PAEDIATRICS

# A NATIONAL MODEL OF CARE FOR PAEDIATRIC HEALTHCARE SERVICES IN IRELAND

## CHAPTER 15: PAEDIATRIC CARDIOLOGY

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ROYAL COLLEGE OF PHYSICIANS OF IRELAND

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# 15.0 INTRODUCTION

The national paediatric cardiology service is based in Our Lady's Children's Hospital Crumlin (Crumlin) and has 27 inpatient beds in a new, purpose-built facility known as the Children's Heart Centre. In addition, there are currently eight paediatric intensive care unit (PICU) beds specifically assigned for cardiac patients in the PICU2 with frequent additional admission of cardiac PICU (medical) patients into PICU1. Currently, there are six paediatric cardiologists and three paediatric cardiothoracic surgeons. A seventh paediatric cardiologist with a subspecialty interest in cardiac catheterisation is due to start in July 2015 (Dr. Damien Kenny). There are six clinical nurse specialists whose role is critical to the overall success of the service. There are 51WTE nurses, 8WTE echocardiography technicians and 12 non-consultant hospital doctors (NCHDs) between paediatric cardiology and cardiothoracic surgery. In addition health and social care professionals (HSCPs) have an integral role in the multidisciplinary team (MDT) for cardiology (see Current Service Provision below).

On the 14th of October 2014, a joint policy statement was published by the Minister for Health and Social Services in Northern Ireland, Jim Wells, and the Minister for Health in the Republic of Ireland, Leo Varadkar, referencing the expert International Working Group (IWG) report on the Assessment of Cardiology and Cardiac Surgery for Congenital Heart Disease in Northern Ireland and the Republic of Ireland ( http://health.gov.ie/wp-content/uploads/2014/10/International-Working-Group-Report-North-South-Congenital-Heart-Disease.pdf ). The report made clear and explicit recommendations on the development of a single congenital cardiac service for Northern Ireland and the Republic of Ireland. A total of 14 recommendations were presented which together form the IWG's proposed model for a service that would meet the needs of the entire population with congenital cardiac conditions, whether adult or child, in Northern Ireland and the Republic of Ireland. All paediatriac surgical and cardiac catheterisation cases for the island of Ireland will be performed at Crumlin and there will be a phased increase in capacity at Crumlin to absorb all of the additional volume by April 2016. Currently, approximately 150-160 children with congenital heart disease are referred per year for cardiac surgery in Northern Ireland, and it is anticipated these patients will now be managed and cared for in a shared-care model with medical care being delivered in the Royal Victoria Hospital in Belfast and Crumlin and surgical care at Crumlin. Similarly, all cardiac catheterisation cases that, to date, have been performed in Northern Ireland, will now be undertaken at Crumlin. With the establishment of a cross-border collaborative implementation commission, work has commenced on increasing capacity and resources to accommodate the integration of services from Northern Ireland.

Finally, the cardiology group at Crumlin are on track to transition to electronic medical records across the entire care pathway – from outpatient visit, through to peri-operative PICU admission and transfer to post-operative care in the Children's Heart Centre. Health Insights Technologies Ltd. are at an advanced planning stage of installing MedicaPlus, a hospital and clinical information system that will meet the ongoing and future needs of the department. MedicaPlus is due to go live in mid-2016.

# 15.1 CURRENT SERVICE PROVISION

## 15.1.1 Outpatient Clinics

In 2014 5,660 patients attended the cardiology outpatient service.

*The most common reasons for referral included:*

- request for murmur review (52%),
- family history of heart disease (8%),
- chest pain (7%),
- palpitations (4%),
- dizziness (4%), and
- sickle cell disease (1%).

The other 24% of referrals were not otherwise coded. Between 45-65 new referral requests are processed each week. With regards to the commonest outpatient review request – murmurs - 28% are from general practitioners (GPs) in the Dublin area; 40% were referrals from paediatricians, and approximately 20% were from the Dublin Neonatal Intensive Care Units (NICUs) (see Table 1 in appendix). Of note, there were relatively few referrals from Temple Street Children's University Hospital (Temple Street), Cork University Hospital and University Hospital Galway – programmes that operate their own cardiac non-invasive imaging service.

Of those children with murmurs on the cardiology outpatient waiting list, approximately half were under a year of age, and a significant proportion (37.6%) were from County Dublin (see Graphic 1 in appendix). 48% of murmur referrals are on the cardiology outpatient waiting list for 0-6 months, with 87% waiting less than a year (2014). The total number of all patients waiting to be seen by paediatric cardiology (2014, scheduled and unscheduled) is over 5,000. Including the National Treatment Purchase Fund (NTPF) clinics, 80 new patients were being seen per week in 2014. Since the NTPF clinic was not funded for 2015, an additional public outpatient clinic has commenced on Friday to continue to try and address the issue with waiting times for elective appointments. In addition to all outpatient services in Crumlin, cardiology satellite clinics are run in Temple Street, Tallaght, Galway, Sligo, Letterkenny, Limerick and Cork. Inpatients are also assessed in the three maternity hospitals - the Coombe Women and Infants' University Hospital, the National Maternity Hospital, and the Rotunda Hospital. In terms of investigations, 14,133 echoes were performed and read for the calendar year 2014, 793 Holter monitors were reported, and 9,426 electrocardiograms (ECGs) were recorded.

## 15.1.2 Medical Day Unit

In 2014, there were 629 cardiology day cases booked through the medical day unit. Many patients undergo elective sedated echocardiograms cardiac catheterisation and cardiac magnetic resonance imaging. In the autumn of 2015 a 5-bed, dedicated cardiac day unit will open to better serve the medical and surgical cardiac patients in a more efficient manner, decreasing the impact of internal bed availability (or lack thereof) on patient care. This is seen as a cornerstone to the implementation of our commitments to care for surgical and cardiac catheterisation patients from Northern Ireland. The role of an advanced nurse practitioner is being explored to support the medical day unit.

### 15.1.3 Procedures

For the calendar year 2013-2014, using numbers submitted and verified by National Institute for Cardiovascular Outcomes Research (NICOR) there were 284 cardiac catheterisations performed (an increase of 3.5%) – this makes Crumlin the 7th busiest centre between the UK and Ireland (see table 4 in appendix). This volume is likely to further increase with the hiring of a third interventional cardiologist, the collaboration between Northern Ireland and Crumlin, and the opening of a new state-of-the-art cardiac catheterisation hybrid laboratory in late 2015.

Over the same period, 332 cardiac surgical cases were undertaken, making Crumlin the 6th busiest paediatriac surgical centre between the UK and Ireland (see table 3 in appendix). 248 of those cases were performed on cardiopulmonary bypass. 25% of all surgical procedures were in neonates with a further 45% in children under a year of age. There are 284 cardiac catheterisations per year (two thirds are interventional in nature) and 1500 Holter monitors are assessed per year with 20-30 ablations for Wolff Parkinson White (WPW) syndrome. In excess of 300 cardiac MRI scans are performed annually under the supervision of paediatric cardiology (Dr. Colin McMahon or Dr. Orla Franklin).

### 15.1.4 Theatre Activity

There are nine operating days per fortnight and theatre is supported by cardiac anaesthesia, cardiac-trained nursing staff, a perfusionist team and allied support staff.

### 15.1.5 Antenatal Diagnosis

Prenatal cardiac ECHO clinics with joint cardiology and obstetric consultant attendance are provided at the Coombe Women and Infants Maternity Hospital, the Rotunda Hospital and the National Maternity Hospital. There is an increasing emphasis on pre-natal diagnosis of complex congenital heart disease with local delivery of children predicted to require early postnatal intervention.

### 15.1.6 Newborns with Suspected Congenital Heart Disease

Transport of the sick newborn with congenital heart disease is via the national neonatal transport service with telephone advice prior to transfer.

### 15.1.7 Outreach Clinics

Outreach clinics are one of the great strengths of the service and work best if the local paediatrician is also in attendance when the patient is seen (as in Sligo and Letterkenny). Outreach clinics take place in Cork (monthly), Galway (3 clinics per year), Sligo (3 clinics per year), Limerick (5-6 clinics per year) and Letterkenny (5-6 clinics per year). Dr. Orla Franklin provides clinical support for Temple Street with a weekly outpatient clinic along with inpatient consult commitments.

### 15.1.8 Occupational Therapy

Occupational therapists working with children with cardiac conditions aim to enable the child to be as physically, psychologically and socially independent as possible. In addition, the occupational therapist will support the family in the comprehensive care and development of the child or adolescent presenting with performance concerns in all activities of daily life. Children with complex cardiac heart disease have an increased risk of developmental delay and therefore require close developmental monitoring and input. Children with these risk factors may have difficulties in the areas of fine and gross motor skills, executive function, visual-motor integration and learning difficulties. Occupational therapists are also involved in post-surgical care and treatment which includes splinting, equipment provision and postural management, scar management and pressure care management.

This cohort of patients may require 24-hour postural management systems (sleep systems, specialised seating, and bathing equipment), specialist equipment for participation in activities of daily living (ADLs), assistive technology and splinting. There may also be feeding difficulties related to sensory issues. There is close liaison between hospital and community services in a child's efficient, timely and safe discharge home from the hospital. Community occupational therapists are involved in on-going developmental intervention, rehabilitation, postural management and environmental adaptations. There is no dedicated cardiology occupational therapist in Crumlin, and access is currently limited as it is shared between several other specialties.

### 15.1.9 Psychology

The psychologist's work is divided between acute care, which is predominantly inpatient-based, and chronic care, which is predominantly outpatient-based. In the acute setting the psychologist offers support and advice promoting emotional coping to individual families at the time of diagnosis, through hospitalisation and surgical intervention, to recovery and aftercare. The main aim of psychological intervention is to optimise coping and minimise trauma to maximise quality of life in the context of living with a cardiac condition. Family-centred care is valued and the child patient, siblings and parents may all be the focus of psychological support and intervention.

Psychological interventions designed to reduce distress, increase coping and make sense of the experience can significantly reduce the risk of trauma from surgical treatment and PICU admission. The psychology service have been involved in several quality initiatives that aim to improve timely access to services. The psychologist in paediatric cardiology is also involved in palliative care and bereavement, providing support for families and staff.

The psychologist is developing a protocol to promote the successful transfer of adolescents from child/adolescent cardiac services at Crumlin to adult cardiac services at the Mater Hospital. This will involve both group and individual work to ensure that both patients and parents are psychologically prepared for safe and successful transition between services. This has been identified as a highly problematic area in terms of securing continued health care and maintaining health status. Despite the obvious needs of this adolescent group, the competing priorities of the large number of children undergoing acute procedures for cardiac conditions in Crumlin makes it impossible to provide adequately for this transitional group without increasing psychology staff numbers.

### 15.1.10 Physiotherapy

All children undergoing surgery for cardiac conditions are assessed and treated in PICU on the first postoperative day by the respiratory physiotherapist. This includes weekend on-call services where almost 60% of patients seen are cardiothoracic patients. Once discharged to the ward, children will continue to receive physiotherapy until their chest is clear and they are mobilising independently and freely.

*Physiotherapy intervention can involve:*

- Treatment of areas of collapse or consolidation, clearing retained secretions and facilitating early extubation
- Early mobilisation of older children to treat and prevent respiratory complications and encourage pleural drainage, which is of particular importance to optimise the Fontan circulation
- Modified bronchoalveolar lavage for early detection and treatment of viral or bacterial processes including ventilator-associated pneumonia

Respiratory physiotherapy activity has increased by 35% between 2010 and 2013 on weekdays, and by 15% at weekends.

There is currently no dedicated neurology/neurodevelopment physiotherapist for the cardiology/cardiothoracic service despite the large and expanding caseload.

*Referrals to physiotherapy currently include:*

- Blanket referral for all hypoplastic heart conditions following the Glenn procedure, or earlier if neurological/ developmental concerns have arisen.
- Immediate referral of any cardiac patients regardless of heart condition or stage of surgery that have neurological concerns or asymmetries
- Any cardiac patients with documented hypoxic events or changes on cranial ultrasound or MRI
- Cardiothoracic patients subjected to prolonged PICU or ward stays (longer than three weeks) pre- or post-surgery
- Blanket referral for neurodevelopmental follow-up and rehabilitation for all patients treated on extra-corporeal life support (ECLS)

Patients are reviewed as soon as medically stable in the PICU or earlier if there is a concern regarding neurological sequelae. As per the European Life Support Organisation (ELSO) guidelines, it is recommended that patients treated on ECLS have regular neurodevelopmental and physical re-evaluation for at least five years post-surgery. This target is currently not achieved due to lack of dedicated neurodevelopmental physiotherapists.

### 15.1.11 Medical Social Work

The cardiology Medical Social Work team provide support to families in inpatient and outpatient settings. The cornerstone of intervention is the completion of comprehensive psychosocial assessments with families in order to provide appropriate targeted supportive interventions.

*Medical social workers in this setting prioritise the following areas of intervention:*

- Child protection
  Carry out risk assessments in respect of children's welfare and protection needs in conjunction with the hospital and wider community team to proactively promote the welfare of children.
  In this context social work address mental health, addiction, suicidal ideation and domestic violence among other issues.

- Critical illness
  Offer support and assistance to families in finding ways to deal with the impact of illness on their everyday lives, work, school, families and relationships.
  Practical information and advice is provided to parents regarding any travel, accommodation or other issues in these difficult and stressful situations.

- Bereavement
  Work closely with the medical team to support parents who are newly bereaved, providing emotional support and practical assistance around any issues which may arise following the death of their child.
  Medical social workers participate in post-bereavement MDT meetings which are offered to parents following the death of their child, and maintain a link with parents for an extended period following the death of their child continuing to provide emotional and practical support.

- Advocacy around practical and emotional support
  Provide practical and emotional support and advocacy to children and their families, including linking in with the MDT to clarify a diagnosis and its implications for families.
  Medical social workers also provide short-term counselling and emotional support to help families to adjust to illness, make decisions or cope with difficult emotions, as well as providing support and guidance to assist parents to find ways of talking with their children and their siblings about illness and bereavement.
  The team offer practical assistance to support families in meeting the care and other needs of children with complex and often life limiting conditions. This may include assisting families around housing, entitlements and other financial issues.

- End Of Life Care
  In addition to existing involvement by medical social workers in end of life, there is a role for further development with palliative care services.

### 15.1.12 Speech and Language Therapy

*The role of the speech and language therapist (SLT) in the Children's Heart Centre includes:*

- Communication evaluation and intervention, in the context of developmentally supportive and family focused care.
- Feeding and swallowing evaluation and intervention, to include pre-feeding assessment and promotion of readiness for oral feeding, evaluation of breast and bottle feeding ability, and completion of instrumental swallowing evaluations (videofluoroscopies).
- Parent/caregiver education and counselling, and MDT collaboration, which includes information regarding developmental expectations, communication interaction patterns, and feeding and swallowing behaviours.

There are a range of areas for development of the SLT service in cardiology, including quality control/risk management; discharge/transition planning and follow-up care; professional education and supervision; public education and advocacy; and research to advance the body of knowledge relevant to communication development and feeding and swallowing for infants and children with congenital heart disease. All cardiac patients who require tube feeding, are experiencing feeding problems, are at risk of swallowing problems (aspiration) and other speech and language problems need speech and language therapy services. A total of 2.5WTE SLTs are needed for paediatric cardiology.

### 15.1.13 Dietetics

Paediatric cardiology patients are at very high risk of disease-related malnutrition, and nutrition support is universally recognised as an essential component of clinical management of children with congenital heart disease. In the past two decades there has been a more proactive approach to pre-operative nutrition, coupled with a radical increase both in complexity and caseload volume. Dietetic services are provided to inpatients in the Children's Heart Centre and PICU, as well as to a large number of cardiology outpatients.

80% of patients admitted under cardiology receive nutrition support interventions. Approximately fifty patients are discharged on home enteral tube feeding each year, and continue to attend dietetic outpatient services in Crumlin due to lack of available community dietetic follow-up. A tube weaning programme is in place to ensure safe progression to oral feeding for the majority of patients discharged on home enteral tube feeding. This involves frequent dietetic review over a period of months to years, and liaison with the MDT. There are up to sixty cases of chylothorax per year, with the majority exclusively managed through dietary manipulation. The

dietetic service also functions as a national resource for nutritional information for all healthcare professionals managing paediatric cardiology patients.

*There are several areas where service developments are required:*

- Care pathways for the national paediatric hub and spoke model for paediatric cardiology, including patients from Northern Ireland
- Research and audit
- Implementation of growth surveillance similar to international cardiac centres
- Development of a national education programme on nutritional management of paediatric cardiology patients
- Guidelines on the management of complications associated with congenital heart disease such as protein-losing enteropathy

### 15.1.14 Cardiac Nursing

*The NCHC nursing team consists of:*

- 1.4WTE Clinical Nurse Manager 2
- 1WTE Clinical Nurse Facilitator
- 4.5WTE Clinical Nurse Manager 1
- 42WTE Staff Nurses

The nursing team is supported by health care assistants, ward clerks, household staff, and a play specialist.

*Cardiac Clinical Nurse Specialists*

The cardiac CNS works with the cardiologist and cardiothoracic surgeon and the other health professionals in partnership with children, young people and their families. The role of the cardiac CNS in Crumlin involves the planning, negotiating, implementing and evaluating of an agreed plan of care to meet the identified needs of the child/young person and family. Since the role was first established in Crumlin in 1993 it has continued to evolve and develop to promote a cardiac centre of excellence, and to meet the demands and needs of the growing population of cardiac patients and their families in all of Ireland. There are six cardiac CNSs currently in post. Their caseloads are divided between biventricular and univentricular care pathways incorporating other sub-specialties.

*The role of the CNS includes:*

- Antenatal diagnosis of congenital heart disease which allows pre-emptive planning of stabilisation of the baby. The CNS is in position to help parents by providing clear and accurate information, sensitive emotional support and optimising social support systems.
- The provision of practical information, education and emotional support to parents following post-natal diagnoses of cardiac condition is another key role of the cardiac CNS. Good communication skills are required to impart complex and potentially distressing information to the child and family, supporting them through the consent process and ensuring informed decision making.
- Prepare children for medical and surgical interventions and provide updates and support to the parents/family throughout elective procedures.
- Provide support and advice during the transition between home, community and hospital cardiac care through a bleep and email system. A nurse review clinic is run in collaboration with the consultant team where clinically indicated.

- Working across the service as a key advocate for the child/young person and their families by ensuring the partnership they have with their family and health and social care professionals is central to, and focuses on, meeting their needs and expectation within the care pathway. This includes working in a collaborative manner with all members of the multiprofessional team.
- Provide unique and specialist nursing knowledge required to work in partnership with the cardiac transplant and pulmonary hypertension shared care programmes between Crumlin and the specialist centres in the UK.
- Education and training (both structured and impromptu educational opportunities) to facilitate staff and parent education. The CNSs act as a resource for local paediatric and community services., and support and facilitate the transition of young people into adult services.
- Continue to support children and young people and families who are on the palliative care pathway. The CNS also provides ongoing support for parents and families following the death of their child by telephone, and coordinates the post-bereavement clinical follow up meetings in collaboration with the health and social care professionals.

The future of the CNS role is very exciting. Many new changes are occurring including further development of nurse-led clinics; five day cardiac ward facility; research programmes; and the first all-island integration of paediatric cardiology services. A more structured transition care pathway should be developed to support young people into adult services. The cardiac CNS role will continue to adapt and evolve to meet the needs of the growing population of children and young people and their families with congenital and acquired heart disease. In order to achieve this, there should be sub-specialised nursing roles, i.e. inherited cardiovascular and heart failure nurse specialists, ensuring continued delivery of a world class service to this unique cohort of patients.

*Achievements in Paediatric Cardiology and Cardiothoracic Surgery*

- Excellent outcomes with 2% mortality overall
- ECLS programme with clinical excellence award from the ECLS Organisation
- The appointment of two additional cardiologists was approved last year with both in post by mid-2015 (Terence Prendiville and Damien Kenny).

*Future Developments in Paediatric Cardiology and Cardiothoracic Surgery*

- The implementation of the International Working Group report is currently in progress and a phase of increasing both human resources and infrastructure is underway.
- The development of a transitional care programme to adult congenital heart disease services is required. It is anticipated that additional consultant staff will be required to care for adult patients with congenital heart disease and this post is actively being recruited.
- The development of advanced nurse practitioner roles in cardiothoracic surgery (may take up roles currently performed by SHOs) is required.
- Based on the current numbers of newly diagnosed cardiac heart defect patients each year, advances in treatment options, together with the existing caseload and the current gaps in service provision, a significant increase in HSCP staffing is required also for the ongoing management of these children.
- There is potential to develop a fellowship programme between Dublin and a United Kingdom unit for paediatric cardiothoracic higher surgical training. At present there is just one fellowship programme with a year in both Great Ormond Street Hospital and Birmingham. This should be explored.
- A paediatric cardiology training programme is in the advanced phases of being approved by the Royal College of Physicians of Ireland (RCPI). This will provide a high-quality, accredited and approved training experience for SpR trainees going forward (see below under National Children's Cardiology Centre).

- A hybrid cardiac catheterisation lab is due to open in late 2015 (construction underway).
- Increasing the role of CT/MRI with a diminished role for cardiac catheterisation should be explored.
- Adult congenital heart disease surgery is performed by the paediatric cardiothoracic surgeons in various hospitals in Dublin, however a significant number of congenital cases continue to be performed by non-congenital surgeons in other cardiac surgery centres in Ireland. The data from these procedures is currently not being submitted to NICOR due to lack of personnel.
- A fourth congenital cardiothoracic surgeon will be appointed once the Northern Ireland congenital workload transfers to Dublin and as the adult programme is consolidated in the Mater Hospital.

## 15.2 PROPOSED MODEL OF CARE

Surgery and interventional procedures are only two elements of a complex network of services for children with congenital heart disease that begins with prenatal screening and continues through to the transfer to adult services. It is recognised that children and families are entitled to local access for as much other cardiac-related care as is safe and appropriate, however all diagnostic and most ward-based care will continue to be centralised in the National Children's Heart Centre in the medium term.

Within this document the vision for the development of local networks with the appointment of paediatricians with a special interest in cardiology to strengthen the planning and delivery of non-interventional care within local settings. This vision is modelled on the approach set out by the British Congenital Cardiac Association.

### 15.2.1 The National Children's Heart Centre (NCHC)

The NCHC (currently based in Crumlin) is designated to perform surgical and interventional procedures on children. All children requiring a surgical or interventional procedure will be referred for this purpose. Based on current BCCA recommendations, the centre should have a minimum of twelve paediatric cardiologists to accommodate the existing Republic of Ireland population and also the predicted increase in volume from the integration of patients from Northern Ireland. NCHC teams will be able to perform the full range of inpatient (including neonatal) and outpatient non-invasive diagnostic procedures and ongoing management of children with congenital heart disease.

As part of a National Children's Heart Centre, Crumlin is developing a training programme through the RCPI for paediatric cardiology fellows, capitalising on the broad range of tertiary centre experience that can be provided in Ireland. The proposed duration of training is over five years, one of which may be spent in full-time research. Entrants will have completed basic specialist training in approved posts and obtained membership of the RCPI (paediatrics or general medicine) or a UK / European equivalent. The expectation is to train two paediatric fellows at any one time, one cardiology fellow and one paediatrician with a special interest in cardiology (PECSIG).

*The NCHC is responsible for:*

- Managing and developing referral, care, treatment and transfer pathways, policies, protocols, and procedures
- Performance monitoring and audit
- Professional training and development
- Facilitating the development of as much care and treatment as possible close to the child's home and the transition to adult services

The NCHC and regional centres (initially in Cork) will hold regular multi-disciplinary meetings across the network for issues such as agreement of protocols, review of audit data and monitoring of performance. Meetings will be held at least every 6 months.

### 15.2.2 Paediatric Cardiology Peripheral Services (PCPS)

PCPS will be delivered in larger regional hospitals (Cork, Limerick and Galway intially) and will have close working relationships with the national centre and with other local hospitals in close proximity. The team will include a consultant paediatrician with expertise in cardiology, and there will be a named consultant paediatric cardiologist from the national centre so that combined paediatric cardiology clinics are held regularly at the PCPS. Each PCPS should have 1WTE general paediatrician with a special interest in cardiology who will see all paediatric cardiology outpatient referrals and inpatients requiring cardiology input. In order to maintain clinical skills and echocardiogram competence, they should spend four weeks each year at the NCHC. Each PCPS will also require the support of a dedicated paediatric-trained cardiac physiologist (1WTE echo technician) along with a dietitian (0.5 WTE). Paediatric echocardiography services in PCPS sites will require at least one dedicated echocardiography machine with paediatric probes.

As part of the provision of services in regional centres, the department of paediatric cardiology at Crumlin are developing a curriculum with RCPI oversight to formally train paediatricians with an interest in paediatric cardiology, entitled A Framework of Competences for the Special Interest Module in Paediatric Cardiology. The curriculum is based on the PECSIG (Paediatricians with Expertise in Cardiology Specialist Interest Group), formally approved by the Royal College of Paediatrics and Child Health (RCPCH) in April 2008 (http://www.pecsig-nhs.org.uk) and completed over a one year training period. An inclusive and detailed document for Irish trainees is in development. Competency in transthoracic echocardiography is a cornerstone to this training programme with ongoing professional development in non-invasive imaging after appointment to post. Standards for assessment in echocardiography are through accreditation with the European Association of Echocardiography and endorsed by the Association of European Paediatric Cardiology (AEPC). The goal is to attract competitive applicants for dedicated subspecialty training in paediatric cardiology, provided under the supervision of paediatric cardiology at Crumlin, and then to subsequently support them in post at a regional centre with shared patient care, incorporating ongoing non-invasive imaging competency and management algorithms through a network service arrangement. It is anticipated that, once paediatricians with an interest in cardiology are formally trained and employed in designated regional and national centres, outpatient paediatric cardiology clinics at Crumlin will focus more attention on sub-specialty provision of care.

Hospitals providing PCPS will accept referrals for children suspected of having congenital heart disease from local hospitals, general practitioners, area medical officers and others involved in primary care, and they will be supported by echocardiography technicians with specific accreditation and training in paediatric echocardiography. To ensure that children receive as much non-interventional treatment as close to their home as is safe, networks will be supported by the NCHC and PCPS. The PCPS will have telemedicine facilities to link with the NCHC.

Each PCPS will have a formal annual training plan in place, which ensures ongoing education and professional development across the network for all healthcare professionals involved in the care of children with congenital heart problems.

### 15.2.3 Pathways of Care and Management of Congenital Heart Defects

*Pre-natally diagnosed congenital heart defects*

Where a prenatal diagnosis of congenital heart disease that is likely to require early post natal assessment or intervention, the mother will be transferred to a tertiary maternity centre in Dublin. The NCHC will agree and establish protocols with feto-maternal medicine units and tertiary neonatal units for the care and treatment of pregnant women whose fetus has been diagnosed with a major heart condition. The protocols must meet the 'Fetal Cardiology Standards' developed by the British Congenital Cardiac Association and ensure that pregnant women are referred to the relevant specialists as early as possible, for diagnosis, further testing and counselling. In all cases where a baby is likely to require immediate post-natal intervention or surgery, it is advised that the baby is delivered in a maternity unit adjacent to the NCHC.

*Newborns with a murmur and otherwise clinically well*

All referrals for cardiology assessment should first be reviewed and assessed by the on-site paediatrician who has a special interest in paediatric cardiology.

*Neonates and infants diagnosed with congenital heart defects*

Transport of a sick newborn with congenital heart disease is via the neonatal retrieval service with phone advice prior to transfer. Point-of-care echocardiography in the referring units by adult technical or inappropriately trained physician staff can be an issue if it results in misdiagnosis or delay in transfer to the appropriate services and complex heart disease can be missed. International guidelines clearly delineate the role of transthoracic echocardiography in the care of patients with suspected or confirmed congenital and acquired congenital heart disease (ACC / AAP / AHA / ASE / HRS / SCAI / SCCT / SCMR / SOPE 2014 Appropriate Use Criteria for Initial Transthoracic Echocardiography in Outpatient Pediatric Cardiology).
The national centre and PCPS will provide close monitoring for the development of heart failure, cyanosis or arrhythmias, and their initial management by medical treatment, if appropriate.

*New referrals of older infants and children from GPs and paediatricians*

Local hospitals will refer children to the NCHC or PCPS hospital, as appropriate, for the following categories of referrals:

1. *Murmurs*

   The clinical evaluation of an infant or child with a murmur is based on the history, exam and electrocardiogram. Pertinent components of the patient's medical history that may increase the likelihood of a congenital cardiac defect include syndromes or major other organ malformations (including inborn errors of metabolism) associated with congenital heart disease; family history of congenital heart disease, cardiomyopathy, sudden cardiac death or sudden infant death sydndrome; Kawasaki's disease, rheumatic fever; frequent respiratory infections; failure to thrive; maternal diabetes; pre-term delivery; teratogenic exposure in-utero; or chest pain with exertion.

   Pathology on examination includes an active precordium, thrill, heave, displaced apex, hepatomegaly, systolic murmur grade 3/6+, diastolic murmur, rub, click, gallop or weak pulses. Paediatric electrocardiogram interpretation requires experience and training and the cardiology service at NCHC provides assistance with ECG interpretation if requested.

Should an infant or child with a murmur have suspected heart disease, then timely referral to the NCHC is appropriate. Urgent referrals are dealt with best by discussing the patient with the on-call medical staff. Less urgent referrals are triaged to out-patient review based on each individual referral.

2. *Cyanosis*

Pulse oximetry testing for newborn congenital heart disease should be routinely undertaken prior to discharge in all neonatal centres across the country in line with the HSE / RCPI-endorsed guideline: http://www.hse.ie/eng/about/Who/clinical/natclinprog/paediatricsandneonatology/resources/PulseOximetryTesting.pdf

Following the seminal publication on the screening tool's validity by Ewer et al. (2011), pulse oximetry is the first, appropriately simple method to be used for the screening of congenital heart disease (CHD). If the study's findings were applied to the Irish population of 75,000 babies annually, approximately half of all neonates with major CHD would be detected through this simple screening tool. The procedure should be performed by a nurse or midwife who has been trained in the technique.

*Clear instructions must be in place on how to interpret the result (taken from either foot):*

- If the oxygen saturation is ≥95% it is normal
- If the oxygen saturation is 90-94% it is equivocal. Repeat in 1-2 hours. If still abnormal, refer for further assessment and investigation.
- If the oxygen saturation is <90% it is clearly abnormal. Refer immediately for further investigation.

The investigations undertaken when an abnormal result is obtained include repeat oxygen saturation, heart rate recording, 4 limb blood pressures, ECG, chest x-ray, nitrogen washout test, capillary blood gas, cardiac ECHO.

The sensitivity of pulse oximetry screening is 75% for detecting critical congenital heart disease and 49% for detecting all major congenital heart disease cases so clinical judgement is still required if concern remains for potential congenital heart disease despite a normal screen by pulse oximetry. Similarly, the most commonly missed congenital cardiac diagnosis by pulse oximetry is coarctation of the aorta. Weak or absent pulses may not be readily apparent until the ductus arteriosus closes by the fifth day of life. Again, clinical judgement and concern supercedes any screening tool for detection of congenital heart disease.

3. *Chest Pain*

Most chest pain is musculoskeletal in nature, however a history of chest pain induced by exercise forcing the child to stop the activity they are engaged in then congenital heart disease should be considered. If cardiology referral is required, a chest x-ray, ECG, echocardiogram and occasionally treadmill exercise test should be coordinated. A suggested algorithm for investigation is included in the appendix of this document.

4. *Palpitations*

Palpitations are a common referral reason to paediatric cardiology services and the vast majority of paediatric patients with the sensation of palpitations do not have heart disease. A suggested algorithm for investigation is included in the appendix of this document.

5. *Syncope or dizziness*

   Patients should be seen in the general paediatric clinic to determine whether symptoms are cardiac or neurological in nature. Appropriate investigations may include ECG +/- Holter 24 hour tape and, if indicated, an electroencephalogram (EEG). If still doubt or an abnormality found, then the patient should be referred for cardiology review for further investigation. A suggested algorithm for investigation is included in the appendix of this document.

6. *Screening because of family history of congenital heart defect, cardiomyopathy, sudden cardiac death or other syndromes*

   These patients should be referred to cardiology.

7. *Kawasaki disease*

   All patients with suspected or proven Kawasaki disease (KD) should have an echocardiogram performed during the acute admission and then again at 6 weeks. See excellent summary article for KD based on UK management strategy (reference below).

8. *Ongoing care of children and young people diagnosed with congenital heart defects*

   Local hospitals will refer children to the NCCC or PCPS as appropriate, for close monitoring for the development of heart failure or cyanosis, depending on the underlying heart defect. Designated transition nurses should facilitate effective and timely transition from children's to adult services.

### 15.2.4 Levels of Care to be provided

Cardiac care for children in Ireland will be provided in the following way:

*National Children's Heart Centre*

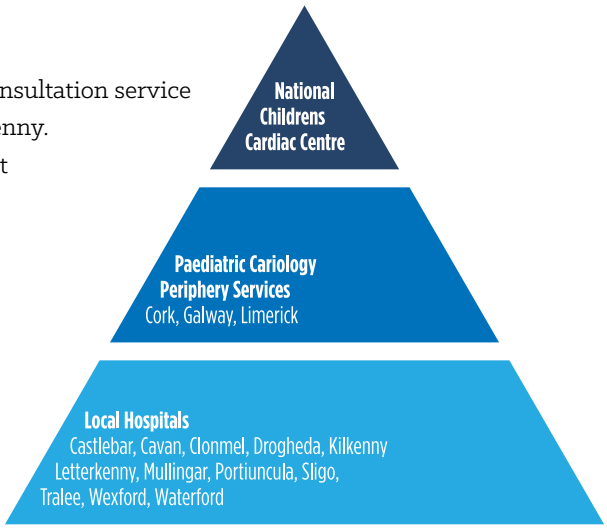
The NCHC is currently located in Crumlin, and in the future it will be based in the new children's hospital. This centre will perform surgical and interventional procedures on children.

*Paediatric Cardiology Periphery Services*

Cork, Limerick and Galway will be designated paediatric cardiology peripheral services.

*Local Hospitals*

All other paediatric units will provide a paediatric consultation service backed up by an outreach clinic in Sligo and Letterkenny.
In essence, paediatricians will assess largely innocent murmurs.



### 15.2.5 End of Life Care

Formal links should be established with the national palliative care service and with local outreach nurses. The cardiac psychologist is currently involved in palliative care and bereavement support for families in preparation for a death, and support for staff in working with the families is provided to as many patients as possible. Psychology support help prevent post-traumatic reaction in families and coming to terms with the loss of a child. Unfortunately due to the limited current resources this service cannot be provided for all families.

### 15.2.6 Service User Involvement in the Development of the Model of Care

Close collaboration with Heart Children Ireland is essential both in terms of service delivery and the development of a national model of care.

## 15.3 REQUIREMENTS FOR SUCCESSFUL IMPLEMENTATION OF MODEL OF CARE

### 15.3.1 Staffing Requirements

*National Children's Heart Centre*

| Staff Category | | Current (WTE) | Proposed (WTE) |
|---|---|---|---|
| Consultant Paediatric Cardiologist | | 5.07 | 12[1] |
| Consultant Congenital Cardiothoracic Surgeons | | 3 | 4 |
| Specialist Registrar – Cardiology | | 2 | 4 |
| Specialist Registrar – Cardiothoracic surgery | | 1 | 2 |
| Registrar – Cardiology | | 1 | |
| Registrar – Cardiothoracic surgery | | 3 | |
| SHO – Cardiology | | 2 | |
| SHO – Cardiothoracic surgery | | 2 | |
| Cardiology trained nurses | | 51WTE staff nurses + 2WTE CNM2 + 1.5WTE CNF + 4WTE CNM1 | |
| Advance Nurse Practitioner | | 0 | 4 – with reduction of SHO numbers |
| Clinical Nurse Specialist | | 6 | 1 Thoracic |
| Echocardiography technicians | Chief<br>Senior<br>Basic | 1<br>1.1<br>6 | |
| Dietitian | | 2 | 4.8[2] |
| Physiotherapist | | 1[3] | 4[4] |
| Pharmacist | | 1 | 1 |
| Occupational Therapist | | 0 | 2 |
| Psychologist | | 0.9[5] | 3.7[6] |
| Social worker | | 2 | 3[7] |
| Speech and Language Therapy | | | 2.5[8] |
| Data Manager | | 1 | 3, To include 1 for adult congenital |

1 Based on current BCCA recommendations, the NCHC should have a minimum of twelve paediatric cardiologists to accommodate the existing ROI population and also the predicted increase in volume from the integration of patients from Northern Ireland

2 Cardiac ICU should be allocated dietetic funding for 0.1WTE per PICU bed = 0.8WTE. A dedicated dietetic post is essential to the NCHC to develop national standards, education and training in nutrition support. Therefore it is recommended that funding for at least 2 additional WTE senior paediatric dietitians is provided for in Crumlin.

3 Currently there is 1WTE senior respiratory physiotherapist dedicated to the tertiary cardiology service covering PICU, NCHC and outpatients. There is no dedicated physiotherapist to neurodevelopmental cardiology, ECLS or post-transplant rehabilitation.

4 Based on the current numbers of newly diagnosed CHD patients yearly and advances in treatment options together with the existing caseload and the current gaps in service provision as discussed above, the paediatric cardiology service requires a total of 4WTE dedicated physiotherapists (2WTE physiotherapists dedicated to respiratory cardiology and 2WTE physiotherapists dedicated to neurodevelopmental/ neurology rehabilitation cardiology)

5 Current Staffing Levels - 0.9WTE (0.5 Lead, Principal Clinical Psychologist and 0.4 Senior Clinical Psychologist).

6 Staffing Requirements
The allocation of 3.7WTE clinical psychologists dedicated to cardiology is recommended:
- Required South of Ireland existing clinical population. In order to cope with the current caseload 1.6WTE additional senior clinical psychologists and 0.2WTE additional principal clinical psychologist are required.
- Required North of Ireland proposed clinical population. In order to cope with the proposed increased service an additional 1WTE senior clinical psychologist would be required.

This staffing complement for the NCHC would ensure that referred children and families could be seen in more appropriate timelines and receive more consistent support in all the aforementioned areas while the wider range of service development tasks could be achieved, consultation, antenatal diagnosis support, research, group work, staff training and supervision. The ability to provide a consistent and complete service to a particular team contributes to professional satisfaction and avoids professional frustration and burn out. This would ensure the retention of experienced staff, benefit the complete MDT's functioning and enhance patient care.

7 With current numbers of open medical social work cases running at approximately 100 at any given point in time and using of 1:40 new cases per year as a guideline (Sargent, 2008 – More than my Illness ), this would infer a need for at least 1WTE additional senior medical social worker. There are currently 2WTE basic grade posts in cardiology, where there was up to 3WTE posts in the past.

82.5WTE SLTs (two at senior level, 0.5 at staff grade level).

*Each Paediatric Cardiology Peripheral Service*

| Staff Category | Current (WTE) | Proposed (WTE) |
|---|---|---|
| Paediatrician with specialist interest | 0 | 1 |
| Cardiac physiologist | 0 | 1 |
| Clinical nurse specialist | 0 | 0.5 |
| NCHD | 0 | 0.5 |
| Psychologist | 0 | 0.5 |
| Dietitian | 0 | 0.5 |
| Social worker | 0 | 0.5 |

# 15.4 PROGRAMME METRICS AND EVALUATION

### Key Performance Indicators

National Institute for Cardiovascular Outcomes Research (NICOR), based at University College London, annually audits outcome metrics for our cardiac surgery and cardiac catheterisation programmes. Data is voluntarily contributed to this UK-based, NHS-endorsed National Congenital Heart Disease Audit (NCHDA). A random selection of medical charts are audited by an external team representing NICOR to validate the accuracy of data submitted. A data coordinator submits data to this registry as it is generated and the centre ranks highly in terms of data quality. Outcome metrics are on a par with the highest performing paediatric cardiac centres internationally. The new hospital consent form for Crumlin incorporates permission to submit patient data to NICOR, in line with regulation from the Office of the Information Commissioner.

*Parameters monitored include:*

- Demographics of patient cohort (age, weight sex, ethnicity, postcode)
- Pre- and post-operative co-morbidities
- Antenatal diagnoses
- Procedures performed (surgical and cardiac catheterisation)
- Intra-operative surgical times (bypass time, cross-clamp time, circulation arrest time)
- Post-operative days of intubation and date of discharge.
- Cardiac catheterisation procedure, fluoroscopy times and radiation dosages received.
- 30 day surgical mortality
- Infective endocarditis (cardiac lesion, infectious organism, dental treatment in prior three months, outcome)

The cardiology service also participates in a dedicated Morbidity and Mortality (M&M) meeting monthly or bi-monthly with the PICU on cardiac patients, and are in the process of setting up a similar M&M for the NCHC.

As per Irish Medical Council requirements, staff participate in additional audit projects across the service, including outpatient services and incidence of necrotising enterocolitis. Guidelines have been developed to standardise management and work-up of particular conditions, such as metabolic cardiomyopathy.

## 15.5 KEY RECOMMENDATIONS

*To implement the cardiology model of care nationally, the following is required:*

- Implementation of the 14 International Working Group recommendations with oversight from the committees set up to coordinate and deliver on the required resources
- Funding of outreach clinics by each hospital group
- The appointment of paediatricians with a special interest in cardiology (two years training in cardiology) in Cork, Limerick and Galway. Dedicated HSCP staffing will also be required in these regional centres to complement the development of these paediatrician posts. There is a requirement for additional funding of 0.5WTE senior paediatric dietitian for each regional centre (Cork, Galway, and Limerick)
- Development of a robust adult transition service
- A clear national strategy must be developed to deal with the child with a likely innocent murmur
- There should be more formalised training of technicians
- The guidelines for trisomy 21 and cardiac screening (sudden cardiac death, pre-propranolol for infant haemangiomas and ADHD) need to be revised
- The addition of advanced nurse practitioners to cardiothoracic surgery should be explored
- The paediatric cardiothoracic and paediatric cardiology higher surgical training fellowship programme should be developed
- A hybrid cardiac catheterisation laboratory whereby the theatre can act both as an operating theatre and catheterisation suite (due to open in late 2015)
- Increasing the role of CT/MRI with a diminished role for cardiac catheterisation to be explored
- Adult congenital heart disease surgery performed by congenital cardiothoracic surgeons to be developed. Further congenital cardiothoracic surgeon (i.e. fourth surgeon) appointee should have an adult focus.

## 15.6 ABBREVIATIONS AND ACRONYMS

| | |
|---|---|
| AEPC | Association of European Paediatric Cardiology |
| ASD | Atrial Septal Defect |
| AVSD | Atrioventricular Septal Defect |
| CHD | Congenital Heart Disease |
| CNF | Clinical Nurse Facilitator |
| CNM | Clinical Nurse Manager |
| CT | Computer-aided Tomography |
| ECG | Electrocardiogram |
| EEG | Electroencephalogram |
| GP | General Practitioner |
| IWG | International Working Group |
| MRCPI | Membership of the Royal College of Physicians of Ireland |
| MRI | Magnetic Resonance Imaging |
| NCHC | National Children's Heart Centre |
| NCHD | Non-consultant Hospital Doctor |
| NICU | Neonatal Intensive Care Unit |
| NTPF | National Treatment Purchase Fund |
| OPD | Outpatient Department |
| PCPS | Paediatric Cardiology Periphery Service |

| | |
|---|---|
| PDA | Patent Ductus Arteriosis |
| PECSIG | Paediatricians with Experience in Cardiology Special Interest Group |
| PICU | Paediatric Intensive Care Unit |
| RCPCH | Royal College of Paediatrics and Child Health |
| RCPI | Royal College of Physicians of Ireland |
| SHO | Senior House Officer |
| SpR | Specialist Registrar |
| UK | United Kingdom |
| US | United States |
| VSD | Ventricular Septal Defect |
| WPW | Wolff Parkinson White Syndrome |
| WTE | Whole Time Equivalent |

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# 15.8 APPENDICES

*Table 1: New referral sources to out-patient clinic (2014) with concern for murmur*

| New murmur referral Jan – Nov 2014 (1,361) | | |
|---|---|---|
| GP referral | 383 | 28.1% |
| 'Referred from another hospital (NOS)' | 201 | 14.8% |
| Coombe WH | 169 | 12.4% |
| Our Lady of Lourdes Hospital | 120 | 8.8% |
| National Maternity Hospital | 70 | 5.1% |
| 'Health clinic' | 60 | 4.4% |
| OLCHC clinic / dept internal referral | 46 | 3.4% |
| Portlaoise General Hospital | 45 | 3.3% |
| St. Luke's General Hospital, Kilkenny | 42 | 3.1% |
| Mullingar General Hospital | 29 | 2.1% |
| OLCHC Emergency Room | 28 | 2.1% |
| Rotunda Hospital | 25 | 1.8% |
| UHG | 23 | 1.7% |
| Castlebar General Hospital | 19 | 1.4% |
| Letterkenny General Hospital | 15 | 1.1% |
| Sligo General Hospital | 9 | 0.7% |
| Cork University Hospital | 8 | 0.6% |
| Waterford Regional Hospital | 5 | 0.4% |
| Tralee General Hospital | 3 | 0.2% |
| Naas General Hospital | 1 | 0.1% |

*Graphic 1: New referrals to out-patient clinic (2014) with concern for murmur: geographic location of patient address*

*Table 2: Surgical procedure data (source NICOR 2014)*

| | |
|---|---|
| Surgical Procedures All Ages | 332 |
| Neonate (1-30 days) | 82 |
| Infant (31-365 days) | 148 |
| Child (1-16 years) | 102 |
| Adult (16+ years) | 0 |

*Table 3: Surgical operative casemix (source NICOR 2014)*

| Lesions operated on (by NICOR Code) | |
|---|---|
| Anomalous coronary artery repair | Isolated Pulmonary artery band |
| Aortic root replacement (not Ross) | Mitral valve replacement |
| Aortic Valve Replacement - non Ross | Multiple VSD Closure |
| Aortic valve replacement - Ross | Norwood procedure (Stage 1) |
| Aortic valvoplasty | PDA ligation (surgical) |
| Aortic valvotomy | Pulmonary atresia VSD repair |
| Aortopulmonary window repair | Pulmonary valve replacement |
| Arterial shunt | Rastelli procedure |
| Arterial switch (for isolated transposition) | Repair of total anomalous pulmonary venous drainage |
| Arterial switch + VSD closure | Senning or Mustard procedure |
| ASD repair | Subvalvar aortic stenosis repair |
| Atrioventricular septal defect and tetralogy repair | Supravalvar aortic stenosis repair |
| Atrioventricular septal defect (complete) repair | TAPVD Repair + Arterial Shunt |
| Atrioventricular septal defect (partial) repair | Tetralogy repair |
| Bidirectional cavopulmonary shunt | Tetralogy with absent pulmonary valve repair |
| Cor triatriatum repair | Tricuspid valve replacement |
| Fontan procedure | Truncus and interruption repair |
| Heart Transplant | Truncus arteriosus repair |
| Interrupted aortic arch repair | VSD Repair |
| Isolated coarctation repair | |

*Table 4: Catheter procedure data (source: NICOR 2014)*

| Surgical Procedures All Ages | 284 |
|---|---|
| Neonate (1-30 days) | 30 |
| Infant (31-365 days) | 157 |
| Child (1-16 years) | 82 |
| Adult (16+ years) | 13 |

*Table 5: Interventional cardiac catheterisation casemix (source: Nicor 2014)*

| Cardiac Catheterisation Procedures Performed | |
|---|---|
| Aortic balloon valvotomy | Pulmonary artery stenting |
| ASD closure (catheter) | Pulmonary balloon valvoplasty |
| Blade atrial septostomy | Pulmonary valvotomy (radiofrequency) |
| Coarctation angioplasty | Radiofrequency ablation for supraventricular tachycardia |
| Coarctation stenting | Recoarctation angioplasty |
| Duct Stenting | RVOT Stenting |
| Implantable Cardioverter Defibrillator | Transcatheter PVR |
| PDA closure (catheter) | VSD closure (catheter) |
| PFO closure (catheter) | |

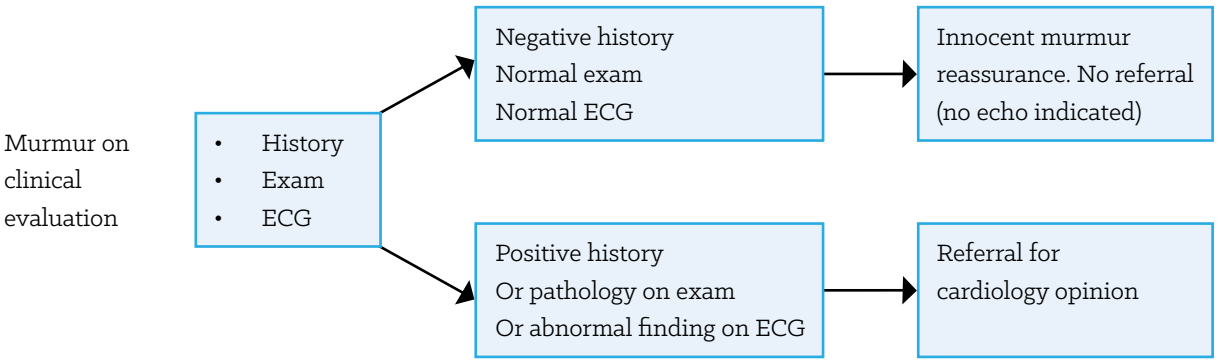
This algorithm has been produced by the National Paediatric and Neonatology Clinical Programmes. It is aimed at medical, nursing and allied health professionals working in both primary and emergency care settings.

| | | | |
|---|---|---|---|
| Algorithm Number | P25 | Document drafted by: | National Paediatric and Neonatology Clinical Programme |
| Revision Number | 1.0 | Document Status | Approved |
| Date of last update | June 2015 | Document approved by | Faculty of Paediatrics Clinical Advisory Group<br>Paediatrics and Neonatology Clinical Advisory Group<br>Irish College of General Practitioners |
| Approval date | November 2015 | Revision Date | November 2017 |

This algorithm has been prepared to promote and facilitate standardisation and consistency of practice, using a multidisciplinary approach.

Clinical material offered in this algorithm does not replace or remove clinical judgement.

## Algorithm for murmur work-up




**Positive history:** Syndromes or major other organ malfunctions (incl inborn errors of metabolism) associated with CHD; Fam history of CHD, CM, SCD or SIDS, Kawasaki's disease, Rheumatic fever; frequent respiratory infections; failures to thrive; maternal DM; preterm delivery; teratogenic exposure in-utero; chest pain with exertion;

**Pathology on exam:** active precordium, thrill, heave, displaced apex; hepatomegaly, systolic murmur grade 3/6+, diastolic murmur, rub, click, gallop, weak pulses

Definition of innocent murmur: soft, short duration, varies with positioning ('sensitive'), systolic, single (no clicks or gallops), sweet (not harsh sounding), small (does not radiate)