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]

[1] Center for Congenital Heart Disease, University Hospital Inselspital, Department of Cardiology, University of Bern, Switzerland; [2] Department of Public Health and Primary Care, KU Leuven, Belgium; [3] Faculty of Medicine and Health Sciences, Centre for Research and Innovation in Care, Division of Nursing and Midwifery, University of Antwerp, Antwerp, Belgium; [4] Department of Cardiology, Hospital Universitario Virgen del Rocio, Seville, Spain;[5] Knight Cardiovascular Institute, Oregon Health & Science University, Portland, USA; [6] Institute of Health and Care Science, University of Gothenburg, Sweden; [7] Department of Paediatrics and Child Health, University of Cape Town, South Africa;
[8] Division of Cardiology, Peter Munk Cardiac Centre; Toronto Congenital Cardiac Centre for Adults, University of Toronto, Canada; [9] Department of Cardiology, University Hospital Basel, University of Basel, Switzerland; [10] European Society of Cardiology Patient Forum Representative, Member of "Cuore Matto" and Global ARCH, Zurich, Switzerland; [11] Department of Cardiology, Medical University of Vienna, Austria; [12] Department of Cardiology, University Heart Center, Zurich, Switzerland; [13] Department of Cardiology, Erasmus MC, Erasmus University, Rotterdam, the Netherlands; [14] Palliative Care Unit and Competence Center, Department of Internal Medicine, Hospital Schwyz, Switzerland

Author of correspondence

Prof. Markus Schwerzmann

Center for Congenital Heart Disease, University Hospital Inselspital, University of Bern, Switzerland

Phone: +41 31 632 78 59; Fax: +41 31 632 80 50

Email academic: markus.schwerzmann@med.unibe.ch

Abstract

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 barriers to initiate ACP discussions and emphasize the importance of a sensitive and situation-specific communication style. Conclusions presented in this paper reflect agreed expert opinions, and include both patient and provider perspectives.

Key words: adult congenital heart disease; advance care planning

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).[1] Improved survival has been driven largely by a reduction in infant mortality, particularly among individuals born with severe forms of CHD.[2] 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.[3, 4] Many now live long enough to acquire typical age-related comorbidities.[5] Over 50% of a contemporary adult CHD cohort receiving follow-up at a tertiary centre had at least one acquired comorbidity and almost a quarter had two or more comorbidities.[6] Although survival beyond the age of 18 years is now >90%,[7] survival prospects for adults with CHD still remain below expectations for the general population.[8, 9] 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.[10] 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 adult CHD (ACHD) practice. ACHD health care providers require skills facilitating the timely and sensitive initiation of ACP and the coordination of holistic care for adults with CHD at all stages of their life.[11, 12] Conclusions presented in this paper are agreed expert recommendations of general rules extrapolated from the ACHD population[12] but 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 advance care planning (ACP) is provided in Box 2 – glossary of terms. ACP is based upon consideration of situations that might arise in the future in a person's life.[13, 14] ACP 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 ACP for people with advanced cardiovascular disease

The American Heart Association,[19] the European Society of Cardiology[20, 21] and the European Association for Palliative Care[15] recommend ACP for people with advanced cardiac disease, including those with heart failure. ACP 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 a marker of excellent care in advanced chronic heart failure.[22] ACP 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.

ACP needs and preferences among adults with CHD

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 health care 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 health care 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 ACP within clinical ACHD practice

Initiation of ACP 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.[11] Although tools like the "surprise question" posed to health care 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.[28] Furthermore, the circumstances below might prompt the initiation of ACP discussions as well as review of previously expressed ACP preferences and documents as appropriate (see also supplementary material: Table 1).[14, 29, 30]

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.[31] Population-based prognoses can be estimated using generic[32, 33] 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.[34] 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 judgment.[15] 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.[30] 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.[30]

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.[35, 36] As part of the pre-pregnancy counselling process,[37] the impact of pregnancy on long-term functioning and future health can lead directly to a comprehensive discussion of ACP.[38]

ACP discussions should not be restricted to adults of a certain age group and can also be appropriate for adolescents and young adults with CHD.[39, 40] Discussion of the impact of CHD on later life and anticipated long-term outcomes can be an integral part of the transition process from pediatric to adult care,[41] 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 ACP process

Table 1 presents known barriers that may hamper the initiation of the ACP process as well as proposed solutions. ACP 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 programs 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 ACP

ACP 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.[42] Assumptions should be avoided and provider are 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.[43, 44] Open and sensitive communication concordant with personal needs and values should include an explanation of how the CHD diagnosis impacts longer term health expectations as well as

anticipated disease progression, prognosis, and the advantages and disadvantages of potential treatment options.[29] 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 (summary illustration).

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. 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.[26] In some of these cases, discussions of long-term health expectations might be interpreted with reassurance and relief.[25]

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 health care representative, and Physician Orders for Life-Sustaining Treatment (health care representative, POLST; see Box 2 – glossary of terms) that should be made accessible to all health care providers involved in the care process.[35] 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.[17, 45,46] The supporting role of palliative care for the family and loved ones goes beyond the person's death and includes bereavement counseling.[47]

Structural requirements for implementing ACP in regular ACHD practice

Effective ACP sometimes requires an interdisciplinary team approach.[48] 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).[49] Within the team, it is important to identify the coordinating 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.[23] 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.[50]

In addition to staffing considerations, a set of programmatic processes is necessary to ensure regular review and accessible storage of advance directives.[13] Legal requirements, such as the binding effects of ACP documents, identification of one's health care representative, and matters related to assisted dying depend upon laws of one's local jurisdiction.[42] At a minimum, ACP documents should be declared as "statements of wishes to be given due respect."[51] A

formal document signed by patients and witnesses is ideal. This document should be easily identifiable and accessible within the electronic and/or paper medical record, and not merely summarized in a clinical progress note. The aim is to ensure access by all inpatient and outpatient health providers.[29] It is also advised that patients provide family members and new health care providers with a copy of their advance directives to ensure at the time of admission, the patient or the persons most involved with its medical support, have a hard copy or an electronic version at hand. ACP preferences and decisions may change over time. It is important for ACHD providers to re-visit this topic over time, and to adjust the corresponding documents accordingly. Providers may also require about the presence of externally-prepared ACP documents (e.g. completed with a lawyer). If available, the recommendation to include these in the medical record should be offered.

Provider training in ACP and end-of-life discussion is associated with increased ease and comfort to initiate ACP discussions.[52] Communication during the ACP process may be challenging for all stakeholders. Practical tools such as the Six Step Protocol for Delivering Bad News (SPIKES) or the 'ask-tell-ask' algorithm can help health care professionals to communicate bad news in an effective but compassionate way.[15, 53] Information on national courses is available through the European Association for Palliative Care (www.eapcnet.eu). Given that ACP discussions can be emotionally burdensome,[54, 55] it is advised that providers receive training in coping strategies and have support systems in place.

Summary

ACP has emerged as an integral part of comprehensive care for adults with CHD. It should be offered during routine clinical practice to all interested persons, and particularly to those at risk for significant deterioration in the near future. We suggest that ACP be delivered as a structured process across different stages, with content dependent upon the anticipated

disease progression. We acknowledge potential barriers to initiate ACP discussions and emphasize the importance of a sensitive, responsive, and adaptable communication style. ACP benefits from a team approach, provider training, adequate staffing and a systematic approach to documentation. Death is an inevitable outcome for all. Supporting patients to be well prepared for their final journey may be one of the most challenging and also rewarding experiences for ACHD providers.

References

1. Marelli AJ, Ionescu-Ittu R, Mackie AS, Guo L, Dendukuri N, Kaouache M. Lifetime prevalence of congenital heart disease in the general population from 2000 to 2010. Circulation 2014;**130**(9):749-56.

2. Khairy P, Ionescu-Ittu R, Mackie AS, Abrahamowicz M, Pilote L, Marelli AJ. Changing mortality in congenital heart disease. J Am Coll Cardiol 2010;**56**(14):1149-57.

3. Tutarel O, Kempny A, Alonso-Gonzalez R, Jabbour R, Li W, Uebing A, Dimopoulos K, Swan L, Gatzoulis MA, Diller GP. Congenital heart disease beyond the age of 60: emergence of a new population with high resource utilization, high morbidity, and high mortality. Eur Heart J 2014;**35**(11):725-32.

4. Baumgartner H. Geriatric congenital heart disease: a new challenge in the care of adults with congenital heart disease? Eur Heart J 2014;**35**(11):683-5.

5. Bhatt AB, Foster E, Kuehl K, Alpert J, Brabeck S, Crumb S, Davidson WR, Jr., Earing MG, Ghoshhajra BB, Karamlou T, Mital S, Ting J, Tseng ZH, American Heart Association Council on Clinical C. Congenital heart disease in the older adult: a scientific statement from the american heart association. Circulation 2015;**131**(21):1884-931.

6. Bracher I, Padrutt M, Bonassin F, Santos Lopes B, Gruner C, Stampfli SF, Oxenius A, De Pasquale G, Seeliger T, Luscher TF, Attenhofer Jost C, Greutmann M. Burden and impact of congenital syndromes and comorbidities among adults with congenital heart disease. Int J Cardiol 2017;**240**:159-164.

7. Erikssen G, Liestol K, Seem E, Birkeland S, Saatvedt KJ, Hoel TN, Dohlen G, Skulstad H, Svennevig JL, Thaulow E, Lindberg HL. Achievements in congenital heart defect surgery: a prospective, 40-year study of 7038 patients. Circulation 2015;**131**(4):337-46; discussion 346.

8. Moons P, Bovijn L, Budts W, Belmans A, Gewillig M. Temporal trends in survival to adulthood among patients born with congenital heart disease from 1970 to 1992 in Belgium. Circulation 2010;**122**(22):2264-72.

9. Oliver JM, Gallego P, Gonzalez AE, Garcia-Hamilton D, Avila P, Yotti R, Ferreira I, Fernandez-Aviles F. Risk factors for excess mortality in adults with congenital heart diseases. Eur Heart J 2017;**38**(16):1233-1241.

10. Oliver JM, Gallego P, Gonzalez AE, Garcia-Hamilton D, Avila P, Alonso A, Ruiz-Cantador J, Peinado R, Yotti R, Fernandez-Aviles F. Impact of age and sex on survival and causes of death in adults with congenital heart disease. Int J Cardiol 2017;**245**:119-124.

11. Troost E, Roggen L, Goossens E, Moons P, De Meester P, Van De Bruaene A, Budts W. Advanced care planning in adult congenital heart disease: Transitioning from repair to palliation and end-of-life care. International Journal of Cardiology 2019;**279**:57-61.

12. Ludmir J, Steiner JM, Wong HN, Kloosterboer A, Leong J, Aslakson RA, Group ARCW. Palliative Care Opportunities Among Adults With Congenital Heart Disease-A Systematic Review. J Pain Symptom Manage 2019;**58**(5):891-898.

13. Sudore RL, Fried TR. Redefining the „planning“ in advance care planning: preparing for end-of-life decision making. Ann Intern Med 2010;**153**(4):256-61.

14. Brighton LJ, Bristowe K. Communication in palliative care: talking about the end of life, before the end of life. Postgrad Med J 2016;**92**(1090):466-70.

15. Kovacs AH, Landzberg MJ, Goodlin SJ. Advance care planning and end-of-life management of adult patients with congenital heart disease. World J Pediatr Congenit Heart Surg 2013;**4**(1):62-9.

16. Bernacki RE, Block SD, American College of Physicians High Value Care Task F. Communication about serious illness care goals: a review and synthesis of best practices. JAMA Intern Med 2014;**174**(12):1994-2003.

17. Sobanski PZ, Alt-Epping B, Currow DC, Goodlin SJ, Grodzicki T, Hogg K, Janssen DJA, Johnson MJ, Krajnik M, Leget C, Martinez-Selles M, Moroni M, Mueller PS, Ryder M, Simon ST, Stowe E, Larkin PJ. Palliative Care for people living with heart failure - European Association for Palliative Care Task Force expert position statement. Cardiovasc Res 2019.

18. Steiner JM, Cooper S, Kirkpatrick JN. Palliative care in end-stage valvular heart disease. Heart 2017;**103**(16):1233-1237.

19. Braun LT, Grady KL, Kutner JS, Adler E, Berlinger N, Boss R, Butler J, Enguidanos S, Friebert S, Gardner TJ, Higgins P, Holloway R, Konig M, Meier D, Morrissey MB, Quest TE, Wiegand DL, Coombs-Lee B, Fitchett G, Gupta C, Roach WH, Jr., American Heart Association Advocacy Coordinating C. Palliative Care and Cardiovascular Disease and Stroke: A Policy Statement From the American Heart Association/American Stroke Association. Circulation 2016;**134**(11):e198-225.

20. Ponikowski P, Voors AA, Anker SD, Bueno H, Cleland JG, Coats AJ, Falk V, Gonzalez-Juanatey JR, Harjola VP, Jankowska EA, Jessup M, Linde C, Nihoyannopoulos P, Parissis JT, Pieske B, Riley JP, Rosano GM, Ruilope LM, Ruschitzka F, Rutten FH, van der Meer P, Authors/Task Force M. 2016 ESC Guidelines for the diagnosis and treatment of acute and chronic heart failure: The Task Force for the diagnosis and treatment of acute and chronic heart failure of the European Society of Cardiology (ESC)Developed with the special contribution of the Heart Failure Association (HFA) of the ESC. Eur Heart J 2016;**37**(27):2129-200.

21. Jaarsma T, Beattie JM, Ryder M, Rutten FH, McDonagh T, Mohacsi P, Murray SA, Grodzicki T, Bergh I, Metra M, Ekman I, Angermann C, Leventhal M, Pitsis A, Anker SD, Gavazzi A, Ponikowski P, Dickstein K, Delacretaz E, Blue L, Strasser F, McMurray J, Advanced Heart Failure Study Group of the HFAotESC. Palliative care in heart failure: a position statement from the palliative care workshop of the Heart Failure Association of the European Society of Cardiology. Eur J Heart Fail 2009;**11**(5):433-43.

22. Fang JC, Ewald GA, Allen LA, Butler J, Westlake Canary CA, Colvin-Adams M, Dickinson MG, Levy P, Stough WG, Sweitzer NK, Teerlink JR, Whellan DJ, Albert NM, Krishnamani R, Rich MW, Walsh MN, Bonnell MR, Carson PE, Chan MC, Dries DL, Hernandez AF, Hershberger RE, Katz SD, Moore S, Rodgers JE, Rogers JG, Vest AR, Givertz MM, Heart Failure Society of America Guidelines C. Advanced (stage D) heart failure: a statement from the Heart Failure Society of America Guidelines Committee. J Card Fail 2015;**21**(6):519-34.

23. Tobler D, Greutmann M, Colman JM, Greutmann-Yantiri M, Librach LS, Kovacs AH. End-of-life in adults with congenital heart disease: a call for early communication. Int J Cardiol 2012;**155**(3):383-7.

24. Tobler D, Greutmann M, Colman JM, Greutmann-Yantiri M, Librach SL, Kovacs AH. Knowledge of and preference for advance care planning by adults with congenital heart disease. Am J Cardiol 2012;**109**(12):1797-800.

25. Greutmann M, Tobler D, Colman JM, Greutmann-Yantiri M, Librach SL, Kovacs AH. Facilitators of and barriers to advance care planning in adult congenital heart disease. Congenit Heart Dis 2013;**8**(4):281-8.

26. Deng LX, Gleason LP, Khan AM, Drajpuch D, Fuller S, Goldberg LA, Mascio CE, Partington SL, Tobin L, Kim YY, Kovacs AH. Advance Care Planning in Adults with Congenital Heart Disease: A Patient Priority. Int J Cardiol 2017;**231**:105-109.

27. Steiner JM, Stout K, Soine L, Kirkpatrick JN, Curtis JR. Perspectives on advance care planning and palliative care among adults with congenital heart disease. Congenit Heart Dis 2019;**14**(3):403-409.

28. Romo RD, Lynn J. The utility and value of the “surprise question” for patients with serious illness. Canadian Medical Association Journal 2017;**189**(33):E1072-E1073.

29. Rietjens JAC, Sudore RL, Connolly M, van Delden JJ, Drickamer MA, Droger M, van der Heide A, Heyland DK, Houttekier D, Janssen DJA, Orsi L, Payne S, Seymour J, Jox RJ,

Korfage IJ, European Association for Palliative C. Definition and recommendations for advance care planning: an international consensus supported by the European Association for Palliative Care. Lancet Oncol 2017;**18**(9):e543-e551.

30. Denvir MA, Murray SA, Boyd KJ. Future care planning: a first step to palliative care for all patients with advanced heart disease. Heart 2015;**101**(13):1002-7.

31. Stevenson LW, O'Donnell A. Advanced care planning: care to plan in advance. JACC Heart Fail 2015;**3**(2):122-6.

32. Rozzini R, Sabatini T, Trabucchi M. Prediction of 6-month mortality among older hospitalized adults. JAMA 2001;**286**(11):1315-6.

33. Walter LC, Brand RJ, Counsell SR, Palmer RM, Landefeld CS, Fortinsky RH, Covinsky KE. Development and validation of a prognostic index for 1-year mortality in older adults after hospitalization. JAMA 2001;**285**(23):2987-94.

34. Diller GP, Kempny A, Alonso-Gonzalez R, Swan L, Uebing A, Li W, Babu-Narayan S, Wort SJ, Dimopoulos K, Gatzoulis MA. Survival Prospects and Circumstances of Death in Contemporary Adult Congenital Heart Disease Patients Under Follow-Up at a Large Tertiary Centre. Circulation 2015;**132**(22):2118-25.

35. Dunlay SM, Strand JJ, Wordingham SE, Stulak JM, Luckhardt AJ, Swetz KM. Dying With a Left Ventricular Assist Device as Destination Therapy. Circ Heart Fail 2016;**9**(10).

36. Clayer MT. Clinical practice guidelines for communicating prognosis and end-of-life issues with adults in the advanced stages of a life-limiting illness, and their caregivers. Med J Aust 2007;**187**(8):478.

37. Regitz-Zagrosek V, Roos-Hesselink JW, Bauersachs J, Blomstrom-Lundqvist C, Cifkova R, De Bonis M, Iung B, Johnson MR, Kintscher U, Kranke P, Lang IM, Morais J, Pieper PG, Presbitero P, Price S, Rosano GMC, Seeland U, Simoncini T, Swan L, Warnes CA, Group ESCSD. 2018 ESC Guidelines for the management of cardiovascular diseases during pregnancy. Eur Heart J 2018;**39**(34):3165-3241.

38. Cauldwell M, Gatzoulis M, Steer P. Congenital heart disease and pregnancy: A contemporary approach to counselling, pre-pregnancy investigations and the impact of pregnancy on heart function. Obstet Med 2017;**10**(2):53-57.

39. Wiener L, Zadeh S, Battles H, Baird K, Ballard E, Osherow J, Pao M. Allowing adolescents and young adults to plan their end-of-life care. Pediatrics 2012;**130**(5):897-905.

40. Informed consent, parental permission, and assent in pediatric practice. Committee on Bioethics, American Academy of Pediatrics. Pediatrics 1995;**95**(2):314-7.

41. Sable C, Foster E, Uzark K, Bjornsen K, Canobbio MM, Connolly HM, Graham TP, Gurvitz MZ, Kovacs A, Meadows AK, Reid GJ, Reiss JG, Rosenbaum KN, Sagerman PJ, Saidi A, Schonberg R, Shah S, Tong E, Williams RG. Best practices in managing transition to adulthood for adolescents with congenital heart disease: the transition process and medical and psychosocial issues: a scientific statement from the American Heart Association. Circulation 2011;**123**(13):1454-85.

42. Jox RJ, Horn RJ, Huxtable R. European perspectives on ethics and law in end-of-life care. Handb Clin Neurol 2013;**118**:155-65.

43. Bosma H, Apland L, Kazanjian A. Cultural conceptualizations of hospice palliative care: more similarities than differences. Palliat Med 2010;**24**(5):510-22.

44. Pereira-Salgado A, Mader P, O'Callaghan C, Boyd L, Staples M. Religious leaders' perceptions of advance care planning: a secondary analysis of interviews with Buddhist, Christian, Hindu, Islamic, Jewish, Sikh and Baha'i leaders. BMC Palliat Care 2017;**16**(1):79.

45. Munoz-Mendoza J. Competencies in Palliative Care for Cardiology Fellows. J Am Coll Cardiol 2015;**65**(7):750-3.

46. Kavalieratos D, Gelfman LP, Tycon LE, Riegel B, Bekelman DB, Ikejiani DZ, Goldstein N, Kimmel SE, Bakitas MA, Arnold RM. Palliative Care in Heart Failure: Rationale, Evidence, and Future Priorities. J Am Coll Cardiol 2017;**70**(15):1919-1930.

47. Gibbs JS, McCoy AS, Gibbs LM, Rogers AE, Addington-Hall JM. Living with and dying from heart failure: the role of palliative care. Heart 2002;**88 Suppl 2**:ii36-9.

48. National Consensus Project for Quality Palliative Care. In. *Clinical Practice Guidelines for Quality Palliative Care*. 4th edition ed. Richmond, VA; 2018.

49. Commission TJ. *Advanced Certification for Palliative Care. Review Process Guide Orgnaization Version*. Oak Brook, Illinios 60523: The Joint Commission; 2015-2016.

50. Dixon J, Knapp M. Whose job? The staffing of advance care planning support in twelve international healthcare organizations: a qualitative interview study. BMC Palliat Care 2018;**17**(1):78.

51. Ward AD. Enabling citizens to plan for incapacity. In: (CDCJ) ECoLC-o, (ed): Council of Europe; 2018.

52. Moir C, Roberts R, Martz K, Perry J, Tivis LJ. Communicating with patients and their families about palliative and end-of-life care: comfort and educational needs of nurses. Int J Palliat Nurs 2015;**21**(3):109-12.

53. Kelemen AM, Ruiz G, Groninger H. Choosing Words Wisely in Communication With Patients With Heart Failure and Families. Am J Cardiol 2016;**117**(11):1779-82.

54. Rizo-Baeza M, Mendiola-Infante SV, Sepehri A, Palazon-Bru A, Gil-Guillen VF, Cortes-Castell E. Burnout syndrome in nurses working in palliative care units: An analysis of associated factors. J Nurs Manag 2018;**26**(1):19-25.

55. Diehl E, Rieger S, Letzel S, Nienhaus A, Escobar Pinzon LC. [Burdens and resources of nurses working in the specialist palliative care: an explorative cross-sectional study]. Pflege 2019;**32**(4):209-223.

56. Rogers JG, Patel CB, Mentz RJ, Granger BB, Steinhauser KE, Fiuzat M, Adams PA, Speck A, Johnson KS, Krishnamoorthy A, Yang H, Anstrom KJ, Dodson GC, Taylor DH, Jr.,

Kirchner JL, Mark DB, O'Connor CM, Tulsky JA. Palliative Care in Heart Failure: The PAL-HF Randomized, Controlled Clinical Trial. J Am Coll Cardiol 2017;**70**(3):331-341.

57. Evangelista LS, Lombardo D, Malik S, Ballard-Hernandez J, Motie M, Liao S. Examining the effects of an outpatient palliative care consultation on symptom burden, depression, and quality of life in patients with symptomatic heart failure. J Card Fail 2012;**18**(12):894-9.

58. Kernick LA, Hogg KJ, Millerick Y, Murtagh FEM, Djahit A, Johnson M. Does advance care planning in addition to usual care reduce hospitalisation for patients with advanced heart failure: A systematic review and narrative synthesis. Palliat Med 2018;**32**(10):1539-1551.

59. Sahlen KG, Boman K, Brannstrom M. A cost-effectiveness study of person-centered integrated heart failure and palliative home care: Based on a randomized controlled trial. Palliat Med 2016;**30**(3):296-302.

60. Metzger M, Song MK, Devane-Johnson S. LVAD patients' and surrogates' perspectives on SPIRIT-HF: An advance care planning discussion. Heart Lung 2016;**45**(4):305-10.

61. Brush S, Budge D, Alharethi R, McCormick AJ, MacPherson JE, Reid BB, Ledford ID, Smith HK, Stoker S, Clayson SE, Doty JR, Caine WT, Drakos S, Kfoury AG. End-of-life decision making and implementation in recipients of a destination left ventricular assist device. J Heart Lung Transplant 2010;**29**(12):1337-41.

62. Siouta N, Clement P, Aertgeerts B, Van Beek K, Menten J. Professionals' perceptions and current practices of integrated palliative care in chronic heart failure and chronic obstructive pulmonary disease: a qualitative study in Belgium. BMC Palliat Care 2018;**17**(1):103.

63. Goldstein NE, Mather H, McKendrick K, Gelfman LP, Hutchinson MD, Lampert R, Lipman HI, Matlock DD, Strand JJ, Swetz KM, Kalman J, Kutner JS, Pinney S, Morrison RS.

Improving Communication in Heart Failure Patient Care. J Am Coll Cardiol 2019;**74**(13):1682-1692.

Figure legends

Figure 1: The ACP process benefits not only the individual, but also their loved ones and care providers. Societal discussions of quality versus quantity of life as well as limitations of care are also important.

Figure 2: In accordance with advanced disease progression, discussion may transition from general health expectations to more specific topics. It is helpful for patients, loved ones and providers to prepare for in "in-the-moment decision making" in acute health situations in ways that reflect patients' known preferences and priorities. Over time, an individual's care needs may shift from active, disease targeted therapies (green), through periods of increasing symptoms and disability requiring more palliative care measures (purple). Active and palliative care strategies are not mutually exclusive (shades of blue).

Summary Figure: ACP is a staged and iterative process rather than a one-time event. ACP 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 ACP, 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 CHD prefer to have discussions about ACP care with their treating ACHD 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 ACHD specialist to mention and initiate ACP, if whished by the patient.

Box 1 – The perspective of adults living with CHD

- In a society that mutes discussions on death and dying and that simplistically lauds improved ACHD outcomes as success stories, health care providers tend to avoid and/or delay ACP discussions with their patients.
- Patient organisations can help health care providers spread information about ACP. They are also irreplaceable for adults with 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 health care providers about future health are typically welcome, even though this is often considered a stressful topic. Adults with CHD as well as ACHD health care 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.

Box 2 – Advance care planning: glossary of terms

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 health care 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) health care 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 health care 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 ***health care representative*** (also referred to as a substitute decision maker, surrogate decision maker, power of attorney for personal care or health care 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 health care 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 health care representative.

Table 1: Barriers to ACP communication and proposed clinical strategies

Barrier	Strategies for health care providers
At patient level	
No/minimal knowledge about ACP	Provide education about the rationale for ACP
Reluctance to begin this discussion	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 Encourage patients to have loved ones present for these conversations 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)
Avoidance of discussion of health deterioration (which might be amplified during acute cardiac events)	Initiate ACP during stable (rather than acute) phase of the disease to allow for a less-pressured patient experience
Desire to protect family and loved ones	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
At health care provider level	
Fear of causing patients to experience unnecessary emotional distress	Acknowledge that emotional reactions to disappointing information are understandable Acknowledge and label emotional reactions as they occur Strive for a balance between preparation for undesired outcomes and maintaining hope; emphasize that these are not mutually exclusive Emphasize that ACP is intended to be an empowering process for patients and their loved ones
Uncertainty about prognosis	Acknowledge challenges associated with an unpredictable disease trajectory Use standardized prognostic indexes (e.g. NYHA class) or results from functional testing (e.g., cardiopulmonary exercise testing) to guide predictions Offer broader time ranges for life expectancy (i.e. decades vs. years vs. months vs. weeks)
Lack of confidence and skills in ACP	Practice discussions with colleagues to develop comfort with this language Consider a standardized approach (e.g., ask-tell-ask) to initiate discussions Use a checklist to guide discussions (see supplementary material)

	Seek continuing education opportunities Liaise with a palliative care team to enhance skills
Low familiarity with specific patient factors that warrant unique attention (e.g., culture, religion, background)	Communicate in a respectful and sensitive manner Inquire about religious, cultural and background factors that may impact patients' decision-making Maintain a respectful curiosity regarding patients' beliefs and practices Avoid assumptions based on patients' culture, religion, or background
Personal discomfort with ACP discussions and end-of-life care	Acknowledge that emotional reactions are understandable Be aware that responding to emotions may enhance rather than diminish the patient-provider relationship Remain cognizant of situations in one's own life that may impact comfort level (e.g., death of a loved one, one's personal health challenges) and seek counselling as appropriate Seek peer/professional consultation to develop strategies to manage emotional distress
At ACHD program/institutional level	
Ambiguity regarding who is responsible for initiating and maintaining the ACP dialogue	Acknowledge that adults with CHD prefer to discuss ACP with clinicians whom they trust Develop a standardized process for identifying the team member who is responsible for overseeing ACP
Lack of time	Advocate for a clinical scheduling practice that allows sufficient time to discuss ACP within a routine outpatient visit Schedule ACP-specific clinical visits Recognize that time might be more flexible within the inpatient setting

Table 2: Staged Implementation of Advance Care Planning*

	Who	Action	Steps
Stage 1	**Anticipated life expectancy: decade(s)** Adult with CHD with any of the following ▪ expressing interest in future health discussion (e.g. during transition to adulthood) ▪ having unrealistic health expectations, particularly when confronted with important life planning decisions (e.g. family planning) ▪ reduced life expectancy, such as - adults with Fontan procedure - adults with cyanotic heart disease - adults with a systemic right ventricle approaching 40 years of age	▪ Invite discussions about future health expectations and preferences ▪ Explain the rationale and advantages of ACP ▪ Discuss future health expectations, while acknowledging challenges with longer-term prognostication ▪ Inquire about personal preferences, goals and personal values ▪ Offer to include relatives or loved ones in the conversation	▪ Schedule dedicated outpatient visit(s) for the purpose, as appropriate ▪ Provide written documentation in medical records of elements discussed ▪ Share information with general practitioner and other health care professionals
Stage 2	**Anticipated life expectancy: years** Adult with CHD with any of the following ▪ expressing interest in ACP discussion ▪ before CRT or ICD implantation ▪ at the time of diagnosis with advanced heart failure, particularly before heart transplant assessment[22] ▪ requiring cardiac surgery, complex catheter-based therapeutic interventions	▪ Revisit the elements discussed at stage 1 AND ▪ Offer more comprehensive ACP discussion ▪ Prepare or review advance directives including the nomination of a health care representative ▪ Inform and discuss about POLST	▪ Schedule dedicated outpatient visit or facilitate ACP discussion during an inpatient stay ▪ Provide an update of written documentation of ACP, if applicable ▪ Document advance directives (including health care representative) and/or POLST and share this information with all stakeholders

Stage 3	**Anticipated life expectancy: weeks to months** Adult with CHD with any of the following ▪ their provider would not be surprised if the patient died within the next year ▪ refractory end-stage heart failure[21, 22] (e.g. a failing Fontan circulation; repeated re-admission for decompensated heart failure requiring inotropic support and/or ICU stay; if temporary or long-term mechanical circulatory support is considered or may arise in due management course)	▪ Revisit the elements discussed at stage 2 AND ▪ Discuss end-of-life preferences, including the location of death ▪ Organize support for family members ▪ Involve palliative care team as appropriate ▪ As appropriate, discuss deactivation of implanted cardiac device functions	▪ Update written ACP documents as applicable ▪ Consider organization of home care ▪ Consider involvement of social work ▪ Consider involvement of palliative care ▪ Consider involvement of psychology and/or religious support providers
Stage 4	**Anticipated life expectancy: days** The dying adult with CHD	▪ Provide end-of-life care reflecting personal preferences and documented directives ▪ Coordinate bereavement care for loved ones, as appropriate	▪ Consider involvement of social work ▪ Consider involvement of palliative care ▪ Consider involvement of psychology and/or religious support providers ▪ Provide support to care team as necessary

*Adapted table from «Staged implementation of advance care planning, anticipatory care planning and integrated end-of-life care planning"[30]

Figure 1: Benefits of ACP in adults with cardiac disease[56-63]

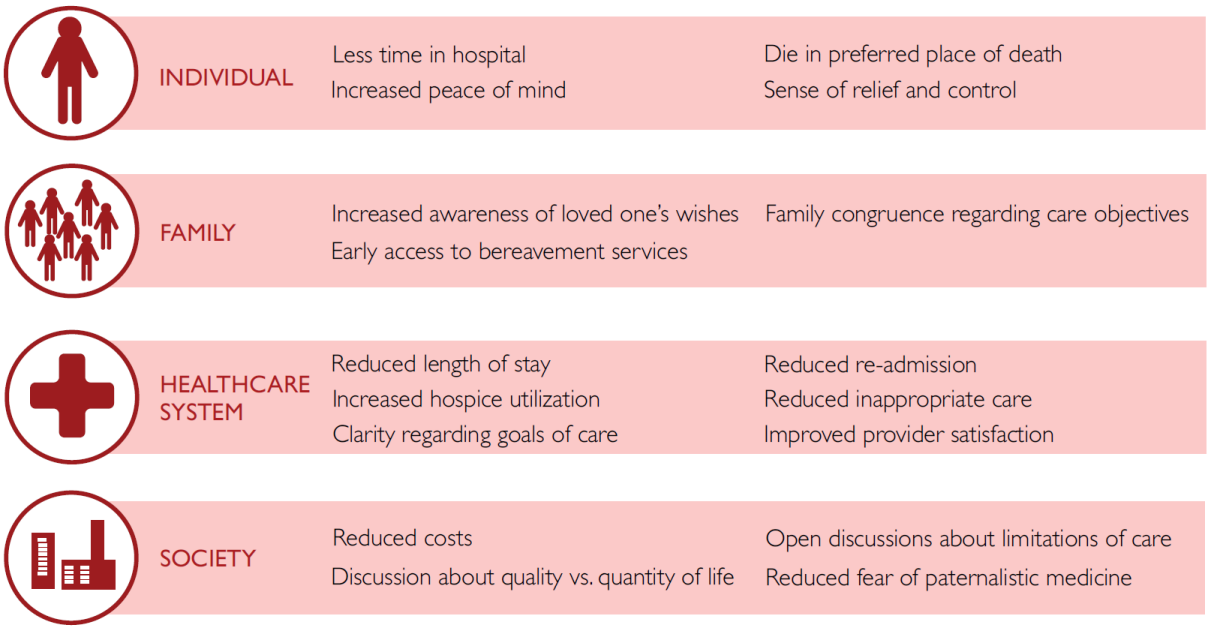
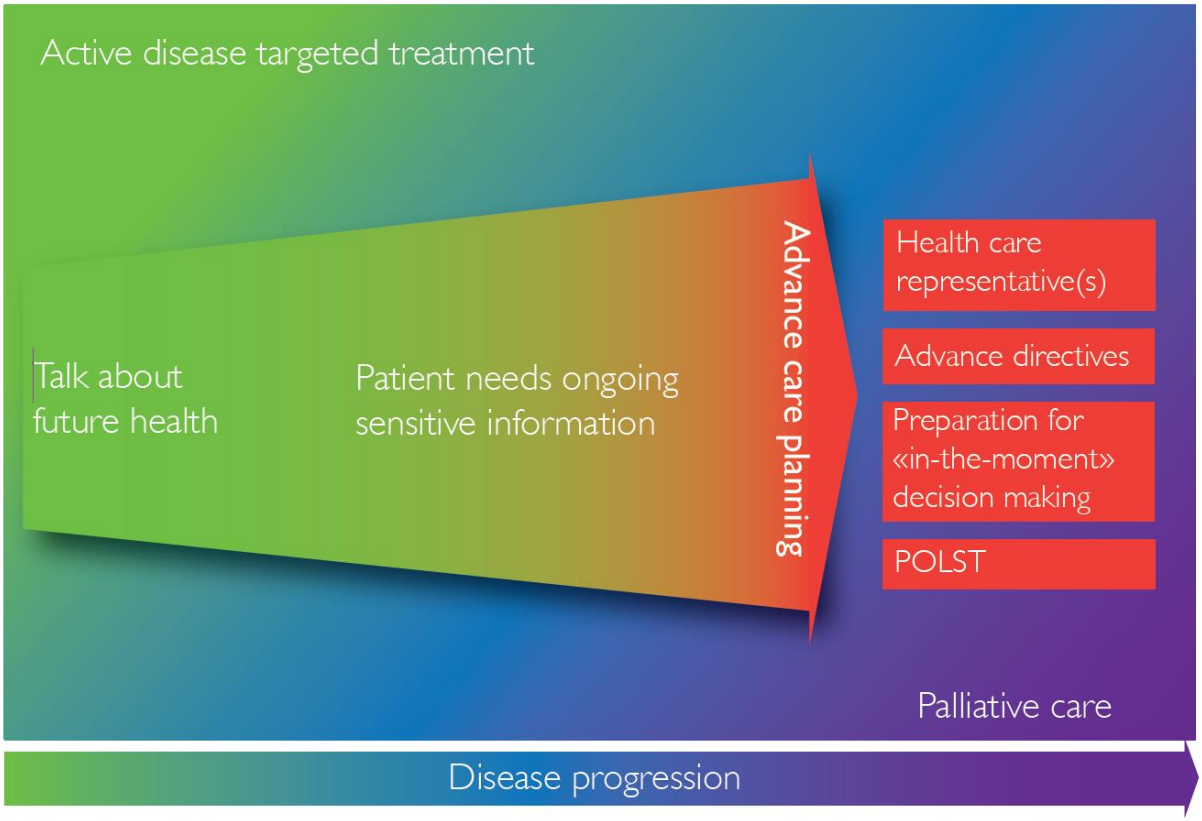


Figure 2: Evolution of the ACP discussion in relation to disease progression



Summary Figure: Staged implementation of ACP