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Preface

The 4th Annual Report of the New Zealand Paediatric Surveillance Unit for 2001 contains an increasing amount of important information.

Surveillance of acute flaccid paralysis (AFP) has been undertaken since October 1997 for the Ministry of Health's National Certification Committee for the Eradication of Poliomyelitis (NCCEP). It has also been undertaken for other conditions introduced since that time, most of which have been for paediatricians with a particular research interest.

Some key findings in this report for 2001 are:

Acute Flaccid Paralysis (AFP)

- 11 cases of AFP were reported in 2001 (1.29 per 100,000).
  - The system successfully captured the rate of AFP expected by WHO.
- All 11 AFP cases have been discarded by the NCCEP as non-Polio.
- Even though WHO believes polio 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.

Bronchiectasis

- 24 cases of Bronchiectasis were reported in 2001.
- CT scans of 88% of these have been received and confirmed as Bronchiectasis.

Haemolytic Uraemic Syndrome (HUS)

- There were 6 cases of HUS notified in 2001.
- The North Island predominance of the cases has remained unchanged from previous years.
- Unlike what has been found in some countries, there continues to be no seasonal pattern in New Zealand.

Kawasaki Disease

- 29 cases of Kawasaki disease were reported in 2001.
- M:F ratio 2.6:1, the majority of which (86.2%) were under 5 years of age.
- Diagnosis and treatment is being made promptly.

Perinatal HIV Exposure

- 7 infants were born in New Zealand to women known during the pregnancy to be infected with HIV, none of these infants were known to be infected at one year follow-up.

Subdural Haemorrhage

- There were 26 cases of Subdural Haemorrhage in children under 2 years in 2001.

- Subdural haemorrhage was due to birth injury in 6 cases, accidental injury in 2, child abuse in 12 and suspected child abuse in 5 cases. One case was due to Vitamin K deficiency bleeding.

Vitamin K Deficiency Bleeding (VKDB)

- There were two cases of VKDB in 2001.

Retinopathy of Prematurity (Stage III and over): Final report

- There were at total of 34 notifications of ROP in 1999 and 2000 of which information has been received on 27.
- Of these 27 reports there were 25 valid cases.
- 14 infants have received treatment, 9 with laser therapy, 2 with cryotherapy only (to one eye only in one infant), and 3 infants were treated with a combination of laser and cryotherapy.
- The longer term outcome is unknown at this time but one infant is bilaterally blind and at least 3 other infants have significant cicatricial disease.

Fetal Alcohol Syndrome (FAS): Final report

- There were a total of 72 notifications of suspected or definite FAS during the 2.5 year study period (July 1999 to December 2001).
- Of these 72, 62 were valid reports, giving an incidence of 2.9 per 100,000 children <15 years of age per year.
- Average age of diagnosis was 5.5 years.
- The majority of affected children are in foster or extended family care.
- There is an increased awareness of the behavioural and learning difficulties as manifestations of FAS.

The ongoing success of 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.

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

Barry Taylor Nigel Dickson Melissa Carter

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. 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 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 (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). 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 ten rare childhood conditions.

The NZPSU is a member of the International Network of Paediatric Surveillance Units (INoPSU). Further information can be viewed on our website at: http://www.paediatrics.org.nz/nzpsu/nzpsu1.html

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 the World Health Organisation.

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

How the Surveillance System Works

Specialist 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 a case of any of the conditions under surveillance. However, cases of AFP are also required to be reported immediately by telephone to the NZPSU. When a case of any of the conditions are 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 2 or 3 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 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.

There were ten conditions under surveillance in 2001, one of which was removed at the end of the year and another was added.

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
Dr 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 2001

In 2001 there were 179 clinicians who participated in the system. The average response rate to the monthly report card/email was 95%, with no consistent set of non-responders. Table 2 shows the response rate per area.

Table 2: Response rate per health locality (as defined by DHB) 2000 & 2001

Health Locality	2000	2001
	%	%
Northland, Auckland, Waitemata, Counties Manukau	93	91
Waikato, Bay of Plenty, Taranaki, Lakes	97	94
Capital and Coast, Hutt, Wairarapa, Whanganui, Tairawhiti, Hawkes Bay, MidCentral	91	94
Nelson Marlbourgh, Canterbury, South Canterbury, West Coast	94	96
Otago, Southland	97	99
TOTAL	94.4	94.8

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 need to be kept under review.

Figure 1 shows the percentage of clinicians on the mailing list that reported cases during 2000 and 2001. The figure shows that in 2001 over half (53.9%) of the participants did not report any cases, with 40% reporting between one and three cases. The proportion reporting 4 or more cases has halved since 2000, due to the removal of the relatively common diabetes mellitus from the card.

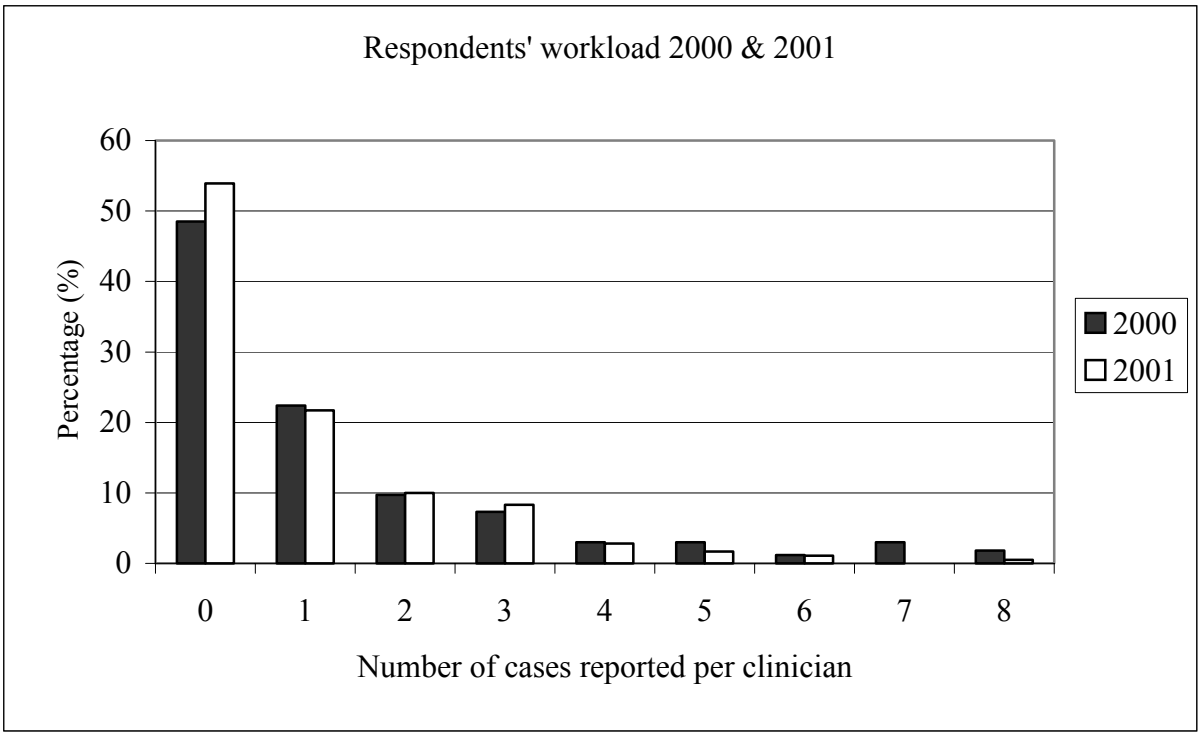

Figure 1: The percentage of clinicians that reported cases during 2000 and 2001

In 2001, NZPSU monitored 10 uncommon childhood conditions (Table 3). Some of the protocols and questionnaires used were adapted from those used by the Australian Paediatric Surveillance Unit.

Table 3: Conditions under surveillance in 2001

Condition (age range included)	Surveillance started	Principal Investigator(s)
Acute flaccid paralysis (<15 years)	October 1997	Dr Nigel Dickson, Dr Paul Shillito
Haemolytic uraemic syndrome (<15 years)	January 1998	Dr William Wong
Congenital rubella syndrome (<15 years)	January 1998	Professor Diana Lennon
Perinatal HIV exposure (<15 years)	January 1998	Dr Nigel Dickson, Dr Lesley Voss
Vitamin K deficiency bleeding (<15 years)	January 1998	Professor Brian Darlow
Subdural haemorrhage (<2 years)	January 1999	Dr Patrick Kelly
Fetal Alcohol Syndrome (<15 years)	July 1999	Dr Alison Leversha
Kawasaki Disease (< 15 years)	January 2001	Dr Paul Heaton
Bronchiectasis (<15 years)	January 2001	Dr Cass Byrnes
Idiopathic Nephrotic Syndrome (<15 years)	July 2001	Dr William Wong

Brief Reports on Selected Conditions

We have not provided a report for Congenital Rubella as there were no cases reported in 2001, or Idiopathic Nephrotic Syndrome as this study was started half way through the year. The 2001 findings for the latter study will be reported in the 2002 Annual Report.

Acute Flaccid Paralysis

Dr Nigel Dickson

Ongoing study started in October 1997

Introduction

To confirm the absence of poliomyelitis the World Health Organization (WHO) requires a surveillance system to be in place:

1. That captures an annual incidence of Acute Flaccid Paralysis (AFP), not due to polio, of at least 1 per 100,000 children <15 years.
2. In which 80% of cases of AFP have 2 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 2001

- There were 14 notifications in 2001, of which, one was a duplicate and 2 did not fit the criteria for AFP.
- Information has been obtained on all but one of these children including follow up information two months after diagnosis.
- 10 cases were from the North Island and 1 from the South Island.
- 8 males, 2 females and 1 unknown. (Ratio 4:1 M:F).
- Age range 2 to 11 years, median age 4.5 years.
- No seasonal variation.
- The overall incidence was 1.29 per 100,000 children <15 years.
- A diagnosis of Guillain-Barre Syndrome (GBS) was made in 8 of these cases, transverse myelitis in one, and in the remaining 2 the diagnosis was unclear, however both children recovered fully.
- All 11 cases have been rejected as polio by the National Certification Committee for the Eradication of Polio (NCCEP).
- However, analysis of stool samples satisfying the WHO criteria was complete for only 6 of the 11 (55%) children (see Table 4).

An audit of all children discharged from hospital with a discharge diagnosis consistent with AFP in 2000 and 2001 has been performed, and a total of 9 possible cases were found that had not been reported to NZPSU. Of these, 7 were not cases of AFP but a result of miscoding, and 2 were actual missed cases of GBS.

Table 4: Percentage of AFP cases with adequate stool samples (or otherwise)

Category	Stool samples	
	n	%
2 stool samples within 14 days of onset of paralysis	6	54.5
2 stool samples, but one or both not within 14 days of onset of paralysis	2	18.2
1 stool sample	1	9.1
No stool sample*	2	18.2

*One of these children had transitory upper motor neurone disturbance, and we have not received any information on the other.

Comment

The system successfully captured the required rate of AFP, however, the rate of stool testing (55%) is not yet meeting the WHO criteria (80%). The NZPSU continue to remind clinicians of the need to make telephone notifications of AFP to ensure that timely stool specimens are sent to ESR for appropriate testing.

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.

Bronchiectasis

Dr Cass Byrnes, Dr Jacob Twiss

First year of a two-year study

In 2001 there were 28 notifications of Bronchiectasis, of which, 24 were valid cases. CT scans were received for 88% of these and all were confirmed as Bronchiectasis. Questionnaires are being sent out 1 year after the notification, of which 89% have so far been returned. There have been some significant delays between the diagnosis being made by CT scan and notification which has meant that some children notified in 2002 were actually diagnosed in 2001. As the date of diagnosis is generally more meaningful than date of notification the data have been presented in this way.

Diagnoses made in 2001		
Received:	42	
Duplicates	2	
Errors	1	
Exclusions	1	
Valid	38	
CT scans received for these notifications		35 (92%)
CT scans confirmation of bronchiectasis		35 (92%)
CT scans outstanding		3 (8%)
Questionnaires (1yr after diagnosis)		
Due	24	
Requested	24	(100%)
Received	20	(83%)
Outstanding	04	(17%)

Given that the questionnaires are not requested until one year after notification, most of those that are outstanding are not long overdue.

Demographic data, aetiological data, investigative data and CT scan findings are being collected and will be reported at the completion of the study.

Haemolytic Uraemic Syndrome

Dr William Wong

Ongoing study started in January 1998

In 2001 there were 7 notifications of Haemolytic Uraemic Syndrome (HUS) in children under 15 years of age, of which one was a duplicate. There were therefore 6 confirmed cases.

The annual incidence of HUS during 2001 has remained stable. The geographic features of the cases remain unchanged with the majority reported from the North Island. There was one patient from the Christchurch region, the second in the series so far. There continues to be no seasonal pattern to the cases. The mean age at presentation was 2.0 years. The median interval from onset of symptoms to diagnosis was 4.5 days (range 2-7). The ease of transmissibility of the infection is highlighted by the occurrence of two cases of HUS in the same family.

While the number of children reported to have *E.coli* associated shiga toxin infections continued to rise over the past 12 months (Fig 2), the number with diagnosed HUS has remained stable.

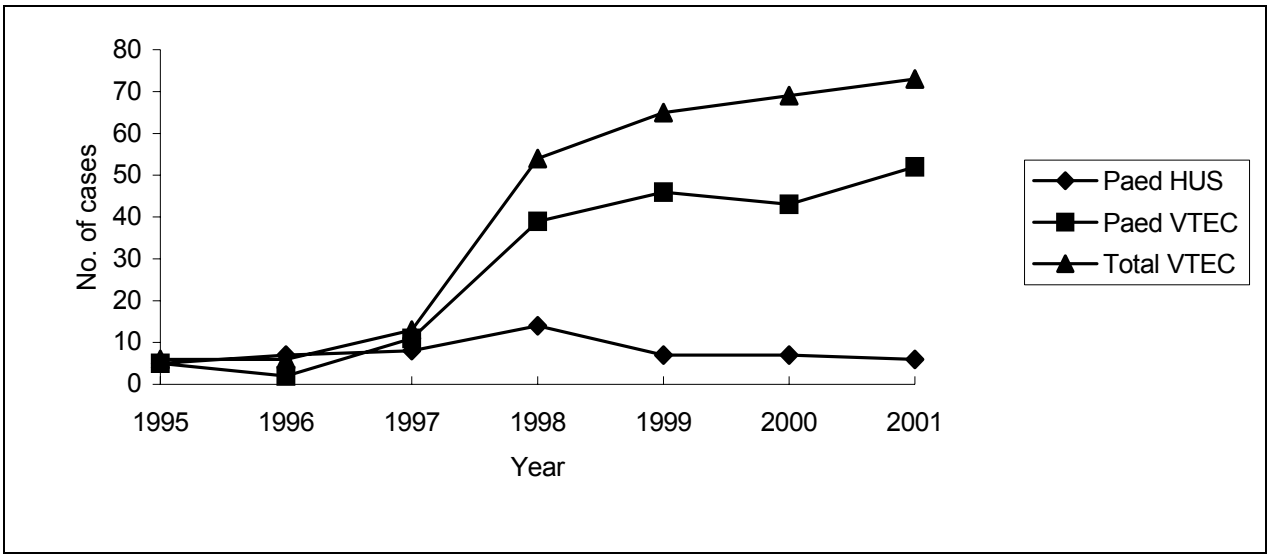

Figure 2: The number of cases of HUS and VTEC infections

Notes: 1. The figures for 1995 to 1998 are Starship admissions for HUS.
2. The "VTEC rates" are from the ESR laboratories.
3. VTEC is also known as STEC (Shigella-like toxin associated *E.coli*)

Kawasaki Disease

Dr Paul Heaton

First year of a two-year study

There were 36 notifications in 2001, the first year of this two-year study. Completed questionnaires were returned in 35 cases. In 26 the diagnosis of Kawasaki Disease (K.D.) was considered to be definite, in two probable, and in one case possible. There were three duplicate notifications and in three cases K.D. was thought not to be present.

Of the 29 new cases:

- 21 were male, 8 female (M:F 2.6:1). Age range was 3 months to 8 years and 2 months. 25 (86.2%) were aged less than 5 years and 7 (24.1%) less than 1 year.
- 20 cases (68.9%) were resident in the North Island and 9 (31%) in the South Island.
- In 12 cases (41%) the onset of illness was in Dec-Feb, 9 (31%) Mar-May, 5 (17%) Jun-Aug and 3 (10%) Sep-Nov.
- Of 25 cases where ethnicity was documented, 12 (48%) were NZ European/European, 9 (36%) were Polynesian (5 Tongan or Samoan, 4 Maori or part-Maori) and 4 (16%) were Oriental (2 Korean, 1 Chinese and 1 Japanese).
- K.D. was diagnosed at a mean of 7.2 days (Range 3-38) from the onset of illness, and a mean of 2.0 days (Range 1-12) from the day of admission.
- Only 1 case (pericardial effusion) had abnormal cardiac symptoms or signs at presentation.
- All cases had echocardiographic studies of which 17 (58.6%) were normal and 12 (41.4%) showed abnormalities. Of the latter, 8 had ectasia or mildly dilated coronary arteries, 2 had pericardial effusions, 2 had valvular regurgitation and there was 1 atrial septal defect.
- 27 patients were treated with IVIG (usually 2g/Kg) and aspirin. This was started on average 1.9 days after admission, which was 6.2 days after the onset of illness. 21 patients were prescribed aspirin on discharge.
- One child died aged 3 months, on day 22 of illness. This was an “atypical” case in which the diagnosis was made post-mortem following death from myocardial infarction due to accelerated vaso-occlusive disease affecting coronary and mesenteric arteries. The case-fatality rate was 3.4%.

Comments

There will be a full report when the study is completed. Current data suggests disproportionate over-representation by children of Oriental or Polynesian extraction. Diagnosis and treatment is being made promptly however there is a high incidence of minor echocardiographic abnormalities affecting the coronary arteries. The fatal case of accelerated vaso-occlusive disease is of particular concern.

Perinatal exposure to HIV

Dr Nigel Dickson

Ongoing study started in January 1998

In 2001, there were 11 notifications of infants/children born to women infected with HIV, this includes 2 infants born in 2001 who were notified in 2002.

- Of these 11, 9 were born in New Zealand.
- 2 were children diagnosed with HIV who were born in 1999 to mothers in New Zealand whose HIV had not been diagnosed until after the pregnancy. One of these mothers was from a part of the world where HIV is particularly prevalent.
- The remaining 7 were born in 2001, 5 in the North Island and 2 in the South Island.
  - 3 of the mothers of these infants were from parts of the world where HIV is particularly prevalent.
  - None of these 7 children have been found to be infected with HIV.
  - Information was provided on measures taken to reduce perinatal transmission for 6 of these 7 women. All were given antiretroviral treatment during pregnancy, were delivered by Caesarian section and did not breast feed.

How these reports and other data on children in New Zealand born to women with HIV can give an indication of the prevalence of undiagnosed HIV among pregnant women in New Zealand is being prepared for publication in the New Zealand Public Health Reports.

Paediatricians need to be aware that there are likely to be a number of undiagnosed perinatally infected babies and children in our community born to mothers whose HIV has not been recognized.

Subdural Haemorrhage in Children Under 2 Years of Age

Dr Patrick Kelly

Third year of a four year study

There were 38 notifications of subdural haemorrhage in children under 2 years of age in 2001, the third year of this study. Completed reports have been received for 35 (92%) notifications. Of these 35 notifications, there were 26 cases, 7 duplicates, and 2 errors in reporting.

Of the 26 new cases:

- 17 were male, 9 female (1.9:1 M:F).
- 23 were from the North Island and 3 from the South Island.
- Most infants were under the age of 1 year (65%).
- In the completed reports, subdural haemorrhage was due to birth injury in 6 cases (23%), accidental injury in 2 (8%), child abuse in 12 (46%) and suspected abuse in 5 (19%). One case was due to Vitamin K Deficiency Bleeding (4%).

Comments

There will be a full report when the study is completed. Neurosurgeons are being included in the reporting of cases for 2002 which will be the final year of this study.

Vitamin K Deficiency Bleeding (VKDB)

Professor Brian Darlow

Ongoing study started in January 1998

There were two notifications of VKDB received in 2001, both were valid reports although in both cases the diagnosis is only "probable".

- One case involved a home birth at 41 weeks, where the mother declined vitamin K for her baby. Bleeding began at 24 hours of age, when i.m. vitamin K was given (with a haematoma forming at this site). Coagulation studies were only performed some hours later when the baby arrived at hospital and were essentially normal.

- The second case was unusual.
  - The mother presented at 32 weeks of pregnancy with decreased fetal movements. An antenatal MRI scan showed bilateral subdural haematoma.
  - The family were Polynesian Islanders and abdominal massage was queried but denied. There was no history of taking any prescribed or herbal medications.
  - At birth (32 weeks) the infant had a haemoglobin of 54 g/l, normal platelets (230) and a coagulation screen was typical of vitamin K deficiency. Factors 2, 7, 9 and 10 were low but factors 5 and 8 were normal. The coagulation screen normalised after i.v. and i.m. vitamin K.
  - This child has severe ongoing developmental delay.

Final Reports for Completed Studies

Retinopathy of Prematurity (Stage III and over)

Professor Brian Darlow

Background

Retinopathy of Prematurity (ROP) is an important cause of morbidity, including blindness, in very premature infants.

An important US multicentre study reported ROP of any stage in nearly 66% of infants with birthweight <1250g, stage III in 12% and "threshold disease" (stage III ROP involving 5 or more continuous or 8 or more cumulative clock hours with "plus" disease, which has a 50% chance of poor visual outcome) in 6% (Arch Ophthalmol 1988;106:471).

The last national New Zealand audit of ROP (in infants <1500g) was in 1986. Since them there has been considerable improvements in survival of the most premature infants (approximately 85% of <1000g infants survived in 1996 compared to 66% in 1989) and cryotherapy has become available to treat threshold disease (reducing adverse visual outcomes by nearly 50%). Overseas data give conflicting information of the impact of improved survival of these smallest infants on the incidence of ROP. Whilst studies from Australia have suggested that the incidence of blindness from ROP has fallen, one study from Manchester, UK, reported it had increased greatly.

Objectives

To assess:

1. The annual incidence of severe (Stage III and beyond) ROP in New Zealand.
2. Regional differences in the incidence of Stage III ROP.
3. what proportion of eyes (and infants) reaching Stage III ROP receive cryotherapy or laser therapy and what is the outcome (anatomic and visual) of treated and untreated eyes.

Case definition and reporting instructions

Any infant newly diagnosed with Stage III (or beyond) ROP in either eye by an ophthalmologist following the recommendations of the Committee for the Classification of Retinopathy of Prematurity (Pediatrics 1984;74:127-33).

Results

There were 34 notifications of Retinopathy of Prematurity (Stage III or over) in 1999 and 2000. Information has been received for 27 of these, of which 25 were valid reports.

Of these 25 cases:

- The mean gestation was 25.4 weeks (range of 23-31weeks), with just one infant having a gestation of over 28 weeks.
- The mean birthweight was 743g (range 440-1171), however this excluded one infant with non-immune hydrops who weighed 3075g. Four infants had a birthweight between 1000g and 1200g.

- Stage III disease was unilateral in 4 cases, and in 15 infants staging was clearly at less than threshold.
- Fourteen infants received treatment, 9 with laser therapy, 2 with cryotherapy only (to one eye only in one infant), and 3 infants were treated with a combination of laser and cryotherapy.
- The longer term outcome is unknown at this time but one infant is bilaterally blind and at least 3 other infants have significant cicatricial disease.

Comment

- The incidence of severe (stage 3 or more) acute ROP in New Zealand has probably not increased over the last decade despite increased numbers of very preterm infants and increased survival. In 1986 there were 12 infants with stage 3 or more ROP and, of these, 6 became bilaterally blind (cryotherapy was not available until 1987 in New Zealand) (Darlow *Arch Dis Child*; 1988; 63: 1083-6). From 1986 to 1998/9, infants with birthweight <1500g admitted for intensive care annually increased by 31%, and survival increased from 81% to 90% (Darlow *Arch Dis Child*; 2002: in press). Absolute numbers of infants with Stage 3 or more ROP increased by 33% from 1986 to 1999/2000.
- Treatment with laser or cryotherapy has contributed to reduced numbers of infants with severe loss of vision.
- Screening criteria of <1250g birthweight or <31 weeks gestation would appear to continue to capture all infants with stage 3 or more ROP (although this study can not prove this to be the case).
- There continue to be a number of significant problems with the screening programme:
- The results of eye examinations are sometimes poorly documented and do not follow the international classification
- Back transfer to level II units means some infants who qualify for screening are not examined or are not examined at an appropriate time. Better systems need to be in place to see that multiply transferred infants are examined on time.
    - There is a lack of information on outcome following severe ROP and follow-up procedures are both uncertain and likely to vary around the country.
- There should be an ongoing register of cases of stage 3 or more ROP, with each level III centre responsible for maintaining a 100% screening record of eligible infants and for knowing visual outcome for these infants.

Fetal Alcohol Syndrome

Dr Alison Leversha, Dr Simon Rowley, Dr Rosemary Marks
Starship Children's Hospital and National Women's Hospital

Introduction

Fetal Alcohol Syndrome (FAS) is one of the most common causes of mental retardation. Studies from North America and Europe suggest a prevalence of 0.3-1.9 per 1000 births. Based on these overseas statistics one would predict between 20 and 112 children with FAS are born each year in New Zealand.

However, little information exists about the incidence or prevalence of FAS in NZ. FAS is not a notifiable condition, and there are no central databases of affected children. Prior to 1995, there were no reported cases and no available statistics on the frequency of ARND in NZ. In 1993, we undertook a national survey of Paediatricians to determine the prevalence of FAS. The study estimated there were 63 children with FAS in paediatric care less than 10 years of age in 1993. This figure is considerably less than the expected rate from overseas figures. It is unclear whether New Zealand truly has a lower prevalence than overseas, or whether the estimate is spuriously low, and thus falsely reassuring.

The potential significance of Fetal alcohol Syndrome and Alcohol Related Birth Defects (ARND) in New Zealand becomes clear when we consider the prevalence of other common/well recognised conditions that cause disability or substantial morbidity. FAS/ARND could potentially be the most common cause of disability in New Zealand. Accurate incidence and prevalence information is essential for resource allocation, policy development, and the development of prevention strategies.

Disability	**Prevalence** (live births)	**Estimated prevalence** (per annum)
FAS	0.3-1.9 : 1000	20-114
ARND	5.9:1000	354
Cerebral Palsy	2 : 1000	120
Down syndrome	1 : 1000	60
Cystic fibrosis	1 : 3000	20

The New Zealand Paediatric Surveillance Unit (NZPSU) was established in 1997 to facilitate national surveillance and improve the knowledge of uncommon childhood conditions in New Zealand. As the NZPSU uses an active surveillance system, it is thought to be able to achieve reasonably complete reporting of a range of uncommon paediatric conditions. FAS was added to the surveillance in 1999, with the aim of providing a more reliable estimate of the frequency of this disability.

The specific aims of this project were to:

1. To estimate the incidence of newly diagnosed FAS among children in paediatric care in New Zealand.
2. To examine the diagnostic criteria used by Paediatricians regarding reported cases.
3. To provide data for targeting specific groups for further public health education.

Case definition and reporting instructions

Any child up to 15 years of age who in the opinion of the notifying Paediatrician has a new diagnosis of Fetal Alcohol Syndrome (definite or suspected).

Methods

Fetal Alcohol Syndrome was added to the Paediatric Surveillance Unit report card in July 1999. The 2½ year study finished in December 2001. Questionnaires were sent to all notifying paediatricians requesting details of the reported case. Specific data included reason for referral to the paediatrician, demographic data, diagnostic criteria, and other agencies involved. Initials and DOB were used to cross check for duplicate cases.

Results

Over the 29 month study period, there were 72 notifications, including 3 duplicate reports, and 7 unknown reports (due to missing data). There were thus 62 valid reports of new cases of definite or suspected Fetal Alcohol Syndrome. Thirty-nine percent of cases were New Zealand European, 45% Maori, 8% Pacific, 8% unknown and 8% Other (predominately Russian).

Table 5: Frequency of newly diagnosed cases

	# new cases	New cases per 1000 population*
NZ European	24	0.04
Maori	28	0.14
Pacific	5	0.06
Other	5	0.06
Total	62	0.07

* Denominator =children 0-14 years from the 2001 census

The cases were reported by 35 paediatricians from all around New Zealand, however, the majority of cases (97%) were from the North Island. Three paediatricians reported over a third of cases (23/62).

Ninety percent of children were New Zealand born. The children ranged from birth to 14 years, with the average age at diagnosis being 5½ years (see Figure 3). Children tended to present either in the neonatal period with growth disturbance, dysmorphic features or a maternal history of alcohol abuse, or at school age with behavioural or learning difficulties.

In the majority of instances, the diagnosis was first suspected by the notifying paediatrician (60%). A smaller number of diagnoses were suspected by parents (8%), education services (8%), mental health services (8%), and Child, Youth and Family Services (CYFS) (13%). No General Practitioner suspected the diagnosis and referred on for a paediatric assessment.

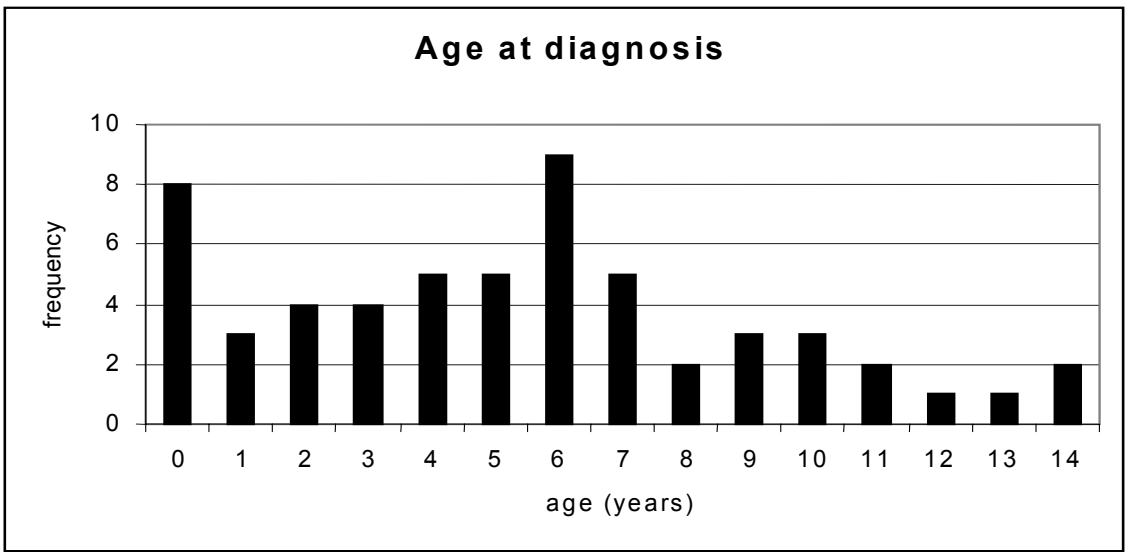

Figure 3: Age at diagnosis

Reasons for referral to paediatricians included were predominately neurobehavioural problems, with 73% referred for learning and/or behavioural difficulties (see Table 6). A proportion were recognised at birth following a recorded maternal history of alcohol abuse or excessive use during pregnancy. Seven children were referred for a confirmation of the diagnosis.

Table 6: Reason for referral to the paediatrician

Reason	Frequency %
Behaviour	58
Learning/school difficulties	57
Maternal history of alcohol abuse	19
Dysmorphic features	13
Other	13
Suspected FAS	11
Growth	8
Adoption assessment	5

% add up to more than 100% as many children were referred with several concerns.

The majority of children were adopted or in foster care (44%). Twenty-six percent of children lived with their biological parents, and the remainder were living with their extended family.

Most children already had several agencies involved in their care when the diagnosis was made.

Antenatal history

The average gestational age at delivery was 37.8 weeks. The average maternal age at delivery was 27.7 years (17-39 years). A similar proportion of mothers were less than or equal to 20 years, or greater than 36 years at delivery. Thirty-nine percent of mothers were known to have consumed other drugs during the pregnancy, most commonly marijuana. Fifty-eight percent of mothers smoked cigarettes.

Table 7: Agencies involved

Agencies involved	%
Special education services	58
Child development services	53
Mental health services	24
Justice department	24
CYFS	24

Alcohol exposure

The gestational alcohol exposure was most often reported by someone who saw the mother drink during pregnancy or another reliable source. As only a modest proportion of affected children remained in their birth families, the history of antenatal alcohol exposure was provided directly by the mother in only 23%. The exact quantity and frequency of alcohol consumption was therefore frequently not known.

CNS involvement

The majority of children had several features of CNS involvement. The most commonly reported were behavioural difficulties, developmental delay, ADHD, language delay and intellectual impairment (see Table 8).

Table 8: CNS involvement

Abnormality	Frequency present %	Frequency absent %	Unknown* %
Behavioural dysfunction	79.0	11.3	9.7
Developmental delay	53.2	35.5	11.3
ADHD	53.2	27.4	19.4
Language disorder	48.4	35.5	16.1
Mild intellectual impairment	40.3	29.1	30.6
Head circumference ≤ 2SD	22.6	74.2	3.2
Hypertonia / hypotonia	14.5	80.7	4.8
IQ ≤60	9.7	51.6	38.7
Seizure disorder	9.7	85.5	4.8
Tremors / marked incoordination	8.1	85.4	6.5
Myopia / hyperopia	3.2	67.8	29.0
Abnormalities on CNS imaging	1.6	12.9	85.5
Sensorineural hearing loss	0	69.4	30.6

*Unknown: diagnostic test not yet performed or too young to be able to determine whether it was going to be a problem.

Growth

Fifty-one percent of children were known to have a birth weight less than the tenth centile, and 45% were known be less than the 10^{th} centile for length at birth. Paediatricians rarely adjusted growth parameters for parental centiles.

Facial features
Eight-nine percent of children had facial features consistent with Fetal alcohol syndrome (short palpebral fissures, smooth philtrum, thin upper lip, flat midface). The remainder did not have palpebral fissure lengths recorded or were in an age range where the facial features are known to be less characteristic.

Discussion

- There were 62 new cases of definite or suspected FAS identified during the study period. The reported incidence is considerably lower than overseas estimates.
- The use of the PSU surveillance system has provided the most accurate estimate of the frequency of FAS in New Zealand to date. However, it depends on affected children being referred to paediatricians, individual paediatricians asking about alcohol exposure during pregnancy, considering the diagnosis of FAS, and then making the diagnosis. Difficulties with each of these factors will contribute to an underestimate of the frequency of FAS in the childhood population.
- There appears to be different thresholds for making the diagnosis. It is likely that some paediatricians do not consider the diagnosis and thus do not make the diagnosis.
- Although the diagnostic criteria for FAS are clear, the diagnosis is elusive to many clinicians as the signs are not specific to alcohol exposure and there are no confirming laboratory tests.
- Individuals with FAS are a subset of all individuals who are affected by in-utero exposure to alcohol. The variable outcomes in children exposed to alcohol in-utero are due to variations in gestational timings and volumes of exposure, differences in maternal metabolism of alcohol, dietary intake, concurrent exposures to other substances, and genetic factors in the mother and child. Individuals who have been exposed to alcohol in-utero and present with cognitive/ behavioural disabilities, but do not have the characteristic FAS facial appearance or growth deficiency are classified as having Alcohol Related Neurological Defects (ARND). These children may have difficulties just as severe and debilitating as those with the complete FAS. There are no data on the frequency of ARND in New Zealand.
- There is an increased awareness of the behavioural and learning difficulties as manifestations of FAS.
- The majority of affected children are in foster or extended family care.
- Children with FAS are born to women of all childbearing ages.
- FAS occurs in all ethnic groups, however, Maori children have disproportionately higher rates. It is unclear whether this represents a greater risk of FAS per se or whether there are ethnic-specific differences in referral and/or diagnosis rates.
- Many agencies are involved in the care of these children, and a notable proportion had involvement of justice and CYFS.

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Thank you to all participating Paediatricians!

Conditions ever monitored by NZPSU

A total of thirteen conditions have been monitored by the NZPSU since it began in 1997.

Table 9: Conditions ever monitored by NZPSU

Condition	Abbreviation	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	VitK	January 1998	Ongoing
Neonatal herpes simplex infection	HSV	January 1998	December 2000
Subdural haemorrhage (<2 years)	SH	January 1999	Ongoing
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	Ongoing
Bronchiectasis		January 2001	Ongoing
Idiopathic nephrotic syndrome	INS	July 2001	Ongoing

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

International Network of Paediatric Surveillance Units

In August 1998, at the 22nd International Congress of Paediatrics in Amsterdam, ten national paediatric surveillance units met to discuss a proposal that would link pre-existing units and improve international collaboration and discussion. Together, they formed the International Network of Paediatric Surveillance Units (INoPSU) (see Table 10). A secretariat, consisting of representatives from the United Kingdom (UK), Australia, Canada and the Netherlands, was set up to carry out the aims and direct the activities of INoPSU.

Founding members included units from Australia, Canada, Germany, Latvia, Malaysia, Netherlands, New Zealand, Papua New Guinea, Switzerland and the UK. More recently, the Welsh unit, which was formed in 1995, and concentrates on less rare disorders, became the eleventh unit to join INoPSU. Several countries such as Portugal, Belgium and the Czech Republic have also expressed an interest in developing national paediatric surveillance units.

A further meeting was held in Ottawa in 2000 sponsored by the Canadian Government. This meeting was attended by Dr Nigel Dickson and Nicola Dow.

INoPSU is currently chaired by Dr Elizabeth Elliot of the Australian PSU.

Aims of INoPSU
The aims of INoPSU are:

- To facilitate 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.

Table 10: Members of INoPSU

Country	Unit	Email	Website
Australia	APSU	apsu@nch.edu.au	www.racp.edu.au/apsu
Britain	BPSU	richard.lynn@rcpch.ac.uk	http://bpsu.inopsu.com
Canada	CPSP	joanne_doherty@hc-sc.gc.ca	www.cps.ca/english/proadv/CPSP/CPSP.htm
Germany	ESPED	heinrich@med.uni-duesseldorf.de	www.esped.uni-duesseldorf.de/
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/nzpsu/nzpsu1.html
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 11: 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	2100
Canada	6.3	1996	2300
Germany	14	1992	500*
Latvia	0.4	1996	22
Malaysia	7.6	1994	400
Netherlands	2.9	1992	390
Papua New Guinea	1.9	1996	40
Portugal	1.6	2000	?
New Zealand	0.8	1997	179
Switzerland	1.3	1995	38*
Wales	0.6	1995	134*

* Heads of Paediatric Centres

Table 12: Conditions under surveillance worldwide 2001

Conditions under surveillance	Country
Abdominal injury due to child abuse	Britain
Acute flaccid paralysis	Australia, Canada, Netherlands, New Zealand, Papua New Guinea, Switzerland
Acute rheumatic fever	Switzerland
Adverse effects from complementary or alternative medicine	Australia
Aplastic anaemia	Latvia
Anaphylaxis	Canada
Atypical mycobacterial infections	Netherlands
Bronchiectasis	New Zealand
Cerebral edema in diabetic ketoacidosis	Canada
Cerebral vascular disease/stroke and like illness	Britain
CHARGE association/syndrome	Australia, Canada
Childhood tuberculosis	Wales
Childhood inflammatory bowel disease	Netherlands
Congenital adrenal hyperplasia	Netherlands
Congenital cytomegalovirus infection	Australia, Britain
Congenital heart disease	Malaysia
Congenital hypothyroidism	Latvia, Papua New Guinea
Congenital rubella	Australia, Britain, Canada, New Zealand, Switzerland
Congenital syphilis	Latvia
Diabetes mellitus/insulin-dependent/<5 years	Germany, Latvia, Netherlands, Papua New Guinea, Portugal, Wales
Duchenne muscular dystrophy	Malaysia
Fetal alcohol syndrome	Australia, New Zealand
Fatal/near fatal asthma	Malaysia
GLUT-1 deficiency/de vivo disease	Germany
Group B streptococcal infections (neonatal)	Netherlands, Portugal, Germany
Haemolytic Uraemic Syndrome	Australia, Canada, Latvia, New Zealand, Portugal, Switzerland
Haemorrhagic disease of the newborn (vitamin K deficiency Bleeding)	Australia, Britain, Germany, New Zealand
Hepatitis C virus infection	Canada
Histiocytosis X	Latvia
HIV/AIDS +/- perinatal exposure to HIV	Australia, Britain, Malaysia, Netherlands, New Zealand
Hodgkin's disease	Latvia
Hospitalized pertusis	Australia, Netherlands
Idiopathic juvenile osteoporosis	Germany
Idiopathic and congenital nephrotic syndrome	Australia
Imported tropical diseases: malaria, schistosomiasis, leishmaniasis	Germany
Ingestion of lamp oil (intoxications)	Germany
Intersexuality and severe genital malformations	Germany
Invasive *Haemophilus influenzae* infection	Germany
Ischaemic stroke in infants (neonatal sinus venous thrombosis)	Germany
Kawasaki Disease	New Zealand, Portugal
Kernicterus	Germany
Leukaemia: ALL, AML	Latvia

Munchausen by proxy syndrome	Australia
Necrotizing fascitis	Canada
Neonatal alloimmune thrombocytopenia	Netherlands
Neonatal herpes simplex virus infection	Australia, Canada
Neonatal liver failure/perinatal hemochromatosis	Canada
Neonatal meningitis	Malaysia
Neural tube defects	Netherlands, Switzerland
Neurological endemic cretinism	Papua New Guinea
Non-Hodgkin's lymphoma	Latvia
Paediatric malignancies	Papua New Guinea
Palliative Care	Wales
Phenylketonuria	Latvia
Pigbel	Papua New Guinea
Pneumococcal sepsis/meningitis	Germany
Progressive intellectual and neurological deterioration/CJD	Britain, Canada
Renal tubular acidosis	Papua New Guinea
Rett syndrome	Australia
Reye syndrome	Latvia
Rotavirus infection	Netherlands
RSV disease requiring intubation and artificial ventilation; severe	Germany, Switzerland
Septo-optic dysplasia	Wales
Severe combined immunodeficiency	Australia
Severe complications of medical therapy	Netherlands
Smith-Lemli-Opitz syndrome	Canada
Steroid resistant nephrotic syndrome	Germany
Splenectomy and hyposplenism	Wales
Subacute sclerosing panencephalitis	Papua New Guinea
Subdural haemorrhage	New Zealand, Wales
Tick-borne encephalitis	Switzerland
Transient myeloproliferative syndrome in newborns with Down syndrome	Germany
Varicella/zoster infection	Switzerland
Venous thromboembolic complaints	Britain

List of (Possibly Obsessive) Clinicians With 100% Return Rate 2001 (& 2000)

Thank you to those clinicians who returned all of their cards in 2001

Abbott	George	Grimwood	Keith	Newman	David
Aickin	Richard	Gunn	Alistair	Newman	John
Aiken	Geoff	Hall	Kate	Nicholson	Ross
Ansarian	Mohammad	Harding	Jane	Nicolls	Wayne
Armishaw	Jeremy	Heaton	Paul	Nobbs	Peter
Austin	Russell	Heron	Peter	Palmer	Penny
Austin	Nicola	Hewson	Michael	Pattemore	Philip
Baker	Nicholas	Jamison	David	Radcliffe	Marlon
Barry	David	Jankowitz	Peter	Ramadas	Ram
Barry	John	Kelly	Andrew	Richardson	Vaughan
Bates	Giles	Kelly	Patrick	Rudge	Susan
Battin	Malcolm	Kerr	Archie	Segedin	Elizabeth
Beasley	Spencer	Kuschel	Carl	Selby	Roslyn
Bourchier	David	Lear	Graeme	Shaw	Ian
Bradley	Stephen	Lees	Hugh	Shaw	Robyn
Bremner	Catherine	Leversha	Alison	Shillito	Paul
Brown	Jeff	Liang	Allen	Simpson	A. K.
Buchanan	Leo	Lourens	Roelof	Sinclair	Jan
Calder	Louise	Macfarlane	Scott	Skeen	Jane
Campbell	Moira	MacKenzie	Neil	Smith	Warwick
Caseley	Terry	Maikoo	Rajesh	Stanley	Thorsten
Chin	Simon	Malcolm	Stuart	Sullivan	Michael
Clarkson	John	Manikkam	Noel	Taylor	Barry
Corban	Jenny	Mao	M.	Thiagarajan	Prakash
Corbett	Robin	Marks	Rosemary	Tomlinson	Paul
Corrie	Joan	Maxwell	Fraser	Trenholme	Adrian
Cutfield	Wayne	McArthur	John	Tyrrell	Vicki
Daniell	Alison	McGill	Fiona	Vogel	Alison
De Sylva	Tony	McIlroy	Peter	Watt	Mike
Dixon	Joanne	McKie	Jill	Webster	Diane
Doran	John	Mildenhall	Lindsay	Wesley	Alison
Drage	Alan	Mitchell	Ed	Weston	Phillip
Elder	Dawn	Moore	Philip	Wilson	Callum
Farrell	Alan	Morreau	Johan	Wilson	Nigel
Fleming	John	Morris	Max	Wilson	Ross
Ford	Rodney	Morrison	Philip	Wiltshire	Esko
Gavin	Raewyn	Moyes	Chris	Wong	Maisie
Goldsmith	John	Nagel	Fred		

Congratulations to Scott Macfarlane who was selected at random to win a $50 book token to be presented at the AGM of the Paediatric Society of New Zealand.