Recommendations for advance care planning in adults with congenital heart disease: a position paper from the ESC Working Group of Adult Congenital Heart Disease, the Association of Cardiovascular Nursing and Allied Professions (ACNAP), the European Association for Palliative Care (EAPC), and the International Society for Adult Congenital Heart Disease (ISACHD)

Markus Schwerzmann 1*, Eva Goossens 2,3, Pastora Gallego 4, Adrienne H. Kovacs 5, Philip Moons 2,6,7, Lorna Swan 8, Daniel Tobler 9, Noémi de Stoutz 10, Harald Gabriel 11, Matthias Greutmann 12, Jolien W. Roos-Hesselink 13, Piotr Z. Sobanski 14, and Corina Thomet 1,2

1Center for Congenital Heart Disease, University Hospital Inselspital, Department of Cardiology, University of Bern, Freiburgstrasse 15, 3010 Bern, Switzerland; 2Department of Public Health and Primary Care, KU Leuven, Kapucijnenvoer 35, box 7001, 3000 Leuven, Belgium; 3Faculty of Medicine and Health Sciences, Centre for Research and Innovation in Care, Division of Nursing and Midwifery, University of Antwerp, Universiteitsplein 1, 2610 Antwerp, Belgium; 4Department of Cardiology, Hospital Universitario Virgen del Rocio, Avenida Manuel Siurot s/n, 41013 Seville, Spain; 5Knight Cardiovascular Institute, Oregon Health & Science University, 3181 SW Sam Jackson Park Rd, UHN-62 Portland, Oregon, USA; 6Institute of Health and Care Science, University of Gothenburg, Arvid Wallgrens Backe, Box 457, 405 30 Gothenburg, Sweden; 7Department of Paediatrics and Child Health, University of Cape Town, Klipfrontein Road Mowbray, Cape Town 7700, South Africa; 8Division of Cardiology, Peter Munk Cardiac Centre, Toronto Congenital Cardiac Centre for Adults, University of Toronto, 585 University Avenue; Toronto ON M5G 2N2, Canada; 9Department of Cardiology, University Hospital Basel, University of Basel, Petersgraben 44031 Basel, Switzerland; 10European Society of Cardiology Patient Forum Representative, Member of “Cuore Matto” and Global ARCH, Dorfstrasse 64, 8126 Zumikon, Switzerland; 11Department of Cardiology, Medical University of Vienna, Waehringer Guertel 18-20, 1090 Vienna, Austria; 12Department of Cardiology, University Heart Center, Rämistrasse 100, 8091 Zurich, Switzerland; 13Department of Cardiology, Erasmus MC, Erasmus University, P.O. Box 2040, 3000 CA Rotterdam, The Netherlands; and 14Palliative Care Unit and Competence Center, Department of Internal Medicine, Hospital Schwyz, Waldeggstrasse 10, 6430 Schwyz, Switzerland

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Survival prospects in adults with congenital heart disease (CHD), although improved in recent decades, still remain below expectations for the general population. Patients and their loved ones benefit from preparation for both unexpected and predictable deaths, sometimes preceded by a prolonged period of declining health. Hence, advance care planning (ACP) is an integral part of comprehensive care for adults with CHD. This position paper summarizes evidence regarding benefits of and patients’ preferences for ACP and provides practical advice regarding the implementation of ACP processes within clinical adult CHD practice. We suggest that ACP be delivered as a structured process across different stages, with content dependent upon the anticipated disease progression. We acknowledge potential

* Corresponding author. Tel: +41 31 632 78 59, Fax: +41 31 632 80 50, Email: markus.schwerzmann@med.unibe.ch

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Introduction

Advances in the diagnosis and treatment of congenital heart defects over the past few decades have led to unparalleled changes in the patient demographic profile, such that adults now outnumber children with congenital heart disease (CHD). Improved survival has been driven largely by a reduction in infant mortality, particularly among individuals born with severe forms of CHD. With decreasing mortality in the young, the CHD population is not only growing but also aging. The prevalence of adults with CHD older than 60 years increased 10-fold from 2000 to 2013 and is currently estimated at 5–10% of the entire CHD population. Many now live long enough to acquire typical age-related comorbidities. Over 50% of a contemporary adult CHD (ACHD) cohort receiving follow-up at a tertiary centre had at least one acquired comorbidity and almost a quarter had two or more comorbidities. Although survival beyond the age of 18 years is now >90%, survival prospects for adults with CHD still remain below expectations for the general population. The most common cause of death among young people living with CHD is sudden cardiac death, while heart failure supersedes it in the aging CHD population. Therefore, we should prepare adults with CHD and their families for both unexpected and predictable deaths, sometimes preceded by a prolonged period of declining health.

The aims of this position paper are to summarize current evidence regarding benefits of and patients’ preferences for advance care planning (ACP) and to provide practical advice regarding the implementation of ACP processes within clinical ACHD practice. Adult congenital heart disease healthcare providers require skills facilitating the timely and sensitive initiation of ACP and the co-ordination of holistic care for adults with CHD at all stages of their life. Conclusions presented in this paper are agreed expert recommendations of general rules extrapolated from the ACHD population and also other adults with cardiovascular disease. This position paper includes both patient and provider perspectives and contributions. Box 1 summarizes the patient perspective in the words of a representative living with CHD.

Terminology and concepts related to advance care planning

A definition of ACP is provided in Box 2—glossary of terms. Advance care planning is based upon consideration of situations that might arise in the future in a person’s life. Advance care planning helpfully begins with identifying personal values and goals in an effort to align future medical treatment and care with these convictions.
Knowledge of a person’s personality and her or his social supports may be helpful for tailoring discussions. The process can be presented to patients and families as ‘preparing for the worst, while hoping for the best’.15 During this reflective process a person may decide which treatments and care measures she or he would prefer in specific situations. Achieving the best possible quality of life within the context of advanced heart disease may entail timely involvement of palliative care (see Box 2—glossary of terms), of which key elements include symptom relief, psychosocial support, and spiritual care.16 There are existing position papers and reviews related to the delivery of palliative care in heart disease patients.17,18 The primary focus of this position paper is the extended process of ACP rather than palliative or end-of-life care.

Benefits of advance care planning for people with advanced cardiovascular disease

The American Heart Association,19 the European Society of Cardiology,20,21 and the European Association for Palliative Care17 recommend ACP for people with advanced cardiac disease, including those with heart failure. Advance care planning would ideally become a routine part of heart failure care, reviewed annually and more often as needed in accordance with disease progression. Benefits of timely initiation of an ACP process are summarized in Figure 1. Despite a limited quantitative and qualitative evidence base, ACP is considered as a marker of excellent care in advanced chronic heart failure.22 Advance care planning is also indicated for ACHD patients with advanced cardiac disease. However, the disease trajectory, patient characteristics, and social situation may differ substantially in ACHD compared to acquired heart failure. Specific recommendations for adults with CHD directed the preparation of this document.

Advance care planning needs and preferences among adults with congenital heart disease

For adults with CHD, the following empirical findings regarding ACP experiences and preferences have been reported:23–27

- Discussions about ACP occur infrequently in routine ACHD outpatient visits.
- Most healthcare providers report that they reserve discussions about ACP for adults living with complex defects and/or at advanced stages of their condition.
- The majority of adults with CHD report interest in ACP independent of underlying defect severity and prefer that such discussions be initiated early in the disease course, before life-threatening complications occur. In one study, 18 years of age was identified as the most appropriate age to initiate an ACP dialogue.26
- Although most adults with CHD report interest in ACP and in receiving information about the general life expectancy of individuals with their type of CHD, some prefer not to have such conversations.
- Although the majority of adults with CHD favour having an advance directive (see Box 2—glossary of terms) available if they are...
Unable to speak for themselves, few have previously completed advance directives or appointed a healthcare representative.

- Most adults with CHD would prefer to have discussions about ACP care with their treating ACHD physician. The trusting relationship between a patient and their ACHD physician was identified as a facilitator for the initiation of such discussions.
- Factors associated with greater interest in ACP discussions include being married, perceiving a shorter lifespan compared to peers, and having more pronounced anxiety symptoms.

Implementation of advance care planning within clinical adult congenital heart disease practice

Initiation of advance care planning process

The process of ACP may be initiated at any moment in life, independent of a person’s health status, and ACP should thus be addressed with all adults with CHD at some time in their lives. Although tools like the ‘surprise question’ posed to healthcare providers (i.e. ‘Would you be surprised if this person died in the next year?’) are not validated as prognostic markers in adults with CHD, they can help identify individuals who may benefit from deliberate and thoughtful reflection regarding future health expectations. Furthermore, the circumstances below might prompt the initiation of ACP discussions and review of previously expressed ACP preferences and documents as appropriate (see also Supplementary material online, Table S1).

Disease progression and worsening prognosis are milestones in the disease course that may initiate the process of ACP in a way that seems most natural. Population-based prognoses can be estimated using generic or CHD-specific tools. Although estimating prognoses in individuals with CHD remains challenging due to less predictable disease trajectories, certain CHD diagnoses have been associated with markedly reduced life expectancy, such as Fontan circulation, a systemic right ventricle, and cyanotic heart disease. For example, the estimated 5-year mortality of a 40-year-old person with Fontan physiology is comparable to that of a 75-year-old person from the general population. Many individuals with complex CHD are unaware of their prognosis and overestimate their life expectancy, which may result in requests or expectations that are at odds with experienced clinical judgement. Exploration of these inconsistencies may naturally trigger ACP.

A second opportunity to initiate ACP occurs at the time of consideration of interventions including cardiac surgery, complex catheter-based therapeutic interventions, implantable cardioverter defibrillator (ICD) implantation, ventricular assist device implantation, or heart transplantation. A ‘what if?’ dialogue can be incorporated into the discussion of potential complications threatening survival, independent functioning or decision-making capacity. During such dialogue, the option of declining the proposed treatment or intervention if incompatible with personal goals and alternative management options should be presented. The discussion can be expanded to include longer-term health expectations in addition to potential immediate complications of the intervention. The occurrence of an acute event (e.g. acute heart failure, ventricular arrhythmia requiring cardioversion) or any other unplanned hospital admission can also trigger the reflective process of ACP. If not relevant or possible early in the course of a hospitalization, ACP can be initiated before discharge by exploring patient’s wishes should another acute event occur.

Changes in a person’s social system, such as death of a close family member, death of an ACHD peer, and family planning, may trigger ACP due to acute awareness of mortality. As part of the pre-pregnancy counselling process, the impact of pregnancy on long-term functioning and future health can lead directly to a comprehensive discussion of ACP.

Advance care planning discussions should not be restricted to adults of a certain age group and can also be appropriate for adolescents and young adults with CHD. Discussion of the impact of CHD on later life and anticipated long-term outcomes can be an integral part of the transition process from paediatric to adult care, particularly for patients with complex defects. As with patients of all ages, the potential emotional impact for patients and their families must be acknowledged.

Overcoming barriers to the advance care planning process

Table 1 presents known barriers that may hamper the initiation of the ACP process as well as proposed solutions. Advance care planning has become a frequent theme in ACHD peer groups and patient organizations, highlighting that people with CHD are increasingly identifying this as an important topic of discussion. As such, ACHD programmes are encouraged to partner with national or international patient organizations (e.g. www.echdo.eu, www.global-arch.org, www.achaheart.org) to provide educational ACP activities, such as webinars. Some patients may appreciate an introduction to ACP concepts in this more general forum prior to having personal discussions in the clinical setting.

Staged implementation of advance care planning

Advance care planning is a gradual process that is initiated by exploring a person’s understanding of the aims and potential benefits of ACP and discussing their personal readiness for decision-making. Information about a person’s health-related experiences, values, psychosocial resources, concerns, and expectations should be sought. Given the diversity in cultural and religious approaches to death and dying both between and within countries, a culturally sensitive approach is essential. Assumptions should be avoided and provider is encouraged to approach discussions with a respectful curiosity. It should be noted, however, that core principles of ACP, such as the expression of respect and compassion, and alleviation of unnecessary distress, are largely similar across ethnic and cultural groups.

Open and sensitive communication concordant with personal needs and values should include an explanation of how the CHD diagnosis impacts longer-term health expectations and anticipated disease progression, prognosis, and the advantages and disadvantages of potential treatment options. This allows providers to individualize ACP to the expected timing of health deterioration. As such, we suggest a staged implementation of the ACP process as outlined in Table 2.
Within every stage of ACP, patients should be given the opportunity to involve family members or loved ones and also to reflect and clarify previously documented wishes; as such, ACP is an iterative process (Take home figure).

When speaking with an adult with CHD with minimal anticipated major health problems in the upcoming years, it may be sufficient for the ACP discussion to include predicted long-term health outcomes and to explore a person’s values, goals, and concerns for the future.

<table>
<thead>
<tr>
<th>Table 1</th>
<th>Barriers to advance care planning communication and proposed clinical strategies</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Barrier</strong></td>
<td><strong>Strategies for healthcare providers</strong></td>
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<tr>
<td><strong>At patient level</strong></td>
<td></td>
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<tr>
<td>No/Minimal knowledge about ACP</td>
<td>Provide education about the rationale for ACP</td>
</tr>
<tr>
<td>Reluctance to begin this discussion</td>
<td>Introduce and normalize this topic in broader terms during a routine clinic visit and offer the opportunity to discuss in more detail at a follow-up visit</td>
</tr>
<tr>
<td>Encourage patients to have loved ones present for these conversations</td>
<td>Introduce ACP during situations when it seems to occur more naturally (e.g. death of a close family member or an ACHD peer, family planning, prior to cardiac intervention)</td>
</tr>
<tr>
<td>Avoidance of discussion of health deterioration (which might be amplified during acute cardiac events)</td>
<td>Initiate ACP during stable (rather than acute) phase of the disease to allow for a less-pressured patient experience</td>
</tr>
<tr>
<td>Desire to protect family and loved ones</td>
<td>Educate about the advantages of ACP communication and advance directives for family members should they be faced making treatment or care decisions for the patient in the future</td>
</tr>
<tr>
<td><strong>At healthcare provider level</strong></td>
<td></td>
</tr>
<tr>
<td>Fear of causing patients to experience unnecessary emotional distress</td>
<td>Acknowledge that emotional reactions to disappointing information are understandable</td>
</tr>
<tr>
<td>Strive for a balance between preparation for undesired outcomes and maintaining hope; emphasize that these are not mutually exclusive</td>
<td>Emphasize that ACP is intended to be an empowering process for patients and their loved ones</td>
</tr>
<tr>
<td>Uncertainty about prognosis</td>
<td>Acknowledge challenges associated with an unpredictable disease trajectory</td>
</tr>
<tr>
<td>Use standardized prognostic indexes (e.g. NYHA class) or results from functional testing (e.g. cardiopulmonary exercise testing) to guide predictions</td>
<td>Offer broader time ranges for life expectancy (i.e. decades vs. years vs. months vs. weeks)</td>
</tr>
<tr>
<td><strong>Lack of confidence and skills in ACP</strong></td>
<td>Practice discussions with colleagues to develop comfort with this language</td>
</tr>
<tr>
<td>Consider a standardized approach (e.g. ask-tell-ask) to initiate discussions</td>
<td>Use a checklist to guide discussions (see Supplementary material online)</td>
</tr>
<tr>
<td>Seek continuing education opportunities</td>
<td>Liaise with a palliative care team to enhance skills</td>
</tr>
<tr>
<td><strong>At ACHD programme/institutional level</strong></td>
<td></td>
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<tr>
<td>Ambiguity regarding who is responsible for initiating and maintaining the ACP dialogue</td>
<td>Acknowledge that adults with CHD prefer to discuss ACP with clinicians whom they trust</td>
</tr>
<tr>
<td>Lack of time</td>
<td>Develop a standardized process for identifying the team member who is responsible for overseeing ACP</td>
</tr>
<tr>
<td>Advocate for a clinical scheduling practice that allows sufficient time to discuss ACP within a routine outpatient visit</td>
<td>Schedule ACP-specific clinical visits</td>
</tr>
<tr>
<td>Recognize that time might be more flexible within the inpatient setting</td>
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</tr>
</tbody>
</table>

ACHD, adult CHD; ACP, advance care planning; CHD, congenital heart disease; NYHA, New York Heart Association.
During these discussions, the provider might discover that patient’s expectations are in fact more pessimistic than would be predicted from long-term follow-up studies. The results of one study indicated that adults with CHD of low complexity were more likely to want information about the average life expectancy compared to those with CHD of moderate or great complexity. In some of these cases, discussions of long-term health expectations might be interpreted with reassurance and relief.
For adults with CHD facing health deterioration, discussions about preferences for interventions and life-sustaining treatments become more pertinent and would ideally result in advance directives, including the naming of a healthcare representative, and Physician Orders for Life-Sustaining Treatment (healthcare representative; see Box 2 — glossary of terms) that should be made accessible to all healthcare providers involved in the care process. Figure 2 illustrates how the content of ACP discussions can transition from general information about future health expectations to more specific topics in accordance with disease progression.

As a person’s condition deteriorates further such that the possibility of death increases, discussions can be extended with specific questions related to end-of-life wishes, including issues related to the modification of cardiac devices (e.g. deactivation of shocking function of an ICD), specific treatments to consider and avoid, and palliative care measures.

Interprofessional collaboration (e.g. palliative care professionals, social workers, psychologists, spiritual advisors) may be particularly helpful in situations in which (i) patients and/or loved ones are struggling with the disease trajectory, (ii) disagreements exist between the patient, family, and care teams, or (iii) challenges arise in managing physical and/or psychological symptoms. It is important to emphasize to patients and loved ones that active disease-specific care can continue in parallel with palliative care if consistent with patients’ preferences and goals. The supporting role of palliative care for the family and loved ones goes beyond the person’s death and includes bereavement counselling.

**Structural requirements for implementing advance care planning in regular adult congenital heart disease practice**

Effective ACP sometimes requires an interdisciplinary team approach. In addition to the ACHD team (i.e. typically cardiologists and nurses), other members of the interdisciplinary team reflect needs of the person and his/her family (e.g. general practitioners, other subspecialists, social workers, spiritual care providers, psychologists, legal counsellors, palliative care specialists). Within the team, it is important to identify the co-ordinating lead who may thus assign specific tasks to other team members. This is often the patient’s identified primary ACHD cardiologist, who most patients consider the most appropriate person to initiate ACP discussions. Advanced practice nurses (e.g. nurse practitioners) are also well suited for this oversight role, given their holistic approach and often long-term relationships with patients.

In addition to staffing considerations, a set of programmatic processes is necessary to ensure regular review and accessible storage of advance directives. Legal requirements, such as the binding effects of ACP documents, identification of one’s healthcare representative, and matters related to assisted dying, depend upon laws of one’s local jurisdiction. At a minimum, ACP documents should be declared as ‘statements of wishes to be given due respect’. A formal document signed by patients and witnesses is ideal. This document should be
Advance care planning is a staged and iterative process rather than a one-time event. Advance care planning supports and empowers individuals to consider and communicate preferences for future health care based on their personal values and beliefs, should they be unable to express their wishes at that time. Within every stage of advance care planning, patients should be given the opportunity to involve family members or loved ones and also to reflect and clarify previously-documented wishes. Most adults with congenital heart disease prefer to have discussions about advance care planning care with their treating adult congenital heart disease physician. A trusting relationship is a facilitator for the initiation of such discussions. The notion of disease progression and specific triggers should raise awareness of the adult congenital heart disease specialist to mention and initiate advance care planning, if wished by the patient. CHD, congenital heart disease.

**Box 1 The perspective of adults living with congenital heart disease**

- In a society that mutes discussions on death and dying and that simplistically lauds improved adult congenital heart disease (ACHD) outcomes as success stories, healthcare providers tend to avoid and/or delay advance care planning (ACP) discussions with their patients.
- Patient organizations can help healthcare providers spread information about ACP. They are also irreplaceable for adults with congenital heart disease (CHD) to have conversations about ACP among peers.
- Documents for ACP available on the Internet through various organizations are usually tailored to elderly people and not to adults with CHD. Adults with CHD would benefit from assistance when using such documents.
- Discussions with healthcare providers about future health are typically welcome, even though this is often considered a stressful topic. Adults with CHD and ACHD healthcare professionals may be challenged by the art of finding the right words at the right moment.
- When an adult with CHD asks what can be done for them, the answer should be realistic and may include comfort measures rather than (or in addition to) strategies to extend life: ‘all that can be done when nothing more can be done’.
- Many individuals have clear ideas about what they consider the worst possible way to die. It is helpful to discuss strategies for preventing such dreaded situations.
- Hope is not just about medical outcomes. Rather than lingering in discomfort, adults with CHD may hope for a sudden death, for effective comfort measures, and for emotional, spiritual, and interpersonal support during the last chapters of their biography.
Advance care planning is a process that supports and empowers individuals, at any stage of their lives or the disease process, to consider, communicate, and document preferences for future health care to their loved ones and healthcare providers. During this process, individuals have the opportunity to make decisions in advance about treatment they would and would not want should they be unable to express their wishes at that time. This process benefits (i) patients (by increasing the likelihood that their wishes will be followed), (ii) loved ones (by helping them more confidently express patients’ wishes if necessary), and (iii) healthcare providers (by providing information sufficient to align treatment plans with patient goals and preferences).

Advance directives are paper and/or electronic documents that specify a person’s preferences and decisions (including the naming of a healthcare representative) for medical treatments or care in advance of a potential serious medical event/situation when they may be unable to communicate this information. Specific documentation requirements may vary depending upon where a patient lives.

A healthcare representative (also referred to as a substitute decision maker, surrogate decision maker, power of attorney for personal care or healthcare proxy) is a named individual that a person entrusts to express their wishes and make medical decisions on their behalf should the individual be unable to speak for themselves.

Palliative care has traditionally referred to care focused on alleviating symptoms and enhancing quality of life and quality of the dying experience rather than curing disease or extending life. The modern definition has been expanded to describe comprehensive care that addresses the physical, psychosocial, and spiritual needs of individuals with diseases unresponsive to curative treatments. Such care is ideally introduced early in the course of the illness and meets the needs of patients and their loved ones.

Physician Orders for Life-Sustaining Treatment (POLST) are medical orders to be honoured by healthcare workers during a critical event. Unlike advance directives, POLST document the patients’ wishes in the form of binding orders (i.e. no referral to the intensive care unit) but do not include other more general preferences such as identifying a healthcare representative.

Supporting patients to be well prepared for their final journey may be one of the most challenging and also rewarding experiences for ACHD providers.

Supplementary material

Supplementary material is available at European Heart Journal online.

Conflict of interest: none declared.

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