



Adult congenital heart disease nurse coordination: Essential skills and role in optimizing team-based care a position statement from the International Society for Adult Congenital Heart Disease (ISACHD)



Christina Sillman ^{a,*}, Joanne Morin ^b, Corina Thomet ^c, Deena Barber ^d, Yoshiko Mizuno ^e, Hsiao-Ling Yang ^f, Theresa Malpas ^g, Serena Francesca Flocco ^h, Clare Finlay ⁱ, Chi-Wen Chen ^j, Yvonne Balon ^k, Susan M Fernandes ^l

^a Lucile Packard Children's Hospital Stanford, Palo Alto, CA, United States

^b University of Ottawa Heart Institute, Ottawa, Ontario, Canada

^c Center for Congenital Heart Disease, Inselspital, Bern University Hospital, Bern, Switzerland

^d Akron Children's Hospital, Akron, OH, United States

^e Chiba Cardiovascular Center, Japan

^f National Taiwan University, Taipei, Taiwan

^g The Prince Charles Hospital, Adult Congenital Heart Disease Service, Brisbane, Australia

^h Pediatric and Adult Congenital Heart Centre, IRCCS Policlinico San Donato, San Donato Milanese (MI), Italy

ⁱ Royal Brompton Hospital, London, United Kingdom

^j National Yang-Ming University, Taipei, Taiwan

^k Peter Lougheed Hospital, Calgary, Alberta, Canada

^l Stanford University, Palo Alto, CA, Departments of Pediatrics and Medicine and Divisions of Pediatric Cardiology and Cardiovascular Medicine, United States

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ABSTRACT

Background: Founded in 1992, the International Society for Adult Congenital Heart Disease (ISACHD) is the leading global organization of professionals dedicated to pursuing excellence in the care of adults with congenital heart disease (CHD) worldwide. Among ISACHD's objectives is to "promote a holistic team-based approach to the care of the adult with CHD that is comprehensive, patient-centered, and interdisciplinary" (<http://www.isachd.org>). This emphasis on team-based care reflects the fact that adults with CHD constitute a heterogeneous population with a wide spectrum of disease complexity, frequent association with other organ involvement, and varied co-morbidities and psychosocial issues.

Methods: Recognizing the vital role of the adult CHD (ACHD) nurse coordinator (ACHD-NC) in optimizing team-based care, ISACHD established a task force to elucidate and provide guidance on the roles and responsibilities of the ACHD-NC. Acknowledging that nursing roles can vary widely from region to region based on factors such as credentials, scopes of practice, regulations, and local culture and tradition, an international panel was assembled with experts from North America, Europe, East Asia, and Oceania. The writing committee was tasked with reviewing key aspects of the ACHD-NC's role in team-based ACHD care.

Results/conclusion: The resulting ISACHD position statement addresses the ACHD-NC's role and skills required in organizing, coordinating, and facilitating the care of adults with CHD, holistic assessment of the ACHD patient, patient education and counseling, and support for self-care management and self-advocacy.

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1. Organizing, coordinating, and facilitating care for ACHD patients

Teams caring for adults with CHD may involve ACHD program administrators, psychosocial support services, advanced practice

providers (nurse practitioners, physician assistants), ACHD physicians and congenital heart surgeons, and other medical consultants. As the liaison for care coordination, organization, and facilitation, the ACHD-NC can play a key role in bridging gaps in patient care. Care coordination involves managing patient load, facilitating partnerships within the medical facility and community, assisting in the organization of active patients, and ensuring timely communication of medical results. A common model is for the ACHD-NC to provide the first line of care.

* Corresponding author at: Lucile Packard Children's Hospital, 700 Welch Road, Palo Alto, CA 94304, United States.

E-mail address: CSillman@stanfordhealthcare.org (C. Sillman).

Consistent patient interactions are ideal in helping to orchestrate care in a cohesive manner, with the objective of improving patient safety and satisfaction.

1.1. Managing patient calls

To triage patient calls, it is helpful to have a call-back line, receptionist, or phone tree. Calls regarding acute, active symptoms should be directed to the ACHD-NC. Ideally, voicemail greetings should let patients know when calls will be returned (e.g., end of the business day) and provide options regarding paging an ACHD provider or presenting to the emergency room if patients feel their symptoms require prompt attention. The ACHD-NC utilizes critical assessment to prioritize phone calls, as described in the triage section. Questions or issues that require adjustment to the established care plan (e.g., medication changes, urgent appointments, or testing) should be discussed with the appropriate practitioner (e.g., advanced care provider [APP], ACHD fellow, or attending ACHD cardiologist). The ACHD-NC should educate clerical staff taking calls from patients regarding symptoms that require punctual assessment by ACHD team members or emergency services. For example, if the ACHD service utilizes an automated phone tree, the first part of the message could state, “If you are having a life-threatening emergency, you should hang up and call emergency services.”

1.2. Care coordination

The ACHD-NC plays an essential role in facilitating communication with ancillary and subspecialty services. The ACHD-NC should develop a comprehensive contact and subspecialty list that includes cardiac subspecialists such as electrophysiologists, interventionalists, congenital heart surgeons, and heart failure and transplant specialists, as well as internal medicine subspecialists. At a minimum, specialty services typically include a pulmonologist, gastroenterologist, hepatologist, hematologist, nephrologist, endocrinologist, obstetrician/gynecologist, geneticist, psychiatrist, and psychologist. A contact for dental issues, including a general dentist and oral surgeon should be included, as well as a contact for resources such as social services. The ACHD-NC should also attend multidisciplinary meetings that discuss patient care.

Care coordination can be enhanced by the development of an “active patient” worksheet or database, as illustrated in Fig. 1. This list can include inpatients, recently discharged patients, those with upcoming procedures, outpatients with active issues (including those who require close follow-up for heart failure), pregnant women, and patients

awaiting transplantation. The “active patient” list should be updated and reviewed with the ACHD team on a regular basis. It can also serve as a tool to promote discussion with the ACHD team as a whole, especially when there are multiple providers, and can be a useful resource for ACHD providers when on call or when urgent concerns arise.

Coordination of study scheduling, results review, ACHD team discussion, and patient notification are also among the responsibilities of the ACHD-NC. Initiation of this coordination process can begin shortly after the outpatient visit is completed by the ACHD provider for follow-up testing or referral to other subspecialists. A study tracking sheet, database or electronic reminder system is useful in assisting this process. Regardless of the type of tracking system, it should include basic information such as patient demographics, the study or subspecialist referral requested and its urgency, and relevant dates (e.g., when studies or referrals were ordered, scheduled, performed, and available for review, and when results were communicated to the ACHD provider). A system for urgent notification of critical results must be in place. Providing guidelines for urgent notification and a pager or contact number can help expedite clinical care. Clinical discussion of non-urgent results should be undertaken with the ordering ACHD provider.

1.3. Cardiac symptoms: triage

The role of the ACHD-NC includes the front-line triage of patients either via telephone, electronic messaging, or in person. Quick assessment of the urgency of medical attention required is imperative in optimizing patient outcomes. In triaging patients, it is recommended that the ACHD-NC first determine if the questions/concerns/requests are medical or non-medical in nature. Non-medical issues such as scheduling, insurance, or billing should be delegated to the appropriate ACHD team member (e.g., clerical or social work staff). Patients with non-cardiac medical issues should be referred to their primary care provider or appropriate caregiver. Cardiac symptoms should be assessed by the ACHD-NC and the urgency of treatment determined. A list of common symptoms and suggested nursing actions are provided in Table 1.

2. Holistic assessment of the ACHD patient

Holistic nursing has been described as a “form of healing that gives attention to all aspects of an individual, including physical, mental, emotional, sexual, cultural, social, and spiritual [1].” The holistic assessment

Adult Congenital Heart Active Patient List										
05/28/2015										
Inpatient/Surgical Patients										
Patient	DOB	Age	MRN	Dx	MD	Srg/Admt	Comments	Expected discharge	To do	SW needs
XXXXXX	XXX	XX	XXXXXXXXXX	XXXX	XX	XXX	XXX	XXX	XXX	
Recently Discharged/ED										
Patient	DOB	Age	MRN	Dx	MD	Surgery/Admis	Hx	Date of Discharge	Follow-up date	To Do
XXXXXX	XXX	XX	XXXXXXXXXX	XXXX	XX	XXX	XXX	XXX	XXX	
Upcoming Surgical/Procedure										
Patient	DOB	Age	MRN	Dx	MD	Date	comments	Plan	To do	SW Needs
XXXXXX	XXX	XX	XXXXXXXXXX	XXXX	XX	XXX	XXX	XXX	XXX	
Outpatient Patients										
Patient	DOB	Age	MRN	Dx	MD	Last Visit	Current Status	Plan	To do	SW Needs?
XXXXXX	xxx	xx	XXXXXXXXXX	XXXX	xx	xxx	xxx	xxx	xxx	xxx
Pregnant Patients										
Patient	DOB	Age	MRN	Dx	MD	EDD	Current Status	Plan/ Delivery locatio	To do	SW Needs?
XXXXXX	xxx	xx	XXXXXXXXXX	XXXX	xx	xxx	xxx	xxx	xxx	
Heart Failure Patients										
Patient	DOB	Age	MRN	Dx	MD	Last Visit	Current Status	Plan	To do	SW Needs
XXXXXX	xxx	xx	XXXXXXXXXX	XXXX	xx	xxx	xxx	xxx	xxx	
Transplant Listed patients										
Patient	DOB	Age	MRN	Dx	MD	Last Visit	Comments	Plan	Cath status	SW Needs
XXXXXX	xxx	xx	XXXXXXXXXX	XXXX	xx	xxx	xxx	xxx	xxx	

Fig. 1. Sample of an ACHD program Active Patient List utilized by the nurse coordinator to track active patients.

Table 1
Common ACHD Symptoms and suggested nursing action.

Patient-reported symptom	Nursing action
Syncopal episode or pre-syncopal symptoms	Call for emergency services (ambulance) or urgent family transport to nearest hospital if emergency services are unavailable
Sustained arrhythmia with accompanying symptoms (dyspnea, chest pain, lightheadedness)	Call for emergency services (ambulance) or urgent family transport to nearest hospital if emergency services are unavailable
Sustained arrhythmia with no accompanying symptoms	Urgent appointment in ACHD clinic
Chest pain with accompanying symptoms (dyspnea, diaphoresis, lightheadedness)	Call for emergency services (ambulance) or urgent family transport to nearest hospital if emergency services are unavailable
Chest pain with no accompanying symptoms	Urgent appointment in ACHD clinic
Change in neurological status (decreased level of consciousness, sudden confusion, weakness or numbness on one side)	Call for emergency services (ambulance) or urgent family transport to nearest hospital if emergency services are unavailable
Respiratory distress (difficulty breathing, increased cyanosis)	Call for emergency services (ambulance) or urgent family transport to nearest hospital if emergency services are unavailable
Profuse bleeding (surgical site, post-injury for patients on anticoagulation)	Call for emergency services (ambulance) or urgent family transport to nearest hospital if emergency services are unavailable
Symptoms of infection/endocarditis (fever, diaphoresis, surgical site infections)	If patient stable, offer an urgent same-day appointment if possible. If patient unstable, call for emergency services (ambulance) or urgent family transport to nearest hospital if emergency services are unavailable. <u>Blood cultures to be drawn prior to any administration of antibiotics.</u>
Heart failure exacerbation (weight gain, increased edema, orthopnea)	Urgent appointment in ACHD clinic

provided by the ACHD-NC consists of a global patient appraisal that serves as the basis for integrated nursing care.

2.1. Psychological assessment

Psychological issues are common in ACHD patients, with mood or anxiety disorders prevalent in approximately 50% of the population [2]. The ACHD-NC can play an active role in identifying patients at increased risk for psychological disorders and should be aware of characteristics that may suggest the need for specialized mental health care. These include non-adherence with care and self-harming behavior or statements. Patients with known or suspected suicidal or homicidal ideation require emergent care; the ACHD-NC should adhere to local policies in coordinating care as needed. If such policies are not in place, the ACHD-NC can work with the clinical team to develop protocols. For patients with stable psychological disorders, the ACHD-NC can assist with establishing access to appropriate psychological care and work with social support services in addressing outstanding psychological care needs. The ACHD-NC should maintain a collaborative relationship with psychological practitioners, coordinate patient care to accommodate psychological needs, and help identify community resources for patients seeking care.

2.2. Social assessment

Social adjustment is a predictor of depression and anxiety in ACHD patients [2] and should be assessed by the ACHD-NC. Social assessment includes appraisal of interpersonal relationships, family dynamics, living arrangements, home environment (for safety), career/work, educational level, financial stability, quality of life, and health literacy. Considerations regarding supportive relationships should be assessed, especially for patients unable to independently care for themselves. In assessing financial stability, the ACHD-NC should identify any financial burden, such as health care costs, travel expenses, housing/hotel accommodations, and medical supplies that might be a barrier to care. The ACHD-NC should also assess each patient's health literacy, defined as, "the degree to which individuals have the capacity to obtain, process, and understand basic health information and services needed to make appropriate health decisions [3]." Social issues identified by the ACHD-NC that might impact the delivery of care should be discussed with social services staff in order to develop a comprehensive plan to address these issues.

3. ACHD patient education

3.1. Medication education

The ACHD-NC can play an important role in improving patient adherence to complex pharmacological treatment regimens via medication education, follow-up, and promotion of adherence. The ACHD-NC may discuss the rationale for starting new medications, side effects, and potential barriers to adherence. The latter can include financial constraints, limited access to a pharmacy, fear of side effects, lack of understanding of the rationale for medical therapy, cognitive deficiencies that preclude self-administration, and medication organization. To help manage complex treatment regimens, the ACHD-NC may assist the patient in developing a weekly medication schedule that is coordinated with other life responsibilities, such as work, family, and school. Follow-up within a few weeks of initiation of a new medication or after major dose adjustments of existing medications is essential in assessing compliance, side effects, barriers to adherence, and need for further dose titration.

3.2. Pre- and post-procedural education

The ACHD-NC can serve a key role in establishing relationships with intra-hospital services and in reviewing patient pre-procedural instructions for appropriateness and relevance to the unique needs of ACHD patients. Pre-procedural information includes patient arrival date and time, arrival location, fasting instructions, medication adjustments, anticipated length of procedure, expected patient experience, skin preparation needs, and items that should accompany the patient. The ACHD-NC can help identify barriers to the procedure, such as transportation issues, financial concerns, patient fears, or family concerns. Verbal, written, and demonstrative educational techniques could be utilized. Post-procedural instructions include follow-up appointment dates, signs and symptoms of post-procedural complications to monitor, physical restrictions or precautions, post-procedure medication regimen and potential side effects, and self-care guidelines. Table 2 summarizes common post-procedural instructions for ACHD patients.

3.3. Heart failure education

Heart failure is increasingly common in the aging population with CHD, with a highly variable trajectory. The ACHD-NC must be attuned to the indicators of heart failure in order to identify patients with new or worsening signs or symptoms. Symptoms such as unexplained

Table 2
Common ACHD Post-Procedural Instructions for ACHD Patients.

Procedure	Signs/symptoms to watch for	Physical precautions	Dietary restrictions	Self-care guidelines
Cardiac imaging (CT/MRI)	Allergic reaction to contrast dye	None	None	If given an anxiolytic, patient will need to be driven home
Cardiac interventional procedures: hemodynamic catheterization and electrophysiology catheter interventions	Bleeding from site, increased bruising at site, or leg numbness/perfusion issues. s/p intervention (closure device, stent, transcatheter valve placement) watch for worsening chest pain, dyspnea, or emergent symptoms	Rest for first 2 days, and then slowly increase activity. For femoral catheterization: do not lift >10 lbs. for first 5 days, avoid strenuous activity for 5–7 days	Avoid constipating foods	Monitor catheterization site for bleeding or signs of infection (increasing redness, hot, oozing pus, or fever)
Arrhythmia devices: pacemaker, defibrillators, and cardiac resynchronization therapy	Unusual swelling, redness, or drainage at incision site; watch for increased dyspnea, pain at site, or fever	Keep incision clean and dry for 5 days; Do not submerge incision in water, raise arm above shoulder level, or lift heavy objects (>15–20 lbs) for 4 weeks	Avoid constipating foods	Steri-strips should fall off spontaneously in 10–14 days No driving for 2–3 days (or longer if syncope or ventricular arrhythmia prior to device implantation)
Cardiac Surgery	Increasing shortness of breath, chest pain, numbness or tingling, edema/weight gain, uncontrolled pain, constipation greater than 5 days, and wound infections	Sternal precautions for 6 weeks: no lifting, pushing, pulling >10 lbs.; avoid excessive twisting or reaching Slowly increase activity	Low sodium (<2 g/daily) Fluid restrictions (<2 L/Day)	Monitor incision for signs of infection (increasing redness, hot, oozing pus, or fever) Medication management No driving for 6 weeks and while on narcotic medications. Do not submerge incision in water for 6 weeks Avoid crowds or infectious people for 2–3 weeks post-surgery

weight gain, lower extremity or abdominal swelling, early satiety, dyspnea with exertion or even at rest, decreased functional capacity, worsening palpitations, progressive fatigue, orthopnea, and paroxysmal nocturnal dyspnea should alert the ACHD-NC to the potential for heart failure. Once patients with heart failure are identified and the ACHD provider establishes a treatment plan, the ACHD-NC can work closely with the patient to ensure adherence. The ACHD-NC should tailor patient monitoring based on the degree of symptoms and intensity of therapy. In addition to close surveillance, strategies for optimizing care include educating patients regarding the physiology of heart failure, the need for fluid (e.g., <2 l per day or <1.5 l per day) and salt (e.g., <2 g per day) restriction, medication adherence, anticipated symptoms, and means of self-monitoring (e.g., daily weighing). Such a strategy can improve patient outcomes, including quality of life, need for hospitalizations, and health care costs [4].

3.4. Anticoagulation education

Depending on local practices, the ACHD-NC may coordinate anticoagulation management within the ACHD care team. Understanding indications for anticoagulation, common international normalized ratio (INR) ranges for different indications, educational needs, and precautions are critical in providing direction. Patient education should include dietary guidance (impact of vitamin K-containing foods on INR), adherence and interactions with other medications, and the importance of timely INR testing and requesting the result (the patient should be aware of the latest INR reading). In addition, the ACHD-RN should provide guidance around activities that may involve impacts, falls, or other injuries, and should assess risk of injury related to work duties, exercise activities, and recreational pursuits that may place the patient at risk of severe bleeding.

In some instances, ACHD patients on anticoagulants may need to temporarily discontinue therapy or transition to heparin for invasive procedures. In other circumstances, the procedure may be safely performed without interrupting anticoagulation. The ACHD-NC can

provide patient guidance, with support from the ACHD provider and/or from the Anticoagulation Clinic. For example, if oral anticoagulation must be stopped, bridging therapy may or may not be required with intravenous or low-molecular weight heparin. When indicated, the ACHD-NC may assist in teaching the patient how to self-administer subcutaneous injections, arrange for home visits, and help organize outpatient management for re-institution of anticoagulation therapy post-procedure [5].

3.5. Prevention and lifestyle education

ACHD patients are not immune from developing adult comorbidities, the impact of which may be substantial considering the prevalence of associated residual hemodynamic abnormalities and organ dysfunction [6]. Lanz et al. documented an increased risk of cardiovascular events [7], and Afilalo et al. reported an increased prevalence of myocardial infarction in older adults with CHD compared to the general population in Canada [8]. Billett et al. from the United Kingdom suggest that the ACHD population has an increased prevalence of renal dysfunction and systemic hypertension [9]. Although obesity (BMI ≥ 30 kg/m²) in the ACHD population appears less prevalent than in the general population, approximately 10% of ACHD patients are obese and 30% are overweight (BMI ≥ 25 kg/m²), suggesting that this is a health concern for this population as well [10,11]. A lack of physical activity may be a contributing factor, as at least 25% of patients with CHD do not meet international recommendations of ≥ 150 min of moderate-intensity aerobic physical activity per week, and muscle-strengthening activities at least twice a week [12]. Cancer risk may also be a concern for ACHD patients. A recent study demonstrated that cancer was one of the leading causes of mortality in a large cohort of ACHD patients (7%), [13], while cancer screening (e.g., mammogram, pap test, and colonoscopy) rates are lower than the general population [14]. The ACHD-NC has a real opportunity to impact on the future health of ACHD patients by providing education on the value of preventative screening and early detection, educating patients about current and

future risk, and offering guidance on lifestyle changes. In addition, the ACHD-NC can ensure that preventative screening tests are ordered, reviewed, and acted upon.

ACHD patients engage in high-risk behaviors at rates similar to the general population [15]. Unfortunately, ACHD patients as a group appear to lack awareness of the negative influences that these high-risk behaviors—such as alcohol use and tobacco or illicit drug use—have on their hearts and overall health [16,17]. Adolescents and young adults who display increased risk-taking behavior should be a priority for intervention, since they are at risk for developing a pattern of lifelong unhealthy lifestyle choices [16]. The ACHD-NC can help identify patients participating in high-risk behaviors. This tendency might be recognizable by non-adherence, missed appointments, and/or symptoms related to substance abuse. The ACHD-NC can provide education, resources, and referral for support as needed. A list of lifestyle choices and suggested nursing education can be found in Table 3.

3.6. Dental and skin care education

The 2015 European Society of Cardiology (ESC) guidelines for the management of infective endocarditis (IE) emphasize the importance of hygiene measures in dental and skin care [18]. Endocarditis is a life-threatening disease, with a 20% risk for mortality or significant morbidity [19]. Clinical symptoms include fever, chills, night sweats, and a general feeling of discomfort. The ESC recommends that prophylactic antibiotics be reserved for patients at high-risk for infective endocarditis-related complications, such as those with any prosthetic valve, including percutaneous valves and valves repaired with prosthetic material, a history of IE, and cyanotic CHD. Patient with CHD repaired utilizing prosthetic material (surgical or transcatheter) require 6 months of prophylaxis post-procedure or longer if a residual shunt or valve regurgitation is present. Antibiotic prophylaxis is recommended before dental procedures that will likely cause bleeding (e.g., gingival manipulation, perforation of oral mucosa) or, if invasive procedures are performed to treat infection. The ACHD-NC could help educate patients on the importance of having blood cultures drawn prior to antibiotic administration for diagnostic assessment. All ACHD patients should have their dental health assessed and the ACHD-NC should educate patients to brush their teeth twice a day, floss daily, and have regular check-ups with a dentist. Regarding skin care, the ACHD-NC can help educate patients on the risks associated with piercings and tattoos and the importance of ensuring that such procedures are performed under sterile conditions.

4. Contraception, family planning and pregnancy

Studies indicate that contraceptive and pregnancy counseling are not consistently delivered to women with CHD [20], with time constraints of the ACHD provider being a common limiting factor. The ACHD-NC is optimally positioned to ensure that this education is systematically provided to women of childbearing age and ability.

4.1. Contraception counseling

Although studies suggest that adolescents and young adults with CHD engage in sexual activity at rates lower than their peers [21], unplanned pregnancies are not uncommon in this patient population [22]. Contraception discussions should ideally be initiated prior to sexual activity, in the pre-teen years, in the pediatric setting. Nevertheless, it is common for patients to have never received contraception education before transitioning from pediatric to adult care [23]. While barrier methods are unreliable due to high failure rates, the importance of condoms should be emphasized for their role in preventing sexually transmitted infections. In assessing methods of contraception, considerations for women with CHD include the risk of thrombosis with estrogenic products (Table 4), the risk of endocarditis and vagal reactions with insertion of an intrauterine device, and potential maternal risks in the event of contraceptive failure [24]. Women with thromboembolic risk factors such as cyanosis with an obligatory right-to-left shunt, pulmonary hypertension, Fontan circulation, sustained arrhythmias, mechanical heart valves, and ventricular dysfunction should generally avoid contraceptive agents containing estrogen [25].

4.2. Family planning and pregnancy

Pre-pregnancy counseling is recommended for all women with CHD, considering the increased risk of maternal-fetal and obstetrical complications [26,27]. ACHD experts and maternal-fetal medicine specialists should provide maternal-risk assessment and counseling. In addition, genetic counseling should be an integral part of pre-pregnancy care given the increased and highly variable risk of anomalies in offspring. The ACHD-NC can be actively involved in these discussions, help to coordinate consultations, and be available to answer questions. In women for whom pregnancy is contraindicated, the ACHD-NC may provide resources and assist in discussions surrounding gestational surrogacy, ensuring that risk assessment and candidacy for egg stimulation and retrieval have been addressed. The adoption pathway may also be

Table 3
Lifestyle choices for ACHD patients and suggested nursing education.

Assessment	Educational needs
Smoking	<ul style="list-style-type: none"> - Promote smoking cessation - Refer to cessation assistance as available
Alcohol	<ul style="list-style-type: none"> - Encourage limited alcohol intake - Evaluate for alcohol contraindication with medications
Drug abuse	<ul style="list-style-type: none"> - Encourage cessation of all recreational drugs - Refer to drug cessation resources as available
Heart-healthy diet	<ul style="list-style-type: none"> - Encourage a balanced, nutrient-rich diet: <ul style="list-style-type: none"> • Maintain low sodium (<2 g/day) • Avoid heavily processed food • Avoid saturated fat and trans fatty acids • Limit sugar intake • Limit caffeine intake
Obesity	<ul style="list-style-type: none"> - Encourage a healthy BMI (<25 kg/m²) - Refer to nutritionists, exercise physiologists, and/or supportive safe weight loss organizations
Physical activity	<ul style="list-style-type: none"> - Assist patients with limited functional capacity with appropriate exercise counseling - Refer to cardiac rehabilitation or other medical exercise programs - Reinforce the exercise prescription provided by the ACHD clinician
Adequate rest	<ul style="list-style-type: none"> - Assess for sleep dysfunction - Refer to sleep specialist for a sleep study - Educate on healthy sleep techniques: minimize caffeine, maintain a consistent bedtime, create a bedtime relaxation routine - Evaluate need for sleep aid medication

Table 4
Combined hormonal contraception use in women with CHD.

Class 1: Always useable	Class 2: Broadly useable	Class 3: Caution if used	Class 4: Use contraindicated
Minor valve disease (e.g. bicuspid aortic valve with normal function or mild pulmonary stenosis)	Tissue prosthetic valve lacking class 3 or 4 features	Thrombotic risk (even on warfarin): Mechanical valve (bileaflet)	Thrombotic risk (even on warfarin): Mechanical valve (Starr-Edwards [†] , Bjork-Shiley [‡])
Repaired coarctation of the aorta with no aneurysm and no hypertension	Uncomplicated mild aortic or mitral valve disease	Previous thromboembolism	Any mechanical tricuspid valve
Simple lesions repaired in childhood with no residual sequelae	Most arrhythmias other than atrial fibrillation or flutter	Atrial arrhythmia	Pulmonary hypertension of any cause
	Uncomplicated Marfan syndrome	Dilated left atrium >4 cm	Left ventricular dysfunction (ejection fraction <30%)
	Congenital heart disease lacking any Class 3 or 4 features	Risk of paradoxical embolism: Potential reversal of left-to-right shunt (e.g. unoperated atrial septal defect)	Fontan circulation
	Small left-to-right shunt not reversible with physiological maneuvers (e.g. small ventricular septal defect)		Previous coronary arteritis (e.g. Kawasaki disease)
	Previous cardiomyopathy with full recovery		Risk of paradoxical embolism: Cyanotic heart disease
			Pulmonary arteriovenous malformations

[†]Edwards Lifesciences, USA (discontinued); [‡]Pfizer Inc., USA.

explored. Whether surrogacy or adoption is chosen, providing support, resources, and assistance with preparation of required documents is vital. When pregnancy does occur, the ACHD-NC can provide clinical care and emotional support, including anticipatory guidance. Allocation of time and resources should be based on pregnancy risk and patient needs. The ACHD-NC should participate in multidisciplinary (cardiology/OB) meetings to discuss and plan pregnancy management and delivery, and serve as the liaison between services. For the subset of high-risk ACHD patients who require anticoagulation during pregnancy, often in the form of subcutaneous or unfractionated heparin [28], the ACHD-NC can provide education and close monitoring, and ensure adherence.

5. Self-care management and self-advocacy

Self-care management becomes a priority as healthcare becomes patient- rather than parent-focused. In the pediatric setting, parents are often the primary focus of disease education and medical management discussions. With the transition to adulthood, the ACHD-NC should focus on enabling the patient to function independently in managing his or her own care. The ACHD-NC can provide education at office visits, phone appointments, electronic communication, or via the development of educational resources. Education should begin with addressing the need for lifelong specialized ACHD care. Accessibility helps build the trusting nurse-patient relationship. The ACHD-NC can provide patients with business cards that include the ACHD clinic's direct phone line and email address, and should encourage the use of patient passports or electronic medical record tools, if available. The ACHD-NC should assess the patient's current level of autonomy and tailor a plan to help encourage the patient to assume responsibility for his/her care. Assigning small tasks at each visit, such as test scheduling, refilling and organizing medications, personally contacting the office with new symptoms or concerns, and preparing questions prior to scheduled visits helps build patient autonomy. The ACHD-NC should continuously reinforce self-care management by providing positive reinforcement, while recognizing the individual's capabilities and limitations. Patients with developmental or cognitive impairment may live at home with aging parents, in a group home setting, or in other living arrangements. The ACHD-NC should elicit the patient's story from available family, guardians, and/or caregivers. ACHD-NCs should generally address such patients directly and engage them when possible, while also

encouraging caregivers to be involved in care management and decision-making. Patients and families should be educated about the potential for erroneous, or even unsafe, online medical advice. Patients and families should be provided with safe, reliable, and easily accessible websites and educational material, as well as contact information for institutional, national, and international ACHD organizations.

6. Life expectancy, advance directives, and end-of-life care

ACHD-NCs may be involved in discussions surrounding life expectancy, advance directives, and end-of-life (EOL) care. Advance directives are legal documents that allow patients to determine their medical treatment at EOL and may include the naming of a healthcare proxy and the types of life-sustaining treatments accepted or refused. The ACHD-NC's responsibility is to ensure that EOL wishes are well documented and easily accessible to all members of the care team, and that all documents are available for EOL discussions. Ideally, the timing of an EOL discussion should be standardized with the ACHD team in order to ensure that it is systematically addressed in a proactive fashion (e.g., prior to invasive procedures) and conducted in a supportive manner with a multidisciplinary team. ACHD-NCs are well-positioned to develop consistent, lasting relationships with patients, and foster an environment conducive to EOL discussions.

7. Conclusion

The ACHD population is rapidly growing, aging, and becoming increasingly complex. In order to support the unique clinical needs and challenges encountered by this patient population, ACHD care must be comprehensive, interdisciplinary, and team-based. The ACHD-NC's role is foundational to this approach in providing front-line care to ACHD patients, while working in collaboration with other team members, including ACHD cardiologists, congenital heart surgeons, interventionalists, advanced practice providers, social workers, mental health specialists, and administrators. The ACHD-NC provides programmatic organization, coordination, and facilitation of patient care and comprehensive patient education, while ensuring a holistic approach to care that supports patient autonomy and dignity. As an invaluable member of the ACHD team, the ACHD-NC can play a key role in optimizing patient outcomes and improving overall patient experience and satisfaction.

Conflict of interest

The authors report no relationships that could be construed as a conflict of interest.

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