number of cases of haemoglobinopathies in Australia may be increasing due to changes in the composition of the population<sup>1</sup>. In contrast, it has been suggested that the incidence of these disorders may be decreasing overseas because of widespread screening programs including carrier screening and counselling of couples at marriage, pre-conception or in early pregnancy<sup>2</sup>.

In all states of Australia, selective screening is the current policy to identify carriers of haemoglobinopathies<sup>3</sup>. Thalassaemia carrier testing is recommended on an ad hoc basis to individuals from high-risk ethnic groups in teenage and early adult years. Carriers can also be detected on routine blood films carried out in early pregnancy. Screening with a full blood examination alone will not detect carriers of sickle cell disease, and haemoglobin electrophoresis is necessary. Also, there are many individuals in Australia who may not be recognised as being from high-risk ethnic groups (such as second or third generation Southern Europeans) Consequently they may be unaware of their carrier status or the potential risks of these conditions for their children and missed by targeted screening programs.

#### Study Objectives

In this study we seek to estimate the incidence and types of haemoglobinopathies in Australian children and their distribution amongst ethnic groups. We will also collect information on the timing and method of diagnosis of haemoglobinopathies in Australia. This information will contribute to efforts to improve the detection and outcome of affected children.

#### Case definition

Report all children under 15 years of age seen in the previous month with a newly diagnosed haemoglobinopathy including:

- structural haemoglobin abnormalities resulting from changes in the amino acid sequence of the globin chains

- thalassaemias, in which the synthesis of one or more of the globin chains is decreased or totally suppressed.

Conditions to be reported include:

- Hb SS disease (sickle cell anaemia)
- Hb CC disease
- Hb EE disease
- $\beta$ -thalassaemia major
- Hb E/  $\beta$ -thalassaemia
- Hb S/  $\beta$ -thalassaemia
- Hb SC disease
- Hb H disease
- Hb Barts disease
- Other rarer, severe haemoglobin variants

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## ■ STUDY PROTOCOL

### Hyperinsulinaemic Hypoglycaemia

#### Background

Hyperinsulinaemic hypoglycaemia (HI) is a biochemical profile reflecting hyperinsulinaemic, hypoketotic, hypofattyacidaemic hypoglycaemia with increased glucose requirements<sup>1,2</sup>. HI is the most common cause of persistent hypoglycaemia in the neonatal period after the first few hours of life. In severe cases hypoglycaemia is devastating. It may be difficult to control even in a hospital setting and may be associated with early brain damage<sup>1,2</sup>. Published overseas data suggests an incidence of about 1/40,000 births<sup>4</sup>,

HI includes a continuous spectrum of conditions with differing genetic aetiology. Clinical features range in severity from subtle signs such as 'floppiness', 'jitters', 'twitchiness' and poor feeding, through to overt signs such as seizures. While mild disease may be controlled by frequent feeding, severe disease may require medication (diazoxide) and severe unremitting hypoglycaemia may only be relieved by surgical removal of the pancreas (up to near-total resection)<sup>5</sup>. Pancreatic histology from surgical cases is usually abnormal. Disease may be focal on a background of normal pancreatic tissue or diffusely abnormal tissue may be located throughout the pancreas. It has been suggested that different histological types will require varying degrees of surgical resection, although currently this is unclear<sup>5</sup>.

HI is a well known cause of neurological damage<sup>2,6,7,8</sup> thus it is essential that HI is rapidly diagnosed and managed. Patients who respond well to medical treatment do not need pancreatic resection, however patients who are either non-responsive or unreliably responsive to medical intervention are at risk of brain damage. In the latter group, the decision to undertake pancreatic resection is very difficult because the risk of diabetes must be balanced against the risk of brain damage. Early definitive molecular diagnosis is an important goal because this may help us predict which patients do not respond to medication and require surgery. This is important because early surgery is associated with a reduced risk of diabetes<sup>8</sup>.

The incidence of HI in Australia is unknown. European and middle eastern data suggest that the incidence of HI varies from 1/2,500 in consanguineous populations to 1/50,000 births<sup>3,4</sup>. Our collaboration has identified over 70 children from Australia and New Zealand since 1977 giving an estimated rate of around 1/70,000 births. However these children were identified retrospectively and this is likely to be an under-estimate of the true incidence.

#### Study Objectives

In this study we seek to document the epidemiology of HI in Australian children and to record known risk factors. With the assistance of reporting clinicians, we will also attempt to recruit notified cases to a longitudinal follow-up study. This will allow collection of prospective data about response to treatment and will help us identify factors associated with good or poor outcomes. This information will contribute to efforts to improve the detection and outcome of HI in affected children.

#### Case definition

**Please report all children under 10 years of age seen in the previous month with newly diagnosed hyperinsulinaemic hypoglycaemia. That is:**

Low blood sugar (<2.6 mmol/L) with low blood fats and low ketones AND inappropriately high insulin level AND persistent or recurrent hypoglycaemic episodes *and/or* requiring glucose infusion for more than 10 days

#### Follow-up of reported cases

A brief questionnaire requesting further details will be forwarded to clinicians who report a case of HI to the APSU. Reporting clinicians will also be invited to send a study information sheet to families of affected children to inform them how to contact study investigators should they wish to participate in the longitudinal study of HI in Australian children.

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This study has the support of the Australasian Paediatric Endocrine Group



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## STUDY PROTOCOL

### Non tuberculous mycobacterial Infections

#### Background

Mycobacteria other than *Mycobacterium tuberculosis* (TB) cause a significant burden of disease in children. Non tuberculous mycobacteria (NTM) are free living soil and water organisms known to cause a spectrum of diseases including lymphadenitis, pulmonary disease, skin and soft tissue infections, ear infections, skeletal infections and disseminated infection<sup>1</sup>.

The annual incidence of NTM infections in the developed world is believed to be increasing possibly due to increasing awareness, better identification techniques and changing population groups<sup>2,3</sup>. However, the magnitude of this problem in children is unquantified. Our study aims to expand knowledge recently gained in Australia through laboratory surveillance.<sup>4</sup>

NTM infections are known to be associated with some medical conditions including human immunodeficiency virus (HIV) infection, malignancy, chronic granulomatous disease (CGD) and chronic lung disease, including cystic fibrosis and bronchiectasis. However most often NTM infections occur in otherwise healthy children.<sup>5</sup> Emerging data from recent studies show that even in healthy children, subtle underlying immunodeficiency or genotype differences may exist, contributing to susceptibility to NTM infection.<sup>6,7</sup>

The natural history of NTM infection has not been well described and optimal management remains unclear. There is evidence that a proportion of NTM lymphadenitis will spontaneously resolve.<sup>2</sup> In children requiring intervention (e.g. due to suppurative changes) surgical clearance has been accepted as the therapy of choice. While surgery is curative in most cases, a proportion of children fail initial surgical management and may require repeated surgery or the addition of medical therapy<sup>5,8</sup>. The role of medical therapy as first line treatment is unconfirmed. Consensus on medical treatment regimens has not been reached.<sup>9</sup> Information from this study will contribute knowledge to improve the detection of NTM and the outcome of affected children.

#### Study Objectives

1. To estimate the incidence of newly diagnosed NTM infection in children seen by child health specialists in Australia;
2. To describe the epidemiology and spectrum of disease and document known risk factors;
3. To describe diagnostic investigations used in Australia; frequency of use of skin testing and the clinical utility of the test, including differential skin testing;
4. To describe the management of NTM in Australia and the response to treatment.

<sup>b</sup> Clinical features of NTM lymphadenitis includes typical firm LN consistency +/- overlying skin changes (e.g. violaceous hue), with no associated constitutional symptoms

<sup>c</sup> Constitutional symptoms referable to NTM infection include unexplained lethargy, fever and/or anorexia and weight loss, generally only occurring with disseminated infections or pulmonary infections in chronic lung disease

<sup>d</sup> Avian PPD, manufactured by Commonwealth Serum Laboratories (CSL) Limited. Intradermal dose 10 IU

#### Case definition

Please report any child under 15 years of age seen in the previous month newly diagnosed with:

##### 1. DEFINITE NTM:

Any child in whom a non-tuberculous mycobacterium species has been identified either by isolation on culture or by polymerase chain reaction (PCR) from a sample from a sterile site. **OR**

##### 2. PROBABLE NTM:

A child who presents with any clinical features compatible with NTM (see below)

AND has undergone one or more of the supportive investigations (see below)

AND in whom *Mycobacterium tuberculosis* (TB) infection is unlikely.

Compatible clinical features	Supportive investigations (one or more)
<ul style="list-style-type: none"> <li>• lymphadenopathy (any site)<sup>b</sup></li> <li>• pulmonary disease with or without constitutional symptoms<sup>c</sup></li> <li>• skeletal infection</li> <li>• cutaneous infection</li> <li>• ear disease</li> </ul>	<p><b>Microbiology:</b> Acid fast bacilli (AFB) seen on sample or biopsy specimens <b>or</b> AFB grown from non-sterile site sample <b>or</b> positive AFB PCR on non-sterile site sample</p> <p><b>Histopathology:</b> Granulomatous inflammation <b>or</b> caseous necrosis <b>or</b> AFB seen</p> <p><b>Skin testing:</b> Tuberculin PPD skin testing <math>\geq 5\text{mm}</math> and less than <math>15\text{mm}</math> <b>and/or</b> Avian PPD<sup>d</sup> <math>\geq 10\text{mm}</math></p>

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## Peer and Editorial Review

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Thank you to all clinicians for your ongoing involvement and contribution to the APSU. We appreciate your support.

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 Dr Mary Paradisis (1)  
 Dr J P Pendergast (2)  
 Dr Susan Phin (1)  
 Dr Elizabeth Pickford (98)  
 Dr Melvyn Polon (1)  
 Dr Christopher C Poon (1)  
 Dr Alison Poulton (1)  
 Dr Keith M Power (1)

## Clinicians Reporting Cases 2002-3

Dr J S Preddy (1)

A/Prof P G Procopis (2)

Dr Denise Riordon (3)

Dr Marilyn Rochefort (1)

Dr L G Roddick (2)

Dr Greg Rowell (3)

Dr John W Ruhno (9)

Dr Monique Ryan (1)

Dr Gary F Sholler (1)

Dr J K H Sinn (2)

Dr Robert Smith (4)

Dr V Soutter (20)

Dr BJ Springthorpe (1)

Dr David R Starte (1)

Dr Glenn Stephens (1)

Dr Lee Sutton (1)

Dr Paul R Tait (1)

Dr Rhonda L Ticehurst (1)

Dr Susan J Towns (1)

Dr Dimitra Tzioumi (2)

Dr Peter Van Asperen (1)

Prof Graham V Vimpani (1)

Dr Mary-Clare Waugh (8)

Dr Mark A Westphalen (2)

Dr Catherine R Wiles (1)

Dr Helen F Wilkinson (1)

Dr Gary Williams (1)

Dr George L Williams (5)

Dr Meredith Wilson (2)

Dr Catherine Wiltshire (7)

Dr Melanie Wong (1)

Dr Ian Wright (2)

Dr Andrea Zalan (1)

A/Prof John B Ziegler (12)

Dr K Zwi (1)

### SA

Dr K Abbott (2)

Dr Brian Coppin (1)

Dr Richard T L Couper (2)

Dr Philip R Egan (3)

Dr David S Everett (5)

Dr Janice M Fletcher (1)

Dr Peter J Flett (2)

Dr Michael S Gold (5)

Dr Paul N Goldwater (3)

Dr Andrew W Grieve (2)

Dr Eric A Haan (2)

Dr Ross R Haslam (1)

Dr Hilary A Holmes (1)

Dr Simon L James (1)

Dr Jon Jureidini (44)

Dr David B Ketteridge (3)

Dr Christopher M Lamb (1)

Dr P Lang (2)

Prof David R Lines (2)

Dr Andrew J McPhee (1)

Dr M A Measday (1)

Dr David J Moore (3)

Dr Scott Morris (2)

Dr Josie Nozza (1)

Dr CC Pearson (3)

Prof Don M Robertson (1)

Dr A J Slater (2)

Dr Michael J Smiley (3)

Dr Nicola J Spurrier (3)

Dr Rima E M Staugas (1)

Dr Heather Tapp (1)

Dr Mark A Thesinger (1)

### WA

Dr L D Adams (4)

Dr D L Baker (2)

Dr David P Burgner (2)

Dr G M Chaney (1)

Dr Annette M Finn (2)

Dr Noel P French (2)

Dr A M Franzmann (6)

A/Prof Ronald Hagan (1)

Dr John D Hobday (2)

Dr Lawrence T H Hu (1)

Dr C A Jeffries-Stokes (1)

Dr A F Kehoe (1)

Dr Rolland Kohan (4)

Dr Richard Loh (9)

Dr Cherry Martin (1)

Dr Catherine F Mews (3)

Dr Lakshmi Nagarajan (3)

Dr Rama Naidoo (3)

Dr Mark Parker (2)

Dr Aida Partridge (6)

A/Prof Susan L Prescott (3)

Dr P C Richmond (11)

Dr Jacqueline M Scurlock (4)

Dr J Silberstein (2)

Prof Karen N Simmer (4)

Dr Harinder A Singh (1)

Dr Colin Somerville (6)

Dr Jeffrey R Tompkins (1)

Dr Jack B Vercoe (2)

Dr A Wilkins-Shurmer (1)

Dr Frank Willis (2)

### QLD

Dr Donald B Adsett (6)

Dr D B Appleton (4)

Dr K L Armstrong (2)

Dr Deborah Bailey (1)

Dr Ruth Barker (4)

Dr R P B Brown (2)

Dr Susan Cadzow (1)

Dr John Coghlan (1)

Dr M G Crawford (3)

Dr Tim J Donovan (3)

Dr A M Easterbrook (1)

Dr Loui Ee (3)

Dr F A Fiumara (2)

Dr W Frischman (1)

Dr John B Gavranich (1)

Dr Bruce Goodwin (3)

Dr Leonie M Gray (6)

Dr MA Harris (1)

Dr Wayne A Harris (2)

Dr G J Harte (1)

Dr Phillip J Harvey (1)

Dr R Heazlewood (14)

Dr T E G Hassell (2)

Dr Thomas M Hurley (3)

Dr J Anne Kynaston (4)

Dr Peter J Lewindon (1)

Dr Helen Liley (1)

Dr Bruce G Lister (1)

Dr James J McGill (2)

Dr Julie McEniery (7)

Dr J McGaughran (1)

Dr R A L McGregor (1)

Dr Lynne McKinlay (2)

Dr Hilary P Mercer (1)

Dr Malcolm N Miller (1)  
 Dr Susan Moloney (11)  
 Dr Anthony Morosini (2)  
 Dr Clare Nourse (6)  
 Dr Brian R Patten (1)  
 Dr J T Pelekanos (1)  
 Dr David R Pincus (11)  
 Dr W Robert Pitt (1)  
 Dr Jose A Prado (2)  
 Dr Jeffrey J Prebble (3)  
 Dr N Previtera (3)  
 Dr Ian F Robertson (4)  
 Dr Peter Roddenby (1)  
 Dr Patrick J Ryan (6)  
 Dr Phil Sargent (3)  
 Dr D C Shelton (3)  
 Dr C Y Skellern (2)  
 Dr B D Slaughter (1)  
 Dr Mark Stretton (2)  
 Dr Kerry O Sullivan (1)  
 Dr F K Y Tan (1)  
 Dr Susan Thornton (2)  
 Dr David I Tudehope (7)  
 Dr Vander Westhuyzen (1)  
 Dr John Varghese (3)  
 Dr T H Warnock (2)  
 Dr J H N Waugh (1)  
 Dr Judy A Williams (2)  
 Dr M L Williams (1)  
 Dr Sue Wilson (3)  
 Dr Geoffrey Withers (1)  
 Dr N F Woolfield (1)  
 Dr L Ah Yui (1)

### VIC

Dr Roger C Allen (3)  
 Dr David Amor (1)  
 Dr Kym P Anderson (1)  
 Dr Gordon Baikie (1)  
 Dr John M Bishop (1)  
 Dr J R Carapetis (2)  
 Dr Bronwyn A Cathels (1)  
 Dr Caroline Clarke (3)  
 Dr Chris Cooper (3)  
 Dr Noel E Cranswick (1)  
 Dr Nigel Curtis (1)  
 Dr N M Davis (1)  
 Dr Martin B Delatycki (1)  
 Dr Peter S Dewez (5)  
 Prof Richard R Doherty (7)  
 Dr Kevin B Dunne (1)  
 Dr Peter J Eastaugh (2)  
 Dr Maurice K Easton (1)  
 Dr Daryl Efron (2)  
 Prof Henry Ekert (1)  
 Dr Adrian M Elderhurst (1)  
 Dr P D Francis (2)  
 Dr Simon Fraser (1)  
 Dr Paul D Fullerton (2)  
 Dr Michael Gordon (2)  
 Dr Peter W Goss (2)  
 Dr Winita Hardikar (3)  
 Dr Simon Hauser (1)  
 Dr K L Heathershaw (1)  
 Dr Peter H Hewson (2)  
 Dr Nigel W J Hocking (2)  
 Dr Rod W Hunt (1)

Dr David I James (1)  
 Dr Bernard M Jenner (1)  
 Dr Lilian Johnstone (1)  
 Dr Hugh Kelso (1)  
 Dr David Krieser (5)  
 Dr Renata Kukuruzovic (2)  
 Dr Thomas J Lee (1)  
 Dr Anthony Lewis (1)  
 Dr Lionel Lubitz (1)  
 Dr Mark T Mackay (2)  
 Dr Catherine McAdam (1)  
 Dr Joanna McCubbin (2)  
 Dr Peter N McDougall (1)  
 Dr William J D McKellar (3)  
 Dr John A McLennan (1)  
 Dr Leslie J Markman (1)  
 Dr R B McNeill (1)  
 Dr Michael K Marks (1)  
 Dr Catherine Marraffa (1)  
 Dr R John H Massie (3)  
 Dr J F Mills (2)  
 Dr Marilyn L Murnane (1)  
 Prof Colin Morley (1)  
 Dr M J Nowotny (2)  
 Dr Gillian Opie (2)  
 Dr Greg M Pallas (1)  
 Dr Julian H Paxton (1)  
 A/Prof Campbell Paul (4)  
 Dr R Neil D Roy (1)  
 Dr Christine Sanderson (2)  
 Dr Kerryn R Saunders (1)  
 A/Prof Susan Sawyer (2)  
 Dr Ingrid E Scheffer (2)

Dr David Sholl (1)  
 Dr Robert A Sloane (2)  
 Dr Lloyd K Shield (2)  
 Dr Anne Smith (1)  
 Dr Arnold L Smith (1)  
 Dr Lindsay J Smith (2)  
 Dr T G Stubberfield (1)  
 Mr R Stunden (1)  
 Dr Mimi Tang (1)  
 Dr Nick H Thies (1)  
 A/Prof Garry L Warne (1)  
 Dr Andrew M C Watkins (4)  
 Dr Margaret Zacharin (2)  
 Dr Harry Zehnwirth (3)

### NT

Prof David R Brewster (1)  
 Dr R D Diplock (3)  
 Dr Anne Jaquiere (1)  
 Dr Andrew White (6)  
 Dr Annie Whybourne (4)

### TAS

Dr Christopher J Bailey (4)  
 Dr Evelyn Funk (4)  
 Dr S J Parsons (1)  
 Dr Mark M Pascoe (2)  
 Dr A W Shugg (2)  
 Dr Michelle Williams (2)  
 Dr Michelle Williams (2)

Thank you for your notification. We apologise to anyone inadvertently left off the list.



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