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Preface

Welcome to the 2003 Annual Report of the New Zealand Paediatric Surveillance Unit (NZPSU). This is the sixth Annual Report since the Unit was established in 1997.

The NZPSU was established with funding from the Ministry of Health in order to undertake surveillance of acute flaccid paralysis (AFP) for the Ministry of Health's National Certification Committee for the Eradication of Poliomyelitis (NCCEP). The opportunity was taken for the study of other uncommon high impact conditions, most of which has been undertaken by paediatricians with a particular research interest.

Time spent in this activity (responding monthly, responding with case material and reading protocols and the annual report) is considered by the RACP as eligible for Maintenance of Professional Standards (MOPS) points. Please see Appendix 1 for a letter from the College outlining the details.

Ongoing Studies

Acute Flaccid Paralysis (AFP)

- Even though the World Health Organisation (WHO) believes poliomyelitis to have been eradicated from the Western Pacific region, it requires New Zealand to continue surveillance of AFP with a phone notification of every case to the NZPSU.
    - There were seven true cases of AFP in New Zealand in 2003, giving an incidence of 0.9 per 100,000 children <15 years.
    - None of the cases notified were evaluated as being due to polio.

Haemolytic Uraemic Syndrome (HUS)

- There were 18 notifications and 12 confirmed cases of HUS in 2003. The annual incidence of HUS during 2003 is similar that of 2002.

Idiopathic Nephrotic Syndrome (INS)

- There were 19 notifications and 14 confirmed cases of INS in 2003. The annual incidence is 1.6 cases per 100,000 children < 15 years.

Inflammatory Bowel Disease (IBD)

- There were 26 notifications of IBD in 2003. The annual incidence was 3.1 cases per 100,000 children < 15 years.

Perinatal HIV Exposure (HIV)

- There were 12 notifications and ten confirmed cases of HIV exposure in 2003. The annual incidence rate was 1.25 cases per 100,000 children <15 years.

Vitamin K Deficiency Bleeding (VKDB)

- There were four notifications of VKDB received in 2003 with two of these cases turning out to be valid cases.

Congenital Rubella (CR)

No cases were reported in 2003.

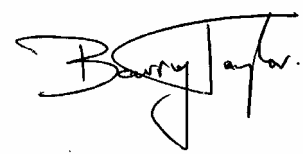
The ongoing success of the NZPSU is largely due to the high level of support from New Zealand paediatricians who have taken the time to provide information on the conditions under surveillance.

There has been a change of personnel at the NZPSU over the year. Melissa Carter spent three years working for the NZPSU, and many thanks to Melissa for all her hard work over that time.

Thanks also to Shirley Jones who worked for the NZPSU during 2003, and to Amanda Phillips who is the current administrator.

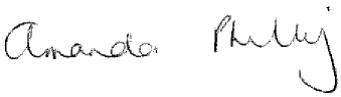
Amanda's contact details are: phone: (03) 474-7825
email: amanda.phillips@stonebow.otago.ac.nz

We would like to acknowledge the ongoing funding from the Ministry of Health.



Barry Taylor

Nigel Dickson



Amanda Phillips

Introduction

Surveillance is important to monitor both the incidence of emerging conditions and the effectiveness of prevention measures. The Paediatric Society of New Zealand (PSNZ) had for some years promoted the establishment of a unit that could regularly request specialist paediatricians to report on a number of conditions and this led to the establishment of the New Zealand Paediatric Surveillance Unit (NZPSU) in October 1997.

The aim of the NZPSU is to facilitate and improve the knowledge of uncommon high-impact childhood conditions in New Zealand. These conditions are of sufficiently low incidence or prevalence that case ascertainment on a national scale is needed to generate adequate numbers for meaningful study. The method was developed in the United Kingdom by the British Paediatric Surveillance Unit (BPSU) and has been used there since 1986. Subsequently, it has been introduced into several other countries including Australia, and is used by some other specialist groups.

The core activities of the NZPSU are funded through a contract with the Ministry of Health to provide active surveillance of acute flaccid paralysis (AFP). WHO, as part of the global eradication process, requires such surveillance to confirm New Zealand is free of poliomyelitis. Since the NZPSU's establishment, the number of conditions under surveillance has increased and now includes seven high impact childhood conditions.

The NZPSU is a member of the International Organisation of Paediatric Surveillance Units (INoPSU).

Aims

The aims of the NZPSU are:

- To operate a system for monitoring acute flaccid paralysis, as part of the global certification of eradication of poliomyelitis, required by WHO.

- To facilitate national surveillance and improve the knowledge of uncommon high-impact childhood conditions in New Zealand.

How the Surveillance System Works

The method of surveillance is based on that developed in the United Kingdom in 1986 by the British Paediatric Surveillance Unit (BPSU). It has subsequently been used for the monitoring of rare childhood conditions in several other countries including Australia, and also by other specialist groups.

Paediatricians in New Zealand gave their support to the surveillance system after the concept was discussed at an annual meeting of the Paediatric Society of New Zealand. A database of eligible clinicians, which included all paediatricians and other specialists working predominantly with children, was developed using the specialist register and the membership list of the Paediatric Society. All eligible clinicians were contacted and invited to participate. Those who agreed were provided with study protocols, which included definitions of the conditions under surveillance, specific reporting instructions, and a contact telephone number. Efforts are made to keep up-to-date with the paediatric specialist work force.

Every month participants are sent either a reply-paid card or an email (depending on their preferred method of reporting) to report whether in the previous month they have seen any cases of the conditions under surveillance. However, cases of AFP are also required to be reported immediately by phone to the NZPSU. When a case of any of the conditions is reported, the reporting clinician is sent a short questionnaire to complete on the case. The case's identity remains anonymous. Duplicate notification is recognised by a code derived from the child's initials and date of birth.

Where possible, cases are regularly compared with other data sources such as hospital discharge data, notifications to the local Medical Officer of Health, and the New Zealand AIDS Epidemiology Group.

It is envisaged that some of the conditions under surveillance will be ongoing while others will be on for a finite period, usually two or three years.

Regular surveillance reports are made to the Ministry of Health, specifically updating the progress with AFP surveillance.

Inclusion of New Conditions

A Scientific Review Panel (SRP) has been established primarily to consider the inclusion of new conditions into the scheme (see *Table 1* for details on members of the SRP). A study is eligible for consideration in the scheme if the condition of interest is:

- a relatively uncommon high-impact childhood condition (or an uncommon complication of a more common disease); and
- of such a low incidence or prevalence as to require ascertainment of cases on a national scale in order to generate sufficient numbers for study; and
- the SRP may also consider inclusion of short-term or geographically limited studies of comparatively more common conditions.

It is important for the success of the scheme that the workload of the mailing list is kept to a minimum. Accordingly, the SRP must be certain that studies conducted through the NZPSU are well designed and worthwhile. The SRP will take into consideration the scientific interest and public health importance of the proposed study, its methodology, and the suitability of the condition for ascertainment through the NZPSU scheme. Studies depending on immediate reporting and/or sample collection, or requiring the participation of other specialties, are less likely to be suitable.

***Table 1*: The Members of the NZPSU Scientific Review Panel (SRP)**

Member	Institution
Professor Barry Taylor	Dunedin School of Medicine
Dr Nigel Dickson	Dunedin School of Medicine
Dr Alison Roberts	Ministry of Health
Professor Elizabeth Elliot	Australian Paediatric Surveillance Unit
Dr Jeff Brown	Palmerston North Hospital
Professor Brian Darlow	Christchurch School of Medicine
Professor Diana Lennon	University of Auckland

Surveillance Activities in 2003

In 2003, 191 clinicians participated in the system. The average response rate to the monthly report card/email was 97%, with no consistent set of non-responders. *Table 2* shows the response rate per area for 2002 and 2003.

We are very pleased with the ongoing high response rate from the whole of the country.

***Table 2*: Response Rate Per Health Locality (as defined by DHB) 2002 & 2003**

Health Locality	2002	2003
	%	%
Northland, Auckland, Waitemata, Counties Manukau	94	96
Waikato, Bay of Plenty, Taranaki, Lakes	93	98
Capital and Coast, Hutt, Wairarapa, Wanganui, Tairawhiti, Hawkes Bay, Mid Central	97	98
Nelson Marlborough, Canterbury, South Canterbury, West Coast	97	96
Otago, Southland	95	98
TOTAL	**95**	**97**

Respondent Workload
Minimising the extra workload that the system imposes on paediatricians is a key factor for its success. The range of conditions under surveillance and their incidence needs to be kept under review.

Table 3 shows the percentage of clinicians on the mailing list that reported cases during 2002 and 2003. The table shows that in 2003 75% of the participants did not report any cases, with none reporting five or more, compared to 51% and 8% respectively in 2002. Overall, less notifications, were required for the conditions under surveillance in 2003 than in 2002.

***Table 3*: Respondents Workload 2002 & 2003**

Notifications	2002		2003	
	No.	**%**	**No.**	**%**
None	97	51%	143	75%
One	42	22%	26	14%
Two – Four	35	19%	22	11%
Five or more	15	8%	0	0

In 2003, the NZPSU monitored seven uncommon childhood conditions *(Table 4).* Some of the protocols and questionnaires used were adapted from those used by the Australian Paediatric Surveillance Unit.

***Table 4:* Conditions Under Surveillance in 2003**

Condition	Surveillance started	Principal Investigator(s)
Acute flaccid paralysis	October 1997	Dr Nigel Dickson Dr Paul Shillito
Haemolytic uraemic syndrome	January 1998	Dr William Wong
Congenital rubella syndrome	January 1998	Professor Diana Lennon
Perinatal HIV exposure	January 1998	Dr Nigel Dickson Dr Lesley Voss
Vitamin K deficiency bleeding	January 1998	Professor Brian Darlow
Idiopathic nephrotic syndrome	July 2001	Dr William Wong
Inflammatory bowel disease	January 2002	Dr Alison Wesley

Brief Reports on Selected Conditions

ACUTE FLACCID PARALYSIS (AFP)

Dr Nigel Dickson

Ongoing study started in October 1997

Introduction

To confirm the absence of poliomyelitis WHO requires a surveillance system to be in place:

1. That captures an annual incidence of acute flaccid paralysis (AFP), not due to poliomyelitis, of at least one per 100,000 children < 15 years.

2. In which 80% of cases of AFP have two stool samples taken at least 24 hours apart, within 14 days of onset tested negative for wild polio virus in a WHO-accredited laboratory.

Telephone notification of all cases of AFP is required by the NZPSU to ensure that the necessary stool containers are dispatched in time to the notifying paediatrician.

Key Results for 2003

- There were nine cases notified to the NZPSU in 2003, of which two cases did not fit the criteria for AFP.
- Information has been obtained on all of these children including follow up information two months after diagnosis.
- All seven true AFP cases were from the North Island.
- Two males, five females
- Age range 23 months to 13.4 years, median age 9.9 years.
- No seasonal variation.
- The overall incidence was 0.9 per 100,000 children <15 years.
- A diagnosis of Gullain-Barre syndrome (GBS) has been made in four of these cases, transverse myelitis in two, with the remaining case unknown, but confirmed not polio.
- All seven cases have been discounted as Polio by the National Certification Committee for the Eradication of Polio (NCCEP).
- Analysis of stool samples satisfying the WHO criteria was only complete for six of the seven children (see *Table 5).*

***Table 5*: Percentage of AFP Cases With Adequate Stool Samples (or otherwise)**

Category	Stool samples	
	No.	**%**
2 stool samples within 14 days of onset of paralysis	4	57
2 stool samples, but one or both not within 14 days of onset of paralysis	2	29
1 stool sample	0	0
No stool samples	1	14

Comment
The system captured a rate of AFP (not due to polio) of 0.9 per 100,000 children < 15 year, just below what WHO believes is the expected rate of 1.0 per 100,000. While the overall rate of stool testing was 85%, in two cases these were collected more than 14 days after the onset of paralysis, the time limit required by WHO. The rate of stool testing within this time period was therefore 57%, less than the 80% required by WHO.

Ongoing surveillance of AFP, even though the WHO believes Polio to have been eradicated from the Western Pacific region, is likely to be required for some years. This will require the continued telephone notification of all cases of AFP, including those with a definitive diagnosis such as Guillain-Barre syndrome etc. A challenge has always been to utilise a non-specific case definition – such as 'acute flaccid paralysis' – in a health system where a more definitive diagnosed for children with such symptoms is likely to be made.

CONGENITAL RUBELLA SYNDROME (CRS)

Dr Diana Lennon

Ongoing study started in January 1998.

We have not provided a report for congenital rubella as there were no cases reported in 2003.

PERINATAL EXPOSURE TO HIV (HIV)

Dr Nigel Dickson

Ongoing study started in January 1998

In 2003, there were 12 notifications to the NZPSU of infants/children born to women infected with HIV. Of these, two were notifications of perinatally infected children born overseas.

Of the remaining ten, two were reports of babies infected with HIV born before 2003 in New Zealand diagnosed with HIV during 2003. One (Child A) was born to a mother from a Pacific Island who had had a partner from a country of relatively high HIV prevalence. This child was diagnosed because of symptoms and led to the mother's diagnosis. The other child was born in 2002 to a mother whose partner was an intravenous drug user.

Of the eight children reported in 2003 born in New Zealand in that year, one is known to be infected with HIV. This child is the sibling of the Child A. He was born before his sibling was diagnosed with HIV, the event which led to his mother being diagnosed.

One of the uninfected children was born to an African woman whose HIV was diagnosed after this child's birth.

Of the remaining six babies born in New Zealand in 2003, all to women whose HIV was diagnosed prior to delivery, none are believed infected with HIV (although some are still awaiting final conformation):

- Five were born to mothers whose HIV had been diagnosed before the pregnancy. One mother (African) was diagnosed during her pregnancy.
- Three of the mothers were African, and the remaining three of European ethnicity.
- All the mothers were given antiretroviral treatment during pregnancy, four of the six were delivered by caesarean section, and none of the babies were breastfed.

HAEMOLYTIC URAEMIC SYNDROME (HUS)

Dr William Wong

Ongoing study started in January 1998

Eighteen notifications of HUS were made to the NZPSU of which 12 were confirmed cases. The annual incidence of HUS during 2003 is similar that of 2002. The geographic distribution of the cases remains unchanged with all of the cases from the North Island. There continues to be no seasonal pattern to the cases. The mean age at presentation is 3.3 years (range 0.8 to 7.8 years). The median interval from onset of symptoms to diagnosis was seven days (range 7-25). Two of the children lived on a farm. Nine of 12 cases required peritoneal dialysis. All children survived the acute illness.

VITAMIN K DEFICIENCY BLEEDING (VKBD)

Professor Brian Darlow

Ongoing study started in January 1998

There were four notifications of VKDB received in 2003; two reports were not valid, hence there were two valid cases.

Both cases were of classic type with neither infant receiving any vitamin K prophylaxis because of a medical error. In both infants bleeding resolved after i.m. vitamin K and there were no long-term complications.

- One infant was born postmature in a Level I hospital and was compromised at birth with meconium aspiration syndrome and pulmonary hypertension, necessitating transfer to a Level III hospital. Vitamin K prophylaxis was omitted in error. Gastrointestinal haemorrhage occurred on day five, the coagulation profile was typical of VKDB and i.m. vitamin K was given with good effect.

- One infant was born in hospital at 38 weeks gestation and due to a misunderstanding the medical record was signed as vitamin K had been given i.m., when in fact none had been administered. On day two bruising was noted particularly over the scalp, and there was oozing from the Guthrie heel-prick site. VKDB was diagnosed, vitamin K was given i.m. The infant developed an extensive subaponeurotic haemorrhage and disseminated intravascular coagulation before resolution of all symptoms.

IDIOPATHIC NEPHROTIC SYNDROME (INS)

Dr William Wong

Three year study started in July 2001

The surveillance study for childhood INS has now reported 40 cases from July 2001 till 31 March 2004. The mean age of presentation is 5.7±3.6years (range 1.8-14.1 years). All had normal renal function at time of presentation and 10 of the 40 were hypertensive at presentation. Microscopic haematuria was detected in 28 patients. All patients, except for one, were treated with 1.5-2.0mg/kg/day of prednisone, with the most common regimen being 2mg/kg/day given once daily until remission. The majority (30/40) were given antibiotic prophylaxis, with only a minority (14/40) had administered, or were planning to administer, pneumococcal vaccine. Thirteen of 40 children were treated with diuretics and 11 children required albumin infusions.

In the 12 month follow up, provisional results show five of 18 have been classified as steroid resistant. Invasive sepsis was documented in two patients, and one patient with focal segmental glomerulosclerosis has progressed to end stage renal failure.

INFLAMMATORY BOWEL DISEASE (IBD)

Dr A Wesley, Dr S Mouat, Dr J Yap, Dr S Chin

Summary after two years of reporting

Over the two year period of the study there have been 52 notifications, 26 in each year, five of these were duplicate notifications. Hence there were 47 children notified. To date 40 completed questionnaires have been received.

	No.	**%**
Notified cases of IBD	47	100
Completed questionnaires to date	40	85
Diagnosis		
Crohns Disease	22	55
Ulcerative Colitis	9	23
Indeterminate Colitis	8	20
Sex		
Female	17	43
Ethnicity		
European	30	75
Indian	3	8
Maori/Pacific Islander	2	5
Other ethnicity	3	8
Area		
North Island	27	68
South Island	11	28
Histologic diagnosis undertaken	38	97
Specialist involved (more than one possible)		
Seen by Paediatric Gastroenterologist	20	51
Seen by General Paediatrician	22	56
Seen by Paediatric Surgeon	8	21
Seen by Adult Gastroenterologist	14	36
Seen by Adult Surgeon	2	5

A more complete analysis, as well as further chasing up of non-returned questionnaires, is planned.

Final Reports for Completed Studies

CHILDHOOD BRONCHIECTASIS (BE)

Dr Jacob Twiss, Dr R. Metcalf, Dr C.A. Byrnes
Starship Children's Hospital and University of Auckland

Introduction

Bronchiectasis (BE) is the abnormal dilatation of bronchial airways usually associated with recurrent lower respiratory tract infection and productive cough. Incidence may have fallen in the 20th century with improved living conditions, immunisation and antibiotics, but the disease still causes significant morbidity and mortality in New Zealand and many other countries. In Maori and Pacific Island Peoples the BE mortality is as high, or higher than, asthma and has been reported as the seventh leading cause of death in Pacific Island women (Public Health Commission Report, 1994, NZ Health Information Service, 2001). Even in the community at large it results in more than half as many deaths as asthma, a far more common disease, and four times as many deaths as cystic fibrosis, a condition with much higher profile.

Bronchiectasis prevalence increases with age, but a significant proportion is thought to arise in childhood. Edwards *et al* estimated the prevalence amongst Auckland children to be one per 6,000 overall, but as high as one per 1,800 in Pacific children, based on a 2001 tertiary clinic population. This clinic has doubled in size since. Other case series have reported high rates in the central Australian Aboriginal population, among southwest Alaskan Natives, and in southern Turkey. Only one national incidence study has been published reporting an annual incidence of 4.9 per million children (Finland, 1998).

The aim of this study was to prospectively estimate the national incidence of BE diagnoses in New Zealand and evaluate regional variation, aetiology and severity.

Methods

BE was included on the NZPSU report card during 2000 and 2001.

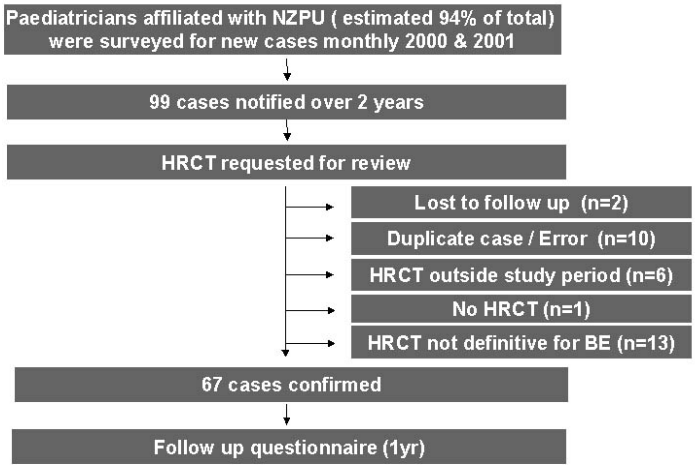
Case definition (all the following criteria had to be met):

- under 15 years of age at diagnosis,
- productive cough daily for >6 weeks or for 3 months per year for 2 consecutive years,
- persistent chest x-ray abnormalities,
- a high resolution computer tomography (HRCT) consistent with bronchiectasis and
- not due to cystic fibrosis.

The HRCT of each notified case was reviewed by a single paediatric radiologist with no clinical data. Diagnosis was confirmed and the HRCT was scored for severity and extent. Confirmed cases were followed up 12 months after diagnosis by questionnaire collecting demographics data, investigation results and ascribed aetiology. The delay was to allow time for investigation. Ethnicity data collection allowed multiple responses and prioritization as per the New Zealand standard classification.

Results

Notifications

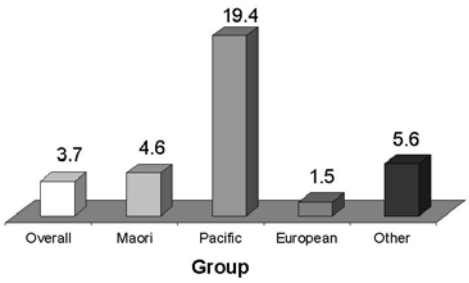


Characteristics of confirmed cases

Female:male ratio	1.2 : 1		
Median age at diagnosis	5.5 years		
Median age at chronic symptoms	2.4 years		
Median age at first respiratory admission	2.0 years		
Ethnicity (multiple choices permitted)	Maori	29%	(23% of population)*
	Pacific Peoples	53%	(11% of population)*
	European	27%	(73% of population)*
	Other	5%	(3% of population)*
Reported aetiology	Unknown / Idiopathic		58%
	Post infectious		19%
	Post oncology		10%
	Aspiration		7%
	Primary immunodeficiency		6%
Infecting organism (65% of cases had positive cultures)	*Haemophilus influenzae*		75%
	Streptococcus pneumoniae		24%
	Moraxella catarrhalis		18%
	Staphylococcus aureus		11%
	Pseudomonas aeruginosa		4%

Incidence

Incidence varied by ethnicity and region, as shown in the rates per 100,000 children <15 years.



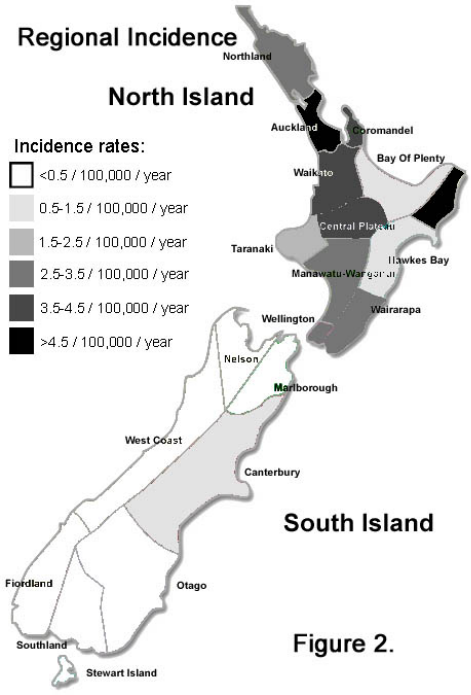
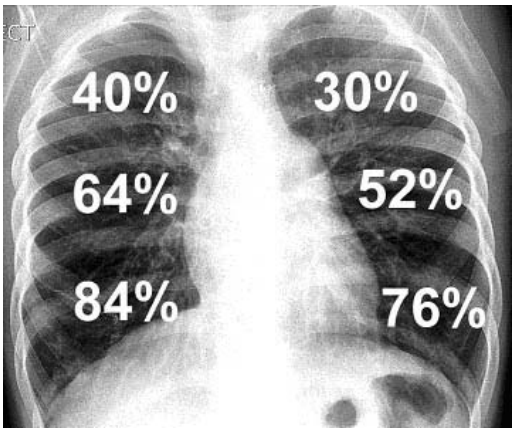


Figure 2.

Severity

In general those identified had extensive, severe disease. Ninety percent of children over six had had spirometry. The mean FEV1 (% predicted Polgar) was 74% (mild obstruction) but ranged from 18 – 116% predicted. Over half had an FEV1 below 80% and a third below 60%. Eighty-three percent had bilateral disease.

FVC (% predicted)	mean 81% (16-121)
FEV1 (% predicted)	mean 74% (18-116)
FEF_{25-75} (% predicted)	mean 73% (16-157)
Bilateral disease	83% of cases
HCRT score (Bhalla)	median 17.5 (4-65)
Worst Lobe	lower lobes (68%)



Conclusions

- The minimum New Zealand childhood incidence of BE is 3.7 per 100,000 children <15 per year, over seven times higher the only other national study (0.49 per 100,000, Finland) and nearly twice the childhood incidence of cystic fibrosis.
- Pacific children are most at risk with five times the national incidence rate and more than ten times the New Zealand European rate. Maori and 'Other' ethnicities also have higher rates.
- Auckland had twice incidence of diagnoses as found elsewhere.
- The median age at diagnosis was 5 years but most had already had symptoms for more than 2 years.
- Those identified had extensive disease, mostly of unknown origin.
- Ten percent of cases were associated with a treated malignancy.

Thanks to:

New Zealand Paediatricians for providing information

The Joan Mary Reynold's Fellowship and Child Health Research Foundation for funding

All the staff of the NZPSU, which is funded by the Ministry of Health

KAWASAKI DISEASE 2001/2 (KD)

Dr Paul Heaton [a] (principal investigator), Dr John Doran [b], Dr Alan Parsons [b], Dr Geoff Aiken [b], Dr Nigel Wilson [c], Dr Ross Nicholson [d]

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[b] Taranaki Base Hospital, New Plymouth
[c] Starship Hospital, Auckland [d] Middlemore Hospital, Auckland

Background

Since the original description of 50 cases in Japan in 1967 KD (KD) has displaced acute rheumatic fever in developed countries as the most frequent cause of acquired cardiac disease in childhood[1]. Thousands of cases have been documented and reports of affected children have come from many different countries [2].

Morbidity and mortality result from cardiac complications of the disease. For many years it has been known that prompt diagnosis and treatment with intravenous immunoglobulin (IVIG) and aspirin improves outcome [3,4].

The aetiology of KD is unknown, though current opinion is that it is an acute inflammatory vasculitis, probably initiated by exposure to a commonly encountered infectious agent but expressed only in those who have certain polymorphisms affecting the regulation of the inflammatory process [5]. There is no diagnostic test to confirm or exclude the diagnosis, and currently the diagnosis is based on the presence of a number of clinical features sufficient to fulfill agreed diagnostic criteria.

The challenge of accurate diagnosis, the urgency of timely administration of appropriate treatment, and the potential for severe acute and long-term cardiac disease, all impose particular difficulties for any clinician faced with a child who may have KD.

KD has been reported from most countries in the world, though there are considerable variations in incidence mainly relating to geographic and ethnic factors [6]. An increasing incidence over time has been observed in some countries [7,8].

There has been limited published data regarding the epidemiology of KD in New Zealand. Gentles *et al* reported the details of 34 children diagnosed with KD in the Auckland region from 1979 to 1988 [9]. These 34 cases were identified retrospectively from the Auckland Hospital Board database for hospital admissions, from the files of Green Lane Hospital Cardiology Department and from the local coroner's pathologist. The recorded incidence of KD for children aged under 5 years was 5.1 cases per 100,000 population, with little variation between ethnic groups. There was one death, of a 3 month male in whom KD was diagnosed at *post mortem*. Two fatal cases of KD were documented by Heaton and Wilson[10], both patients were male European infants who had early occlusive coronary artery disease and died 96 and 110 days from the onset of illness. Four more cases with severe cardiac outcomes were also identified in New Zealand in the late 1990s including another fatal case; a female aged two months who suffered coronary artery rupture during the acute phase of the illness[11].

Objectives

The principal aim of this prospective study was to document the epidemiology of KD in New Zealand during 2001 and 2002. The study was designed to assess clinical aspects of investigation, management and outcome, specifically in relation to cardiac disease.

Methods

The NZPSU monthly reporting card system, used by all pediatricians through NZ, was used to identify cases. The mean monthly return rate was 92%. Questionnaires were then sent to the reporting clinician approximately six weeks following the notification of a suspected case.

Results

Over the 24-month study period there were 58 notifications, of which three were duplicate notifications. In two cases the clinical notes were untraceable and the cases were excluded from analysis. Following review of the data by three investigators and the referring physician the diagnosis was judged not to be KD in 4 cases, either because there were too few diagnostic criteria or there was an alternative diagnosis.

Thus, there were 49 new cases of KD. Of these, 43 were considered definite diagnoses fulfilling all the case definitions. Four cases were considered to be probable cases (not all diagnostic criteria present but clinically considered and treated as KD) and two were possible cases (both late presentations but managed as KD).

Age and Gender

Of the 49 new cases 31 were male and 18 female (M:F 1.7:1).

Age range was 2 months to 11 years. 13 cases (27%) were aged less than one year and 42 (86%) less than five years old. The annual incidence was 8.0 cases per 100,000 population aged less than five years (*Statistics NZ 2001 Census*). For infants aged less than one year the annual incidence was 11.8 cases per 100,000.

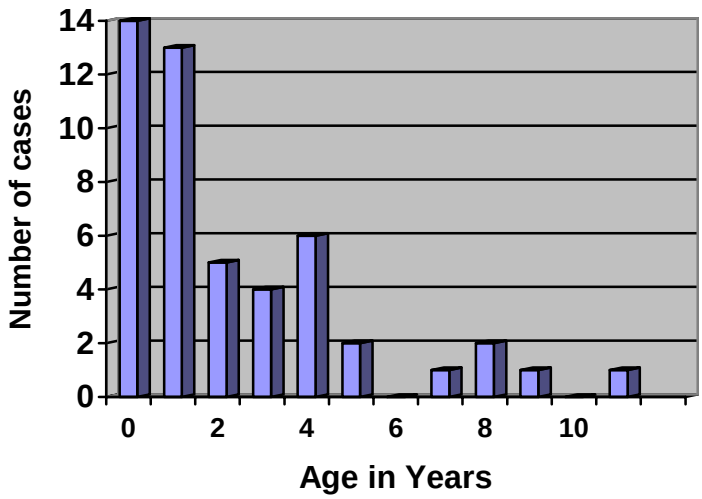

Figure 1: **Age Distribution of Case**

Season and Geography
The month of onset of symptoms was Dec-Feb (summer) in 13 cases (26.5%), Mar-May (fall) in 18 (36.7%), Jun-Aug (winter) in 8 (16.3%) and Sep-Nov (spring) in 10 (20.4%). When documented, the place of residence was North Island in 35 cases (72.9%) and South Island in 13 (27.1%).

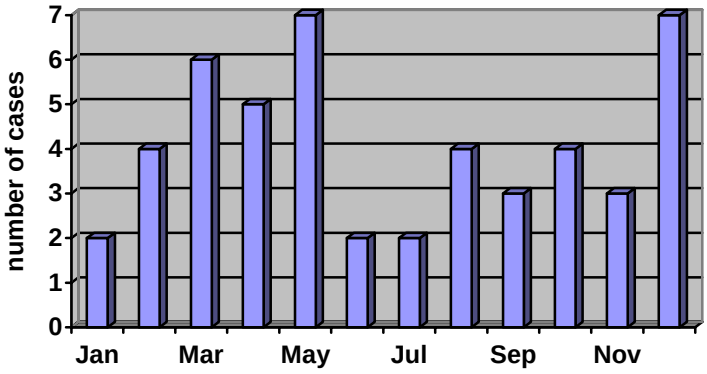

Figure 2: **Month of Onset**

Ethnic Rates
Ethnicity was documented for 45 children (92% of all cases). Of these 49% were identified as NZ European (19 cases) or part-NZ European (3 cases). 29% were Maori (9 cases) or part-Maori (4 cases). 20% were Pacific Islander (8 cases) or part-Pacific Islander (one case); of these four were Samoan or part-Samoan, three Tongan, one Niuean and one Fijian. 13% were of East Asian (5 cases) or part-East Asian origin (1 case); two were Korean, two Chinese, one part-Chinese and one Japanese.

Table 1: **Incidence and Ethnicity *(cases aged less than 5 year)**

Ethnic Group	Number of cases	Population at risk	Annual incidence (95% CI) per 100,000 aged <5 yrs
All cases	42	522,078	8.0 (5.8, 10.9)
NZ European	18 (includes 2 mixed-race)	390,354	4.6 (2.7, 7.3)
Maori	13 (includes 3 mixed-race)	135,120	9.6 (5.1, 16.5)
Pacific Islander	8 (includes 1 mixed-race)	65,350	12.2 (5.3, 24.1)
Oriental	6 (includes 1 mixed-race)	18,501	32.4 (11.9, 70.6)
Not stated	2		

**Note that the sum of the cases in the various ethnic groups add to more than the total as those of mixed ethnicity – for both the numerator and the denominator – are included in all relevant categories.*

Annual incidence rates for the 42 cases per 100,000 population for children aged less than five years were calculated: this analysis included 37 cases described as belonging to one racial group, 2 cases each from 2 racial groups and I case belonging to 3 racial groups. Overall the incidence was 8.0 per 100,000 aged under 5. Analysis by ethnic group shows incidence rates of 4.6 for NZ European, 9.6 for Maori, 12.2 for Pacific Islanders and 32.4 for children of Chinese, Korean or Japanese origin.

Diagnosis and Admission
K.D was diagnosed at a mean of 7.4 days (median of 6 days) from the onset of the illness (range 0-39 days). In 40 cases (82%) the diagnosis was made within the first seven days of the illness[12] In infants less than one year, KD was diagnosed before the eighth day in 12 of 13 (92%). The diagnosis was made in a mean of 2.2 days after admission to hospital (range 0-12 days). Of the 48 who were admitted 38 (79%) were diagnosed within three days of admission.

In 18 of the 49 cases (37%) fever and five diagnostic features were noted and in 25 (51%) fever and four features. Five cases had fever and three other features only. The single case fatality, was an "atypical" case with the diagnosis made at post mortem.

***Table 2:* Diagnostic Features in the 49 Cases**

Diagnostic feature	No. (%)	Comments
Fever for 5 days	43 (88%)	4 days in 1 case,3 days in 2, unknown in1, unspecified in 2 cases
Changes of the extremities	39 (80%)	
Polymorphic rash	46 (94%)	Fatal case had neither rash nor eye involvement
Bilateral conjunctivitis	46 (94%)	
Lip or mouth involvement	45 (91%)	Fatal case had no oral lesions. 1 case had neither oral nor eye changes
Cervical lymphadenopathy	29 (59%)	
Coronary artery dilatation Coronary artery aneurysm Coronary arteritis	13 1 1	

Laboratory Studies
The three late presenting cases were omitted from all the laboratory results.

Haematology
The lowest haemoglobin (Hb) level recorded during the course of the illness was 103 g/L (range 70-135). This trough was noted on Day 6 (range 3-20) from the onset of the illness.

Maximum leukocyte count (WBC) was 18.8×10^9/L (range 5.0-55.2) noted on Day 5.8 (range 3-18) of illness. In 43/46 cases the peak leukocyte count exceeded 10.0×10^9/L.

Peak platelet level was 577×10^9/L (range 266-1497), on Day 8.2 (range 2-19).

Inflammatory Markers

Erythrocyte Sedimentation Rate (ESR) was measured in 31 cases (63%). Excluding the three late presentations the peak level was 78mm/hr (range 10-180) which was noted on Day 6.8 (range 3-19) of illness. In 29 of the 31 cases the peak level exceeded 30 mm/hr.

C-reactive protein (CRP) was measured in 34 cases (69%). Excluding the three late presentations the peak level was 144 mg/dL (range 7-349) noted on day 5.9 (range 2-17) of illness. In 21 of 34 cases the peak level exceeded 100 mg/dL.

Evidence of Streptococcal Infection

Evidence of streptococcal infection was present in 4 of 19 cases where testing was performed. In 11 cases serological tests for ASOT & antiDNAse were undertaken, these were negative in 8 and positive in 3 patients. In the 8 cases where throat swabs were taken there was no growth in 6, no result documented in one and growth of streptococci in one sample.

Other Investigations

Urinalysis was performed on at least one occasion in 39 cases (80%) and was normal in 27. In 8 cases there was sterile pyuria, 3 had proteinuria, 2 ketonuria and in one microscopic haematuria was present.

Abdominal ultrasound was performed in 6 cases and was normal in 5. One patient had mild obstructive changes of the urinary tract.

No tissue biopsies were taken.

Cardiac Symptoms and Signs

In two cases (4%) abnormal cardiac symptoms or signs were present. One child had pericardial effusion and one (the fatal “atypical” case) had cardiac failure. Electrocardiogram (ECG) examination was performed in 35 cases (71%) and showed no abnormality in all 35. One case showed sinus tachycardia.

Echocardiogram Studies

All cases had at least one echocardiogram performed.

The first study was performed at a median of ten days and mean of 21 days from the onset of illness (range 3-106 days). Coronary artery ectasia or dilatation was seen in 13 cases (27%). One small aneurysm of 4mm was detected after 51 days. Two cases showed pericardial effusion, neither of which had coronary artery dilatation. Two had valvular regurgitation, neither had coronary artery dilatation. One study (the fatal “atypical” case) was reported as showing as an atrial septal defect with an aneurysmal fossa ovalis, mild mitral regurgitation but no coronary artery dilatation.

A second echocardiogram was performed on 22 patients (45% of all cases). This echocardiogram was performed at a mean of 37 days from the onset of illness (range 6-82 days). For the 17 cases who had an abnormality on the first scan, 10 (59%) had a second scan performed, 3 failed to attend for their second scan, two were awaiting the second scan at the time of report, one had moved abroad and one had died. Of the 32 children with a normal first scan 11 (34%) went on to have a second scan. The second scan showed an abnormality in 7 cases, all of whom showed coronary artery dilatation, none severe.

A third scan was performed in 5 cases (10% of all cases), at a mean of 91 days from the onset of illness (range 51-188 days). The third scan showed no abnormality in three cases, two of whom had shown coronary artery dilatation on their previous scan. The third patient had two previous normal studies. The two remaining patients both showed continued but improving coronary artery dilatation.

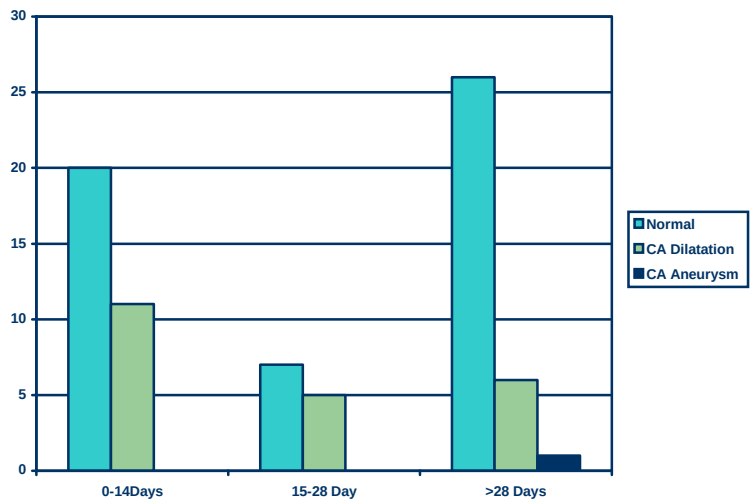

Figure 3: **Echocardiogram result timed from onset of illness**

Overall 43 scans were performed within the first 28 days after the onset of illness. Eleven of these "early" scans demonstrated coronary artery dilatation, at a median of seven days. Only six of these patients had a "late" follow-up scan, defined as greater than 28 days from onset of illness. Three of these 6 had improved or resolved. Seventeen (35%) of cases did not have any scan after 28 days. Only one aneurysm was identified, this was detected on day 51.

In total, 15 patients (31% of all cases), all aged less than five years, had coronary artery abnormalities noted at some point during their illness. 13 had dilatation, one a small LCA aneurysm and the fatal case had luminal occlusion but not dilatation.

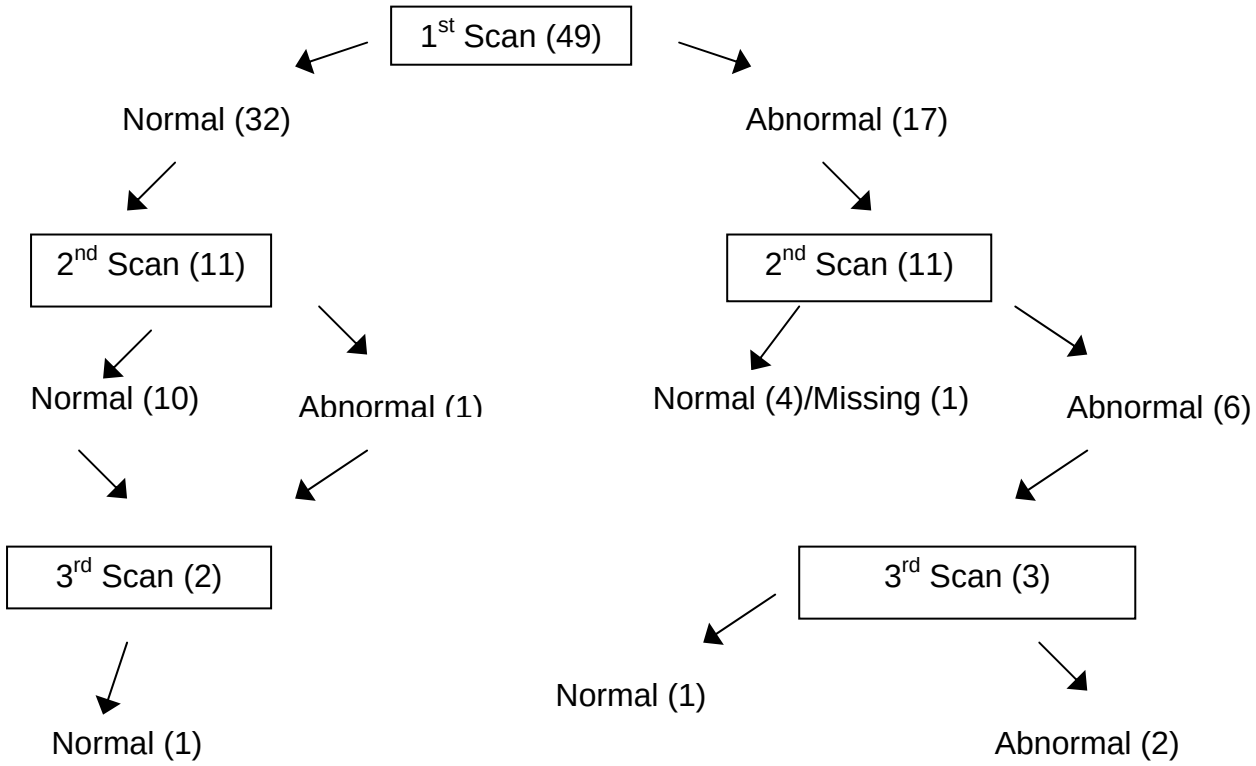

Figure 4: **Echocardiogram Studies**

Treatment
Forty–five patients (92%) were treated with intravenous gammaglobulin (IVIG), 2g/Kg, and aspirin. Of the 4 patients who did not receive IVIG 3 were the late presenters and one was the fatal "atypical" case. IVIG was administered at a median of day six, mean of 5.7 days (range 3-12) from the onset of symptoms and in 39 cases (87% of all those receiving IVIG) the treatment was given within the first 7 days of illness. IVIG was given at a mean of 1.8 days (range 1-8) from admission to hospital.

Three patients had a second course of IVIG on account of continuing symptoms. This was given 2, 4 and 7 days after the first course.

Forty-six patients (94%) were treated with aspirin. Of the three untreated cases one was a late presenter, one was the fatal "atypical" case and in the remaining case no reason was specified for aspirin being not given. The initial dosage of aspirin prescribed varied between 5 and 100 mg/Kg/day, as shown in Table 3. One child was treated with methylprednisolone and four received antibiotics.

Table 3: **Initial dosage of Aspirin**

Mg/Kg/day Aspirin	No. Cases
100	19
80	7
50	1
30	5
20	3
5	11

The Fatal Case
One child in this series died, giving an overall case fatality rate of 2% (Confidence limits 0-11%).

This child was a 3 month old part-Maori male infant admitted on the 12th day of fever and diarrhoea. No rash or mucosal changes were noted. Peak Hb was 94g/L, WBC 37.5×10^9/L, and platelets 735x10 /L, CRP was 309 g/L. There was progressive vomiting and diarrhoea, then a seizure.

Echocardiogram showed a small atrial septal defect with an aneurysmal fossa ovalis, mild mitral regurgitation. At laparotomy for severe abdominal symptoms there was enterocolitis and vascular thrombosis. Six days later a second laparotomy revealed progressive necrosis of the entire ileum and jejunum. Three days later, 22 days after the inset of the illness he developed cardiac failure and died. Post mortem examination showed generalized arteritis affecting coronary and mesenteric and other arteries. There was infarction of the myocardium, spleen, kidney and entire small bowel. No coronary artery dilatation or aneurysms were reported. This case was published emphasizing the overlap between KD and infantile polyarteritis nodosa [26].

Discharge from Hospital
The mean duration of admission was 5.1 days (range 1-12 days), median 4 days. 10 patients (22% of all cases) had a length of stay greater than 7 days. Forty children (80% of all cases) were noted to be receiving (mainly low-dose) aspirin at the time of discharge from hospital.

Discussion

KD in New Zealand is primarily a disorder affecting preschool children. The age distribution and male preponderance from this study are similar to reports from Australia [12], the U.K [13], USA[14], Japan [15] and Jamaica [16]. Cases were reported every month throughout the year with no clearly defined seasonal trend. There was no evidence of an epidemic pattern as observed in Japan [17]. There was no clear geographical trend, and cases were reported affecting children resident from Whangarei to Dunedin. The incidence of KD approximated to the distribution of the population throughout the country with 73% of cases occurring in the North Island, home to 76% of the population, and 23% in the South.

With approximately 24 new cases per year, the incidence of 8.0 cases per 100,000 population of children aged less than five years is almost double the incidence of 5.1 noted by the retrospective study of Gentles *et al* in Auckland children during 1979-1988 [9]. This may reflect the greater awareness of KD among medical practitioners, and the prospective methodology of the current study. The incidence of KD in New Zealand is significantly greater than observed in Australia 1993-5 when a prospective survey under the auspices if the Australian Paediatric Surveillance Unit found an incidence of 3.7 [12]. Reports from the United Kingdom in 2002 indicate an incidence of 8.1 for children under five admitted to hospital and diagnosed with KD[8], more than double the incidence of 3.4 cases identified in the British Paediatric Surveillance Unit prospective study in 1990 [13]. Likewise a rising incidence has been observed in Japan, from 73.8 in 1987 to 111.7 in 1998 [7]. The possible reasons for this apparent increase include raised awareness of the diagnosis or a true increase in incidence.

It is noteworthy that despite KD having such a characteristic and dramatic presentation in its classic form, the condition was only recognized and described in 1967, suggesting that it is a true emerging condition. Differential diagnoses include scarlet fever and viral exanthems, especially measles.

Table 4: **National Incidence of KD (cases per 100,000/ population less than 5 years)**

Incidence	Country	Year	Reference
2.7	Jamaica	1986-1998	**16**
3.7	Australia	1993-1995	**12**
4.0	England	1991-1992	**13**
5.1	N.Z.(Auckland)	1979-1988	**9**
8.0	**N.Z.**	**2001-2002**	**Present study**
8.1	England	1999-2000	**8**
17.1	U.S.A.	2000	**18**
17.6	U.S.A.	1997	**18**
73.8	Japan	1987	**7**
108.0	Japan	1997	**7**
111.7	Japan	1998	**7**

There were clear differences in the incidence of KD between ethnic groups. European children suffered about half the incidence compared with Maori children. Though the total number of cases were small the incidence for Pacific island children and East Asian children were respectively three-fold and over seven-fold that seen in the European population. Studies from the United Kingdom and United States have shown that within a defined geographical area children of Asian and Pacific Island origin suffer higher rates than their European peers, even when socioeconomic factors are taken into account [6,19]. These

observations lend substance to the theory that genetically determined factors play an important part determining the susceptibility to KD of children from differing ethnic populations.

Cardiac complications can be minimized by the timely administration of intravenous immunoglobulin (IVIG), therefore it is essential that the diagnosis is considered as soon as possible following the onset of symptoms[3,4]. Potential sources of delay during the diagnostic process can occur when seeking help from primary health care, during referral to secondary care, and when there is delay in the consideration of the diagnosis of KD or in the delivery of appropriate treatment. The critical time when IVIG should be administered is unknown. Meta-analysis of clinical trails has shown that treatment within the first ten days of the onset of illness will reduce the incidence of cardiac complications [20], however the study of Zhang *et al* indicated that the benefits of IVIG declined after the eighth day of illness [21]. Our study indicates that KD in New Zealand is diagnosed and treated rapidly with 80% of cases, and 85% of under-one year olds, being diagnosed and treated with IVIG within eight days of onset of illness. In the UK study only 61 % of cases received IVIG and the median interval from first symptom to treatment was nine days (range 1 to 27 days) [13]. The Australian survey found that 64% of cases were diagnosed within 10 days of whom 81% received IVIG in that time; thus only 52% of all Australian cases were both diagnosed and treated with IVIG within the first ten days of illness [12].

The spectrum of physical findings observed in the study group was similar to those seen in the national studies from the U.K., Australia and Jamaica[12, 13, 16] (Table 5).

Table 5: **Clinical Features**

Clinical Feature	New Zealand No. (%)	Australia No. (%)	U.K. No. (%)	Jamaica No. (%)
Fever for more than 5 days	43 (88)	N/A	146 (90)	N/A
Changes of the extremities	39 (80)	127 (93)	143 (88)	34 (60)
Rash	46 (94)	130 (95)	156 (96)	43 (75)
Oral changes	45 (91)	128 (93)	140 (86)	48 (84)
Conjunctivitis	46 (94)	123 (90)	127 (78)	46 (81)
Cervical glands	29 (59)	59 (43)	136 (83)	24 (42)
Coronary artery abnormalities	15 (31)	36 (26)	39 (24)	16 (28)

Abnormalities of haematological profile and inflammatory markers were most prominent towards the end of the first week of illness. Though most cases had significant changes there were a number in whom elevation of white cells, platelets and inflammatory markers were not striking.

In our study there was limited evidence of streptococcal infection, though specific testing was only performed in 39% of cases. Of these 19 individuals, a total of four (<20%), had either throat swab or serological evidence of streptococcal infection. We were concerned specifically about streptococcal infection on account of the frequency of streptococcal illness in New Zealand, also on account of the symptomatology shared by both KD and streptococcal scarlet fever.

Cardiac symptoms, cardiac signs and ECG abnormalities were rarely encountered in the study group. However there was a relatively high incidence of echocardiographic abnormalities, specifically relating to coronary artery dilatation, and 31% of all cases were thus affected. Many of these cases were detected on early scanning within 28 days of illness onset. “Transient” coronary artery (CA) dilatation is detected in 40% of KD at a median of 10 days more than half of which will resolve before six weeks. It is concerning that one third of cases did not have a scan after 4 weeks as is recommended

The design of the NZPSU reporting system is such that individual case notes cannot be identified or reviewed by the study group; therefore individual echocardiograms could not be reviewed by any one investigator. It is likely that some variation of interpretation of what constitutes coronary artery dilatation or ectasia occurred at different centres. However, it is also possible though unlikely that large proximal coronary artery aneurysms greater than 4mm were missed. The incidence of coronary artery changes is similar to studies from the UK where 24 % [12], Australia 26 % [13], and Jamaica 28 % [16] were so affected. In most instances the coronary dilatation was relatively minor, and had resolved by the time of the second or third scan. It may be that the comparatively high rate of coronary artery abnormalities was a reflection of the predominance of early scanning with 43 of 76 studies being performed in the first 4 weeks. By comparison only 21% of late scans were abnormal and only one demonstrated a definite aneurysm. Most of the remainder showed resolving CA dilatation

The timing of echo studies was very variable with the first study being performed at any time from three to 106 days from the onset of illness. The first echo was performed at a median of 10 days (mean 21 days) from the onset of illness, despite the diagnosis being made on the seventh (mean) day. We suspect that the heterogeneity of timing of echo studies largely reflects variation throughout the country in recommendation for the first echo study. Some centres have not routinely performed an echocardiogram on admission as it is not required for diagnosis, and echocardiography has not been shown to alter outcome in standard cases where the patient responds promptly to the IVIG administration. We believe that it is desirable for there to be greater uniformity of practice regarding the timing of scanning.

The best practice evidence supports that all patients are scanned at one to two months from the onset of illness, but it remains unproven that early presenters who respond promptly to IVIG will benefit from early echocardiography. A selective policy of early scans is appropriate for those with abnormal cardiovascular findings, abnormal ECG, failure of response to IVIG, late presentation > 14 days, and atypical cases where there is diagnostic uncertainty. It is essential that prompt echocardiography be performed in non-responders for these individuals have a high chance of coronary abnormalities including aneurysms and coronary thrombus for which newer anti-thrombotic treatments may be indicated. Some authorities recommend that studies be performed at diagnosis, at 10-14 days and six to eight weeks after the onset of disease in all cases [22,23]. There is no evidence that further scanning beyond 6-8 weeks is indicated if studies have shown normal coronary arteries [25].

In the acute phase children with coronary artery aneurysms >4mm in diameter should have at least weekly studies to monitor for aneurysm progression and thrombus formation. Depending on the size of the aneurysms scanning should be repeated 6-12 monthly [22].

There was also considerable variation in the quality of detail documented in the echo report. Some were reported as “normal echocardiogram” whilst others provided great detail about precisely what had been seen, including the coronary artery measurements related to *z* scores adjusted for body surface area. If a standard pattern of reporting was adopted this could improve the quality of care.

Most patients (92% of all cases) were treated with a single course of IVIG 2g/Kg, administered as soon as the diagnosis of KD had been considered. Only the remaining 8% who were late presenting cases and the atypical case did not receive IVIG. In the Australian study there was greater variability in the IVIG dosing regime with 78% of those receiving IVIG (constituting 79% of all cases diagnosed) having 2g/Kg [12]. In the British study[13] no patient received 2g/Kg of IVIG administered as a single dose as the survey had been undertaken prior to studies indicating the preferred dosage regime [20, 24].

There was greater variation in the dose of aspirin administered in the acute phase of the illness. The trend was for most patients to be treated with a high dose of aspirin (80-100mg/Kg/day) rather than a moderate dose (30-50mg/Kg/day) regime that may be better tolerated. The Terai and Schulman meta-analysis showed no benefit of high dose (80-100mg/kg/day) aspirin in reducing the incidence of coronary artery abnormalities[24]. For this reason, we and others recommend using 30mg/Kg/day in the acute illness then 3-5mg/Kg/day once the fever has settled[22].

The median duration of admission was 4 days though almost a quarter of total cases had a length of stay greater than 7 days. The US study noted a median length of stay of three days, with no significant difference between those aged less than five years and older children. [18]

The fatal case raises a number of issues and illustrates the problem of "atypical" KD. Such cases will of necessity be less likely to receive rapid diagnosis and treatment. Table 5 indicates that such fatal cases are often diagnosed only at post mortem, and contribute disproportionately to case fatality rates observed across the globe. Of the 9 cases diagnosed post mortem noted in the studies referred to in Table 5, at least 4 were "atypical". Six were documented as being white males (no details for the remainder), and at least three had vaso-occlusive disease (pathological detail not documented in all the remainder). It has been noted that accelerated vaso-occlusive disease poses specifically a particular diagnostic problem as there may be severe coronary artery luminal narrowing despite the unremarkable echocardiographic appearance of coronary artery diameters[10, 11]. We speculate that these fatal cases, which could be considered as falling within the Infantile polyarteritis nodosa range of the KD spectrum, may follow a significantly different pathophysiological pathway to that of "typical" KD Such cases warrant further study in order to permit more rapid identification and to develop more effective management.

Table 6: **International Case Fatality Rates**

Country	Year of study	Total no. cases	Total no. deaths	Diagnosis of KD made post mortem	Case fatality rate	Confidence limits
New Zealand [9]	1979-88	34	2	1	5.9%	1-20%
New Zealand	2001-2	49	1	1	2%	0-11%
U.K.[13]	1990	163	6	5	3.7%	1-8%
Australia [12]	1993-5	139	2	2	1.4%	0-5%
Jamaica [16]	1986-98	98	0	0	0%	0-4%
U.S.A. [18]	1997 & 2000	4248	0	0	0%	0-0.08%

Practice Points

1) The incidence of KD in New Zealand 2001-2 was 8.0 per 100,000 aged less than 5 years.
2) There is a notable ethnic influence on incidence. The risk to Maori being two fold, Pacific Island three fold, and East Asian children being seven to eight fold that of European children.
3) Preschool children account for most cases, and are more prone to coronary artery abnormalities.
4) Diagnosis and treatment is usually provided rapidly in New Zealand, a reflection of the high standard of paediatric care. Best evidence based practice is followed with respect to IVIG with patients uniformly receiving 2g/kg of IVIG
5) While there is no evidence to support giving greater than 30-50 mg/kg aspirin as initial 'high' dosage higher doses are still frequently used.
6) Coronary artery dilatation is common in the first month. This is usually mild and resolves without cardiac sequelae.
7) There is no uniformity in timing and method of reporting echocardiograms; a selective policy in the acute phase and mandatory scanning at one to two months is recommended.
8) The mortality rate appears high due to an "atypical" case, diagnosed *post mortem*, occurring in the study time period.

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Appendix

CASE DEFINITION

(Based on Rowley and Shulman, Pediatric Clinics of North America, April 1999)

Fever persisting five days or more (in the presence of other diagnostic criteria the diagnosis may be made before the fifth day of fever)

AND at least four of the following:

1. Changes of peripheral extremities:
   a. Initial stage: Reddening of palms and soles, indurative oedema.
   b. Convalescent stage: Membranous desquamation from fingertips.
2. Polymorphous exanthema
3. Bilateral conjunctival congestion
4. Changes of lips and oral cavity: Reddening of lips, strawberry tongue, diffuse injection of oral and pharyngeal mucosa
5. Acute non-purulent cervical lymphadenopathy.

AND

Disease not explained by other disease process (in particular streptococcal infection or measles)

Patients with fever and 3 other criteria may be diagnosed as Kawasaki disease if coronary abnormalities develop.

Conditions Ever Monitored by NZPSU

Table 6: **All Conditions Ever Monitored by NZPSU**

Condition	Abb.	Commenced	Concluded
Acute flaccid paralysis	AFP	October 1997	Ongoing
Haemolytic uraemic syndrome	HUS	January 1998	Ongoing
Congenital rubella syndrome	CRS	January 1998	Ongoing
Perinatal HIV exposure	HIV	January 1998	Ongoing
Vitamin K deficiency bleeding	Vit K	January 1998	Ongoing
Neonatal herpes simplex infection	HSV	January 1998	December 2000
Subdural haemorrhage (<2 years)	SDH	January 1999	December 2002
Retinopathy of prematurity (stage III)	ROP	January 1999	December 2000
Diabetes mellitus	DM	January 1999	December 2000
Fetal alcohol syndrome	FAS	July 1999	December 2001
Kawasaki disease	KD	January 2001	December 2002
Bronchiectasis	BE	January 2001	December 2002
Idiopathic nephrotic syndrome	INS	July 2001	Ongoing
Inflammatory bowel disease	IBD	January 2002	December 2003

Note: all conditions monitored for children <15years unless otherwise stated.

International Network of Paediatric Surveillance Units (INoPSU)

Establishment of INoPSU

The network was formed in August 1998 at a meeting of 10 Paediatric Surveillance Units expressing a desire to link with each other. This took place at the 22nd International Congress of Paediatrics in Amsterdam, The Netherlands. The first INoPSU conference was held in 2000 in Canada and was attended by representatives of the existing units. Subsequent meeting have been help in York, England in 2002 and Lisbon, Portugal in 2004. Dr Nigel Dickson has attended these recent meetings.

Mission

The mission of INoPSU is the advancement of knowledge of uncommon childhood infections and disorders and the participation of paediatricians in surveillance on a national and international basis so as to achieve a series of benefits.

Aims

- facilitating communication and cooperation between existing national paediatric surveillance units;
- to assist in the development of new units;
- to facilitate sharing information and collaboration between researchers from different nations and scientific disciplines;
- to share information on current, past and anticipated studies and their protocols, and on conditions that have been nominated for surveillance but are not selected;
- to encourage the use of identical protocols to potentially enable simultaneous or sequential collection of data on rare paediatric disorders in two or more countries;
- to share and distribute information of educational benefit to constituent units, notably on study and surveillance methodologies;
- to share school techniques and models of evaluation for units;
- to peer review and evaluate existing and proposed units;
- to identify rare disorders of mutual interest and public health importance for cooperative surveys through each national unit;
- to collaborate with and provide information to other groups interested in rare childhood diseases such as parent support groups;
- to respond promptly to international emergencies relating to rare childhood conditions where national and international studies can make a contribution to science or public health.

Members of INoPSU

Founding members:

- Australian Paediatric Surveillance Unit (APSU)
- British Paediatric Surveillance Unit (BPSU)
- Canadian Paediatric Surveillance Programme (CPSP)
- German Paediatric Surveillance Unit (ESPED)
- Latvian Paediatric Surveillance Unit (LPSU)
- Malaysian Paediatric Surveillance Unit (MPSU)
- Netherlands Paediatric Surveillance Unit (NSCK)
- New Zealand Paediatric Surveillance Programme (NZPSU)
- Papua-New Guinea Paediatric Surveillance Unit (PNGSU)
- Swiss Paediatric Surveillance Unit (SPSU)

Additional Members

Welsh Paediatric Surveillance Unit (2000)
Portuguese Paediatric Surveillance Unit (2001)
Irish Paediatric Surveillance Unit (2001)
Greece and Cyprus Paediatric Surveillance Unit (2004)

Associate Members

Trinidad and Tobago Paediatric Surveillance Unit (2004)
British Ophthalmological Surveillance Unit

Administration of the Association

In order to carry out the aims and direct the activities of INoPSU a secretariat has been set up. From 2004 Professor Rudi von Kries (ESPED) will act as convenor, taking over from Professor Elizabeth Elliott (APSU), Dr R Pereira (NSCK) will act as deputy convenor. Richard Lynn (BPSU) will act as communications liaison.

International Collaboration

New Zealand paediatricians who are interested in undertaking international studies, or compare the rates of uncommon disease between countries are encouraged to consider using INoPSU for this purpose. Please contact Nigel Dickson for further information.

Table 7: **Members of INoPSU**

Country	Unit	Email	Website
Australia	APSU	apsu@chw.edu.au	http://apsu.inopsu.com
Britain	BPSU	enquires@rcpch.ac.uk	http://bpsu.inopsu.com
Canada	CPSP	cpsp@cps.ca	www.cps.ca/english/cpsp
Germany	ESPED	heinrich@med.uni-duesseldorf.de	www.esped.uni-duesseldorf.de/
Ireland	IPSU	gilld@iol.ie	
Latvia	LPSU	aspedlat@com.latnet.lv	
Malaysia	MPSU	jho@pc.jaring.my	
Netherlands	NSCK	r.pereira@pg.tno.nl	
New Zealand	NZPSU	nzpsu@stonebow.otago.ac.nz	www.paediatrics.org.nz
Papua New Guinea	PNGPSU	hopepng@datec.com.pg	www.hopeww.org/where/png/png5.htm
Portugal	PPSU	ana.moreira@sb.com	
Switzerland	SPSU	hans-peter.zimmermann@bag.admin.ch	
Wales	WPSU	John.Morgan@eglam-tr.wales.nhs.uk	

INoPSU website: www.inopsu.com

Table 8: **Characteristics of the Paediatric Surveillance Units**

Country	Population ($x10^6$<15years)	Established	Approx. no respondents
Australia	3.9	1992	1000
Britain/Eire	12.8	1986	2200
Canada	7.5	1996	2400
Germany	12.0	1992	500*
Ireland	1.3	1996	150
Latvia	0.4	1996	22
Malaysia	7.6	1994	400
Netherlands	3.0	1992	640
Papua New Guinea	1.9	1996	40
Portugal	1.8	2000	2000
New Zealand	0.83	1997	200
Switzerland	1.3	1995	45*
Wales	0.65	1995	135*

* Heads of Paediatric Centres

Table 9: **Conditions Under Surveillance Worldwide 2003**

Conditions Under Surveillance	Country
Acute encephalitis	Portugal
Acute flaccid paralysis	Australia, Canada, New Zealand, Papua New Guinea, Switzerland
Acute fulminant liver failure	Malaysia
Acute rheumatic fever	Switzerland
Adverse effects from complementary or alternative medicine	Australia, Wales
Alcohol & Children	Ireland
Anaphylaxis following food injestion	Australia
Asthma deaths	Malaysia
Asthma	Malaysia
Autism under 5 years	Ireland
Atypical mycobacterial infections	Netherlands
Ataxia	Netherlands
Atypical tuberculsis infection	Netherlands
CHARGE association/syndrome	Canada
Childhood conversion disorder	Australia
Childhood tuberculosis	Wales
Childhood inflammatory bowel disease	New Zealand
Children hospitalised for early onset eating disorder	Australia
Complicated pneumonia including empyema	Wales
Congenital cytomegalovirus infection	Australia
Congenital hypothyroidism	Papua New Guinea
Congenital rubella	Australia, Britain, Canada, New Zealand, Switzerland, Netherlands
Congenital toxoplasmosis	Britain
Diabetes mellitus/insulin-dependent/<5 years	Germany, Latvia, Papua New Guinea, Wales, Ireland
Down's Syndrome	Netherlands
Duchenne muscular dystrophy	Malaysia
Endocrinology	Latvia
Facial palsy	Wales
Fetal alcohol syndrome	Australia
Fragile X	Ireland

Group B streptococcal infections (neonatal)	Portugal, Germany
Haemolytic Uraemic Syndrome	Canada, New Zealand, Portugal, Switzerland
Haemorrhagic disease of the newborn (vitamin K deficiency bleeding)	Australia, Britain, New Zealand
Hereditary periodic fever syndrome	Germany
Hepatitis C virus infection	Canada
Haematology/oncology	Latvia
Hemoglobinopathy	Netherlands
HIV/AIDS +/- perinatal exposure to HIV	Australia, Britain, Malaysia, Netherlands, New Zealand, Papua New Guinea, Latvia
Hypernatrenia	Netherlands, Wales
Hypophosphatasia	Germany
Idiopathic nephrotic syndrome	New Zealand, Netherlands
Idiopathic thrombocytopenic purpura	Netherlands
Imported tropical diseases: malaria, schistosomiasis, leishmaniasis	Germany
Ingestion of lamp oil (intoxications)	Germany
Inherited hypocalemic salt-losing tubulopathies/Bartter-like syndromes and narcolepsy	Germany
Insufficient breast-feeding	Netherlands
Internal abdominal injuries in children under 14 years	Britain
Intussuception	Switzerland
Invasive *Haemophilus influenzae* infection	Germany
Invasive group b streptoccal disease	Portugal
Ischaemic stroke in infants (neonatal sinus venous thrombosis)	Germany
Junevile Idiopathic Arthitis	Wales
Kawasaki Disease	Portugal
Kernicterus	Germany
Lap-belt syndrome	Canada
MCADD	Netherlands
Malaria	Netherlands
Meningoencephalitis	Portugal
Munchausen by proxy syndrome	Australia
Necrotising enterocolitis	Papua New Guinea
Necrotising fascitis	Canada
Neonatal abstinence syndrome	Wales
Neonatal congenital heart disease	Malaysia

Neonatal herpes simplex virus infection	Canada, Switzerland
Neonatal HSV	Australia
Neonatal hyperbilirubinaemia	Canada
Neonatal meningitis	Malaysia
Neonatal sinus venous thrombosis	Germany
Nephrology	Latvia
Neural tube defects	Switzerland, Ireland
Neurological endemic cretinism	Papua New Guinea
Newly diagnosed diabetes	Wales
Pallative Care	Wales
Physical Child Abuse	Wales
Pneumococcal sepsis/meningitis	Germany
Prader-Willis Syndrome	Canada
Progressive intellectual and neurological deterioration/CJD	Canada
Renal tubular acidosis	Papua New Guinea
Reye syndrome	Latvia
RSV virus	Switzerland
Septo-optic dysplasia	Wales
Severe complications to varicella	Britain
Shaken Baby Syndrome	Switzerland
Splenecotomy and hydrosplenism in childhood	Wales
Small bowel insufficiency	Netherlands
Status epilepticus	Ireland
SSPE	Germany
Systematic pneumococcal infection & meningitis	Germany
Subacute sclerosing panencephalitis	Papua New Guinea
Subdural haemorrhage	Wales
Suspected fatal adverse drug reactions	Britain
Tick-borne encephalitis	Switzerland
Type 1 Insulin Dependent Diabetes Mellitus	Ireland, Papua New Guinea, Portugal
Type 2 non-insulin diabetes mellitus	Ireland
Tuberculosis	Britain
Varicella/zoster infection/complications	Switzerland, Britain, Germany
Vitamin D deficiency rickets	Canada

List of Clinicians with 100% Return Rate 2003 (& 2002)

Clinicians who had a 100% return rate in both 2002 and 2003 are underlined

Thank you to those clinicians who returned all of their cards in 2003!

Aftimos	Salim	Grimwood	Keith	Parsons	Alan
Aho	George	Hall	Kate	Pattemore	Philip
Aiken	Richard	Gunn	Alistair	Pinnock	Ralph
Armishaw	Jeremy	Hewson	Michael	Pitcher	Lydia
Asher	Innes	Harding	Jane	Radcliffe	Marlon
Barry	John	Hofman	Paul	Ramadas	Ram
Bates	Giles	Heron	Peter	Rowley	Simon
Battin	Malcolm	Hornung	Tim	Richardson	Vaughan
Beard	Rachel	Hunter	Warwick	Reith	David
Beasley	Spencer	Jackson	Pam	Rudge	Susan
Bourchier	David	Jamison	David	Sanders	John
Bradley	Stephen	Jankowitz	Peter	Segedin	Elizabeth
Breen	Felicity	Kelly	Andrew	Selby	Robyn
Bremner	Catherine	Kolbe	Anne	Scorer	James
Broadbent	Roland	Kerr	Archie	Shaw	Robyn
Brown	Jeff	Kushel	Carl	Simpson	A. K.
Buchanan	Leo	Leadbitter	Philip	Sinclair	Jan
Buckley	David	Lear	Graeme	Sinclair	Lynette
Byrnes	Cass	Lees	Hugh	Skeen	Jane
Calder	Louise	Lennon	Diana	Skinner	Jon
Campanella	Silvana	Leversha	Alison	Shillito	Paul
Campbell	Moira	Liang	Allen	Smith	Warwick
Campbell-Stokes	Priscilla	McArthur	John	St John	Martyn
Caseley	Terry	McIlroy	Peter	Stanley	Thorsten
Clark	Phillipa	MacKenzie	Neil	Swan	Catherine
Clarkson	John	Maikoo	Rejesh	Taylor	Barry
Corban	Jenny	Malcolm	Stuart	Taylor	Paul
Coulter	Belinda	Lourens	Roelof	Teague	Lochie

Cutfield	Wayne	Marshall	Andrew	Tomlinson	Paul
Dalton	Marguerite	Manikkam	Noel	Trenholme	Adrian
Darlow	Brian	Maxwell	Fraser	Tsang	Bobby
De Sylva	Tony	Marks	Rosemary	Tuck	Roger
Dixon	Joanne	Meyer	Michael	Tyrrell	Vicki
Doran	John	Menard	Keith	Vogel	Alison
Dradge	Alan	Mildenhall	Lindsay	Voss	Lesley
Elder	Dawn	Nagel	Fred	Walker	Wendy
Farrell	Alan	Neutze	Jocelyn	Watson	Peter
Fleming	John	Newman	David	Webb	Alan
Ford	Rodney	Nicholson	Ross	Webster	Diane
Forster	Richard	Nicolls	Wayne	Wesley	Alison
Gavin	Raewyn	Mitchell	Ed	Weston	Phillip
Gapes	Stephanie	Morris	Max	Wills	Russell
Gentles	Tom	Montgomery	David	Wilson	Callum
Gillies	John	Moyes	Chris	Wilson	Nigel
Goldsmith	John	Morrison	Philip	Wilson	Ross
Graham	David	Palmer	Perry	Wong	Maisie

Congratulations to David Newman who was selected at random to win a $50 book token to be presented at the ASM of the Paediatric Society of New Zealand.

The Royal Australasian
College *of* Physicians

COPY

24 August 2004

Associate Professor Elizabeth Elliott
Director Australian Paediatric Surveillance Unit
Department of Paediatrics & Child Health
The Children's Hospital at Westmead
Cnr Hawkesbury Rd & Hainsworth Street
WESTMEAD NSW 2145

Dear Professor Elliott

MOPS points for APSU activities

I am writing in response to your letter of 26 May 2004 regarding approval of MOPS points for activities relating to participation in the Australian Paediatric Surveillance Unit activities.

The Board of CPD agreed that participating in the APSU or NZPSU would be a practice-related CME activity under current MOPS guidelines, with participants eligible to claim half a point an hour for contributing to the project. This includes reading the protocol sheets, returning the reporting cards and completing the questionnaires, which should be claimed on an hourly basis.

If multiple-choice questions were generated as a result of the submissions, then points could be attained in the teaching category for formulating the questions at 1 point per hour and under self-assessment for completion of the questions at two points per hour.

As regards quality assurance points, participants would need to demonstrate how participation in the APSU or NZPSU would contribute to enhancing the quality of their practice. This could involve documenting how the evaluation report produced had influenced their clinical practice.

Participants should keep copies of the documentation they submit, a record of the contribution that they have made to the surveillance system or a certificate of participation.

Yours sincerely

Associate Professor Peter Procopis
Co-Chair (Division of Paediatrics & Child Health)
Board of Continuing Professional Development

Associate Professor Geoffrey Metz
Co-Chair (Division of Adult Medicine)

cc. Shirley Jones, NZPSU

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