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Lost in Transition... The Importance of Follow-Up Care for the Pediatric Patient Transitioning to Adult Care in Regard to Congenital Heart Disease (CHD)

Abstract

Lost in transition discusses the importance of follow-up care in regard to the pediatric patient with Congenital Heart Defect (CHD) transitioning to adult-centered care. Congenital heart defect affects approximately 8 out of 1,000 live births. More than 1 million adults are now living with CHD. Advances in medicine and surgery have allowed children born with CHD to live longer, fuller lives, necessitating proper care into adulthood. The American Heart Association as well as the American College of Cardiology has issued guidelines stating that adults with CHD be cared for in "Regional Centers For Excellence" with expertise in Adult Congenital Heart Disease. This concept is not a new one, however there is not a visible presence of a qualified "Center of Excellence" in our region, even though Kansas City is home to the only pediatric heart specialty hospital in a 150 county region. There are 11 centers participating in a two-year study hoping to make an impact on the education and treatment of this patient population, while learning why there are lapses in care for these adults. We must start by providing a place to go for resources and continue to be a support network for the transitioning patient.

It was a surprise to me when I discovered an interest in congenital heart disease (CHD). After all, throughout my childhood I was told I would lead a "normal" life with few foreseeable problems and may or may not need further intervention as I aged. I would undoubtedly grow up, go on to have children and likely have a "normal" life expectancy. So as I grew up, I played softball, broke my left pinky finger, and even tried track and field. I also experimented (much to my parents chagrin) with tobacco and alcohol. I was a terrible athlete and an even worse delinquent. I never got the hang of running and did outgrow the desire for nicotine. Outside of a couple of impressive scars, I rarely considered my status as a CHD kid. This is in part why; as I headed toward adulthood I was lackadaisical in regard to my own care as a young person with a complex medical history.

"Congenital heart disease is defined as "a gross structural abnormality of the heart or intrathoracic great vessels that is actually or potentially of functional importance" (JALKUT..., 2009).ⁱ About one million adults and 800,000 children are living with CHD in the United States alone (Wood, 2009)ⁱⁱ. With these numbers, it is evident that there are currently *more* adults living with these conditions than there are children being born with congenital heart defects. This fact is due to advances in medicine that have made it possible for babies born with certain defects to be repaired and go on to live into adulthood. The defect I was born with is called Tetralogy of Fallot, which is a diagnosis of 4 defects grouped together. It is a very common cyanotic congenital heart defect and was, more or less, easily repaired when I was a child. This condition consists of VSD or ventricular septal defect, pulmonary artery stenosis, overriding aorta, and enlarged heart (right ventricle). I had two major surgeries as a baby: the first, at 9 months, was a palliative bypass of the faulty ventricle. This procedure would only partially repair the defect (bypassed the improper flow of deoxygenated blood systemically which flowed via the VSD) so that my body would grow larger and be able to tolerate the more invasive corrective open-heart surgery, which for me came at age 3. This surgery corrected the underlying VSD

and stenosed pulmonary valve by placing a Teflon patch over the VSD, opening the stenosed pulmonary artery/valve and correcting the position of the aorta.

The second most common CHD is Transposition of the Great Vessels. (American Heart Association)ⁱⁱⁱ This defect is also present at birth and consists of just what the name suggests; the Great arteries that feed the lungs and body from the heart are transposed or reversed. Normally, the pulmonary artery pumps oxygen *poor* blood that has returned from the body back into the lungs for oxygenation and the oxygen *rich* blood is pumped from the heart via the aorta out to the body. In TGA, these arteries are connected to the heart exactly opposite, the aorta is connected to the right ventricle and oxygen *poor* blood is pumped back to the body, the pulmonary artery is connected to the left ventricle and pumps oxygen *rich* blood back to the lungs. (Lucille Packard Children's Hospital at Stanford)^{iv} With this defect, there may be a VSD and pulmonary valve obstruction, but not always, and in fact this defect may go undetected into adulthood. Surgery is not usually necessary for children with TGA unless there is a large VSD or severe pulmonary valve obstruction. However, in adulthood there could be a myriad of problems such as congestive heart failure, (due to the fact that the right heart is pumping to the body) valve leakage and possible arrhythmias. (American Heart Association)ⁱⁱⁱ

A very serious, rare, cyanotic CHD called HLHS or hypoplastic left heart syndrome leaves children either without a left heart altogether, or with a severely underdeveloped, underfunctioning left heart (ventricle, aorta and mitral valve.) (American Heart Association)^v These children may appear normal at birth, but soon after appear bluish or ashen in color and may have difficulty breathing or eating. (American Heart Association)^v Because the left heart is not functioning correctly, blood returning from the lungs is pumped to the body via the *ductus arterioles* (a patent duct between chambers present at birth, but usually closes within days after birth). (American Heart Association)^v For children with HLHS, this duct is kept open with intravenous medication and surgery is necessary soon after birth or this defect is fatal. (American Heart Association)^v HLHS is not correctable,

however children can survive with multiple surgeries or a heart transplant.^v Adults living with HLHS will need lifelong care, including possible cardiac medicine, future catheterizations and possibly more surgery. (American Heart Association)^v

Corrected heart defects are not without long-term implications. While many of these procedures are beneficial and life-saving, sometimes the corrective procedures bring about a sequelae of new problems. (Allen, 2009)^{vi} These problems can include arrhythmias, valvular insufficiency, ventricular function abnormalities, abnormal vessel structure or resistance as well as non-cardiovascular problems such as, pulmonary complications, decreased CNS function and increased risk with dental procedures to name a few. (Allen, 2009)^{vi} These patients are often left with unique anatomy and physiology, along with the sequelae requiring lifelong specialized treatment and follow-up. (Allen, 2009)^{vi} Recently the American Heart Association along with the American College of Cardiology issued practice guidelines calling for coordination of ongoing care through "Regional centers of excellence" with expertise in Adult Congenital Heart Disease (ACHD). (Wood, 2009)ⁱⁱ This is important because patients tend to want to stay on with their pediatric cardiologist, however this can present problems because these patients need practitioners who are skilled with adult comorbidities such as diabetes and hypertension, and who can also counsel patients on matters such as pregnancy, employment, physical activity, etc. (Wood, 2009)ⁱⁱ

The concept of transition is not a new one; it is familiar to both the adult clinical team and the pediatric team. (Allen, 2009)^{vii} The transition process is not simply switching from pediatric care to adult care at a specific age, but a gradual process when the adolescent patient begins to take on the issues surrounding their own health care.¹ This transition can be described as "a purposeful, planned process that addresses the medical, psychosocial, and educational/vocational needs of adolescents and young adults with chronic physical and mental conditions, as they move from child-centered to adult-oriented health care systems." (Allen, 2009)^{vii}

Some of the barriers associated with transition range from the basic understanding of one's illness/condition to insurance issues. The current healthcare system is complicated to navigate without the added frustration a young adult with CHD must also face. (Allen, 2009)^{vii} Some young adults find they are no longer covered by their parents' insurance, or must face pre-existing laws when applying for their own insurance. (Allen, 2009)^{vii} Another issue for these patients is the lack of qualified adult specialty care centers. Typically providers in this specialty are affiliated with major medical institutions and academic centers which are often in major metro areas and mostly in the Midwest, West and Northeast. This fact presents an issue of travel, expenses and convenience for some patients. (Allen, 2009)^{vii}

On the psychosocial level, children are often more secure with their pediatric clinical team, whom they have grown to trust and have built a very long relationship with. (Allen, 2009)^{vii} They may feel insecure about continuing their care with a new clinical team. Conversely the parents of CHD adolescents may feel uneasy about breaking in a new provider as well as shifting the healthcare responsibility to their child as they mature. (Allen, 2009)^{vii} Adult cardiologists too may feel unsure about their abilities to properly care for adult patients with CHD because of inadequate training and/or experience with transition. (Allen, 2009)^{vii}

There are two lines of thought in regard to the training and role of clinical providers caring for adult CHD patients. Most cardiologists and advanced practice nurses believe that specialty providers should be trained specifically in CHD. (Allen, 2009)^{viii} However, some believe that they should be trained as pediatric specialists with an added specialty in adult congenital heart defects and follow the patients throughout their life. (Allen, 2009)^{viii} This can present problems, however, with the impracticality of seeing adult patients in a pediatric setting. And what if a CHD patient should wish to become pregnant? This would require special monitoring at a collaborative level between trained cardiologists and gynecology/obstetrics providers as well. (Allen, 2009)^{viii} Contrary to this idea is that adult

cardiologists be specifically trained in CHD. However, studies show that, currently, fellowship programs for CHD vary widely. Some fellows were receiving as little as 3 hours of didactic lecture for treating adult patients with CHD. (Allen, 2009)^{viii} This data suggests that there should be some formalized training at major medical institutions to prepare clinical providers to meet demand for this patient population, and at this point there is very little uniformity in the care received. (Allen, 2009)^{viii}

Nurses are often part of the interdisciplinary team and aid the patients with anxiety and education about their disease. (Wood, 2009)ⁱⁱ A staff RN could help the patient understand their medical history, provide paperwork and resources for education and explain what to expect during check ups and procedures. An advanced practice nurse may be more involved with the transition process and could act as a liaison between pediatric care and adult care. Specifically, a PNP (Pediatric Nurse Practitioner) could act as a transition coordinator. (Allen, 2009)^{ix} Current studies suggest this model is most successful for the transitioning patient. (Wood, 2009)ⁱⁱ

The PNP would be assigned to the transitioning patient from the setting of a pediatric cardiology clinic or transition clinic and would be the primary contact for both providers and family to insure that care is not interrupted. (Allen, 2009)^{ix} Because PNP's as a transition coordinator are so closely involved with all aspects of managing these patients, they have the knowledge and skills needed to properly educate and provide family anticipatory guidance. (Allen, 2009)^{ix} The primary role of the PNP would be centered on health education. (Allen, 2009)^x Education of the adolescent patient would be taught one-on-one and through seminars or group learning, support groups, written literature and education materials. (Allen, 2009)^x The PNP would be responsible for networking the patient with social workers, who may help with insurance and vocational needs. (Allen, 2009)^x The PNP would also provide all contact information for the adult care providers as well as documenting all visits and correspondences with the patient throughout transition.

(Allen, 2009)^x A "pre-transfer" visit to the adult center has been recommended as well, with optimally one provider from the pediatric clinic present if possible. (Allen, 2009)^x

To be qualified as a transition coordinator, the PNP must have extensive knowledge of congenital heart defects. The PNP would be expected to begin by educating the patients regarding the specific medical diagnosis in a manner that they can understand as well as reassurance to the child and family of future medical care. (Allen, 2009)^{xi} Establishing trust with family members and patients is important but also the PNP would establish trust with adult providers, because families are more likely to trust the adult care provider if they can perceive a trusting relationship between the pediatric care provider and adult care provider. (Allen, 2009)^{xi} Some abilities that the PNP would be expected to have: Reassure families there will be no interruption of care, build confidence in the adolescent patient, and recognize lack of trust or fear that the young patient may have in regard to transition. (Allen, 2009)^{xi} The PNP would lay a foundation for the adolescent that encourages education and self-care, and explains the difference between pediatric and adult care, stressing the importance of "medical responsibility" for the patient's future. (Allen, 2009)^{xi} Once the time of transfer has been decided, the PNP would prepare all necessary documents and provide copies to both the patient's primary care physician and the adult congenital heart defect clinical provider. (Allen, 2009)^x Ideally, the PNP would have scheduled contact with the patient to insure he or she has followed up with the appropriate clinical providers. (Allen, 2009)^x The PNP should expect some communication early on from the adult cardiac clinical provider regarding the successful transition of the patient. (Allen, 2009)^x

Currently there are 11 U.S. centers participating in a two year, National Institutes of Health-funded Health Education and Access Research Trial in Adult Congenital Heart Disease study known as HEART-ACHD. (Debra Anscombe Wood, 2009)ⁱⁱ The purpose of this study is to learn why there are lapses in care for CHD patients as they age out of pediatric care. (Wood, 2009)ⁱⁱ The study also hopes to impact education, treatment and research for this patient group. (Wood, 2009)ⁱⁱ There is one significant nationwide advocacy group for

adult congenital heart patients. ACHA, or Adult Congenital Heart Association is a national non-profit organization founded in 1988 in Pennsylvania by a group of CHD survivors and their families. (ACHA)^{xii}

Locally, there is a non-profit organization based in Kansas City called "CHD Families" (CHD Families)^{xiii} which aims to support families stricken by congenital heart defects. In Kansas City I found only one provider that listed congenital heart defects as an area of expertise and service. (Kansas University Medical Hospital)^{xiv} This statistic alarmed me, because I know of at least one other provider, however his hospital affiliation/website did not provide any information that he is a specialist in ACHD. This tells me that there is much work to be done not only nationally but locally, considering Kansas City is home to Children's Mercy Hospital: the *only* pediatric heart specialty hospital in our 150 county region. (Children's Mercy Hospital, 2010)^{xv} With the guidelines now being specified by the American Heart Association and the American College of Cardiology that care of patients with congenital heart defects be coordinated between pediatric and adult providers through centers of excellence, it is evident there is a need to insure that this patient population can be served by a proper level of care in their hometown, and not be left to travel great distances or forego care altogether, as has been the trend. (Wood, 2009)ⁱⁱ

My interest lies in assuring that young patients with CHD in Kansas City are afforded the proper guidance and are empowered to take charge of their health and future. In order to accomplish this, we must, as a medical community, provide them somewhere to "land" so to speak, in the adult care arena. We must get on board with the national guidelines and pave the way for appropriate care for this complex population to avoid pitfalls for those who would otherwise be lost in transition.

Bibliography

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- i {JALKUT..., Transition from Pediatric To Adult Health Care For Adolescents with Congenital Heart Disease: A Review Of the Literature and Clinical Implications. *Pediatric Nursing*. Pitman. Nov/Dec. 2009; Vol 35, Issue 6; pp. 381
- ii (WOOD, DA, Follow Your Heart: Congenital Heart Survivors Lack Adequate Care. *Nursing Spectrum*. Mar. 2009; Vol 22, Issue 5)
- iii (American Heart Association)http://www.heart.org/HEARTORG/Conditions/CongenitalHeartDefects/AboutCongenitalHeartDefects/d-Transposition-of-the-great-arteries_UCM_307024_Article.jsp
- iv (Lucille Packard Children's Hospital at Stanford)<http://www.lpch.org/DiseaseHealthInfo/HealthLibrary/cardiac/tga.html>
- v (American Heart Association)
http://www.heart.org/HEARTORG/Conditions/CongenitalHeartDefects/AboutCongenitalHeartDefects/Single-Ventricle-Defects_UCM_307037_Article.jsp
- vi (Allen, 2009) Transition from Pediatric To Adult Health Care For Adolescents with Congenital Heart Disease: A Review Of the Literature and Clinical Implications. *Pediatric Nursing*. Pitman. Nov/Dec. 2009; Vol 35, Issue 6; pp. 382
- vii (Allen, 2009) Transition from Pediatric To Adult Health Care For Adolescents with Congenital Heart Disease: A Review Of the Literature and Clinical Implications. *Pediatric Nursing*. Pitman. Nov/Dec. 2009; Vol 35, Issue 6; pp. 383
- viii (Allen, 2009) Transition from Pediatric To Adult Health Care For Adolescents with Congenital Heart Disease: A Review Of the Literature and Clinical Implications. *Pediatric Nursing*. Pitman. Nov/Dec. 2009; Vol 35, Issue 6; pp. 384
- ix (Allen, 2009) Transition from Pediatric To Adult Health Care For Adolescents with Congenital Heart Disease: A Review Of the Literature and Clinical Implications. *Pediatric Nursing*. Pitman. Nov/Dec. 2009; Vol 35, Issue 6; pp. 385
- x (Allen, 2009) Transition from Pediatric To Adult Health Care For Adolescents with Congenital Heart Disease: A Review Of the Literature and Clinical Implications. *Pediatric Nursing*. Pitman. Nov/Dec. 2009; Vol 35, Issue 6; pp. 386
- xi (Allen, 2009) Transition from Pediatric To Adult Health Care For Adolescents with Congenital Heart Disease: A Review Of the Literature and Clinical Implications. *Pediatric Nursing*. Pitman. Nov/Dec. 2009; Vol 35, Issue 6; pp. 387-Table 2.
- xii (ACHA) <http://www.achaheart.org/about/>

xiii (CHD Families) <http://www.chdfamilies.org/index.html>

xiv (KU Med Hospital) <http://www2.kumc.edu/internalmedicine/cv/kmulhern.html>

xv (Children's Mercy Hospital, 2010) <http://www.childrensmercy.org/cardiology/>