Abstract
Congenital heart disease refers to an anatomical or physiological defect of the heart that is present at birth. Over the past three decades there have been significant advances in surgical, medical and nursing care for infants and children requiring cardiac surgery for congenital heart disease. This has meant that the number of adults with moderate and complex congenital heart disease now exceeds the number of children with the disease. This article details the background and diagnosis of congenital heart disease. It considers the implications for infants and children with congenital heart disease, as well as their parents, such as screening methods, risk factors, parental stress and psychosocial needs. This article also discusses the care of adults with congenital heart disease, including specialist monitoring and patient education.

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Keywords
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Congenital heart disease refers to any physiological anomaly or disease of the heart that is present at birth, and includes anatomical defects in the heart wall, valves or blood vessels, some cardiomyopathies (diseases of the heart muscle) and arrhythmias (irregular heartbeat). Congenital heart disease affects up to nine out of every 1,000 infants born in the UK (Pandya et al 2016). Around one third of these infants will require some form of healthcare intervention such as an echocardiogram, cardiac catheterisation, balloon dilation or angioplasty, or cardiac surgery (National Institute for Cardiovascular Outcomes Research 2019). If congenital heart disease has not been diagnosed antenatally and appropriate treatment has not been planned, emergency cardiac surgery may be required (National Institute for Cardiovascular Outcomes Research 2019).

Patients can be diagnosed with congenital heart disease antenatally or during childhood. In some patients the disease will remain undetected until adulthood, with patients requiring expert care and advice throughout their lives (NHS England 2016). Largely because of improvements in the diagnosis and treatment of infants and children with congenital heart disease, such as successful cardiac surgery, the number of adults known to have congenital heart disease is increasing, with more than four out of every 1,000 adults affected (Marelli et al 2014, NHS England 2016). Adults with congenital heart disease will require ongoing specialist care, and available interventions include cardiac surgery, transcatheter intervention, invasive electrophysiology and pacing procedures, palliative care and transplantation (NHS England 2016). Box 1 outlines some of the common structural anomalies seen in patients with congenital heart disease.

TREATMENT OF CONGENITAL HEART DISEASE IN INFANTS AND CHILDREN

<table>
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<th>Structural Anomaly Description</th>
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<td>Ventricular septal defect (VSD)</td>
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Care of infants, children and adults with congenital heart disease
Kerry Gaskin and Fiona Kennedy
Box 1. Common structural anomalies in patients with congenital heart disease

- Septal defects – a defect between two of the heart’s chambers (left and right ventricles or left and right atria), commonly known as a ‘hole in the heart’
- Coarctation of the aorta – narrowing of the aorta, the body’s largest artery
- Pulmonary valve stenosis – narrowing of the pulmonary valve, which controls the flow of blood from the lower right ventricle to the lungs
- Tetralogy of Fallot – a combination of four defects: pulmonary stenosis (narrowing of the pulmonary valve); ventricular septal defect (a hole between the right and left ventricle); overriding aorta (in which the aorta overrides the ventricular septal defect); and right ventricle hypertrophy (thickening)
- Transposition of the great arteries – where the pulmonary and aortic valves and the connected arteries are reversed (transposed)
- Underdeveloped heart – the atria or ventricles are underdeveloped, leading to an inability to pump enough blood around the body or to the lungs, for example seen in hypoplastic left heart syndrome

(NHS 2018)

- Pharmacological options – for example, mild congenital heart defects can be treated with various medicines, such as diuretics to remove fluid from the body, and digoxin to slow the heartbeat and strengthen the heart’s pumping function.
- Implantable heart devices – for example, arrhythmias can be treated with pacemakers and implantable cardioverter defibrillators.
- Cardiac catheterisation procedures – for example, aortic valve stenosis (a narrowed aortic valve) can be treated by balloon valvuloplasty. This technique involves passing a catheter through the blood vessels to a narrowed heart valve, which is widened using a catheter balloon. Large atrial septal defects can be closed with a device inserted using a catheter.
- Surgery – for example, defective aortic valves can be replaced by valves made from animal or human tissue, or the patient’s pulmonary valve. Similarly, large ventricular septal defects may be repaired with surgery. In infants with critical congenital heart disease (CCHD), surgery may be staged over a period of time because of the complexity of the condition. CCHD is defined as a structural malformation of the heart that is present at birth and requires intervention in the first year of life (Olney et al 2015).

Congenital heart disease in infants and children

Early diagnosis of congenital heart disease during prenatal ultrasound screening can reduce mortality and morbidity, enabling healthcare professionals to prevent clinical deterioration by planning neonatal care. The NHS England (2016) standards for congenital heart disease state that mothers with suspected congenital heart disease in their fetus should be seen by:
- An obstetric ultrasound specialist and a fetal cardiology specialist within three days of referral.
- A specialist cardiac nurse on the day of diagnosis.

Furthermore, early diagnosis of congenital heart disease in infants enables parents to be involved in any treatment decisions, and can enable them to psychologically prepare for the treatment pathway required to treat their child (Reid and Gaskin 2018).

Critical congenital heart disease

CCHD accounts for around 25% of all cases of congenital heart disease in infants (Oster 2019). Most infants with CCHD are either diagnosed antenatally or immediately after birth and before being discharged. However, up to 30% of infants born with CCHD can present normally in the first few days of life when the routine neonatal examination is undertaken, and are then discharged (Khoshnood et al 2012, Ailes et al 2015). Therefore, the underlying CCHD is not identified. Recognising the presenting signs of an infant with CCHD who has not been diagnosed antenatally is an essential skill for nurses. Presenting signs include heart failure, lethargy, hypoxia, cyanosis, cardiorespiratory collapse with shock, weak or absent femoral pulses, abnormal heart rate and/or rhythm, heart murmur, and weight gain or loss (Cook and Langton 2009).

The time at which these infants present with CCHD varies depending on the underlying cardiac defect. For example, those with duct-dependent lesions can deteriorate rapidly when the ductus arteriosus (a blood vessel that supplies the fetus with oxygen from the placenta) begins to close following birth when the infant’s lungs are able to supply the body with oxygen.

A congenital heart defect is termed duct-dependent when, because of the serious nature of the infant’s congenital anomaly, their circulation continues to rely on a patent ductus arteriosus to maintain systemic and pulmonary circulation (NHS 2018). Closure of the ductus arteriosus in a patient with a duct-dependent lesion can be life-threatening if not identified promptly (Oster 2019). For example, patients with left-sided obstructions (hypoplastic left heart syndrome, critical aortic stenosis, coarctation (narrowing) of the aorta) rely on blood flow through the duct to provide systemic circulation, without which the major organs will not be perfused. Patients with right-sided obstructions – such as critical pulmonary stenosis, pulmonary atresia or tetralogy of Fallot – rely on left to right blood flow through the duct to provide pulmonary circulation, without which the blood cannot be oxygenated, resulting in severe hypoxia and cyanosis. However, the patency of the duct can be maintained pharmacologically with dinoprost (prostaglandin E2) therapy until corrective surgery is undertaken (NHS Children’s Acute Transport Service 2018).

Screening

Existing screening methods, including prenatal ultrasound scans and neonatal examination before discharge, do not identify all infants with CCHD. Therefore, a European panel of experts recommended universal screening of oxygen saturations with pulse oximetry, although only some countries have adopted this recommendation (Manzoni et al 2017). For example, in the UK, pulse oximetry is not
on the post-operative period for infants with life-threatening congenital heart disease, and they recommended targeted interventions to reduce the number of adverse events, such as guidelines for the entire discharge process and home monitoring programmes.

Family-centred care
One significant aspect of family-centred care provided by children's cardiac nurses is communication, which includes ongoing support of the child and their family members, provision of individualised information, and involvement of the child and family members in decision-making at every stage of care provision (NHS England 2016). Several studies have identified the stress associated with parenting infants with congenital heart disease at home during the first year of life, and have demonstrated the need for improved discharge preparation to ensure that parents have the appropriate knowledge to respond to deterioration, if required (Gaskin et al 2016, Gaskin 2017). Furthermore, nurses have a responsibility to adequately prepare parents for discharge from hospital to home (Weiss et al 2008, Jones et al 2009).

In response to this need for improved information for parents, and to support discharge planning and home monitoring, a parental early warning tool known as the Congenital Heart Assessment Tool (CHAT) was developed (Gaskin et al 2016, 2018). This tool can be used as part of a discharge preparation strategy, with nurses teaching parents how to assess their infant at home and observe for signs of deterioration such as breathlessness and reduced urine output. CHAT is designed for parents of infants who have undergone surgery for CCHD presentations such as functionally univentricular heart (congenital heart defects where one of the ventricles, either left or right, is not fully developed), and are ready for discharge from hospital to home. However, future versions of the tool will consider all types of congenital heart disease. CHAT aims to empower parents to assess their child and make decisions based on a traffic light system, in which (Gaskin et al 2016, 2018):

- 'Green zone' signs indicate that parents do not need to act. These signs include the infant behaving normally and the absence of changes to their skin colour or temperature.
- 'Amber zone' signs indicate that parents should phone the hospital for advice. These signs include breathlessness and abnormal oxygen saturations.
- 'Red zone' signs indicate that parents should phone for an ambulance immediately. These signs include the infant not responding to normal interaction, difficulty breathing or reduced urine output.

Psychosocial care
It is important that nurses consider the psychosocial needs of children with congenital heart disease, and those of their siblings, parents and other family members (Franich-Ray et al 2013, Fonseca et al 2013, Mackie et al 2004, Fixler et al 2012, Kogon et al 2012).

Identifying at-risk patients
Nurses caring for infants in any setting should be able to recognise the presenting signs of an infant with CCHD during a clinical assessment, and should be confident in referring the patient to specialist services immediately because of the serious risk of avoidable mortality in this group of patients.

Crowe et al (2016) identified several risk factors for adverse events following cardiac surgical procedures conducted on infants with congenital heart disease, including: lower-than-expected weight for age at the time of the surgical procedure; additional acquired cardiac diagnoses and preoperative clinical deterioration; neurodevelopmental conditions; and younger age at surgery. These risk factors were in addition to previously identified risk factors including non-cardiac congenital anomalies, prolonged length of hospital stay, and prematurity (Mackie et al 2004, Fixler et al 2012, Kogon et al 2012). Crowe et al (2016) concluded that there has been a lack of attention

Key points

- Congenital heart disease refers to any physiological anomaly or disease of the heart that is present at birth, and includes anatomical defects in the heart wall, valves or blood vessels, some cardiomyopathies (diseases of the heart muscle) and arrhythmias (irregular heartbeat).
- Patients can be diagnosed with congenital heart disease antenatally or during childhood. In some patients the disease will remain undetected until adulthood, with patients requiring expert care and advice throughout their lives.
- It is important that nurses consider the psychosocial needs of children with congenital heart disease, and that of their siblings, parents and other family members.
- A multidisciplinary team approach is essential to provide holistic care to all patients with congenital heart disease. With appropriate healthcare support and management, most individuals born with cardiac defects can lead active lives and survive into late adulthood.
Jordan et al 2014), in collaboration with clinical psychologists. Adjusting to the various traumatic events and transitions that can occur during a child’s treatment for congenital heart disease can be challenging for parents and family members, who may develop conditions such as acute stress disorder (Franich-Ray et al 2013).

NHS England’s (2016) congenital heart disease standards state that each child or young person must have access to a children’s cardiac nurse specialist who should be responsible for coordinating their care and providing psychological support to the child, parents and family members.

There is growing evidence of neurodevelopmental implications of congenital heart disease, particularly for infants with CCHD requiring intervention in the early neonatal period (Marino et al 2012). These patients may not meet developmental milestones, while speech delays and behavioural issues can become apparent in childhood and adolescence (Ringle and Wernovsky 2016).

CCHD has been associated with suboptimal intellectual functioning (Ryberg et al 2016), while ongoing physical implications include suboptimal exercise tolerance, significant reductions or increases in weight, and mental health issues (Ringle and Wernovsky 2016).

Staff in children’s cardiac centres in the UK include specialist transition nurses who work with young people aged 12-13 years, and their families, to support them to understand their condition and become increasingly independent. The NHS England (2016) congenital heart disease standards outline the management of care for young people transitioning from paediatric to adult services after the age of 16 years, as well as adults newly diagnosed with adult congenital heart disease.

**Congenital heart disease in adults**

A growing number of adults will receive a diagnosis of congenital heart disease late in life, such as those with structural and/or valvular anomalies, including atrial septal defect or pulmonary stenosis. This may be because of a later complication of an asymptomatic lesion such as infective endocarditis (an infection of the endocardium, the inner lining of the heart) in a patient with a bicuspid aortic valve (an aortic valve that only has two leaflets – also known as flaps – rather than the usual three-leaflet tricuspid valve), the development of an arrhythmia such as atrial fibrillation, a paradoxical embolism (an embolus moving from the arterial to the venous side of the body or vice versa) or atrial septal defect-associated stroke (Curtis and Stuart 2005, Kennedy 2008).

While some patients with congenital heart conditions are treated effectively in childhood, several other heart defects, such as pulmonary stenosis, that have been treated with surgery or cardiac catheterisation procedures have the potential to develop complications such as residual structural defects and valve anomalies and ventricular dysfunction some time after the primary intervention is completed. These complications involve lifelong specialist monitoring such as imaging, while further interventions may be required at some point (Kennedy 2007, Pandya et al 2016). Lifelong specialist monitoring is essential because the onset of symptoms such as dyspnoea (breathing difficulties) and fatigue can be subtle and subclinical, with early detection enabling appropriate interventions such as further surgery to reduce progressive myocardial and circulatory deterioration.

As life expectancy for people living with congenital heart disease continues to improve, the effect on acute and community healthcare services will be considerable, with interventions such as advanced heart failure treatment and transplantation required in many cases. A multidisciplinary team approach is essential to provide holistic care to all patients with congenital heart disease, and, with appropriate healthcare support and management, most individuals born with cardiac defects can lead active lives and survive into late adulthood (Pandya et al 2016).

**Long-term complications**

People who have lifelong cardiac complications may not notice or may ignore slight changes in their exercise capacity until these become significant (Burchill 2016). For example, by the time the patient becomes aware of the development of dyspnoea, underlying valve and ventricular dysfunction can be severe and potentially irreversible. Adults with congenital heart disease, particularly those with complex defects, are more likely to attend emergency departments than the general population; similarly, hospital admission rates for this patient group are twice as high as in the general population, particularly among older age groups (Verheugt et al 2010).

The most common reasons for attendance to emergency departments include arrhythmia, heart failure, haemorrhage-related or thrombus-related conditions, and infection (Negishi et al 2015).

Long-term complications for patients with complex cyanotic congenital heart disease include venous thromboembolism, epistaxis (nosebleeds) and increased bleeding following surgical procedures. Other symptoms associated with cyanotic congenital heart disease include iron deficient anaemia, cerebrovascular accident, infectious complications such as endocarditis, and cerebral abscess (Baumgartner et al 2010). The frequency of follow-up for these patients will depend on their clinical status and the severity of any residual complications (Pandya et al 2016). Using effective communication skills to inform and educate patients about possible complications is an important nursing role.

**Patient education**

It is important for nurses to provide patient education to enable adults with congenital heart disease to understand their condition and recognise any clinical changes. Nurses can also educate patients about the requirement for lifelong follow-up and how to access specialist services for support, information and advice. There may be occasions when adults with congenital heart disease may require additional advice and support from the multidisciplinary team, for example to enhance their
understanding of the need for further catheter interventions or surgery for pulmonary valve regurgitation (where the pulmonary valve is incompetent), or on the treatment of atrial or ventricular arrhythmia (Baumgartner et al 2010).

Adult patients with congenital heart disease also require support and information regarding the management of lifestyle issues. These include cardiovascular risk factors such as obesity and extreme sports, and the effects of tobacco, alcohol, stimulant drinks and recreational drugs on the circulation. Information should also be provided to patients about insurance, which adult patients with congenital heart disease may find challenging to obtain (Baumgartner et al 2010). Travel insurance is known to be a particularly challenging area for adult patients with congenital heart disease, and support groups for these patients can offer useful advice in relation to this issue (Pickup et al 2016).

Information about the use of contraception in adults with congenital heart disease is also important; for example, while hormonal contraceptives are highly effective, they should be avoided in women at risk of thromboembolism (Baumgartner et al 2010). Women with congenital heart disease who are pregnant can be classified as high, medium or low risk of obstetric complications, such as premature labour and post-partum haemorrhage, and complications for their fetus, such as restricted fetal growth, prematurity and fetal death (Regitz-Zagrosek et al 2018). Women who have a moderate or high risk of complications during pregnancy should receive pre-conception counselling and management during pregnancy and delivery in a specialist adult congenital heart disease centre, and should receive this from a multidisciplinary team that includes a cardiologist, obstetrician and anaesthetist (Baumgartner et al 2010).

Adult patients with congenital heart disease should be informed that regular dental care and optimal oral hygiene is vital, because it can reduce the risk of infective endocarditis from oral bacteria, which can damage the heart and be fatal. Adult patients with congenital heart disease are at greater risk of infective endocarditis than the general population (Baumgartner et al 2010). While routine antibiotic prophylaxis against infective endocarditis is not routinely recommended during dental procedures or non-dental procedures such as urological, gynaecological and obstetric procedures, or ear, nose and throat procedures (National Institute for Health and Care Excellence 2016), some cardiac specialists have continued to recommend antibiotic prophylaxis to reduce the risk of infective endocarditis in specific patients, such as those with mechanical valves and a previous history of infective endocarditis (Pandya et al 2016).

Providing coordinated care
Ensuring coordinated care is an important aspect of adult services, as detailed in the NHS England (2016) congenital heart disease standards. Ooues et al’s (2018) survey of adult congenital heart disease patients’ experience of outpatient clinics revealed variations in patient care, including in the number of investigations undertaken during a clinic day, the provision of patient information leaflets, and patients’ knowledge of how to contact the adult congenital heart disease team. Ooues et al’s (2018) study raised concerns about the equity of access to care, and recommended standardised management protocols and training days for local cardiologists, obstetricians, emergency department nurses and doctors.

Nurse specialists have an important role in providing information and support to all patients with adult congenital heart disease, as well as preparing patients for cardiology procedures and surgical pre-admission clinics. Patients with congenital heart disease have reported that a service that meets their changing needs over time and which is easily accessible contributes positively to their well-being and provides a source of emotional support (Hatchett et al 2015a).

Hatchett et al (2015b) undertook a service evaluation of adult congenital heart disease patients’ experiences of a nurse specialist service. 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. Hatchett et al (2015b) recommended expansion of the adult congenital heart disease specialist nurse role to include pre-admission assessments, increasing autonomy in clinic management, nurse prescribing and end of life care.

Conclusion
An increasing number of children with congenital heart disease are surviving into adulthood as a result of the success of cardiac surgery, medical interventions and nursing care. This means that nurses practising in adult services will need to continually update their knowledge of the surgery and therapies used to treat congenital heart disease, as well as any associated long-term complications. To provide optimal care for infants, children and adults with congenital heart disease, nurses should consider elements such as identifying at-risk patients, clear and effective communication, psychological support, and patient education in areas such as diet, lifestyle, contraception and dental care.

References


