# Table of Contents

**Introduction**

**Aims**

**How the surveillance system works**

**Inclusion of new conditions**

**Surveillance Activities in 1998**

**Discussion**

**Reports on Individual Studies**

- Acute Flaccid Paralysis
- Congenital Rubella
- Perinatal HIV exposure
- Haemolytic Uraemic Syndrome
- Vitamin K Deficiency Bleeding
New Zealand Paediatric Surveillance Unit

We hope that you will find this 1st Annual Report of the New Zealand Paediatric Surveillance Unit of interest. In it we report on the establishment of the Unit and the findings from the first full year of its operation.

The successful first year was 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. We would specifically like to thank those who have arranged the stool samples that are required to ensure that each child with acute flaccid paralysis (AFP) is investigated to the level required by the World Health Organization. It is important to maintain reporting of AFP and the testing of stool samples to fulfill the WHO requirements to exclude polio.

We anticipate that this high rate of participation will continue through 1999 and beyond, particularly as more studies initiated by New Zealand paediatricians are now included. We appreciate that the conditions included in 1999 are more common than those originally studied and that this will require a greater time commitment from paediatricians. However we believe that this will also make the findings of more relevance to you.

We would like to acknowledge funding from the Ministry of Health to ensure necessary surveillance of AFP. In addition we wish to thank the Australian Paediatric Surveillance Unit for their assistance with the establishment of the Unit and the principal investigators of Australian studies who have helped in the ways explained in this report.

Barry Taylor  
Nigel Dickson  
Nicola Dow
Key Practice Points

**AFP**
- Surveillance of all cases AFP is needed to confirm the absence of polio in New Zealand
- The proportion of cases of AFP that have appropriate stool investigations for polio virus is lower than required by WHO
- Telephone notification to the NZPSU (03 474 7825) must be made as soon as a child is diagnosed with AFP. NZPSU will dispatch containers for stool samples which should be returned to ESR for virological testing
- Guillain-Barre syndrome (GBS) is the commonest cause of AFP among children

**Congenital Rubella**
- Women on child bearing years born outside New Zealand should be screened for rubella antibody at the first health encounter and vaccinated if appropriate
- CRS should continue to be a diagnosis searched for. It is likely there are currently undiagnosed and/or unreported cases
- Rubella virus continues to circulate as documented by virus laboratories

**HIV**
- The Ministry of Health recommends that asking about HIV risk should be a routine part of antenatal care
- The risk of perinatal transmission of HIV can be markedly reduced by anti-retroviral therapy started before delivery and other strategies
- Most, but not all, HIV infected women are from, or have had contact with people from parts of the world where heterosexual transmission of HIV is relatively common
- It is possible that not all HIV infected mothers will have been identified so paediatricians should be aware of the possibility of HIV infection in children they see
- *AIDS – New Zealand*, published quarterly and available from the AIDS Epidemiology Group (Fax 03 479 7298) provides information on HIV/AIDS in New Zealand.
Vitamin K deficiency bleeding

- Lead maternity carers should inform prospective parents of the issues relating to vitamin K deficiency bleeding and of the recommendations for prophylaxis. The current recommendations are that all newborns be given vitamin K by a single intramuscular injection at birth. Alternatively, vitamin K may be administered orally, but multiple doses are required.
- Cases of late-onset hemorrhage may occur, particularly among breast-fed infants, if vitamin K prophylaxis is not given. Infants with late onset disease should be investigated for liver disease.

HUS

- The increasing incidence of *E. coli* 0157 (and *campylobacter*) should alert health professional to the risk of HUS, particularly in young children
- Early culture of stools from children with bloody diarrhoea is necessary to identify the organism responsible for HUS
- Isolates of suspected or proven VTEC, and cases of HUS must be notified to the Medical Officer of Health for appropriate public health investigation and action
- Prevention of *E coli* 0157 infections requires high standards of public health surveillance and control in all stages of the food chain
Introduction

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

The aim of the NZPSU is to facilitate and improve the knowledge of rare 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 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 NZPSU was initially established, under a contract with the Ministry of Health, to provide active surveillance of acute flaccid paralysis (AFP). The World Health Organization (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 nine rare childhood conditions.

Aims

The aims of the NSPSU are:

- to establish and operate a system for monitoring acute flaccid paralysis, as part of the global certification of eradication of poliomyelitis, required by the World Health Organisation
- to facilitate national surveillance and improve the knowledge of uncommon childhood conditions in New Zealand
How the surveillance system works

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

Specialist paediatricians 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 specialist 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.

*Figure 1* shows the main components and information flows of the NZPSU. Every month, participants are sent a reply-paid card to report whether in the previous month they have seen any cases of the conditions under surveillance. However, cases of AFP must 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.

Regular surveillance reports are made to the Ministry of Health, specifically updating the progress with AFP surveillance. A newsletter updating surveillance of all the conditions is sent to the participants.
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 rare childhood condition (or a rare 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.

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.

Applications for new conditions to include in the surveillance system were sought from paediatricians in early 1998. It was decided to include subdural haemorrhage, retinopathy of prematurity and diabetes mellitus under the age of 10 from February 1999. All conditions are kept on the card for a minimum of two years.

Table 1: The members of the NZPSU Scientific Review Panel

<table>
<thead>
<tr>
<th>Member</th>
<th>Institution</th>
</tr>
</thead>
<tbody>
<tr>
<td>Professor Barry Taylor</td>
<td>Dunedin School of Medicine</td>
</tr>
<tr>
<td>Dr Nigel Dickson</td>
<td>Dunedin School of Medicine</td>
</tr>
<tr>
<td>Dr Alison Roberts</td>
<td>Ministry of Health</td>
</tr>
<tr>
<td>Dr Elizabeth Elliot</td>
<td>Australian Paediatric Surveillance Unit</td>
</tr>
<tr>
<td>Dr Jeff Brown</td>
<td>Palmerston North Hospital</td>
</tr>
<tr>
<td>Professor Brian Darlow</td>
<td>Christchurch School of Medicine</td>
</tr>
<tr>
<td>Professor Diana Lennon</td>
<td>University of Auckland</td>
</tr>
</tbody>
</table>
Surveillance Activities in 1998

In 1998, 163 clinicians participated in the system. The average response rate to the monthly mail-out of reporting cards was 96%, with no consistent set of non-responders. Table 2 shows the response rate per area.

Table 4: Response rate per health locality (as defined by the HFA) for 1998

<table>
<thead>
<tr>
<th>Health Locality</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Northland, Auckland</td>
<td>95</td>
</tr>
<tr>
<td>Waikato, Bay of Plenty, Taranaki</td>
<td>96</td>
</tr>
<tr>
<td>Wellington, Wairarapa, Manawatu, Wanganui, Tairawhiti, Hawkes Bay</td>
<td>97</td>
</tr>
<tr>
<td>Nelson, Marlbourgh, Canterbury, West Coast</td>
<td>94</td>
</tr>
<tr>
<td>Otago, Southland</td>
<td>100</td>
</tr>
</tbody>
</table>

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. Confining the system to conditions that are very rare will limit the demand on clinician's time, but conversely it will be less likely to provide useful information.

Figure 2 shows the percentage of clinicians on the mailing list that reported cases during 1998. The figure shows that 70.0% of participants did not report any cases, with 28.4% reporting between one and three cases. 1.2% reported 4 or more cases during 1998.
In 1998, NZPSU monitored 6 uncommon childhood conditions (Table 5). Some of the protocols and questionnaires used were adapted from those used by the Australian Paediatric Surveillance Unit.

### Table 3: Conditions under surveillance in 1998

<table>
<thead>
<tr>
<th>Condition (age range included)</th>
<th>Surveillance started</th>
<th>Principal Investigator(s)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Acute flaccid paralysis (&lt;15 years)</td>
<td>October 1997</td>
<td>Dr Nigel Dickson, Dr Paul Shillito</td>
</tr>
<tr>
<td>Haemolytic uraemic syndrome (&lt;15 years)</td>
<td>January 1998</td>
<td>Dr William Wong</td>
</tr>
<tr>
<td>Congenital rubella syndrome (&lt;15 years)</td>
<td>January 1998</td>
<td>Prof Diana Lennon</td>
</tr>
<tr>
<td>Perinatal HIV exposure (&lt;15 years)</td>
<td>January 1998</td>
<td>Dr Nigel Dickson, Dr Lesley Voss</td>
</tr>
<tr>
<td>Vitamin K deficiency bleeding (&lt;15 years)</td>
<td>January 1998</td>
<td>Assoc Prof Brian Darlow</td>
</tr>
<tr>
<td>Neonatal herpes simplex infection (&lt;15 years)</td>
<td>January 1998</td>
<td>Dr Dawn Elder</td>
</tr>
</tbody>
</table>

### Classification of Case reports

A valid report is one confirmed by the investigator as satisfying the diagnostic criteria set out in the case definition. Invalid reports can be either:
- Duplicate reports of cases already reported to the NZPSU, or
- Reporting errors arising from cases that have been reported but which:
  - Do not satisfy the diagnostic criteria, or
  - Are a result of misdiagnosis, or
  - The wrong box on the yellow card was ticked

An unknown report is one where insufficient follow-up information is available to the investigator or information has not been received by the NZPSU.

Table 4 shows the classification of cases reported to the NZPSU for conditions under surveillance in 1998.

### Table 4: Classification of notified cases for conditions under surveillance in 1998

<table>
<thead>
<tr>
<th>Conditions under surveillance</th>
<th>Total Reports</th>
<th>Valid reports n (%)</th>
<th>Invalid reports</th>
<th>Unknown n (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Acute Flaccid Paralysis (from Oct 1997)</td>
<td>10</td>
<td>8 (78)</td>
<td>1 (11)</td>
<td>1 (11)</td>
</tr>
<tr>
<td>Congenital Rubella Syndrome</td>
<td>1</td>
<td>1 (100)</td>
<td>0 (0)</td>
<td>0 (0)</td>
</tr>
</tbody>
</table>
Incidence can be estimated as the number of newly diagnosed cases in a defined period of time. It is expressed either per 100 000 live births or per 100 000 children in a particular age group. Population statistics for the denominator were obtained from the New Zealand Health Information Service.

Table 5 shows the number of cases reported in 1998 and the estimated incidence rate for each of the conditions under surveillance. No data on neonatal herpes simplex infection are reported, as these data are only available for the second half of 1998.

Table 5: Conditions under surveillance in 1998: the number and estimated incidence rates

<table>
<thead>
<tr>
<th>Condition (age range included)</th>
<th>Confirmed cases in 1998</th>
<th>Number</th>
<th>Incidence rate</th>
</tr>
</thead>
<tbody>
<tr>
<td>Acute flaccid paralysis (&lt;15 years)</td>
<td>6</td>
<td>0.7 per 100 000 under 15 years of age</td>
<td></td>
</tr>
<tr>
<td>Haemolytic uraemic syndrome (&lt;15 years)</td>
<td>14</td>
<td>1.7 per 100 000 under 15 years of age</td>
<td></td>
</tr>
<tr>
<td>Congenital rubella syndrome (&lt;15 years)</td>
<td>1</td>
<td>1.1 per 100 000 pregnant women</td>
<td></td>
</tr>
<tr>
<td>Perinatal HIV exposure (&lt;15 years)</td>
<td>4</td>
<td>7.0 per 100 000 births</td>
<td></td>
</tr>
<tr>
<td>Vitamin K deficiency bleeding (&lt;15 years)</td>
<td>2</td>
<td>3.5 per 100 000 births</td>
<td></td>
</tr>
<tr>
<td>Neonatal herpes simplex infection (&lt;15 years)</td>
<td>2</td>
<td>-</td>
<td></td>
</tr>
</tbody>
</table>

Notes: 1 The child diagnosed with congenital rubella syndrome in 1998 was born in 1989. Rates of CRS are generally presented per 100 000 pregnant women. As this child was diagnosed many years after birth such a rate is inappropriate.
2 Complete data could not be collected until the second half of 1998.
Discussion

The first year of operation of the NZPSU has shown that an active surveillance system of this type can achieve reasonably complete reporting of a range of uncommon paediatric conditions, and therefore it is a sensitive system for diagnosed cases of the conditions under surveillance.

The NZPSU surveillance system does have some limitations. First, the system can only be used for conditions that can reasonably be expected to come under the care of a paediatrician. For this reason, it is only suitable as a method of monitoring or investigating serious conditions. Second, as with any surveillance system, conditions that are under-diagnosed, for example CRS or maternal HIV infection, will be under-reported.

The third limitation is that the monthly reporting frequency is not adequate for diseases that require an immediate public health response. Three of the conditions monitored by the NZPSU are also notifiable to the local medical officer of health: AFP as suspect poliomyelitis, HUS as suspected VTEC infection, and CRS. This allows the medical officer of health to take urgent action to identify sources of infection and prevent further cases. Any suspect poliomyelitis case initially needs to be treated as a potential wild-type poliovirus infection, and assessed without delay to determine whether it is likely to be due to wild-type virus. VTEC infections need to be investigated to determine likely sources of infection and to identify common source outbreaks.

However, the active NZPSU surveillance system is more sensitive for AFP than the system of passive notifications to medical officers of health. Although practitioners have been advised for several years that all cases of AFP should be immediately notified to the medical officer of health, only two cases were notified in 1998 (Galloway Y. Personal communication, 1999), a third of the number reported by the NZPSU system. This current rate of notification of AFP cases to medical officers of health would not meet the WHO’s specified sensitivity for AFP surveillance.

The NZPSU system complements other forms of disease monitoring. Linking information on HUS provided by paediatricians with notifications of VTEC infection, allows the burden of serious disease due to this pathogen to be determined. The system provides an active method of assessing the effectiveness of disease prevention programmes, such as those to eliminate poliomyelitis, vitamin K deficiency bleeding, and CRS. Also, with time, as children perinatally infected with HIV become symptomatic, it will be possible to estimate the effectiveness of antenatal HIV detection.
The information reported during the first full year of the NZPSU’s operation predominantly provides a measure of the effectiveness of control programmes. The data that are now being collected on neonatal herpes infection, childhood diabetes, subdural haemorrhage, and fetal alcohol syndrome will provide national information on the epidemiology of these conditions that has not previously been available. Such increased understanding of these conditions is necessary to guide treatment and control programmes.

In the future we will remove some conditions from the list. Some, such as those that are providing surveillance of control programmes will probably remain, whereas others, such as those that are providing basic epidemiological information, will be included for only as long as is needed to obtain the necessary information.

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. Confining the system to conditions that are very rare will limit the demand on clinician’s time, but conversely it will be less likely to provide useful information. Feedback both to and from clinicians is vital, as their perception of the usefulness of the scheme will influence its functioning.
Principal Investigators

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Dr Paul Shillito, Paediatric Neurologist,  
Department of Paediatrics,  
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Background

There are many causes of acute flaccid paralysis (AFP) in children including trauma, the Guillain-Barre syndrome, transverse myelitis and poliomyelitis. As part of the programme for the worldwide eradication of polio it is important for New Zealand to be registered by the World Health Organization (WHO) as being free from wild polio. There were only 4 notifications of poliomyelitis in the 30-year period 1965-1996. To achieve WHO registration however it is necessary that all children with AFP are adequately investigated for polio virus infection, even when the clinician does not believe that polio is the cause. The necessary investigation includes the testing of two stool samples taken within two weeks of the onset of paralysis by a WHO-accredited laboratory.

The NZPSU was established under contract from the Ministry of Health at the end of 1997 to undertake active surveillance of AFP, which started in October 1997.

Paediatricians are requested to make the initial notification of AFP by telephone to the NZPSU. The Unit then informs the Virus Laboratory at ESR who dispatch stool specimens containers to the paediatrician who return them to ESR for testing.

The effectiveness of the surveillance process is determined by the recording of an incidence of AFP (not caused by polio) of at least 1 per 100,000 children less than 15 years with at least 80% being investigated appropriately.

Objective of study

To determine:
1. the incidence of acute flaccid paralysis (AFP) in children in New Zealand
2. whether any cases of AFP in New Zealand are causes by polio
Case Definition

Any child less than 15 years old with acute onset of flaccid paralysis in one or more limbs, or acute onset of bulbar paralysis.

Telephone notification is requested to ensure that appropriate faecal samples are collected for viral testing.

Results

Surveillance for AFP has been undertaken since October 1997. This report covers the period 1 October 1997 to 31 December 1998.

During this 15-month period the NZPSU received information of 8 children with AFP. The annual incidence rate for children less than 15 was 0.74 per 100,000 (95% confidence interval 0.32-1.5 per 100,000)

Of these 8 children:
- 4 were boys and 4 girls.
- One was aged less than one, 4 were aged between 1 and 4, one was between 5 and 9 years, and 2 were between 10 and 14 years.
- Two were from the Northern health region, 5 from the Midland health region and one from the Central health region.
- Three children were European, 2 Maori, one each of mixed Maori/European and Maori/Pacific island ethnicity and one an African immigrant.

Six of the cases of paralysis were due to Guillain-Barre syndrome, one was related to trauma and one possibly due to oral polio vaccine\(^1\).

Faecal samples were tested at ESR for evidence of wild polio for 6 of the 8 children; one of the children who was not tested was the child whose paralysis was due to trauma. However only 3 of the 6 children whose stools were tested had the specimens collected within two weeks of the onset of the paralysis, the period required by WHO.

Discussion

Ascertainment of children hospitalised with AFP is likely to be complete because of the involvement of all paediatricians in general paediatric practice and high return rate. It is possible that some young people under 15 years might be admitted under adult clinicians and thus not notified. However in a review of hospital discharges between August 1997 and
September 1998 all children less than 15 years identified with GBS from that source had been reported to the NZPSU.

It has been requested that all cases of AFP be notified to the local Medical Officer of Health as suspect polio. However only two cases were reported in this way in 1998. Thus the active surveillance by the NZPSU is more sensitive in detecting cases than passive notification.

The WHO requires a country’s surveillance system to identify one case per year of non-polio AFP per 100,000 children less than 15. The 95% confidence limits around the annualized rate of notified AFP included this. Therefore the system appears to have identified an appropriate rate of AFP.

A WHO requirement that 80% of cases meet the criteria for being appropriately investigated for poliovirus was not met. This was either because they were not collected or were collected too late. This situation has continued into 1999. To improve the situation all pediatricians have been reminded to make notification by phone to the NZPSU (rather than ESR). NZPSU will arrange for the dispatch of the appropriate specimen containers.

The findings so far confirm the clinical impression that New Zealand is free of polio, however it is necessary to continue adequate surveillance to meet the WHO requirements.

One child was believed to suffer from vaccine associated paralytic polio. The case might kindle a debate of the use of oral or injectable polio vaccine, the latter now being recommended in the US.

### Practice Points

- **Surveillance of all cases AFP is needed to confirm the absence of polio in New Zealand**

- **The proportion of cases of AFP that have appropriate stool investigations for polio virus is lower that required by WHO**

- **Telephone notification to the NZPSU (03 474 7825) must be made as soon as a child is diagnosed with AFP. NZPSU will dispatch containers for stool samples which should be returned to ESR for virological testing**

- **Guillain-Barre syndrome (GBS) is the commonest cause of AFP among children**

Congenital Rubella Syndrome 1998

Principle Investigator

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Otahuhu
Auckland

Background

Congenital rubella (CRS) should be preventable by the current New Zealand infant immunisation programme with adequate vaccination levels. However these are not achieved in many areas. In addition pregnant women may have grown up in developing countries eg: the Pacific nations where rubella is not a routine childhood vaccine because of cost. As most infants and children with CRS are seen at some stage by a paediatrician the NZPSU is a useful mechanism to obtain more information.

Details on diagnosis and vaccine status could lead to a targeted approach to those at greatest risk.

Objectives

1. To more accurately define the present incidence of congenital rubella syndrome in New Zealand.
2. To evaluate the reasons why mothers of children with congenital rubella have not effectively vaccinated.
3. To monitor the outcome of the rubella vaccination programme.

Case Definition

Any child or adolescent up to 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.

Diagnosis of congenital rubella is confirmed by:

- the detection of specific IgM antibodies in a serum during the first months of life
the persistence of rubella-specific IgG antibodies in a child aged 6 to 12 months, or in a child up to 2 years who has not been vaccinated
isolation of the virus which may be shed from the throat and urine for as long as a year

Results

There was one report of CRS in 1998 received on the monthly yellow cards. A completed questionnaire was subsequently received. The patient was born in 1989. The child's mother arrived in New Zealand two years before the pregnancy.

Discussion

CRS notification in New Zealand from 1982 to 1995 suggest 0-1 cases per year (i.e.: ~2/100,000 births). In the pre-vaccine era rates were ~10 times higher (eg: UK 46/100,000 births; US 40-80/100,000 births; NZ ~50/100,000 live births). US CRS rates dropped to a very low <0.1/100,000 pregnancies in the late 1980s but these rose to ~0.5/100,000 live births in 1990.

From 1982-1995 in New Zealand, 16 CRS cases were reported through the notification system (~2/100,000 live births). This suggests our rates are importantly higher and more data on such cases may facilitate intervention(s).

The current 2 dose strategy of MMR is likely to be the most effective.

Practice Points

- Women on child bearing years born outside New Zealand should be screened for rubella antibody at the first health encounter and vaccinated if appropriate
- CRS should continue to be a diagnosis searched for. It is likely there are currently undiagnosed and/or unreported cases
- Rubella virus continues to circulate as documented by virus laboratories

References

i Immunisation Handbook 1996. Ministry of Health
ii Plotkin and Mortimer. Vaccines 2nd ed 1994
Perinatal Exposure to HIV 1998

Principal Investigators

Dr Nigel Dickson, Senior Research Fellow
NZPSU & AIDS Epidemiology Group,
Departments of Paediatrics and Child Health & Preventive and Social Medicine,
University of Otago Medical School, Dunedin

Dr Lesley Voss, Infectious Disease Specialist
Starship Children's Health, Auckland

Background

Information on all infants exposed to HIV infection in utero in New Zealand has been sought through the NZPSU network of paediatricians since the start of 1998. This will complement data on children infected with HIV and on those who have progressed to AIDS that is currently collected by the AIDS Epidemiology Group.

The risk of perinatal transmission of HIV from an infected mother to her infant can be significantly reduced by the appropriate use of anti-retroviral drugs and avoidance of breast feeding. However there has been a major concern in New Zealand that women are not being diagnosed with HIV infection at a time that would allow their infants to benefit from these strategies. There have been young children born in New Zealand in recent years with perinatally acquired HIV whose mothers were not diagnosed until after the pregnancies in spite of having known epidemiological risk factors. Although the Ministry of Health has issued guidelines that all pregnant women should have their risk factors for HIV infection assessed, a survey of providers of maternity care in Dunedin found this was not normal practice.

The information collected in this study will, in time, be of use in auditing the diagnosis of HIV among pregnant women and the implementation of preventive strategies.

Objective of study

To determine the extent and outcome of recognised perinatal exposure to HIV infection in New Zealand.
Case Definition

Any infant or child, whether infected or not, newly recognised as being born to an HIV infected woman.

Results

There were 8 reports of perinatal HIV exposure received on the monthly yellow cards. Completed questionnaires were subsequently received for all these cases, which included 2 duplicate reports. Of the 6 children reported, 2 were born prior to 1998.

Thus there were known to be 4 pregnancies to HIV-infected women in New Zealand during 1998.

The rate of identified HIV exposure in 1998 was 4/55,674 = 0.7 per 10,000 pregnancies, 95% confidence interval 0.2-1.8 per 10,000.

Of these 4 pregnancies:
- 3 ended in live births, and one in a late fetal death.
- 3 occurred in the Northern health region and one in the Midland region.
- one of the infected women was Maori, and the other 3 were from parts of the world where heterosexual transmission of HIV is common.
- all of the women were diagnosed before or during their pregnancy
- 3 of the women received anti-retroviral treatment during pregnancy, but this was refused by one family
- none of the 3 live born infants was breast fed
- none of the 3 live born infants have been diagnosed with HIV infection, although longer term follow up is required to be absolutely certain.

Two reports were received of children diagnosed with HIV infection in 1998 who were born in earlier years. Both of these children were over 2 years old at the time of diagnosis. One had been born in New Zealand and the child’s mother, who had epidemiological risks for HIV infection, was not diagnosed as infected during pregnancy.

Discussion

It seems likely that information on all known cases of HIV during pregnancy has been obtained for three reasons. Firstly, all practicing paediatricians are included in the network; secondly, a high response rate has been obtained; and lastly paediatricians would be expected to know about all diagnosed HIV-infected pregnant women either through being consulted during pregnancy or through providing specialist follow up for HIV exposed infants. In addition surveillance of newly diagnosed HIV infection has not so far revealed any such infant born in 1998.
However the proportion of HIV-infected pregnant women diagnosed as such cannot be assumed to be 100%, or even to be high. In London, where routine antenatal testing has been recommended since 1992, less than 30% of HIV infected pregnant women had their infection diagnosed before the birth of their child in 1997. Indeed one perinatally infected child born in New Zealand was diagnosed in 1998 when over 2 years old. The proportion of HIV-infected pregnant women whose infection is diagnosed in this country is not known, and cannot be calculated from data collected in this study, as prevalence studies on HIV infection among pregnant women have not been undertaken. There are legal barriers to the practical implementation of unlinked anonymous prevalence studies using neonatal blood taken for metabolic screening, which is the simplest method of determining this and used in several other countries.

With time most children infected with HIV are likely to become symptomatic and it will be possible to estimate the proportion of HIV infected women diagnosed, however this will be too delayed to be a useful audit of current practice.

That 3 of the 4 pregnant women were from parts of the world where heterosexual transmission of HIV is common is consistent with the epidemiology of diagnosed HIV infection among women in New Zealand. Two of these women were from sub-Saharan Africa and one from South East Asia. The remaining woman was Maori. Although no difference has been found in the rates of HIV infection or AIDS among the main ethnic groups in New Zealand, there is concern that HIV testing may be less frequent among Maori and Pacific Island people, and that, if established, HIV may spread more in these groups.

It is encouraging that all pregnant women were offered appropriate preventive treatment, but a concern that one family refused this. It is also pleasing that none of the surviving children have been shown to be infected with HIV.

**Practice Points**

- The Ministry of Health recommends that asking about HIV risk should be a routine part of antenatal care
- The risk of perinatal transmission of HIV can be markedly reduced by anti-retroviral therapy started before delivery and other strategies
- Most, but not all, HIV infected women are from, or have had contact with people from parts of the world where heterosexual transmission of HIV is relatively common
• It is possible that not all HIV infected mothers will have been identified so paediatricians should be aware of the possibility of HIV infection in children they see

• *AIDS – New Zealand*, published quarterly and available from the AIDS Epidemiology Group (Fax 03 479 7298) provides information on HIV/AIDS in New Zealand.

References


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Vitamin K Deficiency Bleeding 1998
(includes Haemorrhagic Disease of the Newborn)

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Background
Haemorrhagic Disease of the Newborn (HDN) is a rare but potentially fatal disorder which presents with spontaneous bleeding, bruising or intracranial haemorrhage. Three types of HDN are recognised:
- Early - usually on the first day of life and related to maternal drug therapy such as anticonvulsants
- Classical - from the 2nd to 7th day of life
- Late - occurring between one week and six months of age, almost exclusively in breast fed infants and often associated with unrecognised liver disease

Although records are incomplete, Vitamin K prophylaxis was probably given by intramuscular injection at birth to most New Zealand infants at the start of this decade. However, in 1992 after a British study reported an association between intramuscular vitamin K and an increased risk of childhood cancer at 10 years of age, the Fetus and Newborn Committee of the Paediatric Society of New Zealand recommended oral vitamin K, in repeat doses, as routine prophylaxis in other than high risk situations. This recommendation was similar to that in many other countries including Australia, the UK and much of Europe but, in part because there was no preparation of vitamin K intended for oral use available in New Zealand (the Fetus and Newborn Committee recommended use of the intramuscular preparation), the Department of Health continued to recommend intramuscular prophylaxis.

Oral administration is complicated by the need for repeat doses, with New Zealand doses being recommended at birth, at five days of age (at the time of the Guthrie test), and at 6 weeks (with the first immunisation), and compliance with this regime has been shown to be poor. In addition, there is evidence that full compliance with the oral programme may not be as efficacious as intramuscular prophylaxis in preventing late haemorrhagic disease. For these reasons, and because epidemiological evidence from both North America and Europe failed to confirm a link between intramuscular
vitamin K and an increased risk of childhood cancer, the Fetus and Newborn Committee produced revised recommendations in favour of intramuscular injection as the preferred route for prophylaxis in 1995\textsuperscript{vii}. Although expert commentaries endorsed this approach\textsuperscript{viii} more recent evidence will have again raised concerns about intramuscular prophylaxis in the minds of some\textsuperscript{ix}.

Because of these uncertainties several countries, including the UK and Australia, have felt it important to have reporting systems in place to monitor the incidence of HDN.

**Objective of study**

To provide a mechanism for the national epidemiological surveillance of HDN and to determine the morbidity and mortality associated with HDN.

**Case definition**

Any infant under 6 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.

**Results**

Four cases were reported that met the definition, but one case was double reported leaving three cases, one possible and two confirmed, in twelve months.

All three cases were born in hospital, one in the Midland Region and two in the Southern Region.

The possible case was early onset, being manifested as pulmonary haemorrhage at birth following a vaginal delivery at 36 weeks gestation. The mother was not taking any anticonvulsant or other medication. Intramuscular vitamin K was given at one hour of age. The initial coagulation profile was performed 3 hours later and could be compatible with vitamin K deficiency. Subsequent investigations did not reveal any cause for the coagulopathy. The infant has been normal at follow-up.

The other two cases were both late onset, at 30 and 37 days, in infants who were exclusively breast-fed. Neither infant was given any vitamin K prophylaxis at birth. In both cases the coagulation profile was compatible with HDN at presentation and normalised following treatment with vitamin K.
One infant was 34 weeks gestation, born via a caesarean section and presented with skin bruising and an intracranial haemorrhage. There was no liver disease. Subsequent development has been normal.

The second infant was born at 39 weeks gestation, via a vaginal delivery and presented with skin bruising. This infant proved to have liver disease and a peroxisomal disorder.

**Discussion**

In the absence of a national perinatal database the proportion of infants receiving vitamin K prophylaxis and by what route remains unknown. However, all main centres have policies in line with the latest Fetus and Newborn Committee recommendations\(^7\), namely that the preferred route is intramuscular injection. It is not possible to come to any conclusions about the effectiveness of vitamin K prophylaxis from this study, although data on the incidence of HDN in the absence of prophylaxis would suggest that the majority of New Zealand infants are receiving vitamin K.

Many infants are cared for by midwives for the first 6 weeks of life and neither this group nor general practitioners are circulated with reporting cards. Hence it is quite possible that some, probably milder, cases have gone unreported. It seems unlikely that significant bleeding would not lead to admission to hospital and recognition of HDN.

The incidence of late onset HDN is 3.6 per 100,000 (95% confidence interval 1.1-13.0) but would be half this figure if congenital liver disease is excluded.

Both reported cases of late onset disease occurred in infants not given any prophylaxis. In neither case is it clear whether this was the result of an informed decision by the parents. Lead maternity carers must be aware of their duty to inform prospective parents of the issues relating to HDN and the current recommendations for prophylaxis, before the birth when possible.

In common with surveillance in the UK over two periods of time\(^x,x^i\), no cases of HDN were reported following intramuscular vitamin K. However, equally no cases were seen following oral prophylaxis. The number of births in New Zealand is relatively few and it is essential that there is adequate vigilance and reporting of cases that meet the definition for a further year.

**Practice Points**

- Lead maternity carers should inform prospective parents of the issues relating to vitamin K deficiency bleeding and of the recommendations for prophylaxis. The current recommendations are that all newborns be given vitamin K by a single intramuscular
injection at birth. Alternatively, vitamin K may be administered orally, but multiple doses are required.

- Cases of late-onset haemorrhage may occur, particularly among breast-fed infants, if vitamin K prophylaxis is not given. Infants with late onset disease should be investigated for liver disease.

References.

Introduction

In New Zealand, and other developed countries haemolytic uraemic syndrome (HUS) is the single commonest cause of acute renal failure in children needing dialysis. The majority of the cases are associated with infection with verotoxin producing *Escherichia coli* (VTEC) serotype 0157 H7. This most commonly occurs among children age 1 to 5 years. VTEC produces a diarrhoeal illness, which commonly results in haemorrhagic colitis. Approximately 10% of people infected develop HUS. Between 3-5 days after the onset of symptoms, these people become pale and lethargic due to the of haemolytic anaemia, and develop thrombocytopenia and acute renal failure. Twelve to 30% will have severe sequelae and 3-5% will die, mainly from central nervous system or cardiac complications.

Within New Zealand the majority of cases of HUS in recent years have occurred in the upper half of the North Island. This corresponds to reported cases of VTEC infections. VTEC is found in the gastro-intestinal tracts of many animals and birds although cattle are thought to be the predominant reservoir of *E. coli* 0157 in North America and Europe. The first New Zealand case of VTEC, an 11-month-old boy from Whakatane, was identified in October of 1993. In a recent study, *E coli* 0157 was detected in only 2 of 371 dairy cow faecal samples in the Waikato.

Since January 1998, paediatricians have been asked to report cases of HUS to the NZPSU. A questionnaire devised by the Australian Paediatric Surveillance Unit was adapted for local use. Clinical data, microbiological information, and initial outcomes were requested

Purpose of the study

To determine the incidence, demographic characteristics, microbiological factors and initial outcome of children with HUS.
Case definition

Any child under the age of 16 years with HUS defined as:
- Microangiopathic haemolytic anaemia
- Thrombocytopenia, and
- Acute renal impairment

Results

There were 14 children (4 boys and 10 girls) with HUS reported to the NZPSU in 1998. Data on hospital discharges collected by the Zealand Health Information Service suggested one further case. The mean age at presentation was 3.6 years, and most (11/14) were under 5. The overall childhood (<15 years) incidence rate for 1998 was 1.6 per 100,000 with a rate of 4.0 per 100,000 among children aged 1–4 years (Table). Four of the children were from the Northern, 6 from the Midland, 3 from the Central and one from the Southern Health Regions.

<table>
<thead>
<tr>
<th>Number</th>
<th>Popn. (at end of 1997)</th>
<th>Rate per 100,000</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;1yr</td>
<td>2</td>
<td>57,470</td>
</tr>
<tr>
<td>1-4yr</td>
<td>9</td>
<td>226,600</td>
</tr>
<tr>
<td>5-9yr</td>
<td>1</td>
<td>298,880</td>
</tr>
<tr>
<td>10-14yr</td>
<td>2</td>
<td>275,670</td>
</tr>
<tr>
<td>Total</td>
<td>14</td>
<td>860,617</td>
</tr>
</tbody>
</table>

The mean period from onset of symptoms to diagnosis was 5 days (range 3-21).

All of the 14 children had anaemia and bloody diarrhoea, and all but one vomiting. Seven of the 14 (50%) children needed acute peritoneal dialysis for a mean duration of 10.2 days (range 4-28 days). Half required treatment of acute hypertension. There was one death in a 21 month old child who presented with seizures and subsequently died of a massive intra-cerebral haemorrhage. All children who required dialysis recovered renal function and at last follow up, none had evidence of chronic renal insufficiency at last follow up.
Microbiological results

_E. coli_ 0157 H7 was isolated in 7 of 14 children with HUS. All 0157 isolates produced Stx-2 toxin. There were 3 isolates of _E. coli_ 0113. _Campylobacter_ was isolated from one child.

Although there was no obvious source of VTEC in any of the cases reported to the NZPSU, there was a history of living or having visited a farm in 4 children in the few weeks before the development of HUS.

Discussion

The 14 children reported with HUS in 1998 was considerably larger than the 5 or 6 hospitalised in previous years. There has been a concomitant rise in diagnosed VTEC infections over the past few years increased awareness and improved methods of detection of the VTEC are possible reasons for the latter rise. This might have affected the rate of recognition of HUS with clinicians being alerted to its possibility because of the increased rate of isolation of VTEC. However it is probably that been a true rise in incidence as serious anaemia and renal impairment are unlikely to been missed in unwell children with gastrointestinal symptoms.

HUS was most common in children less than 5, and as in previous years almost all of the cases lived in the North Island. There were no recognised outbreaks.

There was an appreciable delay between the onset of symptoms and diagnosis in some children with HUS. The diagnosis must be considered in all children presenting with bloody diarrhoea who become progressively unwell. Acute renal failure develops rapidly and coincides with the onset of anaemia. It is important that family doctors and paediatricians are aware of the signs and symptoms of disease to enable early supportive treatment to be initiated. This is especially important in infants where the diagnosis of oliguria is more problematic particularly in the presence of diarrhoea.

Dialysis and other supportive treatments is the mainstay of therapy. Morbidity from HUS is high with 12-30% developing severe sequelae\textsuperscript{2}. Mortality from HUS is 5-10% and is due mainly to central nervous system, cardiovascular complications and bowel involvement.\textsuperscript{vii,viii} During 1998, one child died at from a massive intracerebral haemorrhage.

Antibiotics are not recommended in the treatment of VTEC as they do not appear to alter the duration of symptoms or the risk of progression to HUS and may increase the severity of sequelae by increasing the amount of shiga toxin released.\textsuperscript{ix}

_E. coli_ serotype 0157 is the most common pathogen causing HUS, and was identified in half of the children in this series. Other organisms implicated include other serotypes of _E. Coli_ (3 of these children affected had _E. Coli_ 0113 isolated), _campylobacter, shigella dysenteriae, yersina_ and a number of viruses. One child had _campylobacter_ isolated.
This is a source of potential concern because *campylobacter* infections in New Zealand are presently at an all time high with a rate of 317 cases per 100,000 total population, at least three times higher than other comparable countries.\textsuperscript{x}

Public health surveillance and control of VTEC is important because major outbreaks overseas have been due to contaminated food.\textsuperscript{xi}

Controlling VTEC infection depends on prompt identification of cases, prevention of secondary transmission and the swift recognition and control of common source outbreaks.\textsuperscript{xii} Clinicians should notify the local Medical Officer of Health of all suspect and confirmed cases of VTEC, including cases of HUS, and the public health departments will undertake further appropriate investigation and action.

The ongoing integration surveillance of VTEC infection and of clinical cases of HUS will provide a critical understanding of the spread of this infection and the impact it has on the health of children.

### Practice Points

- The increasing incidence of *E. coli* 0157 (and *campylobacter*) should alert health professional to the risk of HUS, particularly in young children
- Early culture of stools from children with bloody diarrhoea is necessary to identify the organism responsible for HUS
- Isolates of suspected or proven VTEC, and cases of HUS must be notified to the Medical Officer of Health for appropriate public health investigation and action
- Prevention of *E coli* 0157 infections requires high standards of public health surveillance and control in all stages of the food chain

### Reference


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