Key issues in the care of children, young people and adults with Congenital Heart Disease

Abstract

There have been significant advances in surgical, medical and nursing care for infants and children requiring cardiac surgery for Congenital Heart Disease (CHD) over the last three decades. This means that the number of adults known to have moderate and complex CHD is now in fact greater than the number of children with CHD. This article considers key issues for nurses in relation to the implications of (childhood) healthcare experiences for children and adults with CHD, and an understanding of the biopsychosocial impact of CHD on the individual and their families. It identifies the key issues and standards of care required in the UK (and world-wide) in the care of this unique group of patients.

Key words: To be drawn from the NS taxonomy

Congenital heart disease (CHD) refers to any anomaly or disease of the heart, such as anatomical defects, some cardiomyopathies and arrhythmias that are present at birth. CHD affects between 5-9 babies per 1000 born in the UK (NHS England 2016). Other authors have quoted 9 babies affected in every 1000 (Pandya et al, 2016). Approximately a third of these babies need some form of intervention as an infant and, if not diagnosed antenatally, cardiac surgery may be required as an emergency (NICOR 2018). Early diagnosis of CHD during fetal screening (table 1) can reduce mortality and morbidity by preventing clinical deterioration through identifying plans for the infant's delivery and neonatal care. Furthermore, early

diagnosis also enables parental choice and allows time for parental preparation of the health care journey ahead (Reid & Gaskin 2018).

Table 1. National standard and outcome measures regarding diagnosis (NHS England 2016)

• Mothers with suspected CHD in fetus, should be seen by an obstetric ultrasound specialist and a fetal cardiology specialist within 3 days of referral • Mothers with suspected CHD in fetus, should be seen by a specialist cardiac nurse on the day of diagnosis

Most infants with "critical" CHD (CCHD) are either diagnosed antenatally or immediately after birth and before discharge home. "Critical" CHD has been defined by authors such as Olney et al (2015), as structural malformations of the heart that are present at birth and require intervention in the first year of life. However, up to 30% of infants born with CCHD can present normally during the first few days of life when the routine neonatal examination is undertaken and are discharged home (Ailes et al, 2015; Khoshnood et al 2012). Therefore, the underlying diagnosis is not identified. Furthermore, the time at which these infants present varies depending upon the underlying cardiac defect; those with duct dependent lesions can deteriorate rapidly when the ductus arteriosus begins to close (Ostler et al 2019). A congenital heart defect termed 'duct dependent' means that due to the serious nature of the congenital abnormality, the circulation relies on a patent ductus arteriosus, which is present in the fetal circulation before birth, but will close a few days after birth, to aid oxygenation of the body. Closure in a duct dependent lesion can be catastrophic for the baby. Its patency can be pharmacologically maintained until some form of corrective surgery.

As existing screening methods, including the prenatal ultrasound scans and neonatal examination before discharge, do not identify all infants with CCHD, the use of pulse oximetry is now seen internationally in the early neonatal period. In a systematic review of 21 studies of 457,202 babies, pulse oximetry correctly identified 76.3% of babies who had CCHD and correctly identified 99.9% of healthy babies without cardiac problems (NIHR 2018). However, currently in the UK, pulse oximetry is not included in neonatal screening programmes, despite approximately half of maternity units using it as a screening method. This is because a National Screening Committee Pilot (PHE, 2017) indicated that there would be higher false positives than was identified by the systematic review in 2018 (NIHR 2018). However, it can be argued that a key difference in the PHE (2017) pilot, was that screening was undertaken within 12 hours of birth, whilst the systematic review found greatest specificity was after 24 hours.

For nurses and midwives, the rationale for screening should be underpinned by knowledge of embryological development of the cardiovascular system, fetal circulation, physiological changes occurring at birth, as well as the pathophysiology of cyanotic, and acyanotic CHD. This helps greatly in understanding how the defects can manifest and offers the ability to support and explain more clearly what is happening for parents and carers. Furthermore, recognising that the presenting features of an infant, undiagnosed antenatally with CCHD, include heart failure, cyanosis, collapse, weak or absent femoral pulses, abnormal heart rate/rhythm, murmur and weight gain or loss (Archer & Burch 1998; Cook & Langton 2009) is essential. Nurses caring for infants in any setting should be able to recognise these signs

during a clinical assessment and referral to specialist services should occur immediately due to the serious risk of avoidable mortality for this group of very fragile infants.

A national study of cardiac procedures conducted on infants in England and Wales between 2005 and 2010, explored death and emergency readmission after discharge; 333 (4.2%) of 7976 infants died before discharge and 246 (3.2%) of 7643 discharged died outside the hospital or after unplanned intensive care readmission within 1 year (Tregay et al 2015). An additional 268 (3.6%) infants had an unplanned readmission to intensive care but survived; the total number of adverse events was 514 (6.7%) in the first-year post discharge home.

Risk factors for adverse events identified by Tregay et al (2015) included: lower than expected weight for age at time of procedure; additional acquired cardiac diagnoses and pre-operative clinical deterioration; neurodevelopmental conditions and younger age at surgery. These were in addition to previously identified risk factors including: primary diagnosis, noncardiac congenital anomalies (Kogon et al 2012; Mackie et al 2004); prematurity (Fixler et al 2012); prolonged length of stay (Kogon et al 2012; Mackie et al 2004) (indicating greater complexity and perhaps a surrogate for post-procedural complications Brown et al 2003), and ethnicity (Ghanayem et al 2012). Tregay et al (2015) concluded there had been a lack of attention to improvement initiatives in the post-operative period and recommended targeted interventions to reduce the number of adverse events.

Parental stress

Several recent UK studies have highlighted the stress of parenting these vulnerable infants at home during their first year of life. Better parental discharge preparation, ensuring parents

have the right knowledge and know where to access information in order to respond to a deteriorating infant at home is necessary (Gaskin, Barron & Daniels 2016; Tregay et al 2015; Crowe et al 2016; Gaskin 2017). Furthermore, nurses have a responsibility to adequately prepare parents (Jones et al 2009; Titler and Pettit 1995; Weiss et al 2008), before their first transition from hospital to home.

In response to this need for better information, an early warning tool for parents and carers called the Congenital Heart Assessment Tool [CHAT], was developed as a targeted intervention (Gaskin, Barron & Daniels, 2016; Gaskin, Wray & Barron, 2018). Communication, including ongoing support of the child and family/carers, provision of individualised information and involvement of the child and family in decision making at every stage of care provision (NHS England 2016), is a significant aspect of the family-centred care provided by children's cardiac nurses. Nurses use CHAT as part of a discharge preparation strategy, to teach how to assess their infant at home, looking for signs of deterioration.

The CHAT is specifically for parents of infants who have undergone the first stage surgery for complex CHD, such as functionally univentricular (single ventricle) hearts, and are ready for discharge from hospital to home. CHAT aims to empower parents and carers to assess and make decisions based on a traffic light system, where signs in the green zone indicate parents can carry on as normal; signs in the amber zone indicate parents should phone for advice and signs in the or red zone indicate parents should phone for an ambulance immediately. CHAT has been further evaluated through a Health Foundation grant (unpublished study); and a learning and teaching grant from the University of Worcester has enabled the development of an e-learning resource to teach healthcare professionals how to use the CHAT to prepare parents and carers for discharge.

Meeting psychosocial needs

In addition to supporting parents and carers through educational preparation, another key element of nursing care refers to assessing and supporting the psychosocial needs of children, young people, their siblings, parents/carers and grandparents (Fischer et al 2012; Fonseca, Nazare and Canavarro, 2013; Franich-Ray et al 2013; Helfricht et al 2008; Jordan et al 2014; Wray & Sensky 2004). Adaptation and adjustment to the numerous traumatic events and transitions that occur for these families can be difficult; sometimes maladaptation, such as acute stress disorder or post-traumatic stress disorder, occurs as parents and carers struggle to cope. It is essential that nurses understand the psychological implications and impact of having a child with CHD and know and appropriately signpost children, their parents and siblings to psychological therapies at the right time.

The CHD standards, published in the UK by NHS England (2016), state that each child or young person must have access to a Children's Cardiac Nurse Specialist who will be responsible for co-ordinating their care and providing psychological support to promote family (and the child/young person's) adaptation and adjustment. Furthermore, the importance of a dedicated psychology service is recognised in the standards. Each specialist Children's Surgical Centre are required to provide a minimum of 0.25 whole time equivalent (WTE) practitioner psychologists (with experience of working with CHD) per 100 children and young people undergoing cardiac surgery each year; and 1 WTE practitioner psychologist must be employed for each cardiac network (NHS England, 2016).

The CHD standards refer to healthcare services provided across the patient's journey from the point of diagnosis to palliative and end of life care if required. The Paediatric Cardiac

Service Specification outlines care for all children diagnosed with heart disease (except some form of inherited disease) before their 16th birthday (NHS England 2016). Whereas, the Adult Congenital Heart Disease (ACHD) Service Specification covers the management of care for young people who are known ACHD patients transitioning from paediatric services from 16 years; as well as adults newly diagnosed with ACHD (NHS England 2016).

Key Issues for nurses working with Adults with CHD

The number of adults known to have moderate and complex CHD is now greater than the number of children with CHD (Marelli et al 2014). There are also a growing number of older adults with CHD (ACHD), who will present with emerging, unique needs not previously recognised. Some patients, however, receive a diagnosis of CHD late in adult life, such as those with some structural and/or valve abnormalities, including atrial septal defect (ASD) or pulmonary stenosis. This may be due to a later complication of an asymptomatic lesion, such as infective endocarditis in a bicuspid aortic valve (instead of the usual three leaflet tricuspid valve) or the development of an arrhythmia such as atrial fibrillation (AF), paradoxical embolus (an embolus moving from the arterial to the venous side of the body or vice versa) and stroke associated with an ASD.

As life expectancy for people living with CHD continues to improve, with an estimated 60% increase in the number of adults living with single ventricle physiology by 2024 (Coats et al 2014), the impact on healthcare services will be considerable, both within the hospital and in the wider community. A multidisciplinary team approach is essential to provide holistic care to all patients with CHD. Table 2. identifies a number of key issues to consider when caring for patients with CHD.

Table 2 Key issues in caring for patients with congenital heart disease

- Fetal screening
- Early diagnosis
- Psychological support
- Nursing knowledge and skill
- Discharge preparation
- Education, timely information and support
- Life style information
- Equity of access to care
- Lifelong follow up
- Short and Long term complications – heart failure, arrhythmias, haemorrhage, thrombus, infection

With appropriate healthcare support and management, the majority of those born with cardiac defects lead active lives and survive well into late adulthood (Pandya et al 2016). However, few congenital heart conditions are completely corrected in childhood, perhaps except for conditions such as an early successfully surgically ligated (patent) ductus arteriosus. All other repaired defects have the potential to develop late complications, long after primary interventions are completed. Adults with CHD who underwent surgery or percutaneous intervention (with an introduced cardiac catheter) in childhood may have residual structural defects, valve abnormalities and ventricular dysfunction that need monitoring and may require further intervention at some point (Kennedy 2007, Pandya et al 2016). Therefore, lifelong specialist follow-up is essential due to the potential to develop late complications and

as the onset of symptoms can be subtle and subclinical, early detection of changes enables appropriate intervention to reduce progressive myocardial and circulatory deterioration.

Furthermore, people who have lifelong cardiac problems may not notice or may ignore slight changes in exercise capacity until these become significant. For example, by the time the patient becomes aware of dyspnoea and exercise limitation, underlying valve and ventricular dysfunction can be severe and potentially irreversible. ACHD, particularly those with complex defects, are more likely to attend Accident and Emergency Departments than the general population; and hospital admission rates have been noted as twice as high as the general population, particularly amongst older age groups (Verheught et al 2010). The most common reasons for attendance include arrhythmia, heart failure, haemorrhage related or thrombus related conditions and infection (Negishi et al 2015).

Patients with complex cyanotic CHD are more prone to venous thromboembolism, epistaxis and increased bleeding after surgical procedures. The frequency of follow-up for these patients will depend on their clinical status and the severity of residual problems (Pandya et al 2016). Nevertheless, communicating, informing and educating patients about possible complications is an important nursing role.

Education is important in enabling ACHD to understand their heart condition, the need for follow-up, recognising clinical changes, how to access specialist services and where to find support information and advice. There may be occasions when people living with CHD face challenges in their lives and need additional support from their team. For instance, coming to terms with a change in their condition, understanding complex problems and treatments and the effects and side effects of medications.

Patients also require support and information about managing lifestyle issues, such as cardiovascular risk factors that include diet and healthy eating, physical exercise, including extreme sports, the use and effects of tobacco, alcohol, stimulant drinks and the effects of recreational drugs on the circulation. Information should also be provided about insurance, which can be difficult to obtain due to variation between insurance policies and available outcome data (ESC 2010). Travel insurance is particularly known to be a difficult area (Pickup, 2016) and CHD patient support groups offer invaluable advice and are a useful resource.

Information about the most appropriate type of contraception is important. Generally, women with CHD cope well with pregnancy and can have a normal vaginal delivery. However, a mother with CHD has an increased risk of 4-5% of having a baby with CHD. This risk to the baby is dependent on the specific CHD and can be as high as 10-50% amongst women who have for example, the rare heart defects known as Shone syndrome or 22Q11 deletion (DiGeorge syndrome – the deletion of small part of chromosome 22) (Pandya et al 2016). Pre-conceptual counselling with an experienced ACHD Cardiologist is therefore extremely important.

Regular dental care and good oral hygiene for those with CHD is critically important in reducing the risk of infective endocarditis from bacteria in the teeth and mouth, and which can damage the heart and be fatal. ACHD patients are therefore at greater risk of infective endocarditis than the general population. Recommendations for antibiotic prophylaxis to reduce the risk of this disease, in England and Wales, have changed over the years. Antibiotic prophylaxis should no longer be offered routinely for interventional procedures (National Institute for Health and Clinical Excellence, 2008). However, concerns have been raised about these changes; and many healthcare specialists still recommend prophylaxis for specific

patients, such as those with mechanical valves and with a previous history of infective endocarditis (Pandya et al 2016).

Provision of timely information and support at critical times is essential, such as a visit to the outpatient clinic when therapy may be adjusted, contacting the ACHD Nurse Specialist team by telephone or when there is need for medical or surgical intervention. CHD patients have reported that a service that meets changing needs over time and accessibility to the service, contribute positively to their wellbeing and are a source of emotional support (Hatchett et al 2015).

Ensuring co-ordination of care, effective communication and providing good patient experience across the cardiac networks are key themes of the Adult Congenital Heart Service Specification (NHS England 2016). Service providers are expected to measure patient experience and satisfaction with the quality of care received, with the assumption that areas for improvement will be highlighted, informing subsequent changes to service provision. Ooues et al's (2018) survey of patient experience in all outpatient clinics, including the (Level 1) specialist tertiary centre within an NHS ACHD Outreach Network, revealed variations in patient care that included the number of investigations performed on the clinic day, the provision of patient information leaflets, and knowledge about how to contact the ACHD team. The study raised important concerns about the equity of access to care from within the network and recommended standardisation of management protocols and network-training days.

Hatchett et al (2014) reported several themes identified by patients in a service evaluation of ACHD patients' experiences and satisfaction with the delivery of a nurse specialist service in an ACHD specialist centre. The highest ranked priorities for care were the provision of timely

information and advice, specialist nursing knowledge and expertise, effective care coordination, monitoring and support, accessibility, contact and responsiveness. Expansion of the ACHD specialist nurse role was recommended in relation to pre-admission assessments, greater autonomy in clinic management, nurse prescribing and end of life care.

Conclusion

Caring for children, young people and adults with CHD is multi-faceted and requires nurses to have comprehensive knowledge and understanding of biopsychosocial factors affecting patients and their families/carers across the life course. The increasing number of children with CHD surviving into adulthood, due to the success of cardiac surgery, medical interventions and nursing care, means that nurses working in adult services will need to keep abreast of the ever-changing surgical and medical techniques and associated long-term complications.

For children, young people, adults and their families/carers, quality service provision includes clear communication, education, provision of information and psychological support. The national CHD standards and service specifications (NHS England 2016) aim to promote equality and hopefully will address some of the health inequalities experienced by this patient group.

From a nursing perspective, many exciting opportunities now exist for academic and professional progression within children's and adult CHD networks; there is guidance on roles, career pathways and competence development for nurses interested in a career pathway in

CHD (RCN 2014, 2015). The British Adult Congenital Cardiac Nurses Association founded in 2007, enables nurses to share and develop clinical practice, provides peer support among specialist practitioners, education, audit and research relating to ACHD in order to promote high quality care for adult patients with CHD. Likewise, the Congenital Cardiac Nurses Association, set up in 1990, aims to provide a format for the exchange of information and ideas regarding the work and experience of the relatively small number of nurses involved in this field. Lastly, patient support groups and national CHD charities have a wealth of information for professionals on their websites – some of which can be found below.

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Little Hearts Matter www.lhm.org

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