

Challenges Caring for Adults With Congenital Heart Disease in Pediatric Settings: How Nurses Can Aid in the Transition

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As surgery for complex congenital heart disease is becoming more advanced, an increasing number of patients are surviving into adulthood, yet many of these adult patients remain in the pediatric hospital system. Caring for adult patients is often a challenge for pediatric nurses, because the nurses have less experience and comfort with adult care, medications, comorbid conditions, and rehabilitation techniques. As these patients age, the increased risk of complications and comorbid conditions from their heart disease may complicate their care further. Although these patients are admitted on a pediatric unit, nurses can aid in promoting their independence and help prepare them to transition into the adult medical system. Nurses, the comprehensive medical teams, and patients' families can all effectively influence the process of preparing these patients for transition to adult care. (*Critical Care Nurse*. 2016;36[4]:e1-e8)

Thanks to advances in medicine, both diagnostic and surgical, patients with congenital heart disease (CHD) have longer life expectancies than before and are living well into adulthood.¹ According to statistics from the Centers for Disease Control and Prevention, nearly 1 million adults in the United States are living with a congenital heart defect, and “congenital heart defects are the most common type of birth defect in the United States, affecting nearly 1% of—or about 40 000—births per year.”² Now the number of people more than 20 years old who have CHD is greater than the number of people less than 20 years old who have CHD.³ Because of the complexity of these cases, many adult patients with CHD remain in the care of their pediatric cardiologists.⁴ A major concern for this population is the lack of trained surgeons and adult facilities to care for these patients once they transition from pediatric

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1. Discuss the roles of nurses, the comprehensive medical teams, and patients' families in preparing these patients for the transition to adult care
2. Identify 3 clinical challenges of adult patients treated in pediatric settings
3. Describe the 5 factors that should be included in a program's transition curriculum

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Courtney, a 35-year-old woman with CHD, had pulmonary and aortic stenosis diagnosed at 6 months of age. She underwent her first surgery at 4 years old, a balloon valvuloplasty to open up her valves, in order to allow more blood flow out of the heart and improve heart function. Courtney's pediatric cardiology team managed her care and also became her central support system for the next 26 years, in which she underwent 4 open heart surgeries, including a second balloon valvuloplasty and both pulmonary and aortic valve replacements. She endured complications including endocarditis, sepsis, and organ failure. Completing high school was uncertain, and Courtney not only survived, but surpassed those expectations. She went on to obtain her college degree from a Big Ten university and began a teaching career. Achieving these goals may not have been possible without her positive outlook and continued encouragement from family, as well as her pediatric team of providers and nurses.

Although Courtney felt comfortable with her providers in the hospital, she experienced a level of embarrassment

being an adolescent and adult surrounded by infants and young children in a pediatric unit. A survey by Moons et al⁸ documented that adolescents feel out of place in clinic waiting rooms surrounded by young children and would prefer to be with adults. The caring and sensitive nature of pediatric nursing provided a safe, comforting environment but did not push Courtney to grow in her independence or challenge herself during times of rehabilitation. Her parents made most of her medical decisions, and the hospital team would often round outside of her room, not thinking to include her in plan-of-care decisions. Although at the time she did not give much importance to not being involved in the discussions, looking back, it would have helped prepare her for transition to adult care and would have eased the process to have been given the tools by the people she trusted. Having a formal transition program that begins early in adolescence would improve outcomes, allow patients proper time to prepare, and give nurses structure in how to guide these patients to gain more independence while under their care.

programs.⁵ Only 14 of 189 cardiology fellowships in the United States offer training in the field of adult CHD as of 2015.⁵⁻⁷ Although adult cardiac patients remain in the pediatric health system, challenges arise in providing for these patients, including pediatric nurses and ancillary staff being less familiar with caring for adults and their comorbid conditions. Administering care that is safe, effective, and evidence-based for these patients is a priority, and pediatric hospitals may not be the best place to achieve this goal. This article describes the challenges in caring for adult patients in pediatric settings and how nurses and comprehensive medical teams can effectively influence preparing patients for the transition to adult care.

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Clinical Challenges of Adolescent and Adult Patients

Nursing has a goal of providing patient- and family-centered, safe, effective, and evidence-based care, but there are roadblocks to achieving this goal when pediatric nurses care for adult patients. Many challenges stem from the lack of comfort nurses feel when assigned to an adult patient, because of irregular exposure to this age group and differences in their care.⁴ The dynamic with these patients is also often confusing, because they are legally adults, but yet tend to be dependent on their parents and may not make their own medical decisions, and in some cases overprotective parents do not allow the shift of responsibility.⁹ Adolescents do not have the same decision-making capacity as adults, and this situation has led to the creation of a legal doctrine for the mature minor, creating a partnership between parents and adolescents in medical decision making.^{10,11} Some adolescents do not request to be informed or part of the decision making and desire parental guidance, which leads to a parent-directed medical model of decision making.^{10,11} As health care providers, we should view adolescents as secondary decision makers in collaboration with parents until at least the age of 18, at which

point they are required to be the consenting party regardless of the dynamics between the parent and the patient.^{10,11} This lack of independence is a factor that often limits CHD patients from becoming responsible for their health care and transitioning to the adult medical system.⁹ The comforting and reassuring overprotective atmosphere in pediatric care can lead to lower self-esteem and hinder the adolescent's transition to independence.¹²

Adolescence is a period during which establishing health-related behaviors is vital because those behaviors will follow the person through life.⁸ Adolescent development includes finding one's identity and independence, developing a sexual identity, as well as establishing a peer group, all of which can influence the creation of health-related behaviors.^{4,8,13} With the addition of a layer of complexity with chronic illness, the need for medical follow-up, limitations of physical activity, and chronic health maintenance, an already taxing time period becomes more stressful with the addition of transition.¹⁴ The medical team, while these patients are admitted to the hospital, must be aware of the patient's physical state, as well as the patient's psychological state. Understanding how chronic medical conditions affect completing these developmental milestones can guide each patient's transition needs.⁸ It is important to foster patients' growth and encourage them to meet their developmental milestones in order to avoid hindering patients' independence. Promoting developmentally appropriate skills such as self-care, health care decision making, and self-advocacy while patients are hospitalized allows them to become better prepared in order to transition to adult care successfully.¹⁵

Management Differences Between Pediatric and Adult Care

Another challenge for nurses is the need to be aware of the differences in fluid volumes, appropriate ranges for chest tube output, and medication dosing between pediatric and adult standards. Dearani and colleagues¹⁶ found common examples among nursing staff of decreased comfort with adult medication dosing, highlighting the differences between pediatric weight-based dosing and adult standard dosing. Children are not small adults, and differences in their physiology lead to changes in pharmacokinetics and pharmacodynamics.¹⁷ Factors including variations in gastric pH, gastric emptying, protein binding, immaturity of bile secretion and activity, and glomerular filtration rate require consideration

when the dosage of medications is being determined.¹⁷ Many more medications for adults have been approved by the US Food and Drug Administration, whereas pediatric patients use off-label recommendations adapted from adult studies, owing to the smaller populations of child patients and the limited testing in children.¹⁸ For example, anticoagulation medications used in children often include heparin, enoxaparin, and warfarin, whereas drugs such as dabigatran and rivaroxaban are newer options used in adults that do not require frequent monitoring or dose adjustments.¹⁹ Adult care providers have more knowledge on the latest medications being tried and may provide superior options for adult patients, which is a reason for these patients to transition from pediatric to adult centers.

Differences in Care of Comorbid Conditions

Acquired comorbid conditions in this patient population are other areas of limited pediatric nursing comfort and another reason to transition these patients to adult care.¹⁶ Secondary complications of CHD can include coronary artery disease, myocardial infarction, diabetes mellitus, renal

failure, hypertension, and arrhythmias.³ Pediatric hospitals affiliated with adult centers often allow adult services to consult on older patients who are admitted to a pediatric facility because of their need for a pediatric congenital heart surgeon. Challenges for the nursing staff can often occur because of a lack of clarity on which service to contact should a problem arise, leading to delays in care. Free-standing pediatric hospitals without adult consultation services may be leaving their patients at a disadvantage. Data compiled from 37 free-standing pediatric hospitals revealed 707 adult admissions in a 2-year period.²⁰ Of the adults admitted, more than 30% had comorbid conditions that required subspecialty care.²⁰

The need for adult consultation services will only become more prominent as the adult CHD patients continue to age and have more complex medical conditions develop.²⁰

Because of the complexity of their condition, patients require lifelong follow-up and may require psychosocial or behavioral support in tandem with their medical care.⁸ Compared with the general population where 20%

The comforting and reassuring overprotective atmosphere in pediatric care can lead to lower self-esteem and hinder transition to independence.

of people have a psychiatric disorder, 36.4% of adult patients with CHD may have depression or anxiety.^{21,22} Multiple ancillary services, including social work, financial counselors, registered dietitians, and physical therapy are an important part of their care team.²³ These services will better aid the patient if they have experience with adults in needed areas such as contraception, family planning, genetics, and employment advice, areas in which pediatric nurses and providers may have limited knowledge.¹⁶

Rehabilitation Differences

Last, differences exist in nursing care on adult and pediatric units for intubated patients, patients with a ventricular assist device (VAD), and patients undergoing extracorporeal membrane oxygenation (ECMO) because of the patient population and unit culture. Adult patients are often not sedated if stable, weaned off mechanical ventilation, or given a tracheostomy in order to rehabilitate on a VAD or ECMO.^{24,25} Pediatric culture is more comfortable when patients are sedated and unable to interfere with supportive interventions. Active rehabilitation while on ECMO decreases the risk of deconditioning and improves posttransplant outcomes.²⁶ Ambulation of an ECMO patient requires a multidisciplinary approach, thorough preparation, and team commitment. An adult patient on a pediatric unit may be at a disadvantage in these instances because of lack of staff training and comfort ambulating these patients.

Need for Transition Programs

Currently, not enough adult health care providers are trained to care for the complex population of patients with CHD.⁵ The process of transition for these patients can be traumatic, as they are leaving the familiar medical home they have built.⁸ Taking responsibility for their care may be overwhelming for patients and may impede the transition process,¹² especially when their health care team

Nurse-led education improves self-management skills, knowledge of cardiac disease, and self-advocacy.

has not prepared them appropriately. Studies have shown that

adolescents with CHD have limited knowledge about their condition, self-care needs, prevention and signs of infection, as well as physical limitations.²⁷ Many patients fail to even make the first appointment with their

adult-care team after the transition and get lost to follow-up.²⁸ Although patients' independence is at the center of the transition process, preparing the family for this change should be a focus of transition education. Patients' families often feel a lack of comfort with the transition to the patient-centered approach of adult medicine from the family-centered pediatric style,⁸ especially with those families caring for cognitively impaired children. The American Academy of Pediatrics' Medical Home Initiative states that 90% of children with special needs will reach 21 years old, but 45% of these patients lack access to medical care providers who are familiar with their condition.²⁹ The concept of transition should be introduced early, around 12 years of age according to the American College of Cardiology and the American Heart Association guidelines,¹ and the transition should continue to be addressed throughout adolescence. Early introduction of transition allows both the patient's family and health care providers to advocate for the patient's independence over time until the patient is mature enough developmentally to transition to the adult system or the care provider for the cognitively impaired patient is prepared.³⁰ Introducing transition early also allows time to establish a primary care physician or a medical home that can be at the center of the patient's care and provide preventative, as well as acute and chronic illness management.²⁹ The primary care physician should maintain comprehensive medical records on this patient and coordinate care between consulting services.²⁹ There should be a formal transition curriculum with transition readiness assessments and checklists of things that should be accomplished by the patient and family before the transition can occur.^{8,30}

Nurses Aiding in the Transition Process

In an attempt to limit secondary complications, patients are seen by their cardiologists regularly, maintain a healthy lifestyle with certain physical restrictions, and adhere to personalized medication regimens.⁸ According to the American College of Cardiology and the American Heart Association guidelines,¹ adults with CHD should be seen every 6 to 24 months, depending on the complexity of their condition. In order for patients to comply with these demands, becoming knowledgeable about their disease is necessary, and nurses can begin this communication while caring for these patients. Adolescents have trouble adhering to medication regimens

and often are deemed noncompliant, but this behavior may be due to the lack of abstract and mature thinking during this stage of development.³¹ A comprehensive, developmentally appropriate program to educate and prepare these patients is necessary.⁸ Teaching adolescent patients with CHD about their diagnosis and anatomy should be part of their routine care, including red flags and warning signs so that they can recognize the signals that their condition may be worsening or that it is time to seek medical care.¹⁶ They should be in charge of their own care, including knowing which physician to call and having quick access to their contact information, with the backup support of their families.²³ Berg and Hertz³² created nursing competencies in exercise intolerance, pregnancy and contraception, nutrition, and future risks including endocarditis, and they led an adolescent transition clinic to improve overall independence of CHD patients. Studies by Mackie et al³³ and Berg and Hertz³² show that nurse-led education improves self-management skills, knowledge of cardiac disease, and self-advocacy. These initiatives support the idea that nurse-led education can improve the process of transition.

Once a child reaches the age of maturity with the ability to understand his or her disease, beginning the education process for transition should become a priority for the entire health care team.^{27,34} If a patient has deficits in cognitive functioning, a surrogate should be appointed.¹ The Americans With Disabilities Act amendment of 1997 requires transition teaching to start at the age of 14 as part of yearly education, as well as provision of guidance at each clinic appointment at a developmentally appropriate level.¹⁵ Few cardiology programs throughout the United States and Europe have structured programs to prepare patients for transition.³⁴ Structured education should include counseling on topics such as sexuality, contraception, reproductive issues, and risks of smoking and alcohol use.²³ Information should also be provided about their medications, including the need for the medications, their desired effect, when and how to take them, and adverse effects, leading to improved levels of health knowledge.²⁷ Patients should know their medication schedule and be in charge of asking the nurse for their medications while admitted to the hospital, to aid in building their independence. The nurse becomes their safety net should they forget or not be well enough to do this. The nurse should also encourage the patient's family to allow the patient to vocalize his or her needs and

Table Transition curriculum tool

Can you provide the following information?	Date accomplished
Describe your diagnosis.	
What surgeries have you had?	
What are your allergies?	
What medications are you taking (dose and reason)?	
Do you ask the nurse for your medication when it is due?	
Have you filled your prescriptions (pharmacy information)?	
Are you sexually active? Are you using contraceptives? Do you understand the effects of immunosuppression on contraceptives?	
How often do you need follow-up care?	
Have you made your next follow-up appointment (provider, date)?	
How do you access your primary care provider (name and phone number)?	
How would you access emergency health care?	
Which signs and symptoms should make you call the doctor?	
Are there foods you should not eat because of medication interactions?	
What are your exercise restrictions?	
How much weight can you gain or lose before needing to call the doctor?	

take control of his or her care.⁴ In the presence of their parents before discharge, adolescent patients should obtain their prescriptions with instructions on how to fill them at the pharmacy and information on administration.³⁰ This process will enhance the patient's overall independence.

Nurses can promote patients' independence by speaking directly to the patient while also being inclusive of the whole family when discussing the patient's care.⁸ Nurses and patients should advocate for patient-centered rounds inside the room, involving the patients and allowing them to ask questions about their care. Nurses can be an integral part of preparing these patients for the transition process, by using a preparation tool (see Table), at the times of admission and discharge as well as at outpatient clinic visits to assess patients' level of knowledge. This tool is provided only as a general guideline and can be modified if certain questions are not

relevant to the patient. This information would be the responsibility of the primary cardiologist to gather and analyze. Once patients can prove they have an understanding of their clinical history and are taking responsibility for their care, the provider can gauge when to transition them to an adult center.³⁰ Further education and formal programs are necessary to ensure a safe, efficient, and smooth transition. Currently only one-third of centers provide structured preparation for transition of care.³⁴

Structure of Transition Curriculum

This transition curriculum should include patient education, promotion of self-management, family support, coordination and collaboration of care between pediatric and adult providers, and regular follow-up and screening.^{23,35} It is vital that all aspects of the transition model be established and used in order to promote a successful transition of care. Many patients with CHD do not have a true understanding about their diagnosis, how transitioning can affect their medical care, and what to expect when visiting an adult clinic.²⁷ The program should ensure that young adults understand their diagnosis and medical history and are able to recognize important and concerning signs and symptoms.³⁴ In a study by Van Deyk et al,²⁷ only 21% of adolescents could define

Patients should transition to specialized centers for adult CHD, of which there are currently 70 in the United States.

endocarditis and only 1 patient could decipher the signs and symptoms

of endocarditis. The transitioning adolescents should have a good understanding of the rationale for previous interventions and options for future medical, surgical, and catheterization laboratory therapies. With the help of their pediatric cardiology team, patients need to create a passbook where their health information is easily accessible and comprehensive, giving the adult providers a roadmap of their condition.²³ Finally, patients must be taught to navigate the adult health care system: how to access an adult CHD specialist, how often they need to follow up, how to access routine health care and emergency health care, and how to navigate the insurance system.^{8,34}

Family Support During the Transition

Patients' families can assist in this process by allowing their child to become more independent and

autonomous.³⁶ There should be a forum for families to vocalize their concerns and fears, as well as education to help families understand the role they play.²³ In a qualitative study,³⁷ 41% of parents were apprehensive about their child going to the adult system and only 45% thought that their child was ready to take on the responsibility of their own health care. Ideas for families to support their child include having child patients schedule their own visits and be responsible for administering and refilling their medications. Providers should have them practice independence in the pediatric office, by directing questions to the patient, not family members, or having portions of the visit where the provider speaks with the patient alone.²³ The goal is for child patients to have a full understanding of their allergies, medications, medical history, and chronic conditions; to know their baseline health status so they can gauge threatening signs and symptoms; and to have access to their specialists, service providers, and the pharmacy.²³ Patients should carry their information with them, including the health insurance card, providers' phone numbers, and a medication list, should they need to access health care when away from family.²³ Being developmentally ready for the transition process is extremely important for the success of these patients, and it is essential to begin preparing them early to be independent and to have a formal program to educate the health care team, the patient, and the patient's family on how to achieve a smooth transition.

Future Direction

More educational programs for fellows, nurses, and ancillary services are needed to improve familiarity with the pathophysiology of adult patients with CHD to ensure smooth transitions from pediatric care. According to Webb and Williams,³⁸ patients should transition to specialized and comprehensive centers for adult CHD, of which there are currently 70 in the United States. These specialized centers support regionalization of care, providing a combination of both pediatric and adult CHD specialists, as well as a comprehensive team of subspecialists who are comfortable with caring for comorbid conditions of patients with CHD.³⁸ Education for nurses and ancillary staff on supporting the transition process and preparing adolescents should be a part of hospitals' yearly education.

Creation of formal transition programs is necessary to ease adolescent patients and their families into the adult health care world.

Conclusion

Courtney continued to have complications and required a third valve replacement procedure, including replacement of both her aortic and tricuspid valves with mechanical valves. She also needed replacement of her pulmonary valve in hopes of improving right-sided heart failure, which led to development of an arrhythmia, creating the need for an internal cardioverter defibrillator (ICD). The ICD required close follow-up with an adult electrophysiology team, which prompted her transition into the adult system at 30 years old. Courtney describes the process as a breakup and an emotional time, leaving the safety net of her pediatric providers, as well as the family of nurses and staff who had cared for her throughout her lifetime. Once some time had passed in the new adult environment, and she had adjusted to the independent role, Courtney was able to find a group of amazing nurses and providers who have become familiar with her and her history. She has required a total of 4 ICD replacements, has had multiple infections, including endocarditis and replacements of her 3 repaired valves, as she is not a candidate for a transplant. She has come out stronger from each roadblock, with a new team of care providers to support her through each step and to encourage her to remain positive.

Courtney struggled with leaving her pediatric care team, but overall had a good experience transitioning and felt that the idea of transition could have been presented to her much earlier to ease the discomfort of change. She was able to become her own advocate, was educated on her disease and medications, carries appropriate information with her should something go wrong, and joined a support group to meet people in similar circumstances. Although she is doing well, she would have liked a formal process to obtain these skills earlier and ensure a smoother transition. Courtney has accepted that she is a CHD patient and a survivor, but she understands that this diagnosis is not going away and requires lifelong support of a multidisciplinary adult team that can care for her as she continues to move forward. **CCN**

Financial Disclosures
None reported.

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