

THE GREEN LANE PAEDIATRIC AND CONGENITAL CARDIAC SERVICE

STARSHIP CHILDREN'S HOSPITAL

BIENNIAL REPORT

JULY 2009 – JUNE 2011



Collated by Dr Tom Gentles, Director

TABLE OF CONTENTS

1. EXECUTIVE SUMMARY	3
2. BACKGROUND.....	3
3. SERVICE COMPONENTS	4
3.1. SUMMARY	4
3.2. PAEDIATRIC INPATIENTS.....	4
3.3. PAEDIATRIC AND CONGENITAL CARDIAC SURGERY	4
3.4. PAEDIATRIC OUTPATIENTS	10
3.5. PERIPHERAL CLINICS	11
3.6. FETAL CARDIOLOGY	12
3.7. ADULT CONGENITAL HEART DISEASE SERVICE.....	13
3.8. CARDIAC INHERITED DISEASE GROUP (CIDG).....	16
3.9. RHEUMATIC HEART DISEASE	19
4. INVESTIGATIVE SERVICES	21
4.1. ECHOCARDIOGRAPHY	21
4.2. CARDIAC MRI	23
4.3. CARDIAC CATHETERISATION	23
4.4. ELECTROPHYSIOLOGY	26
4.5. PACEMAKER.....	27
4.6. CARDIAC REGISTRY	27
5. NURSING.....	28
5.1. NURSING LEADERSHIP	28
5.2. VISION	29
5.3. INPATIENT	29
5.4. EDUCATION	29
5.5. NURSING LED INITIATIVES	30
5.6. CLINICAL EFFECTIVENESS	31
5.7. FETAL SERVICE.....	31
6. ACADEMIC BIBLIOGRAPHY	32
6.1. PEER REVIEWED ARTICLES	32
6.2. BOOK CHAPTERS.....	34
6.3. INVITED PRESENTATIONS	34
6.4. ABSTRACTS	36
6.5. GRANTS AWARDED	37
6.6. COMMITTEES	38
6.7. OTHER.....	38
7. MEDICAL STAFF.....	39
7.1. CONSULTANT CARDIOLOGISTS.....	39
7.2. CONSULTANT CARDIOTHORACIC SURGEONS	39
7.3. OTHER CLINICAL STAFF.....	39
7.4. CONTRIBUTORS TO THE BIENNIAL REPORT	40

1. EXECUTIVE SUMMARY

The department's performance is dependent on the expertise and dedication of its medical, nursing, technical, and support staff, and also on staff in the intensive care unit, anaesthesia, and involved paediatricians and cardiologists throughout the country. 2009-11 has seen further development of national programmes for Adult Congenital Heart Disease, and the diagnosis and management of Cardiac Inherited Disease. Moves toward national "top-slice" funding for the service were instigated by the National Health Board after a review of paediatric subspecialty services. This initiative aims to address issues of vulnerability in terms of workforce and capital requirements, and ensure equitable service delivery at a national level.

Service development has focussed on subspecialisation with the aim of providing the best possible care for children with heart disease. This has included development of a dedicated paediatric nursing teams in the cardiac catheterisation laboratory and the paediatric cardiac operating rooms, establishment of nurse practitioner positions in the paediatric cardiac and adult congenital heart programme, and the appointment of an additional paediatric cardiologist and paediatric cardiac surgeon. Further development of outreach to the Pacific Islands has been beneficial for clinicians and patients in this region, and has improved the cost effectiveness of treatment.

The service appreciates support from the Starship Foundation and the District Health Board for capital acquisitions including echocardiography machines. The CT facility purchased by the Starship Foundation for the radiology department has further reduced the need for diagnostic cardiac catheterisation. @ Heart (previously Heart Children New Zealand) provides support for the home INR monitoring programme, and professional support to nursing and technical staff through the Advancement Programme. Interaction with funding agencies has resulted in increased availability of medications for the treatment of pulmonary hypertension, and funding for the Melody transcatheter pulmonary valve implantation programme.

Academic activities have benefited grant funded project and salary support from a number of agencies including the National Heart Foundation of New Zealand, the Green Lane Research and Education Fund, Cure Kids and Lottery Health Research.

2. BACKGROUND

The Green Lane Paediatric and Congenital Cardiac Service is a national service based at the Starship Children's Hospital.

It is the sole provider of cardiology and cardiac surgical services for infants and children with congenital and acquired heart disease in New Zealand and also provides a fetal cardiology service and investigation and treatment of those born with congenital heart disease who are now adults. The service provides an extensive network of outreach clinics throughout New Zealand and the South Pacific, and provides consultation and support to clinicians caring for patients within the regional hospital setting.

In addition there is an active clinical research and audit programme that includes collaborative ventures with academic groups nationally and internationally.

3. SERVICE COMPONENTS

3.1. SUMMARY

The service has a number of interrelated components including:

- Paediatric inpatient (medical and surgical)
- Paediatric and congenital cardiac treatment (surgical and catheter based)
- Paediatric Outpatient
- Peripheral Clinics (paediatric and adult congenital)
- Fetal Cardiology
- Adult Congenital Cardiology
- Cardiac Inherited Disease

Investigative Services include:

- Echocardiography
- Cardiac Catheterisation
- Exercise testing
- Cardiac MRI

Ancillary services contracted from Adult Cardiology:

- Electrophysiology laboratory
- Pacemaker diagnostics
- Electrophysiology and electrocardiography technical staff
- Cardiac catheterisation laboratory support staff

Ancillary services contracted from Adult Cardiothoracic Surgery and Operating Theatres:

- Cardiac Perfusion
- Theatre nurses
- Anaesthetists and anaesthetic technicians

3.2. PAEDIATRIC INPATIENTS

There is a dedicated 22 bed ward including a 4 bed Intensive Observation Unit. Nursing resource allows for staffing of 17 of these beds. The service shares a 16 bed paediatric intensive care unit and six bed High Dependency Unit, utilising on average four beds. The intensive care unit is staffed by paediatric intensivists.

All inpatient referrals are tertiary in nature, with the majority originating outside the northern region.

3.3. PAEDIATRIC AND CONGENITAL CARDIAC SURGERY

Paediatric cardiac surgery is undertaken in one of two cardiac operating theatres at Starship Children's Hospital under the leadership of Mrs Kirsten Finucane. Adult congenital cardiac surgery is undertaken by the same surgical team in the Level 4 cardiac operating theatres at Auckland City Hospital (adjoining Starship Children's Hospital). Postoperative patients are transferred to the Paediatric Intensive Care Unit or in the case of adult patients the Cardiovascular Intensive Care Unit. Following the appointment of Mr John Artrip in September 2009, there are three full time surgeons. This additional surgical position not only removes an area of vulnerability, but also allows a more reasonable work/life balance in addition to increased time for database development, research and teaching. The surgical team are supported by two senior registrars, and a physician assistant who has been successfully trained as part of a pilot programme. She has become an invaluable part of the team, assisting in the operating theatre, preparing the patients and educating families preoperatively, and guiding the post-operative, day to day care in the ward. A further appointment is planned for 2012.

Surgical volumes and outcomes

The following statistics are counts of admissions that result in cardiac surgery and exclude patients cannulated for ECMO for non cardiac reasons, and premature neonates who underwent ligation of a patent ductus arteriosus in the neonatal intensive care unit.

The numbers of bypass cases have increased from 275 per year to 350 per year while there has been little change in the number of non bypass cases. (Figures 1-2).

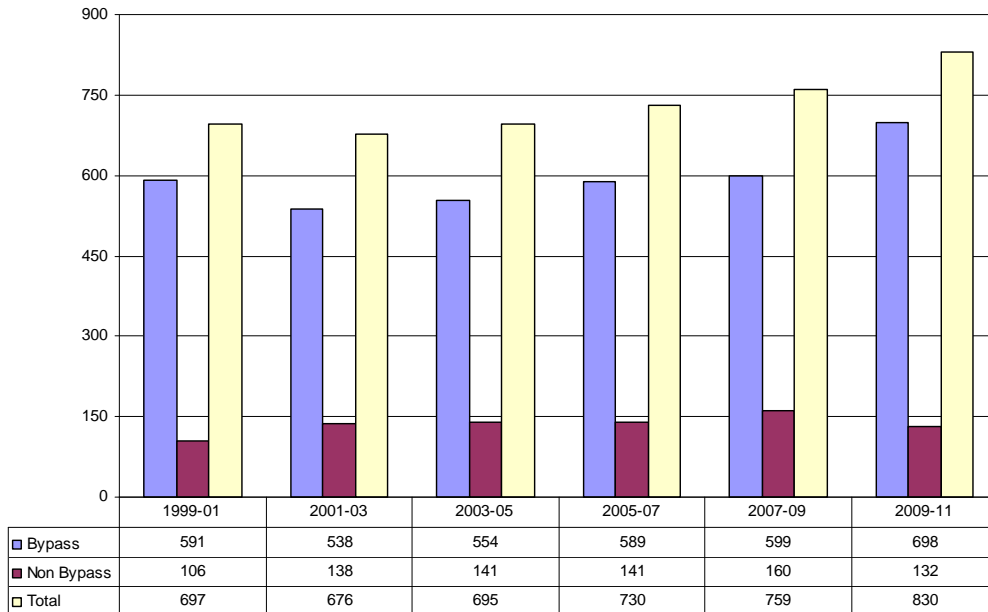


Figure 1. Surgical admissions by year and type of procedure

Approximately one half of surgical admissions continue to be for infants aged <1 year but the adult component over the age of 15 years has increased slightly from 10 to 15% and demographics suggest this increase will continue.

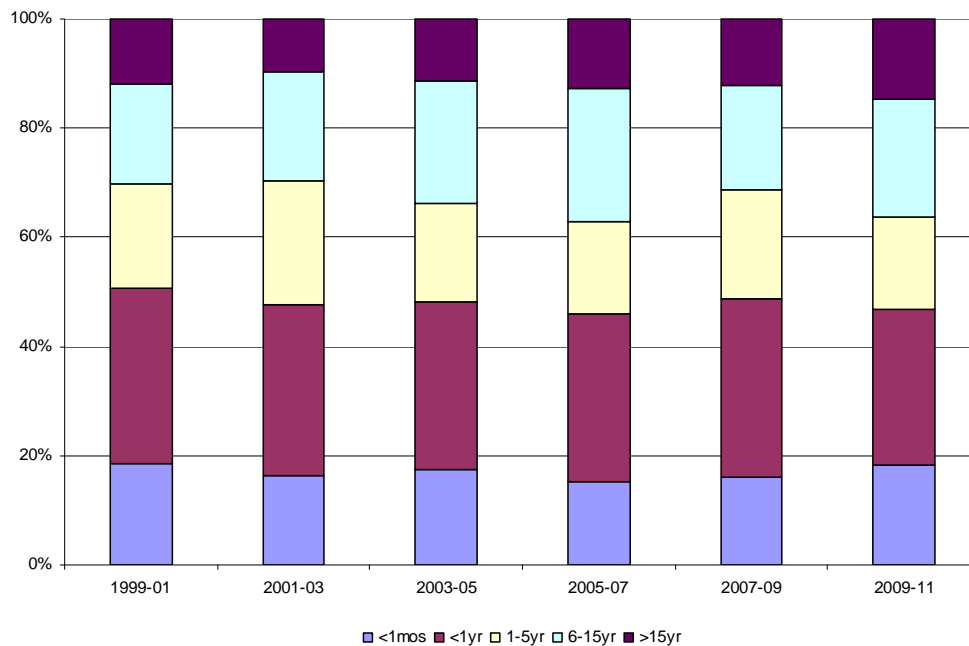


Figure 2. Surgical admissions by age

The early mortality rates quoted in Tables 1-4 and Figures 3-6 relate to deaths during a surgical admission or in the first 30 days after operation.

Surgical mortality is low. However, in the 2009-10 year it increased to 2.5% - the first time it has been over 2% in six years (Figure 3). This increase was primarily related to an increase in deaths in neonates with the neonatal mortality rate increasing from 3.8% (2002-05) to 6.8% (2008-11) (Table 3).

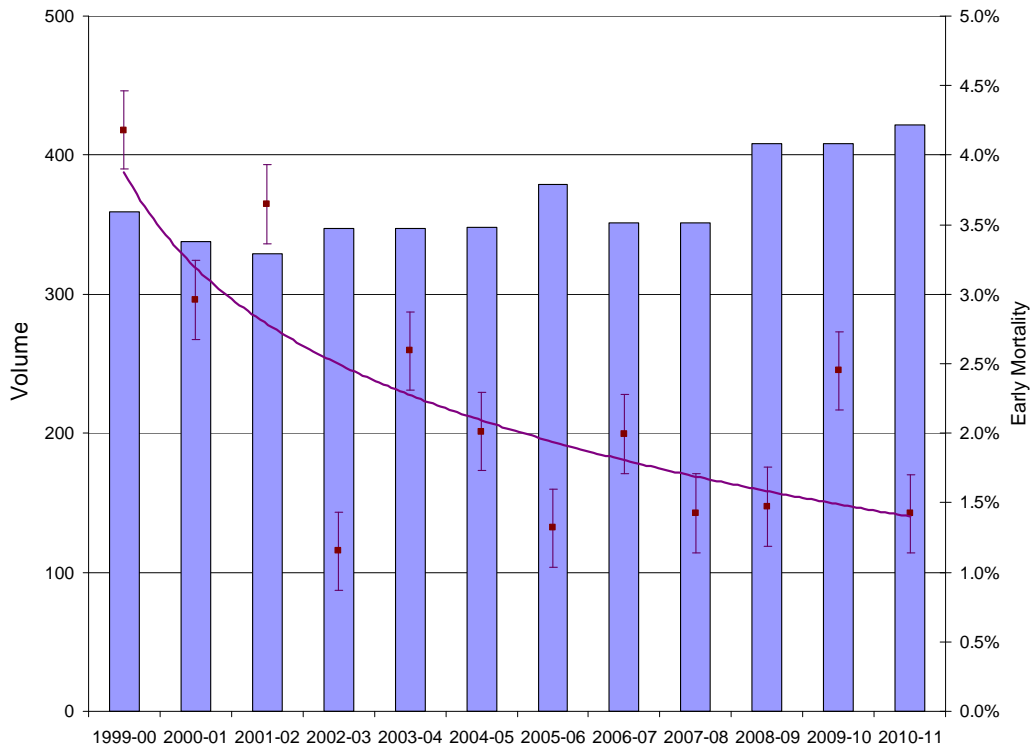


Figure 3. Volumes (surgical admissions) in blue and early mortality (maroon). Bars are +/- SE and lines indicate polynomial trends (black for volume and maroon for early mortality)

The additional mortality was concentrated in those with complex heart disease as evidenced by the increased mortality rate in RACHS-6 patients (Figures 4 and 5). This category mainly contains neonates undergoing a Norwood or Sano procedure. The number of these infants is relatively small with significant year-on-year variation so that mortality trends are difficult to define with any degree of certainty (Figure 6). In addition, infants with a Norwood or Sano procedure often remain in hospital for two to three months so that early mortality (death during hospital admission or within 30 days of operation) is higher than 30 day mortality. Nevertheless, there appears to have been an increase in mortality in these patients and in neonates with single ventricle requiring aortopulmonary shunts, particularly if pulmonary artery plasty is also required.

In the case of hypoplastic left heart syndrome, the Sano procedure is the preferred method for managing pulmonary blood flow. Consequently sudden postoperative death related to shunt failure is uncommon but failure of the systemic right ventricle or tricuspid valve or inadequate pulmonary artery growth may be problematic. A strategy of early bi-directional cavopulmonary anastomosis has been successful in some patients. Analysis of the cohort of Norwood/Sano patients is ongoing as is work with the intensivists and cardiologists to plan a better management strategy for this group.

Infection control is an important issue and mediastinitis was the cause of two of the deaths in the period 2009 to 2011. There has been an increase in infection and re-exploration of sternotomy rates over the last decade. These infections have led to review of management of surgical patients with regard to the root causes of infection in order to reduce the chance of deep sternal wound infection. This continues to be a challenge in the environment of an acute hospital with high rates of resistance to standard antibiotics and the post operative reliance on invasive monitoring in infants who are immuno-compromised following their cardiac by-pass. The

microbiologists have been involved in surveillance of infection patterns and we continue to work closely together on this issue.

Year	Early Mortality	Total	Percent Early Mortality
1999-00	15	359	4.2%
2000-01	10	339	2.9%
2001-02	12	329	3.6%
2002-03	4	347	1.2%
2003-04	9	347	2.6%
2004-05	7	347	2.0%
2005-06	5	378	1.3%
2006-07	7	353	2.0%
2007-08	5	352	1.4%
2008-09	6	408	1.6%
2009-10	10	408	2.5%
2010-11	6	422	1.4%
Total	96	4387	2.2%

Table 1. Early mortality by year

Year	Bypass			Non Bypass		
	Early Mortality	Total	Percent Early Mortality	Early Mortality	Total	Percent Early Mortality
1999-02	33	847	3.9%	4	179	2.2%
2002-05	15	836	1.8%	5	206	2.4%
2005-08	13	868	1.5%	4	213	1.9%
2008-11	18	1018	1.8%	4	220	1.8%
Total	79	3569	2.2%	17	818	2.1%

Table 2. Early mortality by operation type

	<1month	1-12 month	1-5 years	6-15 years	>15 years
1999-02	10.6%	3.1%	0.5%	1.0%	4.3%
2002-05	3.8%	2.7%	0.5%	0.0%	2.7%
2005-08	5.6%	1.2%	0.5%	0.0%	1.5%
2008-11	6.8%	1.9%	0.0%	0.0%	0.6%

Table 3. Early mortality by age

Period	Early Death	Total	% Early mortality	Low Birth Weight % of cases aged <1 year
1999-02	3	23	13.0%	2.2%
2002-05	2	30	6.7%	2.4%
2005-08	1	26	3.8%	2.9%
2008-11	2	32	6.3%	3.6%

Table 4. Early mortality for low birth weight (less than 2500g)

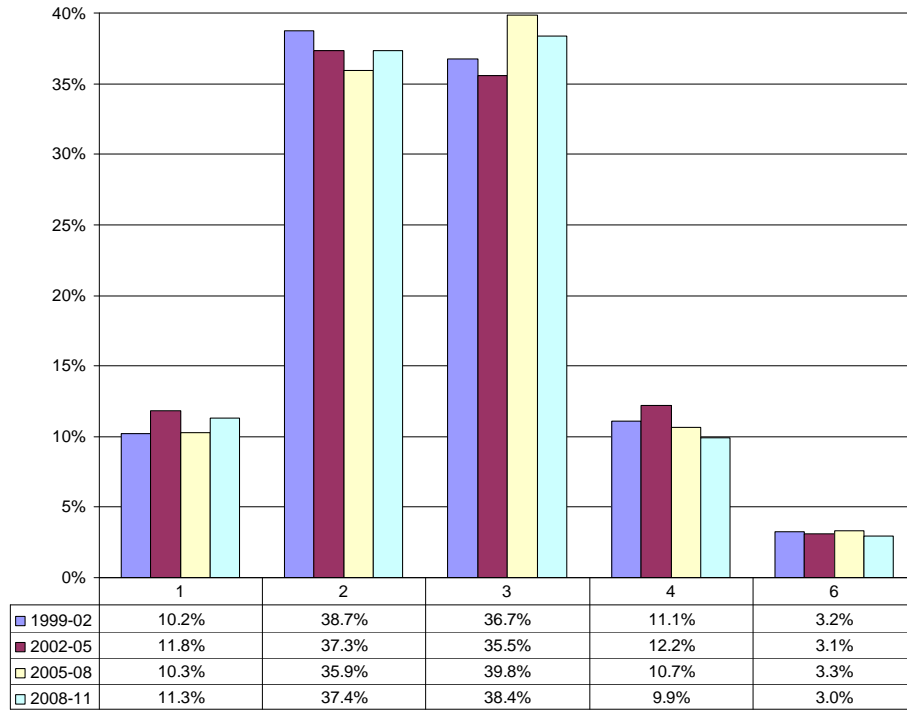


Figure 4. RACHS codes and admissions over three time periods (age <18 years). Class 5 has too few numbers for analysis

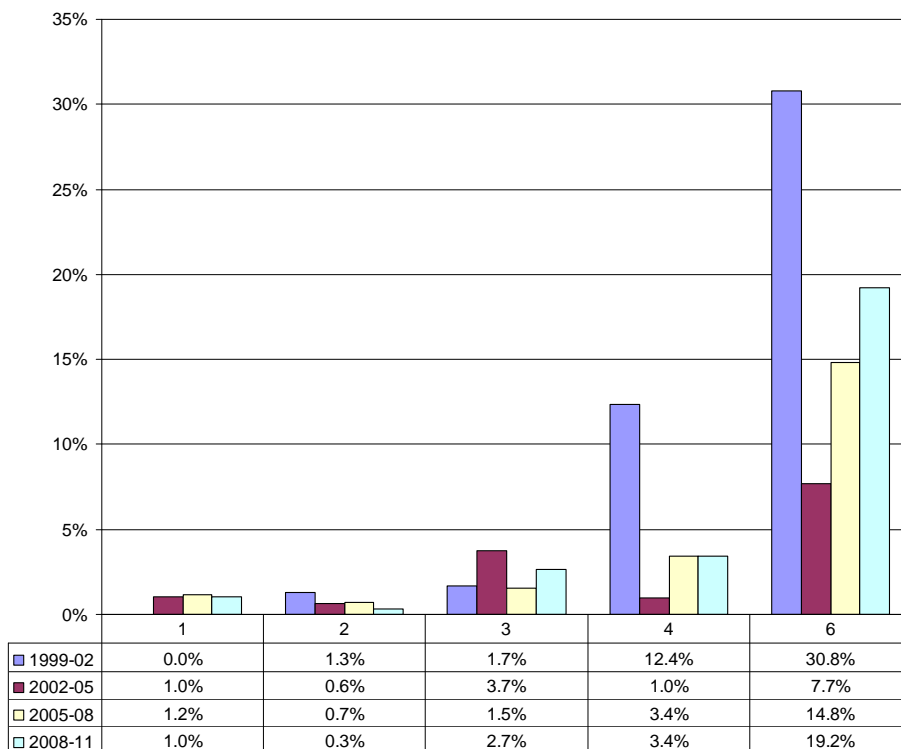


Figure 5. Early mortality by RACHS classification over 4 time periods

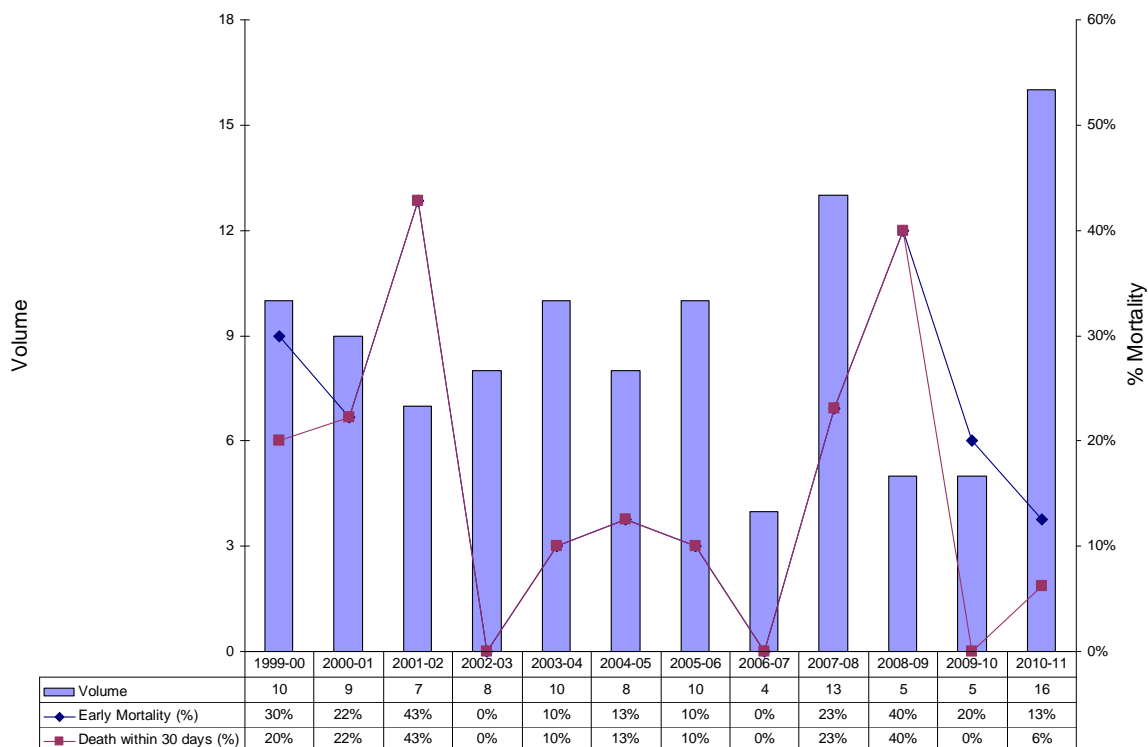


Figure 6. RACHS 6 volume, early mortality and 30-day mortality by year

In addition to New Zealand resident patients, infants and children are admitted from Pacific Islands where paediatric cardiac surgery is unavailable (Figure 7). These children are sponsored by NZ Aid, their local governments, or occasionally non-governmental charitable organisations. This scheme has been successful in most cases, repairing simple valve lesions or defects with a reasonably short hospital stay and low complication rate. The additional volume has allowed the unit to achieve critical mass in a number of staffing and infrastructure areas. Cardiology visits to the Pacific Islands has improved the triage process as well as post-operative surveillance.

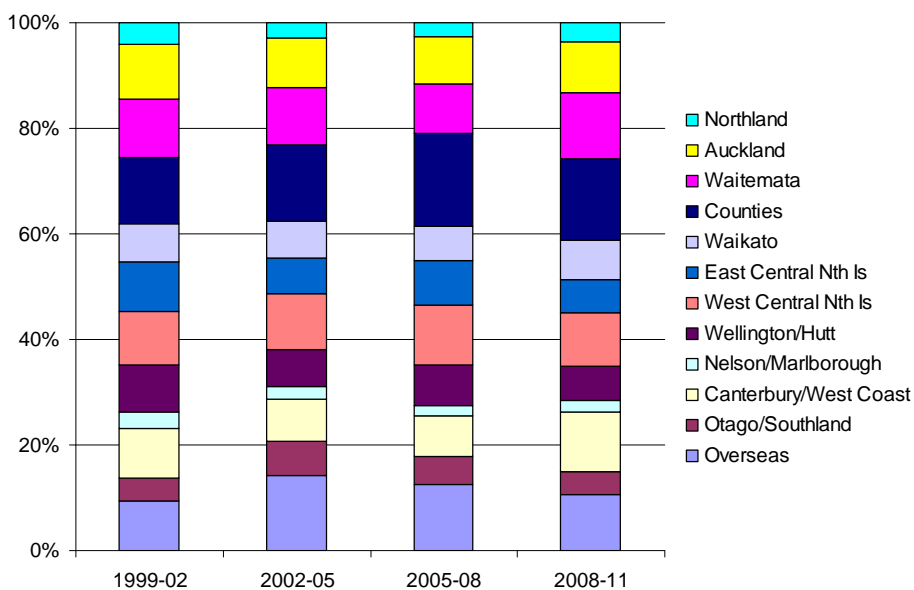


Figure 7. Domicile of surgical patients

The primary focus has moved toward enabling practitioners and administrators in the islands to identify and transfer children early so that they are in a better condition for surgery.

3.4. PAEDIATRIC OUTPATIENTS

There are eight paediatric cardiology outpatient clinics per week, including an Arrhythmia Clinic and two Day Stay sessions. Ninety percent of outpatients are tertiary (referred from paediatricians or cardiologists). Secondary referrals reside almost entirely in the Auckland District Health Board region.

Volumes of patients seen in the clinics at Starship Children’s Hospital have been steady over the past 4 years (Figure 8) despite multiple changes in practice including:

- Primary referrals from out of region have been devolved to paediatricians in West, North and South Auckland
- An increasing trend for heart disease to be diagnosed prenatally or in the new born nursery
- Increased numbers of infants and children surviving complex cardiac surgery
- More intensive surveillance of at risk groups resulting in earlier treatment and reduction in long-term morbidity and mortality
- Increased numbers of outreach clinics and up-skilling of regional expertise

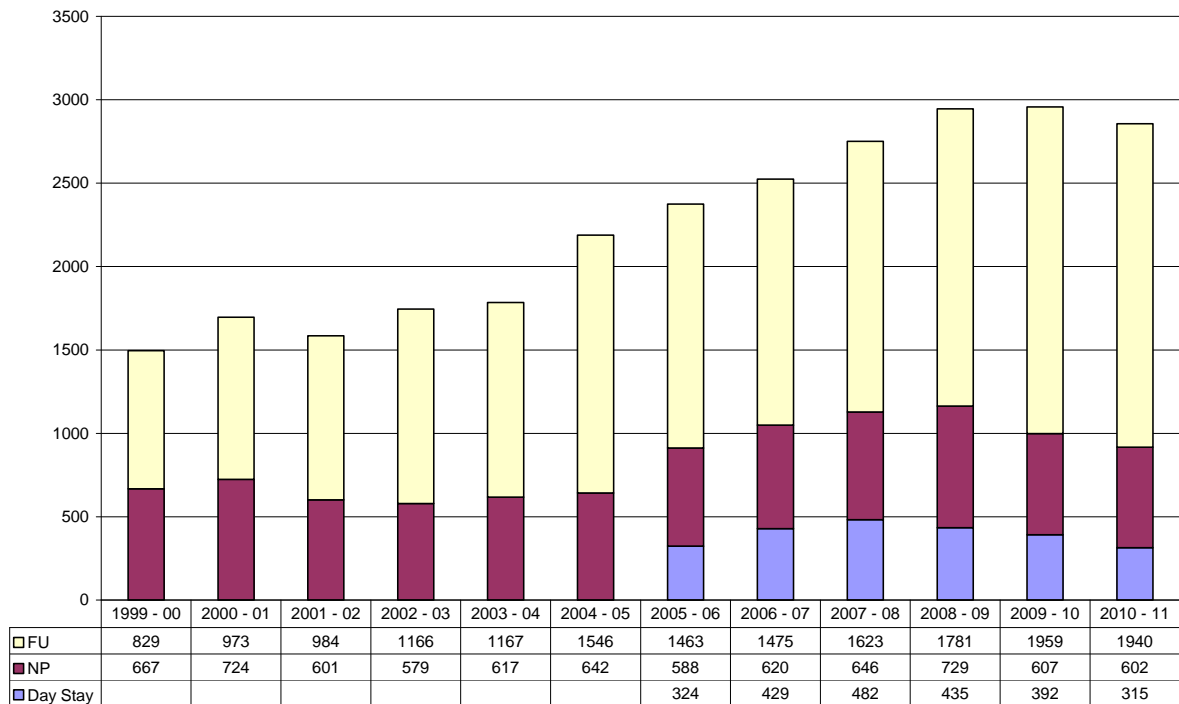


Figure 8. Paediatric Cardiology Outpatient Visits. (NP = new patient, FU = follow-up. Day Stays were included with outpatient visits prior to 2005-06)

3.5. PERIPHERAL CLINICS

Peripheral paediatric cardiology clinics are undertaken in all major metropolitan centres and in most regional centres. There are 118 clinic days per year including eight adult congenital clinics (Table 5). Patients seen in these clinics are solely the result of referral from secondary and tertiary sources. There is a continuing, albeit small, unmet need for visiting clinics and it is likely numbers will increase in 2012.

There is a need to standardise access and quality of service delivery across the regions, and a plan to establish a national Paediatric Cardiac network has been on the agenda of discussions with representative from the National Health Board through 2010 and 2011. Although there is currently an ad hoc network in place there is no consistency across the country in terms of the amount of outreach activity, and local expertise. National planning should result in an equitable spread of clinics and ensure uniform service delivery.



Clinic	2002-2003	2003-2004	2004-2005	2005-2006	2006-2007	2007-2008	2008-2009	2009-2010	2010-2011
Blenheim		1	1	1	2	3	1	1	2
Christchurch	47	44	44	30	32	39	37	42	44
Dunedin		1	4	4	4	6	8	4	7
Gisborne	3	1	2	2	2	4	3	2	3
Hastings	5	3	5	4	5	5	5	4	6
Invercargill	2	2	3	3	3	3	3	3	3
Nelson		1	1	1	1	1	1	3	3
Palmerston North	4	6	5	5	4	7	5	9	7
Rotorua	4	3	4	4	3	2	4	4	4
Tahiti	3	5	5	4	5	5	5	5	5
Taranaki	4	4	4	5	4	5	5	7	5
Tauranga	2	2	4	5	5	6	5	6	4
Waikato	11	8	7	10	6	7	7	7	7
Wellington	5	6	6	6	7	6	6	7	6
Whakatane	2	1	1	1	1	1	2	3	4
Whangarei	5	3	7	5	4	6	5	8	8
Total	97	91	103	90	88	106	102	115	118

Table 5. Outreach Clinics (Paediatric and Adult Congenital)

The service places considerable emphasis on maintaining children in their home regions. Although there are no paediatric cardiologists resident outside Auckland there are a number of paediatricians and cardiologists with subspecialty skills in this area who provide high quality surveillance for cardiac children. There is close liaison between these clinicians and the Green Lane Paediatric and Congenital Cardiac Service. The degree of support is considerable and involves telephone consultation and frequent review of echocardiograms, electrocardiograms and other cardiac investigations. Consultation of this nature that resulted in written response are summarised below (Figure 9). These consultations, together with the peripheral clinics form the basis of an informal clinical network.

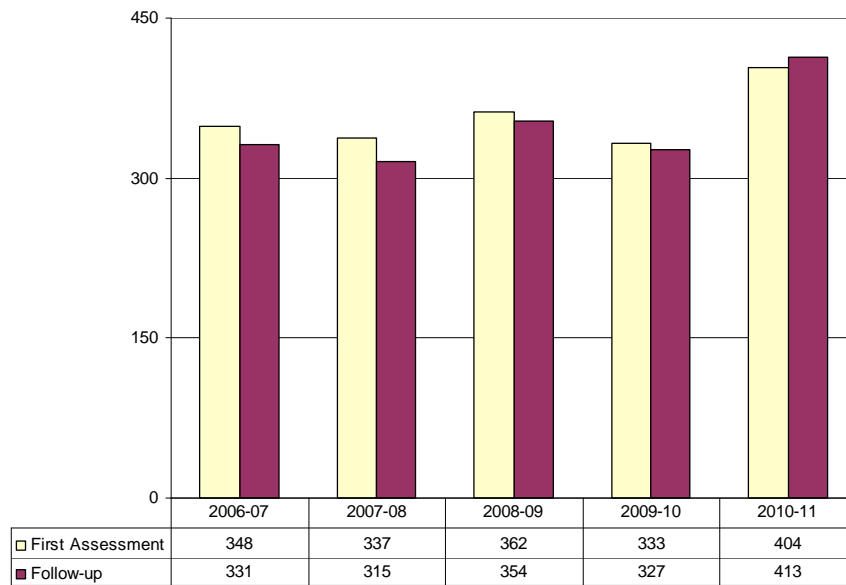


Figure 9. "Virtual" Consultations

3.6. FETAL CARDIOLOGY

Patients are referred from throughout the country for assessment in Auckland, often following diagnosis elsewhere. There are two clinics per week in conjunction with the high risk obstetric service. Fetal cardiology referral patterns changed in 2006 following a decision to restrict referrals to those with suspected fetal heart disease. This decision recognises the expertise of the tertiary obstetric scanning team at Auckland City Hospital. (Figures 10 and 11). In 2009 Dr John Wright joined the fetal cardiology group. A centrally funded National Fetal Maternal Medicine Network was established in 2010, ensuring better coordination of care through the regions. Referral numbers have increased as a consequence of increased rates of fetal diagnosis and more intensive follow-up.

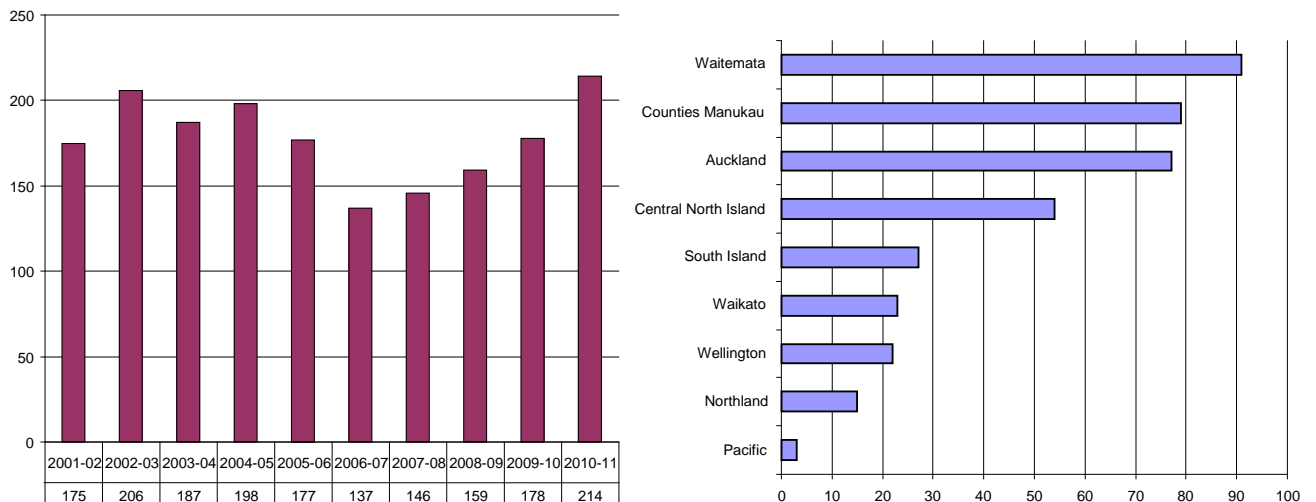


Figure 10. Auckland Fetal Cardiology Clinic volumes (left) and domicile (right)

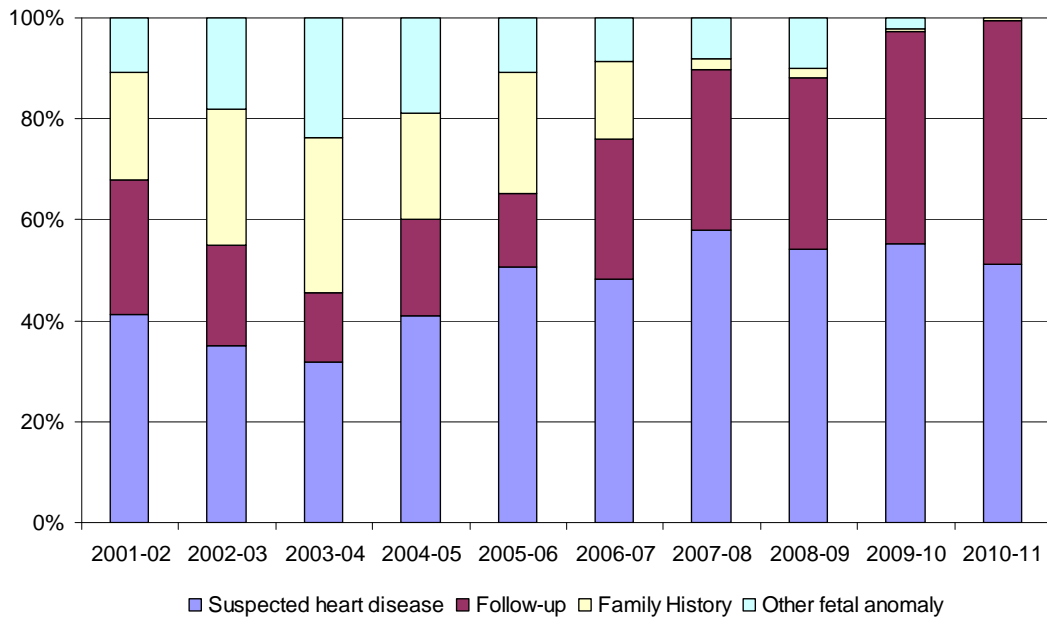


Figure 11. Indications for fetal cardiology outpatient referrals

Distant consultations are provided via digital link and videoconferencing is used for counselling. Figure 12 details those in whom a consultation was undertaken at a distance via review of a scan from another institution and/or counselling via telephone or video link.

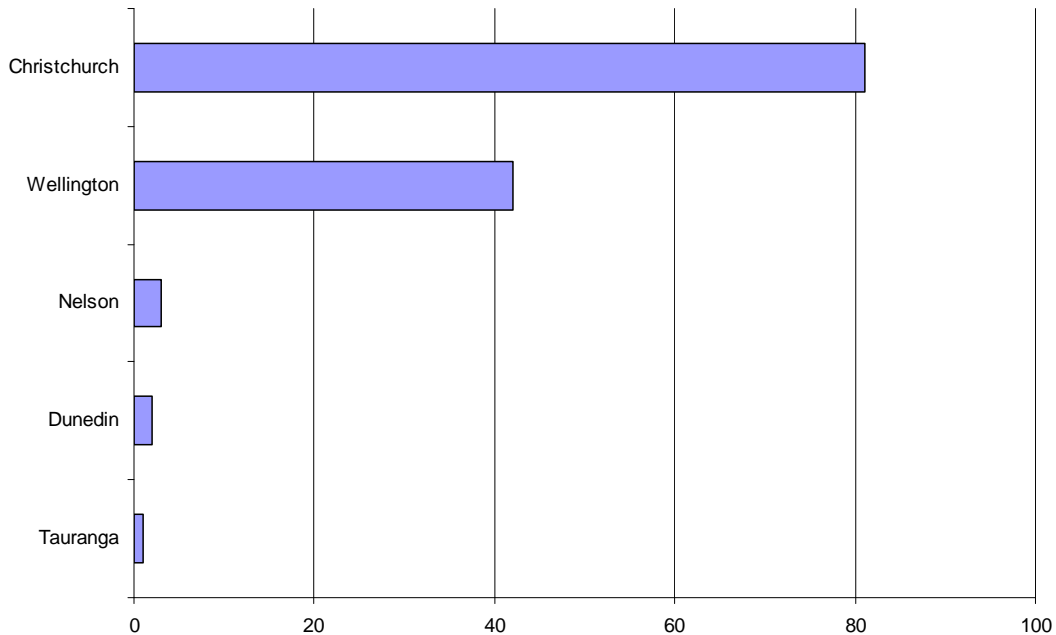


Figure 12. Domicile of fetal "distant consultation" July 2009 – 2011

3.7. ADULT CONGENITAL HEART DISEASE SERVICE

The Adult Congenital Heart Disease (ACHD) Service has continued to develop in response to the needs of a rapidly expanding population. The cardiology team comprises four cardiologists (Drs Tim Hornung and Clare O'Donnell from the Paediatric and Congenital Cardiac Service, and Drs Ivor Gerber and Boris Lowe from the

Adult Cardiac Service), and an ACHD Nurse Practitioner (Annette Rief), who works closely with the Medical Paediatric and Congenital Cardiac Team. Dr Boris Lowe joined the service in 2009, after completing an ACHD fellowship at McGill University, in Montreal. The ACHD cardiologists combine the mix of paediatric and adult cardiology backgrounds to increase the overall standard of care. In addition, an adult cardiology registrar rotated to the Paediatric and Congenital Cardiac Service team spends much of their attachment gaining experience with the ACHD team. The service also has access to a health psychologist, Dr Sue Murray. She joined the service in 2010 and is currently employed for 0.3 FTE.

Cardiac surgeons from the Paediatric and Congenital Cardiac Service undertake surgical procedures on these patients, usually in the adult cardiology operating theatres. Patients convalesce in the Cardiovascular Intensive Care Unit and the Cardiothoracic Ward at Auckland City Hospital. Medical in-patients over the age of 15 are accommodated in the Cardiology Ward at Auckland City Hospital.

There are three outpatient clinics each week. Two of these (one each at the Green Lane Clinical Centre and Starship Children's Hospital) are staffed with consultant cardiologists, the Nurse Practitioner and the ACHD registrar. Up to 12 patients are seen at each clinic. The third is a nurse-led clinic where up to three patients are seen by the Nurse Practitioner. The Health Psychologist attends the clinics and also sees patients on Monday mornings. There are also four Arrhythmia Clinics per year, staffed by a paediatric electrophysiologist (Dr Jon Skinner) and the ACHD consultants, and there are two to three transition clinics at Starship Children's Hospital. The latter are designed to introduce teenagers to the ACHD team and to explain the transition from parent-directed to individual-centred care.

The Starship Hospital clinics and the nurse-led clinic were established in September 2009 to meet the steadily increasing demand primarily from younger patients exiting the paediatric age group. Approximately 120 patients transfer from the paediatric to the adult service each year, and despite the additional clinics there is an increasing waiting time for routine assessment that is now up to five months for routine appointments (Figure 13).

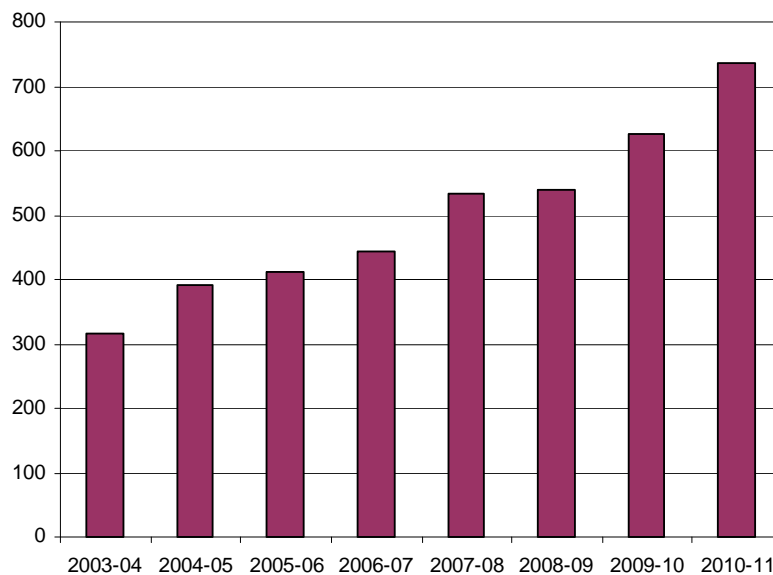


Figure 13. Auckland ACHD Outpatient Volumes

There is an expanding network of Outreach Clinics (Table 6). It is fortunate that there are cardiologists with an interest in congenital heart disease in a number of centres around the country. Close liaison is maintained to assist with evaluation and care of adult congenital patients, with transfer to Auckland as necessary for assessment or treatment. Since 2009 a further two Whangarei clinics and one outreach clinic in Kaitiaki per year have been established. A clinic is shortly to commence in Nelson and hopefully, within the next year in Dunedin, Invercargill and Hawke's Bay. Further development of this network is essential to advance the service and ensure equity of access for patients throughout the country.

Region	2007-2009	2009-2011
Palmerston North	1	1
Taranaki	2	2
Tauranga	1	1
Waikato	2	2
Wellington	1	1
Whangarei	1	3
Kaitaia	0	1
Total	8	11

Table 6. ACHD Outreach Clinics

The ACHD service is frequently involved in consultation with regard to cardiac and non-cardiac medical issues of patients in New Zealand and the Pacific. As most of these individuals had their early care and surgery at Green Lane Hospital, the original surgical and periprocedural documentation is usually available to assist consultation and review.

There is close collaboration with the Paediatric and Congenital Cardiac Service and its subspecialties including imaging, interventional cardiac catheterisation and electrophysiology and with the Adult Cardiovascular Service, including cardiac transplantation. Multidisciplinary involvement including such groups as cardiac anaesthesia, obstetrics and women's health services, as well as dental and general medicine is often required for these complex patients and strong collaborative links have been established with other departments throughout the hospital.

3.7.1. Current Volumes

Inpatient volumes have shown an increase, both in terms of bed days and patient numbers, as have outpatient numbers. (Figure 14). Approximately 120 patients transfer from the paediatric to the adult congenital service each year. Outpatient numbers have been constrained by clinic capacity and there is a clear need for additional consultation time in the Auckland clinics and through the outreach service.

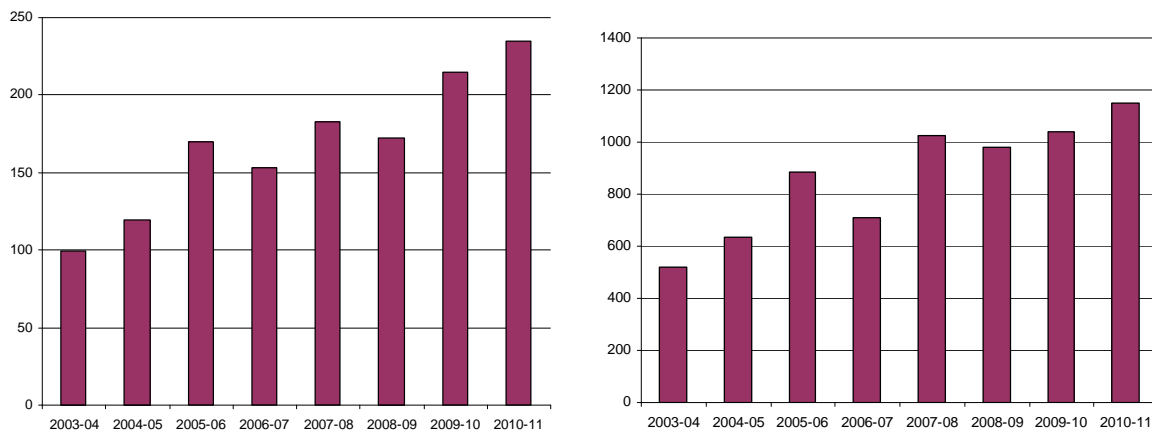


Figure 14. Inpatient numbers (left) and bed-days (right)

3.7.2. Education

The team provides study days, group sessions and individual teaching sessions for nurses in the adult cardiology and cardiac surgical services. Annette Rief facilitates a biannual 8-hour study day to assist staff from these and other departments in the hospital in caring for these complex patients. She also organises resource nurse development in the adult cardiothoracic wards and regular staff teaching on an ongoing basis. Educational material continues to be developed and is available for staff including the now complete ACHD workbook. Policies and recommended best practices related to ACHD patients in the hospital are updated as able.

3.7.3. New initiatives

1. Nurse Practitioner role

Due to increased clinic workload Annette Rief has been seeing a full patient load at both outpatient clinics as well as in her nurse-led clinic, which includes seeing pregnant ACHD patients, who usually attend their obstetric medicine appointments that day. She also travels to several outreach clinics and in most cases is the first point of contact for the national patient population. On average she receives around 50 – 60 referrals per month directly from patients and increasingly from GPs and other Health Professionals; asking for advice regarding ACHD care. The nurse practitioner's busy clinical workload has limited her care coordination and patient education roles. A further business case for additional nursing FTE is envisaged to re-establish this important service.

2. Resource Nurse Development

Due to the busy clinical workload of the Nurse Practitioner the training on the adult cardiac surgical wards (Ward 42/46) has been curtailed and no longer involves regular meetings with education sessions and day-to-day support. Dedicated part-time ACHD resource nurse roles would be helpful to overcome this gap in the future. Christchurch Hospital has successfully employed a part time ACHD resource nurse to work with Dr Lainchbury throughout the year at the Christchurch ACHD clinics, which now requires very little support for the role development from this end. The Tauranga Hospital ACHD service has also developed an ACHD Specialist nurse who works closely with the Cardiac Nurse Practitioner and the ACHD outreach cardiologist. There is nursing interest at Whangarei Hospital for a similar role but funding has not yet been established.

3. Outreach Transition clinics

One outreach transition clinic is held in Christchurch each year and is run by the Auckland ACHD team with local cardiologists and nursing staff in attendance.

4. Educational resources for patients

The "Health Navigator" website for patients and Primary Health Care Providers remains available. For more specific patient education patients are directed to www.pted.org, an American website dedicated to congenital heart patient education.

5. Patient support groups

Heart Children New Zealand employs a national teenage and young adult coordinator, based in Auckland who provides support nationally. Development of a national support network linked to the existing branches is in progress.

6. Out of hours on-call service

Further consideration is being given to establishing an out of hours ACHD cardiologist consultation service with the appointment of a fourth cardiologist.

3.8. CARDIAC INHERITED DISEASE GROUP (CIDG)

The Cardiac Inherited Disease Group (CIDG) is led by Honorary Associate Professor Jon Skinner and includes a salaried 0.8FTE co-ordinator (Jackie Crawford), and 0.8FTE team support. Salary support from Cure Kids is provided 20% for Dr Skinner. The group also includes a number of cardiologists, pathologists and molecular and clinical geneticists throughout the country, and meets by teleconference at a national level bimonthly with a weekly meeting in Auckland.

Long QT Syndrome

The laboratory diagnostics for Long QT Syndrome and Brugada Syndrome are performed at Auckland District Health Board (Lab Plus). Typical turn around for diagnostic screening of LQT genes 1,2,3,5,6,7 is three months, and for family cascade, point mutation screening, about six weeks. New referrals come mostly from adult cardiology services around New Zealand, and from the national forensic and coronial services for the investigation of sudden unexpected death in young people.

Other cardiac inherited disease

A hypertrophic cardiomyopathy (HCM) genetic counselling clinic run by Dr Jim Stewart (recently transferred to Dr Warren Smith) and Jenny Eaton from the clinical genetics department, continues to expand. Samples are sent to Denmark for genetic testing. An inherited arrhythmic syndromes clinic is held monthly, alternately by Jon Skinner and Warren Smith (Adult Cardiology) and Ian Hayes or Jenny Eaton (Clinical Genetics).

CIDGNZ registry

The CIDG database (funded by Cure Kids) is in regular use as a clinical tool for the national consent-based registry. There are 901 individuals, who with informed consent, have their relevant clinical records and genetic material stored with CIDGNZ registry. CIDGNZ has overseen more than 1500 genetic tests for inherited heart disease, mostly Long QT, Brugada and HCM. The registry includes 218 families investigated for Long QT syndrome, 139 with hypertrophic Cardiomyopathy, 40 with arrhythmogenic right ventricular cardiomyopathy, 24 with Brugada Syndrome and 10 with CPVT. (Figures 15 - 16).

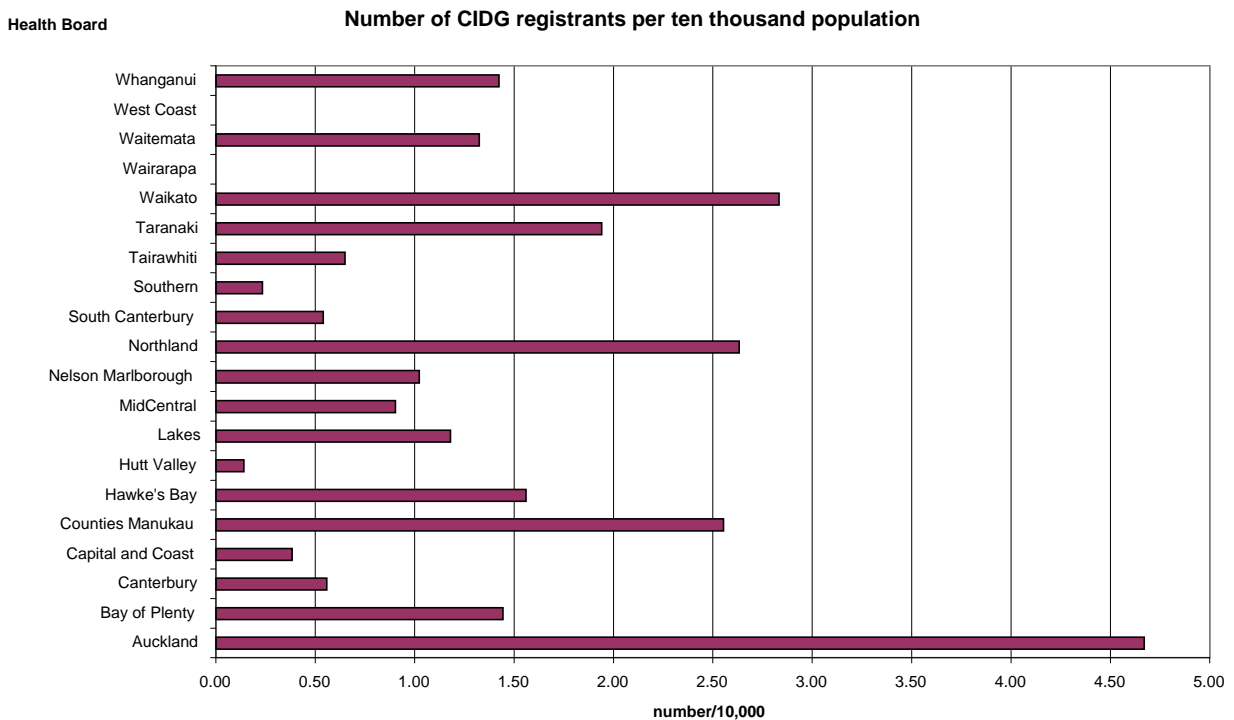


Figure 15. CIDGNZ, consenting registrants by district health board to date. Cases are referred from the whole of New Zealand, but the southern part of the North Island and the South Island refer fewer cases

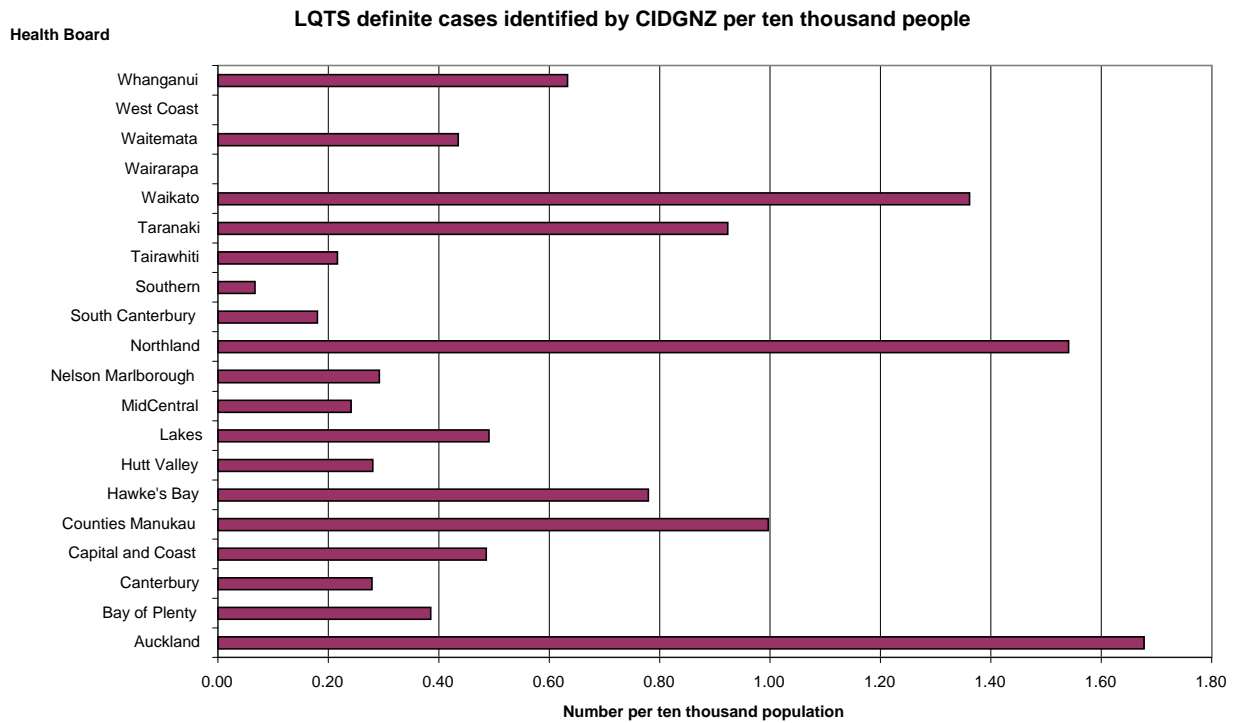


Figure 16. The expected prevalence of Long QT syndrome is 4 per ten thousand. This figure illustrates definite long QT cases, most of which are genotype positive, and a smaller number are clinically definite cases where the genotype has thus far been elusive. For the north part of the North Island the discovered prevalence is over 1 per ten thousand. It is much lower elsewhere though some cases have been identified in the Capital and Coast region independently by the Wellington based clinical genetic service

Investigation of sudden death in the young

There have been 622 cases of young sudden death reported to CIDG for post-mortem genetic investigation or DNA storage since 2001. The Northern Regional collaboration with forensic pathology spread to cover all of New Zealand four years ago. CIDG provides a two stage investigation process in post mortem negative sudden deaths:

1. Family investigation
2. Genetic tests for Long QT syndrome undertaken using DNA stored at post-mortem

Originally funded by Cure Kids, data from this on-going project resulted in a Ministry of Justice decision to fund posthumous LQT genetic testing from July 2008.

A two year collaboration to investigate Sudden Unexplained Death in Young People (SUDY) involving post-mortem LQTS genetic testing and cardiac screening of family members, achieving an overall diagnostic rate of one third for familial inherited heart disease in 1-40 year olds (see bibliography).

Nationalisation of the service

Repeated efforts to obtain some national financial support from the Ministry of Health have been unsuccessful to date, despite collegial support from the major centres and many smaller ones. Efforts are still ongoing. The growth of the service is partly summarised by the illustrations below, and disparities in regional delivery are also made clear (Figure 17). The CIDG team aim for equality of access for all New Zealanders for this service. However the vast majority of registrants are from the North Island.

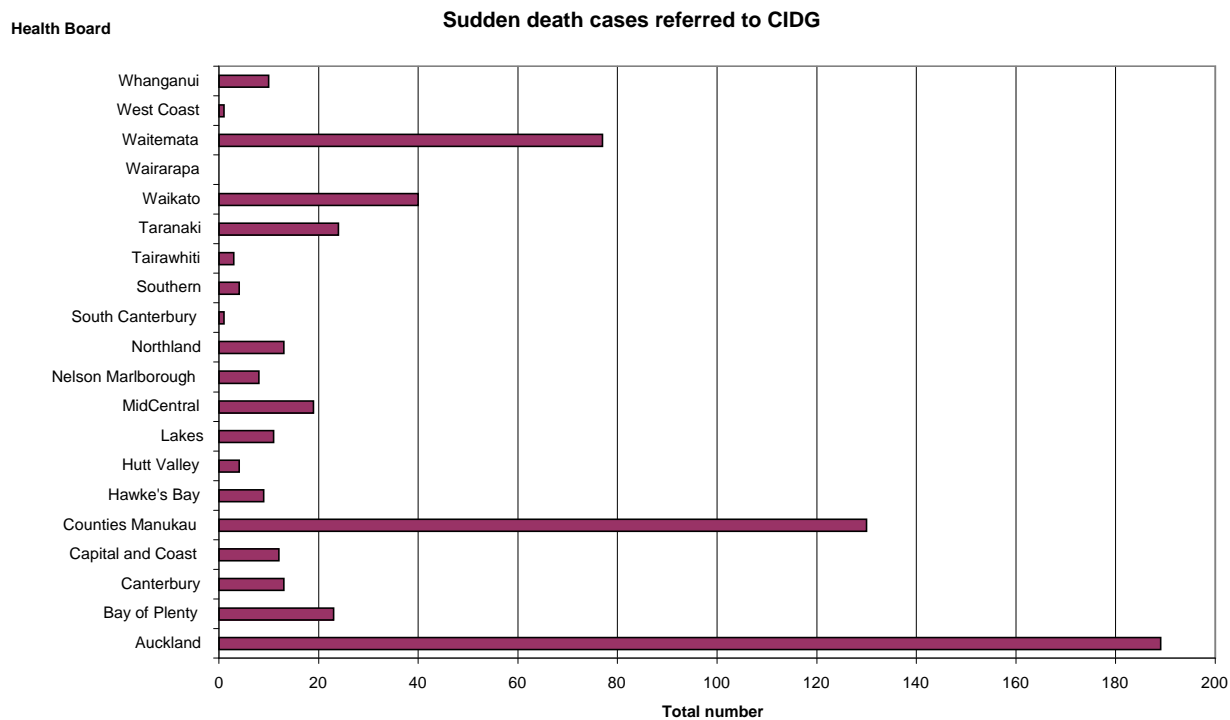


Figure 17. Referral of SUDY cases (0-40 yrs) by region to date. The southern part of the North Island and the South Island refer few cases

Applications for further funding

CIDG applied for development funds from the Ministry of Health through the National Services and Technology Review Process (NSTR) in April 2008 for three liaison nurses and funding for the database development. This process resulted in support for the service, but further support has not been forthcoming.

CIDG research

CIDG has produced nine publications (Bibliography) during these two years in significant journals as a result of national and international collaborations. Dr Skinner continues as chair of the Trans-Tasman SUDY initiative (TRAGADY, recently renamed heart@heart). This group have a three year prospective population based study of SUDY (1-35 years) across Australia and New Zealand funded by the NHMRC. Other international collaborations include Dr Elijah Behr (UK)- sequencing of 50 cardiac genes in SUDY; Dominic Abrams (Harvard), genetic and in vitro evaluation of ARVC; Michael Ackerman (Mayo clinic) deletions and duplications in SIDS and SUDY.

The next two years research will focus on the registry performance to date, interrogation of Long QT gene polymorphisms in coronary disease, reports of the ARVC cohort in New Zealand, and Long QT syndrome in SIDS.

3.9. RHEUMATIC HEART DISEASE

All cardiologists and surgeons are involved with clinical care of children with severe rheumatic heart disease. Clinical activities include:

- Cardiac assessment of children admitted to the Starship Children's Hospital with suspected or confirmed acute rheumatic fever.
- Tertiary opinion for paediatricians and cardiologists managing children with rheumatic heart disease.
- Inpatient management of acute rheumatic fever and severe carditis.

Cardiac surgery was undertaken for chronic and occasionally, acute rheumatic heart disease. Surgical procedures favour valve repair whenever possible and there has been an increasing trend to repair the aortic valve in addition to the mitral valve. From July 2009 to June 2011 there were 79 surgical admissions involving

77 patients aged 3 to 26 years. Twelve were reoperations and there were no early deaths. Double valve surgery was undertaken in 30 and triple in 5. Procedures are detailed in Table 7.

Mitral valve	Repair	50
	Mechanical	12
Aortic Valve	Homograft	13
	Repair	11
Tricuspid valve	Mechanical	8
	Repair	17

Table 7. Rheumatic valve surgical procedures

3.9.1. Future developments and current issues:

The case load is dependent on the local prevalence of rheumatic heart disease, and on the availability of funding for children from the Pacific Islands.

- The number of referrals from the Pacific Islands is dependent on funding and on surgical capacity. Over the past two years there has been significant development in the outreach programme to the Pacific Islands with visiting clinics to Samoa Tonga, Tahiti, and the Cook Islands funded by local governments and/or NZ Aid. An annual clinic to Vanuatu commenced in 2009. These clinics have facilitated communication with local clinicians, and improved follow-up for children who have returned to their homes after cardiac surgery. An ongoing screening programme accompanied by secondary penicillin prophylaxis has the potential to reduce the prevalence of severe rheumatic heart disease. Access to cardiac surgery for Pacific Island peoples is limited by funding issues, excepting for those from the Cook Islands (a New Zealand territory) and Tahiti. The Paediatric and Congenital Cardiac Service case manages children from the Pacific Islands to minimise hospital costs.
- The local prevalence has not been significantly impacted by recent screening projects (see below) as mild cases of RHD are identified with only a few severe cases requiring surgery. Although these projects are likely to identify more cases in the short term, in the longer term penicillin prophylaxis of those with mild disease may reduce the prevalence of more severe disease. Screening is currently a research tool.

3.9.2. Rheumatic Heart Disease Research

Dr Bo Remenyi was the Rheumatic Heart Disease fellow in 2011 and was supervised by Dr Nigel Wilson. Dr Remenyi received a two year research fellowship from the National Heart Foundation of New Zealand, Her projects included:

- International Standardisation of RHD echocardiography – consensus was needed for defining echocardiographic diagnostic criteria when screening for rheumatic heart disease. Dr Remenyi led this project with Drs Nigel Wilson and Jonathan Carapetis with a panel of 20 international investigators. A web-based review of echocardiograms was established in Auckland allowing on-line review of echos. Teleconferences with review of echocardiograms established preliminary definitions with a Kappa agreement of 0.9 in a non-blinded fashion. A workshop was held in Thailand in March 2003 with definitions finalised. A manuscript has been prepared with review of the literature. An inter and intra observer study is now underway with each of the panel reviewing 200 echocardiograms. The project has been endorsed by the World Heart Federation. Dissemination of results with DVDs and an open Web site is planned and will represent the gold standard for RHD diagnosis by echo.
- The RHD research group were involved in further school based RHD screening in the Bay of Plenty (led by John Malcolm) and the Far North (led Roger Tuck and Jonathan Jarman) in 2010. Dr Remenyi supervised and reported the echocardiograms. Drs Wilson, Gentles and Stirling reviewed abnormal scans.
- Dr Remenyi completed a review of long-term outcome following rheumatic heart valve surgery in children that had been started in 2007 by Drs Webb and Wilson. A valuable data base involving 212 patients with over 90% follow-up has been established. Three manuscripts are underway, with one submitted.

Dr Wilson received academic funding for 0.3 FTE for RHD research in 2010 from the Green Lane Research and Education fund and in 2011 from Lottery Health Research Grant.

Future initiatives:

A review of children identified with possible RHD during school screening between 2007 and 2010 from four New Zealand regions is underway. A collaborative case control study to better define risk associated with these types of anomalies is planned in collaboration with Australian centres.

4. INVESTIGATIVE SERVICES

4.1. ECHOCARDIOGRAPHY

The service employs five congenital cardiac sonographers working 4.4 FTE. Megan Burrows stepped down from the role of Charge Sonographer in January 2010 and Fiona Lean was appointed in May 2010. Early 2011 brought challenges with the resignation of one sonographer and another commencing parental leave. We have been unable to recruit into the vacant positions.

Equipment includes 7 cardiac ultrasound systems comprising four Philips iE33's, portable Vivid E and Vivid i systems (purchased with a research grant from the Starship Foundation and used predominantly in the school rheumatic heart disease screening programme) and a Sonosite for use in the catheterisation laboratory. All iE33 systems have live real time 3D transthoracic and transoesophageal capabilities. There are two paediatric, three adult 3D transthoracic probes and two 3D transoesophageal probes. The Starship Foundation has been very generous in their support of paediatric echocardiography.

All inpatient echocardiograms for children and adults with congenital heart disease as well as paediatric outpatient echocardiograms are performed at Starship Children's Hospital. The sonographers staff two Adult Congenital clinics, one held at the Green Lane Clinical Centre and the other at Starship. Also, the sonographers attend a weekly clinic held at Middlemore Hospital. On occasion paediatric cardiac sonographers cover paediatric echocardiography clinics held at other centres including Gisborne, Rotorua, Samoa, Vanuatu and Tonga.

The number of echocardiograms has continued to increase reflecting the growing importance of echocardiography in diagnosis and surveillance and a significant increase in adult congenital and paediatric inpatient and outpatient attendances. (Figures 18 – 20).

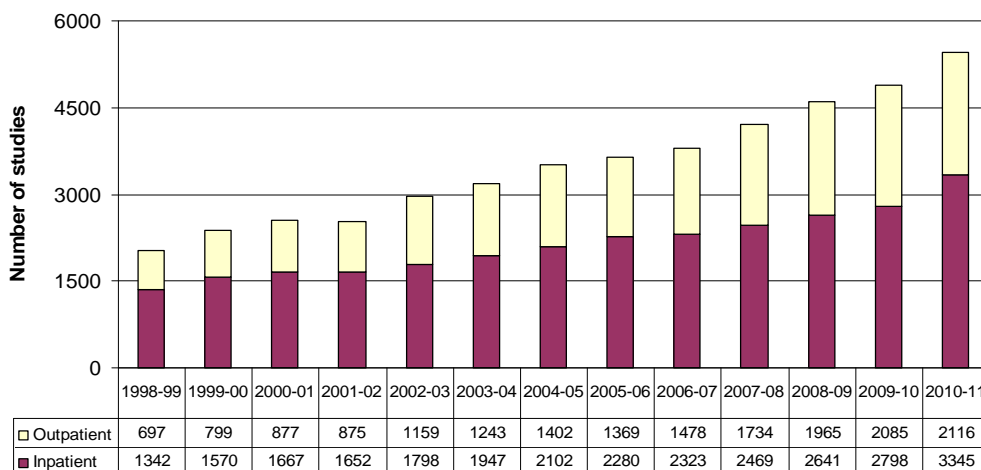


Figure 18. Total number of echocardiograms (paediatric and adult congenital)

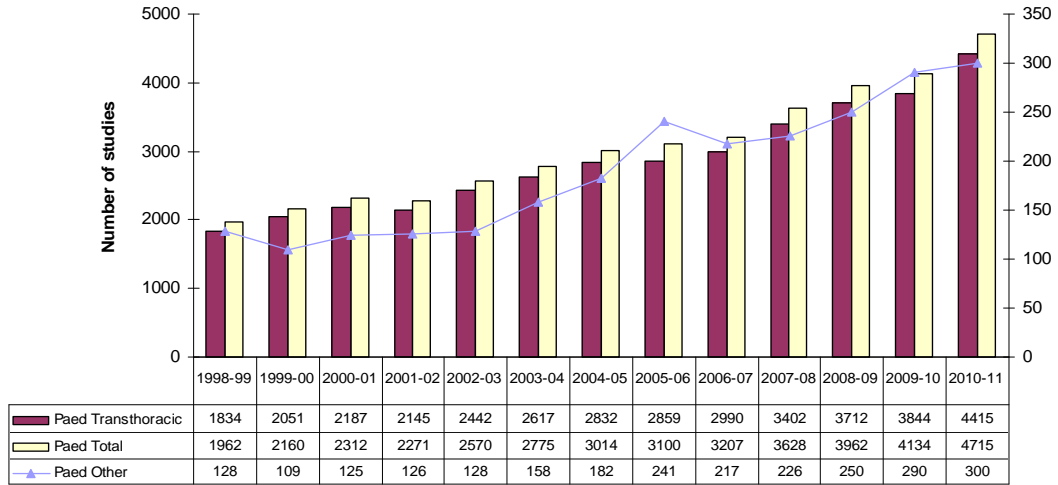


Figure 19. Paediatric echocardiograms: transthoracic and total on left axis and epicardial and transoesophageal ("other") on right axis

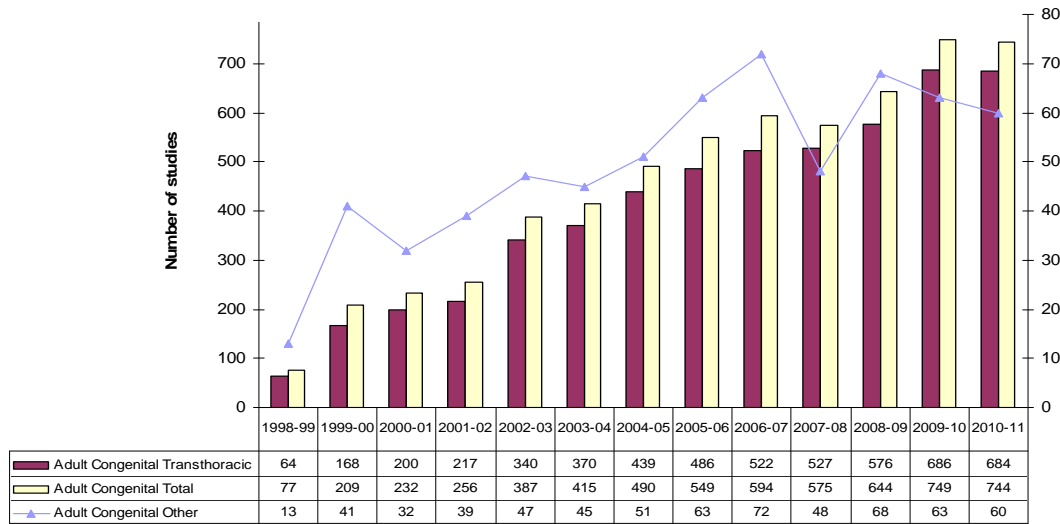


Figure 20. Adult Congenital Heart Disease echocardiograms

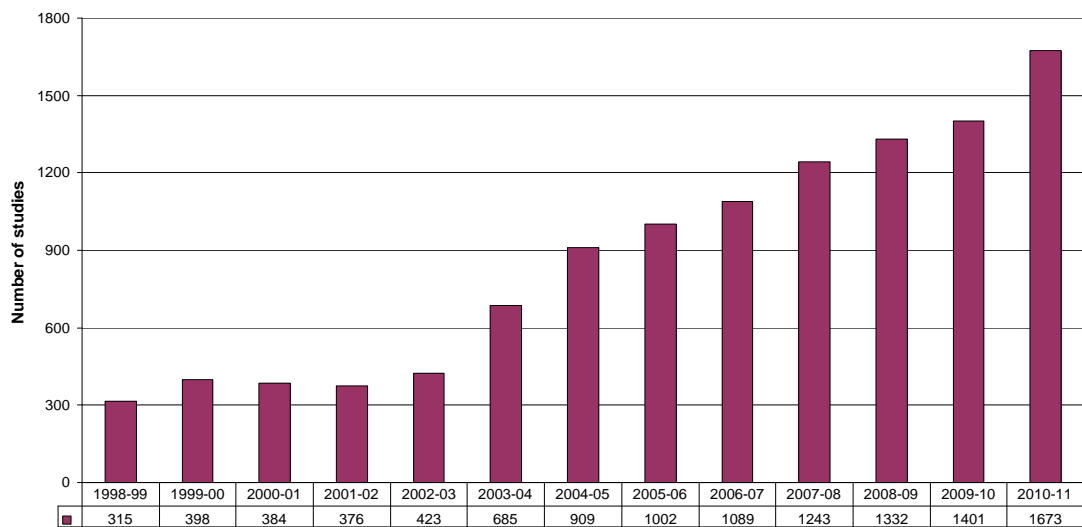


Figure 21. Echocardiograms for other services

The number of echocardiograms undertaken for other services has continued to increase since moving to the Starship Children's Hospital from Green Lane Hospital in late 2003 (Figure 21).

4.2. CARDIAC MRI

The Cardiac MR Service is run by Dr Chris Occleshaw, Dr Tim Hornung, and Dr Boris Lowe. Cardiac MRIs are performed on the 1.5T Avanto and 3T Skyra facilities at the Centre for Advanced MRI (CAMRI) at Auckland University. There was a temporary reduction in scanning capacity when the magnet was moved from Auckland University to Auckland City Hospital, however subsequent to this move there has been increased access to non-GA lists for adults and older children. There have been ongoing problems with access to anaesthesia for younger children and infants, with a resulting long waiting time for these studies. However there are plans to increase the GA capacity from early 2012, which will make for much improved access to cardiac MRI for children.

Congenital cardiac MRI outside of Auckland is supervised by Dr Sharyn MacDonald in Christchurch and Dr Kathy Ferrier at Hutt Hospital. Dr Chris Occleshaw also undertakes a list catering to patients from outside of Auckland.

We will soon be joining a national cardiac MRI registry programme which will facilitate a detailed record of cardiac MRI throughput both in Auckland and elsewhere.

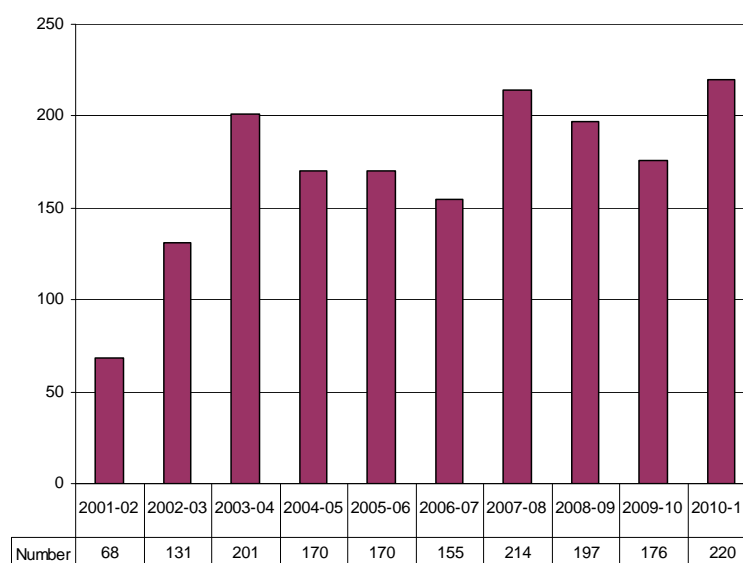


Figure 22. Cardiac MRI in children and adults with congenital cardiac disease

4.3. CARDIAC CATHETERISATION

Paediatric and adult congenital cardiac catheterisation is undertaken in a dedicated catheterisation suite at Starship Children's Hospital under the leadership of Dr Nigel Wilson with Dr Clare O'Donnell in a senior interventional and administrative role. A number of personnel are involved in addition to the catheterising cardiologist including nursing staff, physiology technicians, radiographers, anaesthesia staff and a cardiac radiologist. Beth Tilton leads the nursing staff, successfully introducing, up-skilling and empowering other nurses in the team over the past few years.

Paediatric and adult congenital electrophysiology studies and interventions are undertaken in the electrophysiology laboratory in the adult cardiology catheterisation suite by Dr Jon Skinner and are detailed elsewhere.

The **number of interventional and diagnostic procedures** are detailed in Figure 23.

There has been an increase in the number of high resolution cardiac CTs performed from 2010 and along with a linear increase in MRIs and there was a prediction the number of cardiac caths could fall. Although diagnostic procedure numbers have decreased, interventional procedures have increased at a similar rate.

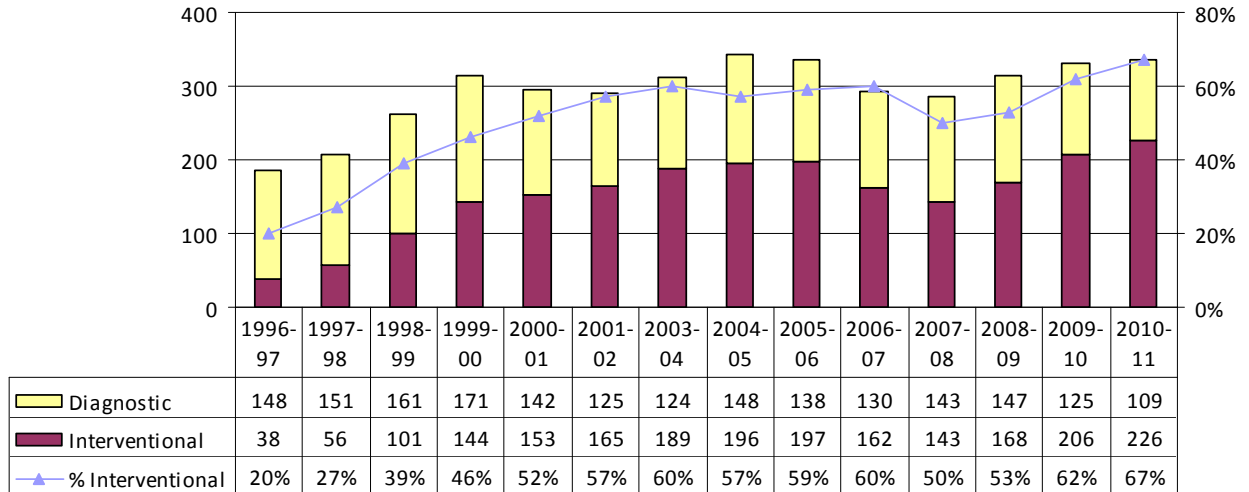


Figure 23. Cardiac catheterisation volumes 1996 to 2011

Cardiologists

There are four paediatric cardiologists who sub-specialise in cardiac catheterisation with the addition of Dr John Wright who joined the team in 2009 from Birmingham Children's Hospital. Dr Peter Ruygrok is also involved with ASD closure in adult patients with assistance from paediatric cardiology staff who provide echocardiography. The case volume by individual cardiologists is detailed in Table 8.

	2009-10	2010-11
Clare O'Donnell	90	109
John Stirling	102	102
John Wright	21	36
Nigel Wilson	87	62
Peter Ruygrok	21	18

Table 8. Cardiac catheterisation procedures by catheterising cardiologist

There were 226 interventional procedures undertaken comprising 67% of all studies. The majority are detailed in Table 9.

Transcatheter pulmonary valve replacement with the Melody valve for adolescents and young adults who have previously undergone pulmonary valve replacement was introduced in 2009. Dr O'Donnell leads this programme which was approved by the Clinical Practice Committee despite a slight cost bias toward cardiac surgery. The surgical team have been very supportive of the programme as most recipients have undergone multiple previous cardiac surgical operations. By end of June 2011 10 patients have received a Melody valve with no significant procedural complications and no early valve failure.

		2007-08	2008-09	2009-10	2010-11
Vessel Occlusion	Coil AP collated	15	26	11	21
	Coronary artery fistula close	2		2	1
	Vein		2	2	2
Angioplasty	Pulmonary artery	16	28	28	29
	Native and Re-coarctation	4	8	6	5
	SVC		1	1	
	Systemic to PA shunt				2
	RV-PA Conduit		3	2	

	Other	1			
Valvuloplasty	Aortic valve	6	3	8	6
	Pulmonary valve	14	11	25	35
	RVOT in Tetralogy of Fallot	1	1	1	
Stent	Pulmonary artery	4	7	4	16
	Native or Re-coarctation	5	4	5	10
	SVC		2		
	Systemic to PA shunt				1
Melody valve implant			5	5	
ASD closure		25	39	44	43
PDA closure		33	38	40	38
Fenestration closure		14	10	14	13
Balloon Atrial Septostomy		21	15	4	11

Table 9. Interventional Procedures

Complications

Cardiologists and fellows are encouraged to record in the log book even the most minor complications. There are two catheterisation morbidity and mortality meetings per year as a protected quality assurance activity. Significant and life threatening complications occur on average in 2-4% of cases (Table 10).

	2008-9	2009-10	2010-11
Major life threatening	2	0	2
Significant	4	9	9
Minor	1	9	13

Table 10. Complications

Death in the cath lab remains a rare event. Analysis of the cath data base for the past 10 years showed four of 3052 cases died with a mortality rate 0.13%, with two deaths related to the catheter procedure; two time-related, but not caused by the catheter procedure.

Complications 2009-10	
Major/life threatening	Pulmonary hypertensive crisis in pt with PPH
Major/life threatening	Post Norwood – low CO
Significant	Adult coarctation: propagating thrombus RFA required surgery
Significant	Embolization of device retrieved by catheter
Complications 2010-11	
Major/life threatening	Neonate cardiac tamponade during RF perforation PV (PA/IVS)
Major/life threatening	Neonate complex Tetralogy with co-morbidity. Unstable pre- and post- PV dilatation, died 3 days later.
Significant	Adult:CVA post coarctation stenting
Significant	Adult: retroperitoneal haematoma
Significant	Embolisation PDA device – successful retrieval and PDA closure
Significant	Fem artery occlusion - neonate
Significant	Pulmonary haemorrhage without sequelae (2)

Table 11. Spectrum of Complications

Echocardiography is frequently performed in the cardiac catheterisation laboratory, either prior for diagnostic information, or during or after an intervention. Overall one third of those undergoing a catheter study have an echocardiogram in the catheterisation laboratory.

Echo exam type	2009-10	2010-11
TOE	61	53
TTE	46	43
Total	107	96

Table 12: Echocardiography in the Cardiac Catheterisation Laboratory

4.4. ELECTROPHYSIOLOGY

Paediatric and congenital electrophysiology utilises a biplane electrophysiology laboratory within the adult cardiac catheterisation suite, and is led by Dr Jon Skinner. Technical staff are drawn from the cardiac physiology department. Complex procedures are done in collaboration with adult electrophysiologist Dr Nigel Lever, and Dr Skinner similarly will assist with some complex adult cases.

Of the 492 ablations over the past 9 years (Figure 24) there have been 39 failures (8%). There was one post-procedure death related to a cerebrovascular accident, and no other major complications.

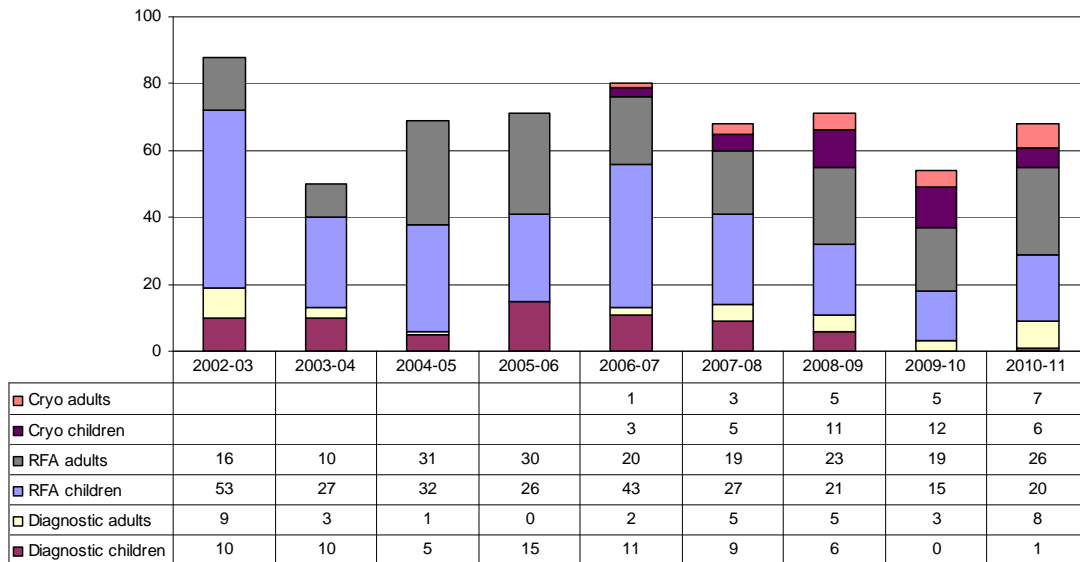


Figure 24. Electrophysiology studies. Numbers were reduced 2003-04 because of move to the new hospital, and in 2009-10 due to Dr Skinner's sabbatical leave

A generous donation from the Starship Foundation allowed purchase of both the Ensite 3D mapping system and cryoablation equipment. 3D mapping has been a major advancement in the treatment of complex cases, particularly post surgical cases, and redo cases. This was used 17 times over the 2008-9 year, and 23 times 2009-10. Cryoablation has reduced the risk of the procedure in young children and young adults with AV node re-entrant tachycardia and in all age groups with parahisian accessory pathways, by avoiding AV node damage and the need for a pacemaker. This combined approach may have contributed to the slightly higher success rate (200 ablations with 12 failures; 6%) over these last four years.

Cryoablation is reported to be associated with a higher recurrence risk than radiofrequency ablation. Of particular interest, 37 patients underwent cryoablation for AVNRT. It was possible to completely abolish slow pathway activity in all but four, where the AV node behaviour was modified. Over a 5 to 28 month follow-up there was only one recurrence, which has been successfully re-ablated. This success rate (97%) is quite remarkable, and certainly better, with a lower recurrence rate, than the same operator achieved in a similar population using radiofrequency ablation for this condition.

4.5. PACEMAKER

Epicardial pacing procedures are performed by the paediatric cardiac surgical team while Drs Lever and Hood place the endocardial systems (older children and adults with congenital heart disease). Supervision and follow-up is provided by Dr Skinner and the cardiac technologists led by Fiona Riddell and Susan Sinclair.

Over 10 years from 2001-2011, 132 epicardial pacemaker devices were placed by the cardiac surgical team. Audit of transvenous systems will be undertaken following improvements in the pacemaker database planned for the New Year.

Staff from the Department of Cardiac Physiology provide technical support during implantation of devices such as permanent pacemakers, implantable loop recorders and implantable defibrillators. Technical staff perform the follow-up checks on these devices within the Auckland, Northland and Hawke's Bay region. They also advise other pacemaker follow-up centres throughout NZ with regard to programming, troubleshooting and planning of further pacemaker surgery.

An important innovation over the last two years has been the advent of remote monitoring, impacting greatly on the cardiac physiologist's work load, both in tracking ICDs and pacemakers.

The number of new pacemaker and ICD implantations for PCCS increased in 2008 and has remained relatively stable since, with a small peak in new pacemakers in 2009-10 related partly to complex post surgical patients (Figure 25).

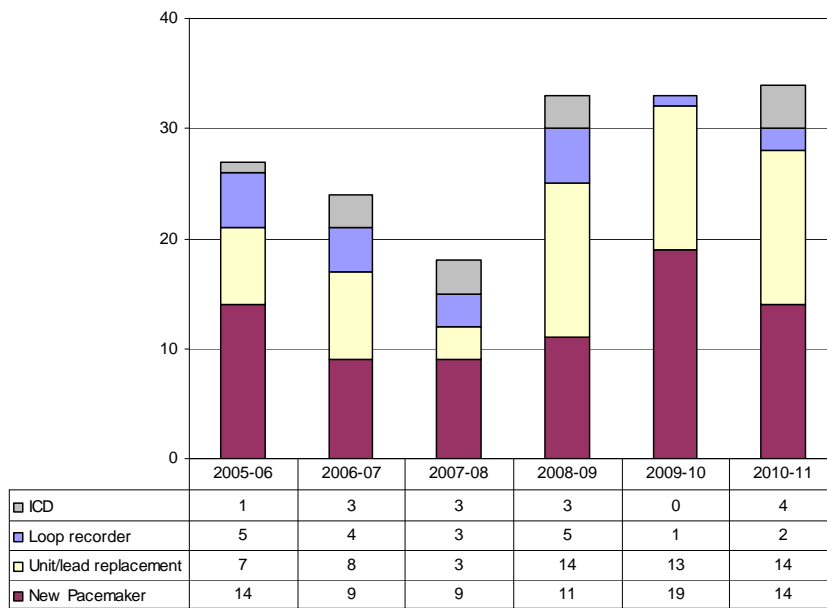


Figure 25. Pacemaker and associated procedures

4.6. CARDIAC REGISTRY

Dr Calder ran teaching sessions for the Cardiology and CTSU Registrars, Fellows and sonographers. These were held annually over nine weeks. These sessions included didactic presentations and practical tutorials, and were 1½ hours duration. The tutorials had a designated cardiologist attend each week to share the teaching with Dr Louise Calder and Chris Occhleshaw. Feedback was extremely positive. The 2011 course will start mid July. There have also been tutorials annually at PICU and Cardiac / Ward 23B Study Days.

There have been a total of 437 organs accessed for teaching and study purposes. There have been two organ returns to families. There have been no organ donations.

The Heart Registry Governance group ensures the organs in the Heart Registry are treated with dignity and respect.

Meetings are held every three months and members include:

- General Manager, Starship Children's and Women's Health
- Parent Representative
- Clinical Director, PCCS (or delegate)
- Chief Advisor, Tikanga ADHB (attends for specific cultural issues)

Ex officio:

- Paediatric and congenital Service Manager
- Cardiac morphologist
- Heart Registry custodian
- Quality Manager (Cardiac)
- Hospital legal counsel (attendance as required)

The HRGG was approached by the Tissue Management Group in 2007 to oversee and provide oversight to Tissue Management and Retention issues within ADHB. HRGG agreed to provide governance. The combined group meet bi-annually and include the following people:

- Director of Surgery, Auckland District Health Board
- Tissue Management Co-ordinator LabPlus
- Nurse Advisor, Operating Rooms
- Quality Department Representative

Report

In 2010 Kay Hyman, General Manager, Women's and Children's Health, commissioned a report on the history of the Green Lane Heart Registry and its role in the evolution of cardiac surgery. This was written by Julie Helean, Manager, Planning and Service Development (funding division). Six members of the HRGG have been meeting regularly during 2011 to complete the report.

5. NURSING

5.1. NURSING LEADERSHIP

The Green Lane Paediatric and Congenital Cardiac Service is a national service based at the Starship Children's Hospital. The Service has a robust nursing infrastructure and nursing leadership is visible and active across the service continuum.

- Stephanie Hlohovsky, Nurse Manager PCCS
- Christine Orchard, Clinical Charge Nurse 23B
- Jane Key Clinical, Charge Nurse 23B
- Elizabeth Tilton, Clinical Specialty Nurse Cardiac Investigations Unit
- Christine Armstrong, Nurse Educator
- Heather Spinetto, Paediatric Cardiac Nurse Specialist
- Julie Stubbs, Paediatric Cardiac Nurse Specialist
- Annette Rief (nee Neugebauer), Nurse Practitioner Adult Congenital Heart
- Ana Kennedy, Nurse Practitioner Acute Care PCCS
- Marion Hamer, Nurse Practitioner Acute Care PCCS

With the diverse number of senior roles the service offers, registered nurses are able to progress into senior roles that are clinical, managerial or educational. This offers a nurse the opportunity to utilise expert knowledge and for some roles practice in an expanded RN scope. These roles make a strong contribution to the multidisciplinary team and the outcomes for children and families who come through the service.

5.2. VISION

"To provide leadership and excellence in care that respects and honours heart children and their families."

Integrity

"We are open, fair honest and transparent in everything we do."

Respect

"We care about and will be responsive to the needs of our diverse people and communities."

Innovation

"We will provide an environment where people can challenge current processes and generate new ways of learning and working."

Effectiveness

"We will apply our learning and resources to achieve better outcomes."

5.3. INPATIENT

Ward 23B has 22 beds, 10 single rooms, two double rooms, one 4-bedded room and a high dependency unit with four beds. The high dependency unit often flexes up to six beds.

Ward 23B is resourced to run at 70% occupancy (16 beds) Monday to Friday and 55% occupancy Saturday and Sunday (12 beds). Average occupancy 2007-2008 was 99.2% of resourced beds and 2008-2009 was 104% of resourced beds (Table 13).

	July	Aug	Sept	Oct	Nov	Dec	Jan	Feb	Mar	April	May	June
2009/2010	102%	96%	107%	109%	109%	103%	84%	100%	107%	90%	105%	104%
2010/2011	100%	118%	108%	106%	89%	100%	109%	110%	112%	113%	109%	100%

Table 13. Ward 23B Occupancy

Ward 23B has a model of care with a strong nurse led focus. In addition to the two nurse practitioners that are responsible for 40% of the inpatient clinical workload there are two clinical charge nurses who manage the clinical operations day to day; manage the daily staffing resource; and are involved in planning, implementing and monitoring operational and clinical process initiatives.

5.4. EDUCATION

Auckland District Health Board has had a well-established Professional Development Programme (clinical pathway) for nurses since 1991. This programme has been based on Benner (1993) *Novice to Expert*. The professional development programme provides a framework, which provides structured support, learning and feedback to assist nurses develop the knowledge and skills necessary to provide safe and effective patient/client care. This programme is a means of recognising and differentiating the skill and competence levels of different nurse clinicians and prepares registered nurses to progress to fill advanced practice roles in patient care delivery and leadership. A nurse may progress through four levels commencing at Novice (level 1) through to Expert (level 4). Level 3 and 4 are currently recognised and linked to a pay incentive and while strongly encouraged are not mandatory.

Consistent with our vision of providing ongoing clinical support and education at the bedside, nursing staff attend weekly education sessions every Wednesday at 1500. In addition to two days of cardiac nursing specific education, nursing staff each attended 3-5 days of ongoing education offered through Learning and Development at ADHB.

A pathway for postgraduate study specialising in Paediatric Cardiac Nursing was developed in partnership with Auckland University.

Senior Nurses from PCCS have been involved with providing education to other clinical areas within ADHB as well as for The University of Auckland, Auckland University of Technology and Massey University.

5.5. NURSING LED INITIATIVES

Neurodevelopment Follow Up For High Risk Cardiac Surgical Patients

The Neurodevelopmental Follow-up Programme started in January 2008 and is coordinated by Ana Kennedy, NP. The goals for this programme are three fold; to provide a service to families so children receive timely intervention; to have accurate local data for the purposes of audit; and to benchmark our cardiac surgical outcomes against international standards.

The criteria for referral to a psychologist for neurodevelopmental assessment at age 2 and 4 years are:

- 1) all neonatal bypass surgery
- 2) infants and children who had aortic arch surgery
- 3) circulatory arrest or regional cerebral perfusion
- 4) pre and or post-operative significant low output and or arrest; need for mechanical support post-operatively, 5) prolonged intensive care stay
- 5) late postoperative signs of neurodevelopmental delay or abnormalities

Neurodevelopment referrals and outcomes for 2009-2011 are as follows. There have been 93 Auckland children under 5-years who met the criteria for referral, 36 have been assessed at two or 4-years, five did not come for follow up, nine died before they could be assessed, three became palliative prior to assessment, four are waiting for appointments and 36 are too young for assessment. Of those assessed, one had severe disability, nine had moderate delay and 26 were within normal range on developmental testing.

Day-stay Cardiac Catheterisation Care Pathway

There is an international trend towards increasing numbers of paediatric procedures being performed on a day-stay basis. Corresponding with this shift in practice, the sole New Zealand provider of Paediatric Cardiac services implemented a pathway to undertake cardiac catheterisation utilising a day-stay approach in select patients. This has been coordinated by Elizabeth Tilton, RN.

A significant increase in the day-stay rates from 2010 to 2011, from 2.9% to 22.6% (p-value 0.000) was reported. A significant decrease in length of hospital stay was identified between 2010 and 2011 (p-value 0.001). In the 2011 sample there were twenty-one day-stay paediatric cardiac catheterisation patients. The mean length of stay for the day-stay patients was 25.49 hours less than those treated as inpatients (p-value 0.000). No patients were re-admitted post discharge on the same day as their cardiac catheterisation. The parental impact questionnaire response rate was 47%. Study participants reported a high level of satisfaction with the day-stay approach, 89% identified they would prefer discharge on the same day as their child's cardiac catheterisation as opposed to overnight hospital stay.

Shaken Baby Prevention Programme

Recognising the large number of neonates admitted through our service and the stress associated with having an unwell infant that may be a poor feeder or difficult to soothe; PCCS volunteered to be part of the shaken baby pilot programme.

In December 2009 Child, Youth and Family invested funds over two years into supporting the Auckland District Health Board's 'preventing shaken baby syndrome programme'. The pilot programme commenced in January 2010 and was initially set up as a two year pilot. The main purpose of the pilot was to create a sustainable programme that educates caregivers of all newborns on how to cope with a crying baby, and the dangers of shaking a baby. The ADHB preventing shaken baby syndrome programme is based on American paediatrician Mark Dias' model, which has shown to reduce Shaken Baby Syndrome rates by almost 50 percent.

Home Monitoring Programme

This programme was established in 2006 and modified in February 2007 to monitor children with single ventricle anatomy that have shunt dependant cardiac conditions between stage one and stage two repair (approximately 0 – 3 months). The objective of the programme is to improve survival for these babies and to detect physiological variations over the period between the Stage 1 and Bi-directional Glenn operations. At home, parents monitor the daily weight (for dehydration) and weekly weight gain and oxygen saturations of the baby. Using this information, parents and local health personnel are able to identify early warning signs of

deteriorating condition and activate early intervention. The programme is coordinated by the clinical nurse specialists.

In the period 2009-2011, 38 patients were on the home monitoring programme. Of these, 20 had the Norwood procedure.

An audit on feeding fragile babies on the home monitoring programme has been carried out which has identified some issues, in particular with infants that are NG fed. A feeding algorithm is being developed with the aim of identifying these issues and prompting early intervention.

Stress in Cardiac Families Study

Recognising a congenital heart diagnosis in a baby is stressful for families. We wished to evaluate what factors impact on stress levels of families and on their perception of the support they received.

A prospective study of 100 babies, 0 – 12 months old, newly diagnosed with a congenital heart defect on first admission to a tertiary children's heart unit was completed. All families were New Zealand residents fluent in the English language. Clinical outcomes were projected for each child and categorised as; no surgery long life, no surgery palliative, single operation repair, multiple operations repair, surgical palliation long term outcome, surgical palliation short term outcome.

The results achieved concur with international data. Most cardiac families experience high levels of stress. Family stress levels do not equate to the child's severity of illness or projected clinical outcome. Staff need to remain alert to levels of stress of all families, however, particular attention should be paid to low income families who may require extra assistance in managing high stress levels. This has been coordinated by Heather Spinetto RN.

5.6. CLINICAL EFFECTIVENESS

Nurse Led Quality Initiatives

Review and updating of current PCCS guidelines and protocols and the development of a secure site for PCCS guidelines on the intranet under Starship Clinical Guidelines - coordinated by M Hamer.

New Guidelines

CT MRI non acute booking guideline
Skin prep for cardiac surgery
ACE inhibitor guideline

New Initiatives

Development of a preparation for cardiac surgery resources for older children
Tracking Outliers folder
23B specific nursing competencies developed
Norwood feeding algorithm- draft algorithm completed
Cardiac Surgical pathway - five day admission pilot completed
Cardiac Catheter Pathway piloted and now in use

5.7. FETAL SERVICE

Two paediatric cardiac service Nurse Specialists are present at diagnosis and involved in consultation and follow up of confirmed complex cardiac cases.

PCCS Nurse Specialist discussions with families are recorded on a designed template and Nurse Specialist follow-up clinic appointments are being organised at each midwifery visit.

The fetal database has been combined and updated.

A family information pamphlet has been reviewed and updated and incorporated in the information pack given to the families.

Nurse Specialists also advise the obstetrician of the delivery and diagnosis of cardiac babies being born out of their local area.

6. ACADEMIC BIBLIOGRAPHY

6.1. PEER REVIEWED ARTICLES

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26. **O'Donnell CP**, Ruygrok PN, Whyte K and **Wilson NJ**. Progressive Pulmonary Hypertension Post Atrial Septal Defect Closure – Early Symptomatic Improvement may not predict Outcome. *Heart Lung and Circulation* 19(12):713-716 Dec 2010.
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6.2. BOOK CHAPTERS

1. Chung S-K, **Skinner JR**, Rees MI. "Molecular Genetics of Arrhythmias" *In: Clinical Cardiovascular Genetics: Principles and Practice*. Ed Kumar D. Oxford University Press, England. 2010.

6.3. INVITED PRESENTATIONS

1. **Calder AL**. A comparison of aortic size and histological findings in cases with bicuspid aortic valves compared to tricuspid or unicuspid ones. 3rd congress of Asia-Pacific Pediatric Cardiac Society (APPCS) Tokyo, July 2010.
2. **Calder AL**. A few examples of congenital heart defects and Cardiac embryology. Neonatal Nursing Science II paper. 17 March 2010.
3. **Calder AL**. Cardiac malformations in Trisomy 13 and Trisomy 18. APPCS 2010.
4. **Calder AL**. Cardiac morphology (talks, demonstrations, hands-on sessions). PCCS: Care of the Congenital Cardiac Child. 9 October, 2009.
5. **Calder AL**. History of Paediatric Cardiology and Prevalence of congenital heart defects. PICU course day 6 April 2011.
6. **Gentles TL**. How Echocardiography Assists in the Timing of Surgery and Suitability for Mitral Valve Repair. 5th World Congress of Paediatric Cardiology and Cardiac Surgery, Cairns, Australia, June 2009.
7. **Gentles TL**. 3-dimensional echocardiography and the mitral valve. Congenital Imaging Symposium, Cardiac Society of Australia and New Zealand Annual Scientific Meeting. Perth, August 2011.
8. **Gentles TL**. Identifying the at Risk Neonate. 5th World Congress of Paediatric Cardiology and Cardiac Surgery, Cairns, Australia, June 2009.
9. **Gentles TL**. Multimodality Imaging. Congenital Imaging Symposium, Cardiac Society of Australia and New Zealand Annual Scientific Meeting. Perth, August 2011.
10. **Gentles TL**. Rheumatic Heart Disease. Is it or Isn't it? American Society of Echocardiography Annual Scientific Meeting. San Diego, June 2010.
11. **Gentles TL**. Stress, strain and hypertrophy. Cardiac Society of Australia and New Zealand Annual Scientific Meeting. Adelaide, August 2010.

12. **Gentles TL.** The role of echocardiography in adult congenital heart disease. Congenital Imaging Symposium, Cardiac Society of Australia and New Zealand Annual Scientific Meeting. Perth, August 2011.
13. **Gentles TL.** Timing and Indications for the Fontan Operation. 5th World Congress of Paediatric Cardiology and Cardiac Surgery, Cairns, Australia, June 2009.
14. **O'Donnell CP.** Asia Pacific Congress of Paediatric and Adult Congenital Cardiology. Invited speaker ACHD and Pulmonary Hypertension Tokyo Japan July 2010.
15. **O'Donnell CP.** NZPAH meeting July 2010 "Update in PAH in congenital heart disease".
16. **Remenyi B.** Cardiac Surgery Best Practice and realities in Pacific Countries. Presented at: Pacific Senior Health Officials Network Rheumatic Heart Disease Policy Partnership Initiative Workshop. 26–29 October 2010 Nukualofa, Tonga.
17. **Remenyi B.** Echo demonstration: live session. Presented at: Pacific Senior Health Officials Network Rheumatic Heart Disease Policy Partnership Initiative Workshop. October 2011 Apia, Western Samoa.
18. **Remenyi B.** Echo Screening RHD Criteria Update. Presented at: Pacific Senior Health Officials Network Rheumatic Heart Disease Policy Partnership Initiative Workshop. October 2011 Apia, Western Samoa.
19. **Remenyi B.** Outcome of RHD Cardiac Surgery Study (NZ) Presented at: Pacific Senior Health Officials Network Rheumatic Heart Disease Policy Partnership Initiative Workshop. October 2011 Apia, Western Samoa.
20. **Remenyi B.** Rheumatic changes on Echo. Presented at: Pacific Senior Health Officials Network Rheumatic Heart Disease Policy Partnership Initiative Workshop. October 2011 Apia, Western Samoa.
21. **Remenyi B.** Rheumatic Heart Disease Echo criteria update. Presented at: Pacific Senior Health Officials Network Rheumatic Heart Disease Policy Partnership Initiative Workshop. 26–29 October 2010 Nukualofa, Tonga.
22. **Remenyi B.** Rheumatic Heart Disease in the Young : an update on management. Presented at: Rural Medicine Australia Workshop. Oct 2011 Alice Springs.
23. **Skinner JR.** "Bicuspid Aortic Valve Disease" Study day in Adult Congenital Heart Disease, Sept 12, Taunton, UK.
24. **Skinner JR.** "Cardiac Inherited Diseases- from the cradle to the grave". New Zealand Pacing and Electrophysiology Group Christchurch Sept 2008.
25. **Skinner JR.** "Cardiac Inherited Diseases: what the adult cardiologist needs to know." CSANZ Wellington June 2009.
26. **Skinner JR.** "Cardiac Sodium channel disease" World Congress Paediatric Cardiology and Cardiac Surgery, Cairns, June 2009.
27. **Skinner JR.** "Cardiac/Genetic Investigation of Sudden Death in the Young". Waikato cardiology and pathology group meeting, Waikato Public Hospital February 19, 2010.
28. **Skinner JR.** "Childhood Brugada: Diagnosis and Therapy". Heart Rhythm Society Meeting May 4, 2011. San Francisco, USA.
29. **Skinner JR.** "ECG masterclass" Heart Rhythm NZ meeting. October 29, Wellington.
30. **Skinner JR.** "Echocardiography on the neonatal unit; Benefits Barriers and Pitfalls", Second Functional echocardiography course (Neonatology), October 19, 2010, Monash Medical Centre, Melbourne.
31. **Skinner JR.** "Epidemiology and genetics of sudden cardiac death". ASEANZ conference. Melbourne. June 4-6, 2010.
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34. **Skinner JR.** "Inherited Heart Diseases" Advanced Cardiac Nurse Training Course. July 18 2011, Auckland University.
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36. **Skinner JR.** "Investigation of Sudden Death in the Young- implications for PICU" Paediatric Intensive Care Unit Staff, Starship Hospital Auckland, March 10, 2010.
37. **Skinner JR.** "Investigation of young sudden death- integration of family investigations" World Congress Paediatric Cardiology and Cardiac Surgery, Cairns June 2009.
38. **Skinner JR.** "Management of infants having LQTS" PACES symposium on "Difficult management issues in the Young Patient Having a channelopathy" May 3, 2011, San Francisco.
39. **Skinner JR.** "Mechanisms of arrhythmia" August 1, 2008. Post Graduate nursing course (730) in paediatric Cardiology and Cardiac Surgery/. Auckland school of nursing.
40. **Skinner JR.** "Pulmonary Hypertension in Children" cardiology/paediatrics CME meeting. September 30, 2010, Mamao Hospital, Papeete.
41. **Skinner JR.** "Sudden Death In the Young- from the lab to the community". Second New Zealand Round Table on Human Genomics (Speaker and panel discussant). The Law Foundation. Wellington. November 24, 2009.
42. **Skinner JR.** "Sudden Death in the Young". Cardiovascular forum. September 8, 2010, Marion Davis Centre, Auckland.
43. **Skinner JR.** "What to do after a near miss-sudden death in intensive care" Paediatric Cardiac Intensive Care Society. Sept 22, 2011. Cambridge. UK.
44. **Skinner JR.** Debate "Mass infant ECG screening is a waste of health service resources" World Congress Paediatric Cardiology and Cardiac Surgery, Cairns June 2009.
45. **Wilson NJ.** Echo Screening for RHD 7th Global Forum on Humanitarian Medicine in Cardiology and Cardiac Surgery, June 2011 .
46. **Wilson NJ.** Cardiology and RHD echo standardization. Workshop Inaugural RHD National Advisory Workshop, Darwin, 2010.
47. **Wilson NJ.** Secondary prevention of Rheumatic Fever 7th Global Forum on Humanitarian Medicine in Cardiology and Cardiac Surgery, June 2011.
48. **Wilson NJ.** Tetralogy of Fallot in the Adult. Catheter Interventional Summit, Istanbul, 30 October 2010.

6.4. ABSTRACTS

1. **Remenyi B, Webb R, Russell P, Lee M, Gentles T, Finucane K, Wilson N.** Long Term Outcomes of Isolated Mitral Valve Surgery for Rheumatic Heart Disease in the Young: New Zealand 1990-2006. Heart, Lung and Circulation. 2010;19:S251.
2. **Remenyi B, Webb R, Russell P, Lee M, Gentles T, Finucane K, Wilson N.** Late Outcomes of Cardiac Surgery for Rheumatic Heart Disease in the Young: The New Zealand Experience 1990-2006. Heart, Lung and Circulation. 2010;19:S250.
3. **Gilbert O, N Wilson.** Early Morbidity of Children Admitted to a cardiac ward with Rheumatic fever. Paed Soc of NZ 24 November 2010.
4. **Webb R, Gentles T, Stirling J, Wilson N.** Echocardiographic findings in NZ children from a low-risk population for ARF: implications for rheumatic heart disease screening. Paed Soc of NZ 24 November 2010.
5. **Bo Remenyi, Rachel Webb, Peter Russell, Kirsten Finucane, Tom Gentles , Nigel Wilson.** Pre-operative risk factors for long-term outcomes of cardiac surgery for Rheumatic Heart Disease in the young: an Oceania cohort. CSANZ Regional Meeting Napier May 2011.

6. **Remenyi B, Webb R, Russell P, Lee M, Gentles T, Finucane K, Wilson N.** Improved Outcomes of Isolated Mitral Valve Surgery for Rheumatic Heart Disease in the Young: World Society of Paediatric and Cardiac Surgery in the Young , Istanbul, June 2011.
7. **Webb R, Gentles T, Stirling J, Lennon D, Trenholme A , Lee M, Wilson N.** Echocardiographic findings in a low risk population for rheumatic heart disease (RHD): implications for screening (Abstract). XVIII Lancefield International Symposium (on-line). Paerlmo, Italy; 2011:http://www.lancefield2011.it/images/stories/pdf/riassunti_definitivo_lancefield.pdf p 283.
8. **Bo Remenyi, Nigel Wilson,** Jonathan Carapetis. International; Standardisation of echocardiographic diagnosis of Rheumatic Heart Disease (RHD). (Abstract) XVIII Lancefield International Symposium (on-line). Paerlmo, Italy. 2011. http://www.lancefield2011.it/images/stories/pdf/riassunti_definitivo_lancefield.pdf p 29.
9. **Remenyi B, Webb R, Russell P, Lee M, Gentles T, Finucane K, Wilson, N.** Late outcomes of Cardiac Surgery for Rheumatic Heart Disease in the young: the New Zealand Experience 1990-2006 Presented at: RACP Congress 2011, Darwin as NZ young investigator.
10. **Remenyi B, Webb R, Russell P, Lee M, Gentles T, Finucane K, Wilson, N.** Long-term outcomes of Cardiac Surgery for Rheumatic Heart Disease in the Young: the New Zealand Experience 1990-2006. Presented at: CSANZ Annual Scientific Meeting 2010, Adelaide.
11. **Remenyi B, Webb R, Russell P, Lee M, Gentles T, Finucane K, Wilson, N.** Long-term outcomes of Cardiac Surgery for Rheumatic Heart Disease in the Young. Presented at the Annual Scientific Meeting of the Paediatric Society of New Zealand 2010 New Plymouth.
12. **Gentles TL, Finucane AK, Mervis J, Wilson NJ.** Outcome of the LV in children and young adults with severe combined aortic and mitral valve regurgitation compared to isolated lesions. American College of Cardiology Annual Scientific Meeting, April 2010.
13. **Gentles TL, Cowan B, Occlshaw C, Beca J, Young AA.** Adverse Remodeling after the Norwood procedure results in progressive ventricular dysfunction. CSANZ Annual Scientific Meeting August 2010.
14. **Gentles TL, Cowan B, Occlshaw C, Beca J, Young AA.** Progressive contractile dysfunction after Norwood procedure compared to biventricular repair and isolated aortopulmonary shunt. AEPC Annual Scientific Meeting, May 2011.

6.5. GRANTS AWARDED

Title:	Investigation of Sudden Cardiac Death in the Young 2009
Principal Investigator:	Ass. Prof Chris Semsarian
Co-investigators:	A/Prof Jon Skinner, Dr Robert Weintraub, Melbourne; Dr Raj Puranik, Sydney; A Prof Jo duFlou, Sydney
Funding Agency:	NHMRC (Australia) – Grant- 632575
Amount:	Aus \$658,500

Title:	RHD Research fellow – 2 years 2009-11
Principal Investigator:	Bo Remenyi
Co-Investigators:	Nigel Wilson
Funding Agency:	National Heart Foundation
Amount:	\$160,000

Title:	Senior fellow 2010
Principal Investigator:	Nigel Wilson
Co-Investigators:	
Funding Agency:	GLREF
Amount:	3/10 th salary

Title:	RHD research
Principal Investigator:	Nigel Wilson
Co-Investigators:	
Funding Agency:	Health Lottery Research Grant – salary support for RHD research 2011-2
Amount:	\$124,200

6.6. COMMITTEES

- **Gentles TL.** Australia and New Zealand Children’s Heart Research Centre (ANZCHRC). Board member.
- **Gentles TL.** Australia and New Zealand Paediatric Cardiac Association. Executive committee member.
- **Gentles TL.** Heart Children New Zealand. Trustee.
- **Skinner JR.** Founder and Chairman of The Trans-Tasman task force the prevention of sudden death in the young – (TRAGADY, since 2005 renamed Heart@Heart 2010).
- **Skinner JR.** Chairman of the Cardiac Inherited Disease Group for New Zealand – (2004 on).
- **Skinner JR.** Committee member of the neonatal section of the CCPU (Certificate in Clinician Performed ultrasound), the Australasian Society for Ultrasound in Medicine (ASUM). April 2008-
- **Skinner JR.** Advisory Board Member- Childrens Research Centre, Starship Childrens Hospital 2009-
- **Wilson N.** Board of GLREF - PCCS trustee (from January 2007).
- **Wilson N.** Heart Registry Governance committee - PCCS - delegate for the Clinical Director (2003-).

6.7. OTHER

Awards and Honours

- **Remenyi, B** Paediatric Society of New Zealand 60th Annual Scientific Meeting - Young Investigator Award 2010.

Courses and workshops convened

- RHD International Echo Standardization workshop Convenor **Remenyi B**, Chaired **Wilson N**.

Current Research Projects

- **Skinner JR.** Audit of quality of autopsy reports in SUDY investigation. Molecular detection of long QT syndrome in SIDS. In vitro evaluation of ARVC (collaboration with St Mary’s Hospital, London). Deletions and duplications in sudden cardiac death genes in SUDY (UK/Australia collaboration) and SIDS (Mayo clinic collaboration). Role of SNPS in Long QT genes in sudden death post myocardial infarction (collaboration with Prof Rob Doughy).
- **Wilson, N.** Screening for rheumatic heart disease in New Zealand.
- **Wilson, N.** Follow up of possible RHD by echocardiography.
- **O’Donnell CP.** Australian and New Zealand Paediatric Pulmonary Hypertension Registry – co-investigator (Lead Dr Weintraub, Melbourne).
- **O’Donnell CP.** Australian and New Zealand Adult Congenital/PAH Registry Co-investigator (lead Drs Weintraub and Prof David Celemajer).
- **O’Donnell CP.** Cardiac MRI During Exercise: Ventricular and Vascular Function – co-investigator (Lead Dr Alistair Young).
- **O’Donnell CP.** Captopril for Infantile Haemangiomas – (Dr Swee Tan lead investigator).

MD student supervisor

- **Wilson N.** FRACP and MD supervisor for Dr Rachel Webb, RHD fellow.

7. MEDICAL STAFF

7.1. CONSULTANT CARDIOLOGISTS

Tom Gentles	Director Paediatric & Congenital Cardiac Service Director, Paediatric Cardiology	Echocardiography Fetal Cardiology Interventional Cardiology
Louise Calder	Paediatric Cardiologist	Cardiac Morphology
Tim Hornung	Co-Team Leader - Adult congenital heart disease	Adult Congenital Heart Disease Cardiac Magnetic Resonance Imaging
Clare O'Donnell	Co-Team Leader Adult congenital heart disease Junior Medical Staff Co-ordinator	Interventional Cardiology Adult Congenital Heart Disease Pulmonary Hypertension
Jon Skinner	Team Leader - Electrophysiology	Invasive and non-invasive electrophysiology Pacing Inherited Cardiac Disease
John Stirling	Paediatric Cardiologist	Interventional Cardiology
Nigel Wilson	Team Leader - Cardiac catheterisation	Interventional cardiology Fetal cardiology
John Wright	Paediatric Cardiologist	Interventional cardiology Fetal cardiology

7.2. CONSULTANT CARDIOTHORACIC SURGEONS

Kirsten Finucane	Director, Paediatric Cardiac Surgery	Paediatric and Congenital Cardiothoracic Surgery
John Artrip		Paediatric and Congenital Cardiothoracic Surgery
Elizabeth Rumball		Paediatric and Congenital Cardiothoracic Surgery

7.3. OTHER CLINICAL STAFF

Paediatric Cardiology Fellows

Danielle Brown
Jonathan Mervis
Kathryn Rice
Hesham Al Saloos
Michael Yeong
Klaus Juul

Cardiac Surgical Fellows

Yuvaraj Davidson
Simon Vari
Steven Thrupp
Anto Sahayaraj
Mark Awori

Paediatric Registrars

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Patrick Yap
Sonja Crone
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Amin Sheikh
Louise Albertella
Smiti Bihari
Silvia Seibt
Anna Tottman
Jan Kelly
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Geetika Badkar
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Marcus Silbery

7.4. CONTRIBUTORS TO THE BIENNIAL REPORT

Cardiac Surgery

Kirsten Finucane

Adult Congenital Heart Disease

Annette Rief

Cardiac Inherited Disease Group

Jon Skinner

Rheumatic Heart Disease

Nigel Wilson

Cardiac MRI

Tim Hornung

Cardiac Catheterisation

Nigel Wilson

Electrophysiology

Jon Skinner

Pacemaker

Jon Skinner / Fiona Riddell

Cardiac Registry

Louise Calder / Christine Orchard

Nursing

Stephanie Hlohovsky

Echocardiography

Sandy Long / Fiona Lean / Tom Gentles

Statistical Assistance

Steve Withy