

Heart Disease Community of Practice Series 3

Non ischemic causes of heart failure



Host: **Holly Finn, Pallium Canada**
Presenter: **Dr. Lynn Straatman, MD FRCPC**

Date: **June 25th 2025**

Territorial Honouring



The Palliative Care ECHO Project

The Palliative Care ECHO Project is a 5-year national initiative to cultivate communities of practice and establish continuous professional development among health care providers across Canada who care for patients with life-limiting illness.

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The Palliative Care ECHO Project is supported by a financial contribution from Health Canada. The views expressed herein do not necessarily represent the views of Health Canada.



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Introductions

Host

Holly Finn, PMP

Senior Manager of Program Delivery, Pallium Canada

Presenter

Dr. Lynn Straatman, MD FRCPC

Clinical Assistant Professor, UBC

Department of Medicine (Cardiology and Palliative Care)

Medical Director, Cardiac Function Clinic

Introductions

Panelists

Dr. Caroline McGuinty, MD FRCPC

Cardiologist, Advanced Heart Failure and Transplantation, Cardiac Palliative Care
University of Ottawa Heart Institute
Assistant Professor, University of Ottawa

Drew Stumborg, RN

Saskatchewan Health Authority

Dr. Michael Slawnych, MD FRCPC

Clinical Assistant Professor
Department of Cardiology, St Paul's Hospital
University of British Columbia

Shannon Poyntz, NP-PHC, MN

Nurse Practitioner, Supportive Care

Dr. Lynn Straatman, MD FRCPC

Clinical Assistant Professor, UBC
Department of Medicine (Cardiology and Palliative Care)
Medical Director, Cardiac Function Clinic

Disclosure

Relationship with Financial Sponsors:

Pallium Canada

- Not-for-profit
- Funded by Health Canada
- Boehringer Ingelheim supports Pallium Canada through an in-kind grant to expand interprofessional education in palliative care.

Disclosure

This program has received financial support from:

- Health Canada in the form of a contribution program
- Pallium Canada generates funds to support operations and R&D from Pallium Pocketbook sales and course registration fees
- An educational grant or in-kind resources from Boehringer Ingelheim.

Facilitator/ Presenter/Panelists:

- Holly Finn: employed at Pallium Canada.
- Dr. Leah Steinberg: Pallium Canada (education material), HPCO (clinical advisory committee, educator).
- Morgan Krauter: Novartis, Alnylam, Pfizer (speaker fees); Allevant (consulting fees).
- Dr. Michael Slawnych: Novartis.
- Dr. Caroline McGuinty: Servier (consulting fees), Novartis (speaker fees).
- Dr. Lynn Straatman: Servier, Novartis, Astra Zeneca, BI, Medtronic, Pfizer, Eli Lilly, Bayer, Merck (clinical trials).
- Shannon Poyntz: None to disclose.
- Drew Stumborg: None to disclose.

Disclosure

Mitigating Potential Biases:

- The scientific planning committee had complete independent control over the development of program content

Welcome and Reminders

- Please introduce yourself in the chat!
- Your microphones are muted. There will be time during this session for questions and discussion.
- Please use the Q&A function to ask questions.
- Add comments or to let us know if you are having technical difficulties via the Chat!
- This session is being recorded and will be emailed to registrants within the next week.
- Remember not to disclose any Personal Health Information (PHI) during the session.
- This 1-credit-per hour Group Learning program has been certified by the College of Family Physicians of Canada for up to **6 Mainpro+** credits.
- This event is also an Accredited Group Learning Activity through the Royal College of Physicians and Surgeons of Canada. You may claim a maximum of **6.00 hours**.

Objectives of this Series

After participating in this program, participants will be able to:

- Describe what others have done to integrate palliative care services into their practice.
- Share knowledge and experience with their peers.
- Increase their knowledge and comfort around integrating a palliative care approach for their patients with advanced heart failure.

Overview of Topics

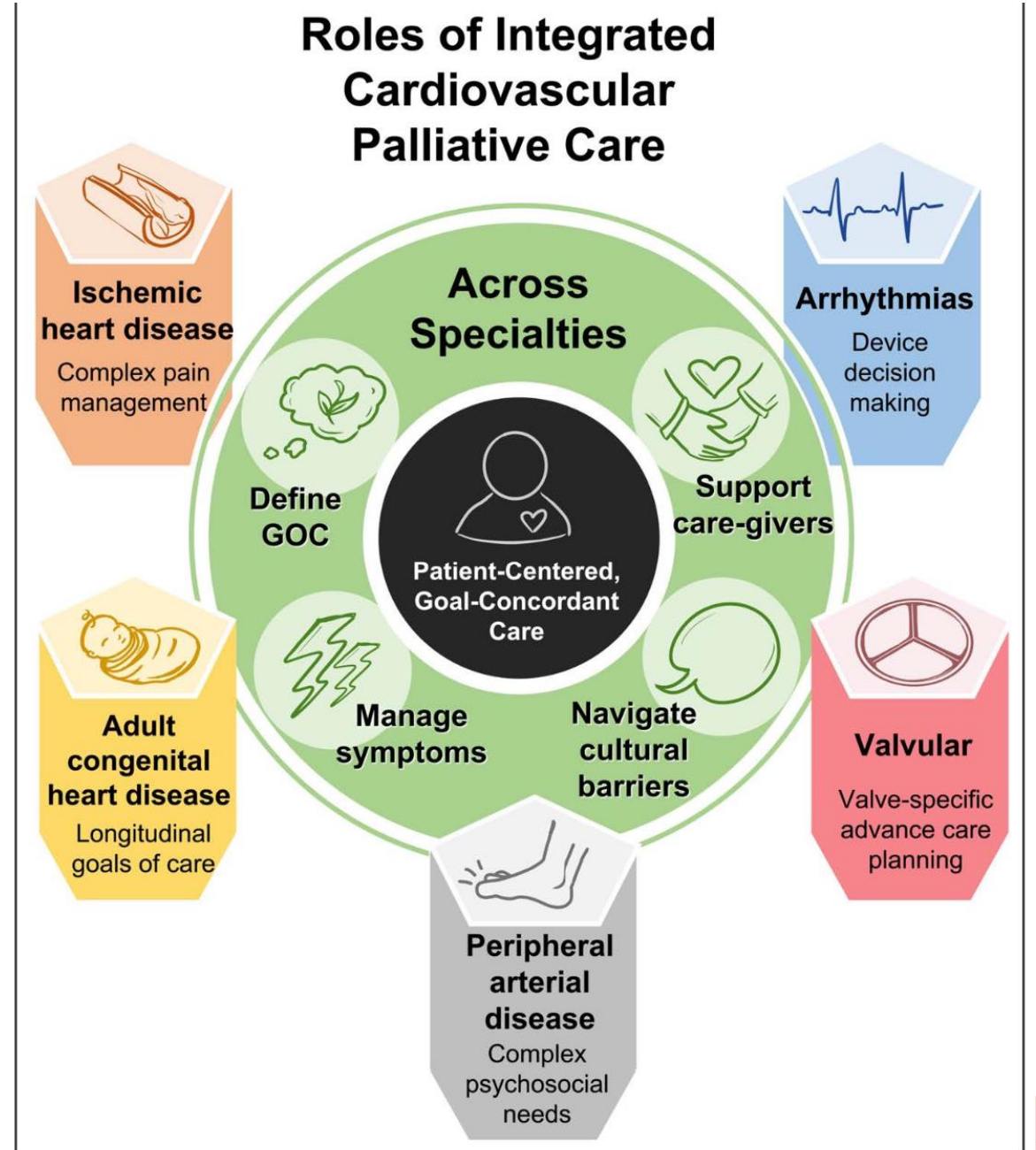
Session #	Session title	Date/ Time
Session 1	Collaboration Building: How to build collaboration with teams in your setting	October 2, 2024 from 12-1pm ET
Session 2	Diuretic management in an outpatient setting	December 11, 2024 from 12-1pm ET
Session 3	Challenging conversations	February 5, 2025 from 12-1pm ET
Session 4	De-prescribing cardiac and other medications: palliative care in people with advanced heart failure	April 30, 2025 from 12-1pm ET
Session 5	Non ischemic causes of heart failure	June 25, 2025 from 12-1pm ET
Session 6	Interaction of heart failure and lung disease	August 20, 2025 from 12-1pm ET

Objectives of this Session

After participating in this session, participants will be able to:

- Identify non-ischemic causes of heart failure (e.g. congenital heart disease, amyloidosis)
- Appreciate the different management techniques for non-ischemic heart failure

Not all Cardiac Palliative Care is Heart Failure



AHA SCIENTIFIC STATEMENT

Palliative and End-of-Life Care During Critical Cardiovascular Illness: A Scientific Statement From the American Heart Association

Circulation. 2025;151:e00–e00. DOI: 10.1161/CIR.0000000000001334

AHA SCIENTIFIC STATEMENT

Palliative Care and Advanced Cardiovascular Disease in Adults: Not Just End-of-Life Care: A Scientific Statement From the American Heart Association

Circulation. 2025;151:e1030–e1042. DOI: 10.1161/CIR.0000000000001323



Table. Proposed Cardiovascular Palliative Care Interventions for Disease-Specific Challenges

Type of CVD	Challenge	Palliative care intervention	Outcome
Ischemic heart disease	Refractory pain	Multidisciplinary angina program with integrated palliative care to address pain control, psychosocial support, mood symptoms	Holistic approach to pain management
Valvular heart disease	Complications after valve procedure	Advance care planning from initial diagnosis through postprocedural period	Valve preparedness plan to establish goal-concordant valve care
Arrhythmia	ICD shocks at end-of-life	Palliative care-facilitated shared decision-making regarding ICD implantation and subsequent ICD shock settings	Alignment of ICD settings with ongoing goals of care
Peripheral artery disease	High rates of in-hospital death	Assistance with end-of-life care including hospice referral when appropriate	Better quality of death in concordance with goals of care
Adult congenital heart disease	Quality of life with chronic illness at young age	Longitudinal goals of care discussions to prioritize quality of life at key milestones (pregnancy planning, career/financial difficulties, caregiving)	Psychosocial support throughout complex chronic illness in early to mid adulthood

• Not all Heart Failure is Ischemic or HFrEF

- Adult Congenital Heart Disease
- Amyloidosis (Infiltrative)-
- Valvular
- Neuromuscular (DMD)
- Restrictive
- HOCM

The Changing Demographics of Congenital Heart Disease

- What is the percentage of children born with congenital heart disease?
- What percentage of children with congenital heart disease live to adulthood in 2024?
- What is the ratio in the population of children to adults living with congenital heart disease?

What is the percentage of children born with congenital heart disease?

- Congenital heart disease (CHD) is the world's leading birth defect. About 1 in 80-100 Canadian children are born with CHD.
- There are an estimated 257,000 Canadians with Congenital Heart Disease.
- There are over 40 types of congenital heart defects

What percentage of children with congenital heart disease live to adulthood in 2024?

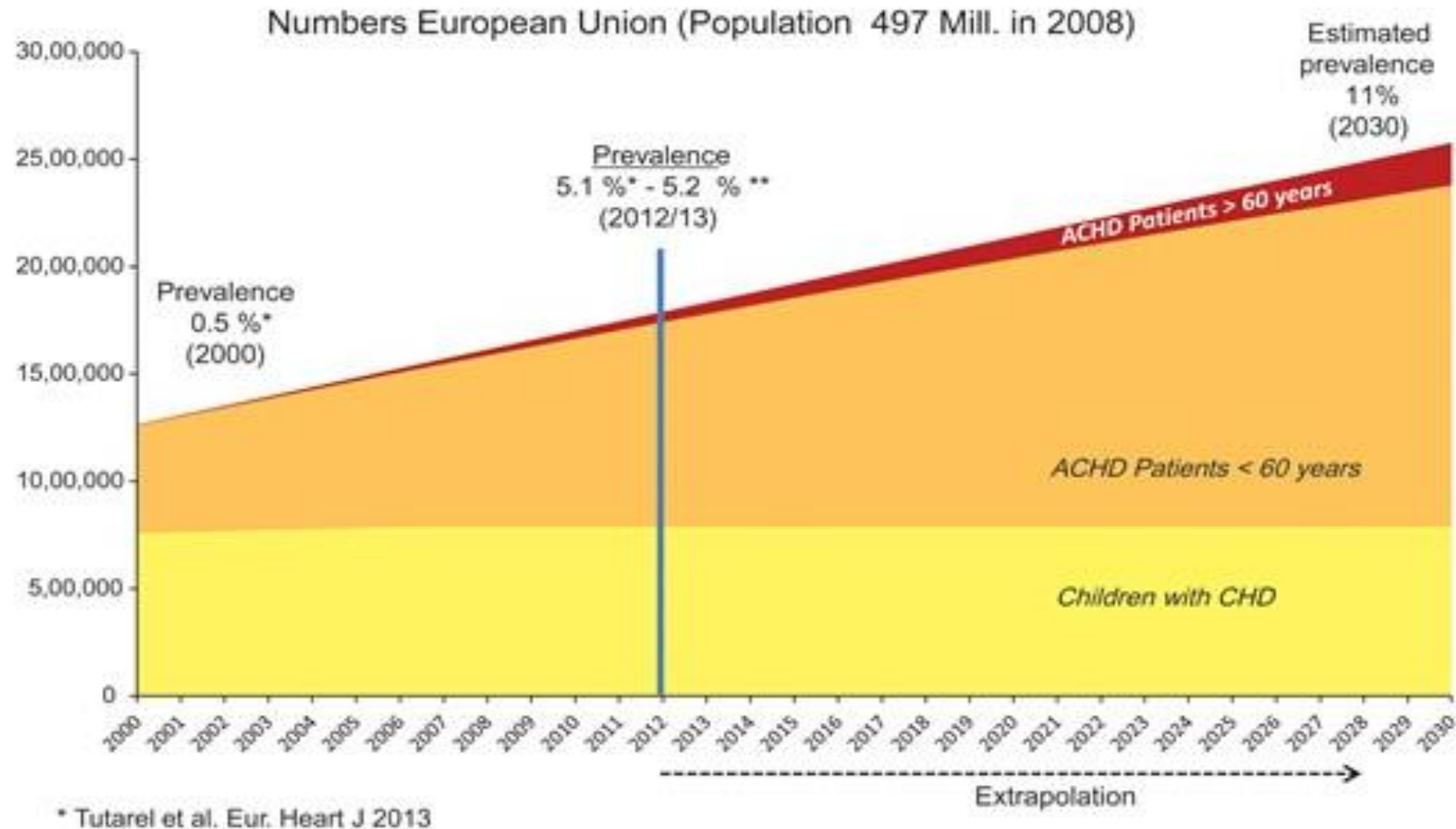
Journal of the American Heart Association

ORIGINAL RESEARCH

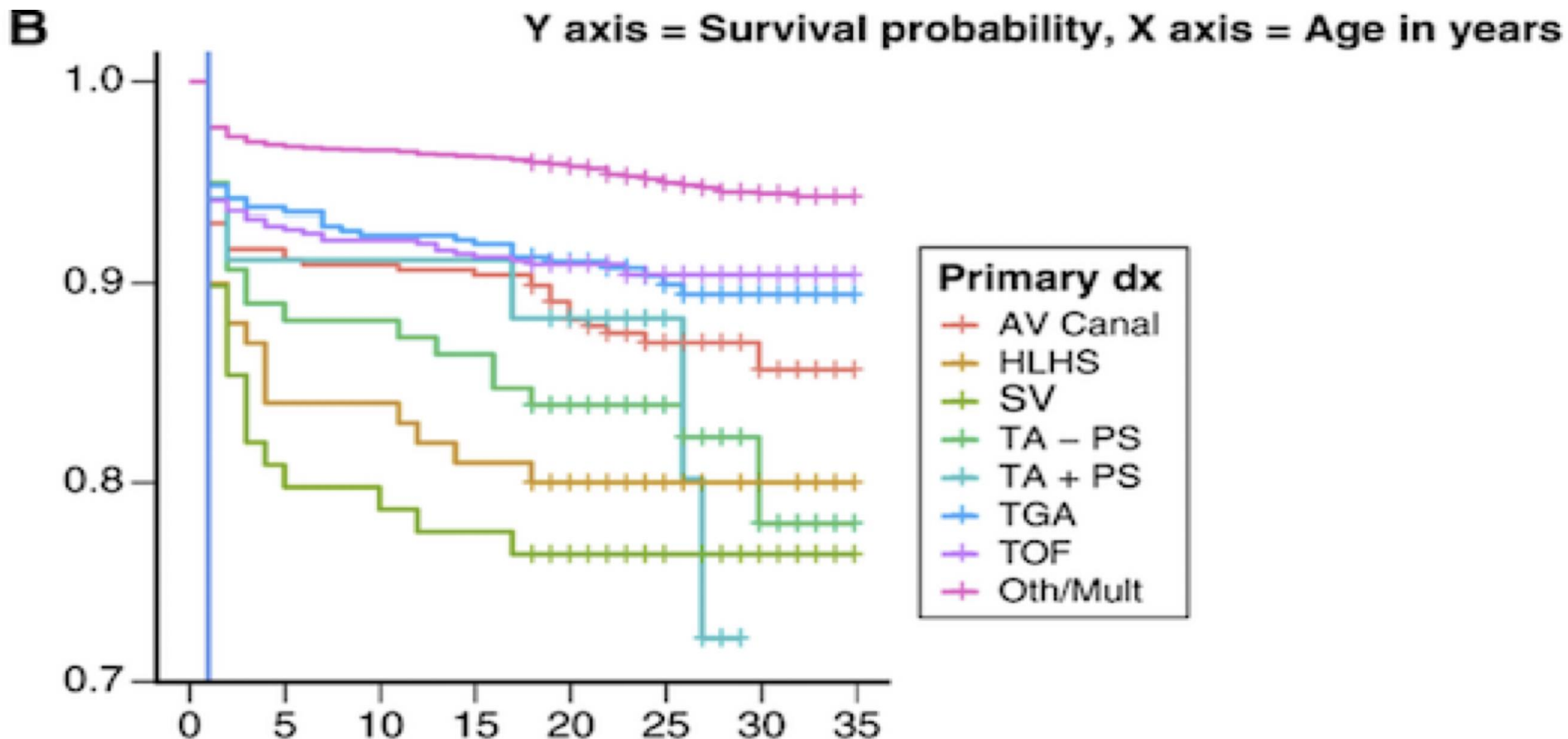
Survival in Children With Congenital Heart Disease: Have We Reached a Peak at 97%?

Zacharias Mandalenakis , MD, PhD; Kok Wai Giang, PhD; Peter Eriksson, MD, PhD; Hans Liden, MD, PhD; Mats Synnergren, MD, PhD; Håkan Wåhlander, MD, PhD; Maria Fedchenko, MD, PhD; Annika Rosengren , MD, PhD; Mikael Dellborg, MD, PhD

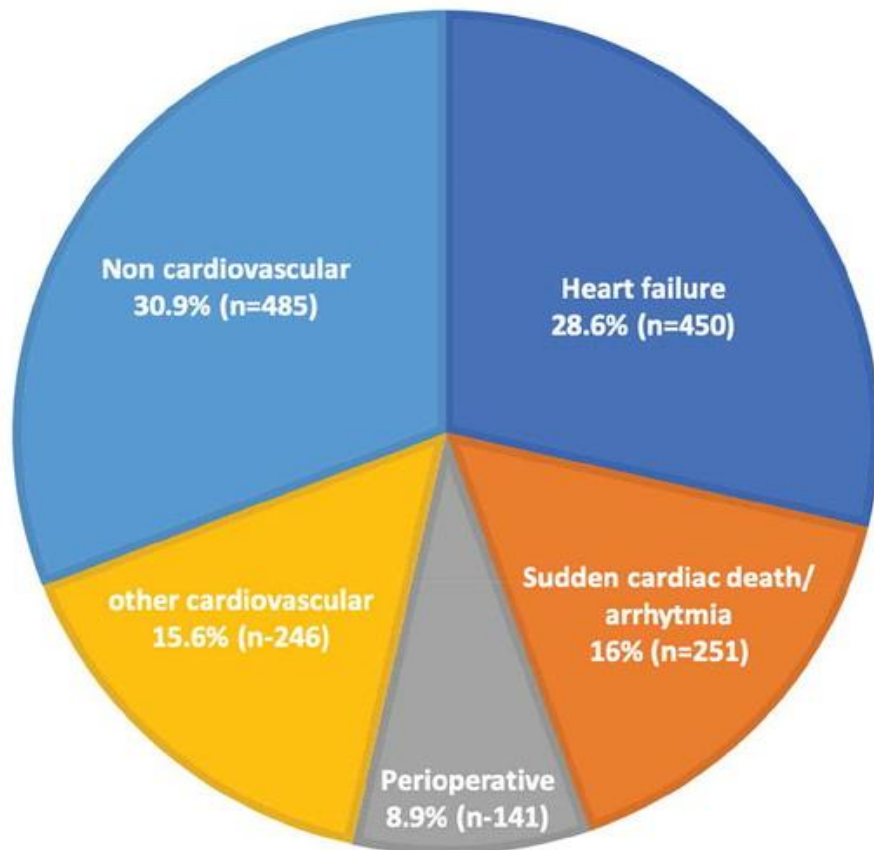
What is the ratio in the population of children to adults living with congenital heart disease?



Changes in Mortality for Congenital Heart Disease



Cause of death in Adult Congenital Heart Disease



Incidence of HF

ToF - 31.7% ages 20 to 29

52.9% over 50

TGA - after atrial switch - 25% age 20,

40% age 30 and 60% age 40

If CHD and admitted to hospital 28x more likely to die than non CHD HF patient

Most HF is HFREF except in Fontan patient - HFREF and HFPEF

Causes of Heart Failure in Patients with CHD

- Volume overload resulting from right to left shunt lesions and valvular regurgitation
- Pressure overload resulting from valvular disease and obstructive lesions
- Ventricular failure resulting from intrinsic myocardial dysfunction
- Pulmonary hypertension caused by CHD lesions, ventricular dysfunction or co-morbidities such as OSA
- System arterial hypertension resulting from coarctation, acquired renal disease
- Coronary artery disease related to CHD or atherosclerosis
- Cyanosis
- Intractable atrial arrhythmias

HFrEF: LVEF $\leq 40\%$ and Symptoms

Initiate Standard Therapies

ARNI or
ACEi/ARB then
substitute ARNI

Beta
blocker

MRA

SGLT2
Inhibitor

Assess Clinical Criteria for Individualized Therapies

HR >70 bpm and
sinus rhythm

- Consider
ivabradine[†]

Recent HF
hospitalization

- Consider
vericiguat^{**}

Black patients
on optimal GDMT,
or patients unable
to tolerate
ARNI/ACEi/ARB

- Consider H-ISDN

Suboptimal rate
control for AF,
or persistent
symptoms despite
optimized GDMT

- Consider digoxin

*Initiate standard therapies as soon as possible and titrate every 2-4 weeks
to target or maximally tolerated dose over 3-6 months*

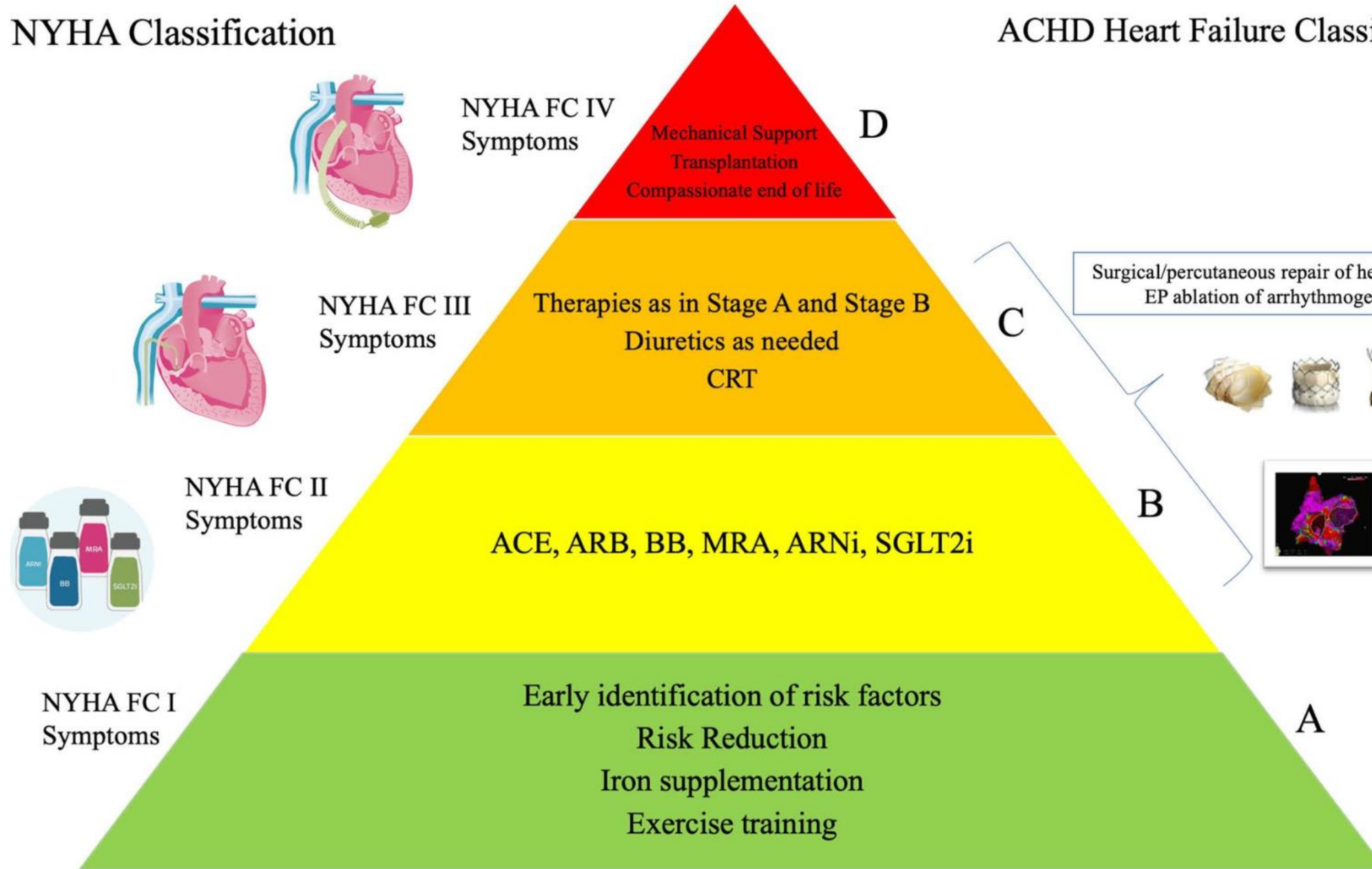
Treat Comorbidities per CCS HF Recommendations
(incl. AF, functional MR, iron def. CKD, DM)

Diuretics to Relieve Congestion (titrated to minimum
effective dose to maintain euvoolemia)

Non-pharmacologic Therapies (teaching, self-care, exercise)

Advance Care Planning and Documentation of Goals of Care

NYHA Classification



ACHD Heart Failure Classification

Surgical/percutaneous repair of hemodynamic lesions
EP ablation of arrhythmogenic substrates

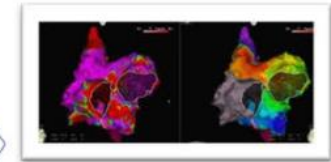
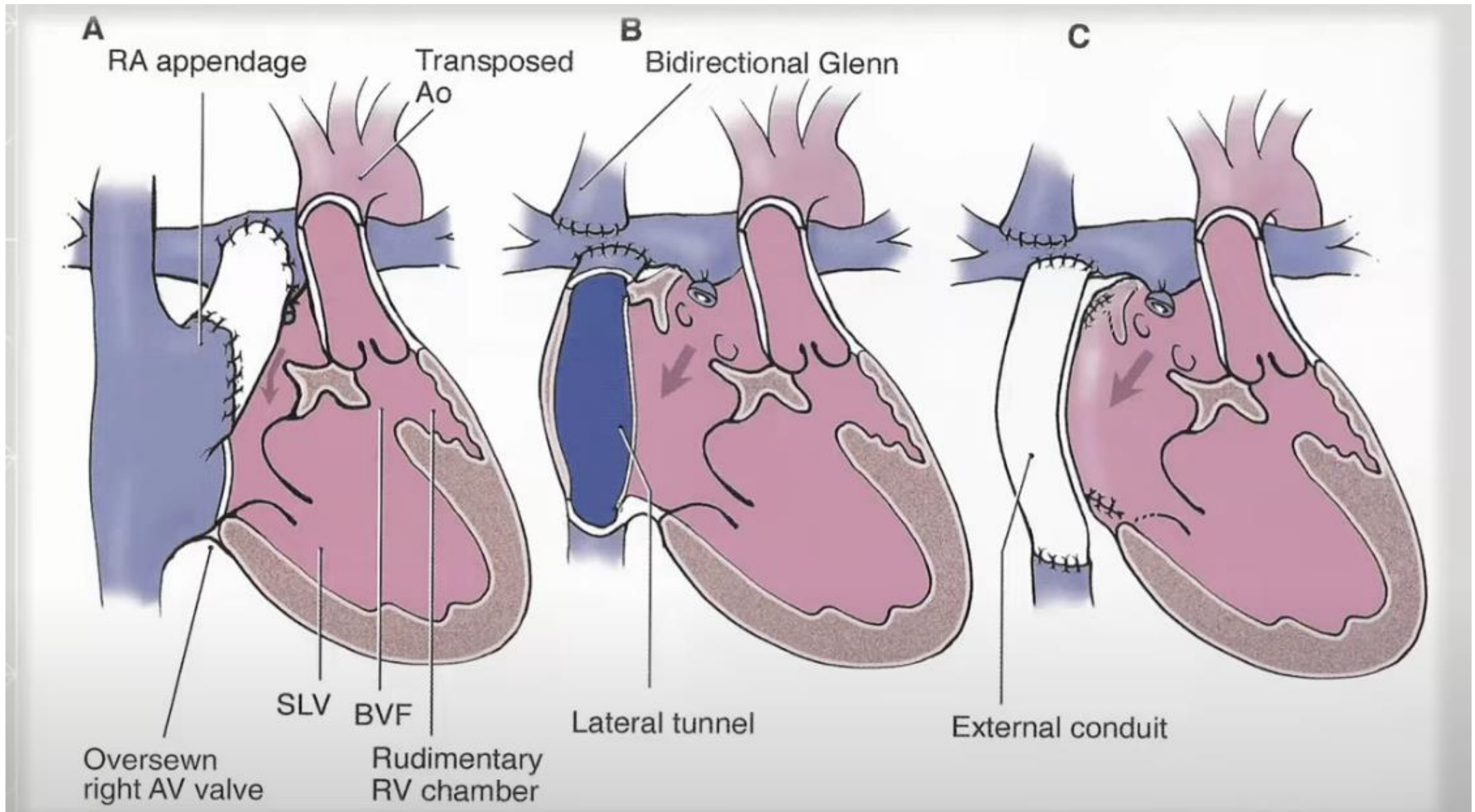


Table 1 ACE/ARB use in adult congenital heart disease

Study	Study type	Population	Intervention	Outcome
van der Bom et al. [14]	RCT ($n=88$)	Systemic RV	Valsartan vs placebo	Improved sRV mass and volumes on CMRI. No change in sRV EF or exercise capacity.
van Dissel et al. [15]	RCT ($n=88$)	Systemic RV	Valsartan vs placebo for longer term	Decreased risk of events. No improved survival at long-term follow-up.
Tutarel et al. [16]	Retrospective cohort ($n=14$)	D-TGA with atrial switch	ACE	Improved NT-proBNP levels.
Lester et al. [17]	RCT with crossover ($n=7$)	D-TGA with atrial switch	Before and after losartan (each patient serving as own control)	Improved EF, decreased AV valve regurgitation, increased exercise time.
Therrien et al. [18]	RCT ($n=17$)	Systemic RV	Ramipril vs none	No change in RV function.
Dore et al. [19]	Multicenter RCT with cross over ($n=29$)	Systemic RV	Losartan vs placebo	No change in exercise capacity or NT-proBNP.
Hechter et al. [20]	Retrospective case control ($n=14$)	D-TGA with atrial switch	ACE vs none	No change in CMRI RVEF. Slight improvement in peak oxygen consumption and exercise time.
Ladouceur et al. [21]	Retrospective cohort ($n=353$)	Systemic RV	ACE or ARB	No change in incidence of heart failure or mortality.

ACE angiotensin converting enzyme inhibitor, *ARB* angiotensin II receptor blocker, *AV* atrioventricular, *CMRI* cardiac magnetic resonance imaging, *dTGA* dextro-transposition of the great arteries, *EF* ejection fraction, *NT-proBNP* N-terminal pro brain natriuretic peptides, *sRV* systemic right ventricle, *RCT* randomized control trial



Symptom Management in Congenital Heart Disease

Heart Failure

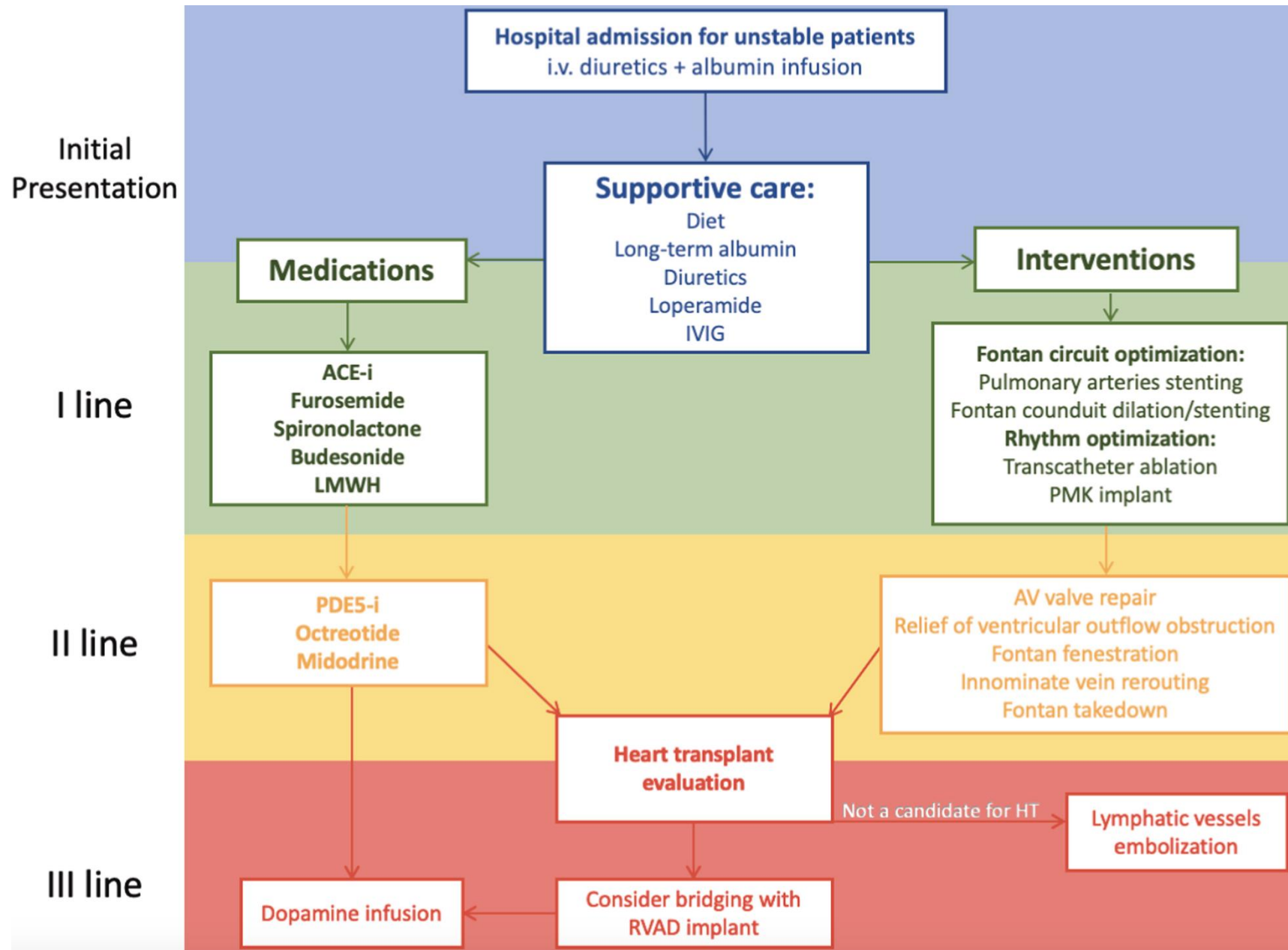
- Need to be careful with diuretics and hemodynamics (understand the underlying anatomy)
- Vasodilation often doesn't help in Fontan
- Hydromorphone can be used to treat dyspnea

Other Common Symptoms

- Gout
- Liver Failure - cirrhosis and portal hypertension(increases risk of procedures and may make ineligible for transplantation)
- Stroke - Among adults with CHD, 1 in 11 men and 1 in 15 women experience a stroke between 18 and 64 years of age

Protein Losing Enteropathy

- occurs in 5% to 12% of individuals after Fontan palliation
- pathophysiology of PLE remains incompletely characterized
- protein loss leads to decrease serum oncotic pressure which leads to systemic edema.
- when the intestinal wall becomes edematous, malabsorption occurs, and protein loss can worsen, leading to a vicious cycle of intestinal symptoms and edema often referred to as a PLE flare.
- Chronic diffuse edema leads to diminished tissue integrity, poor wound healing, coagulation abnormalities, decreases in bone density and immune abnormalities



Deficiencies and Disconnects in ACHD EOL Preparation

Targeting Patients Most Appropriate for Advanced Care Planning Discussion

Identify ACHD Patients at Increased Risk for Premature Mortality

- Eisenmenger Syndrome and severe pulmonary hypertension
- Unrepaired cyanotic heart disease
- Fontan-palliated single ventricle
- Systemic right ventricle

Be aware of life events that may facilitate ACP discussions

- Transition of care to an ACHD specialized provider
- Family planning
- Death of a peer with CHD
- Hospitalization

Recognize triggers that can portend a worsened prognosis

- Progression of symptoms
- Hospitalization for heart failure
- Development of progressive ventricular dysfunction, arrhythmias or pulmonary hypertension
- Need for catheter-based or surgical reinterventions

ndergone formal evaluation



European Society
of Cardiology

European Heart Journal (2022) **43**, 4493–4495

<https://doi.org/10.1093/eurheartj/ehac492>

EDITORIAL

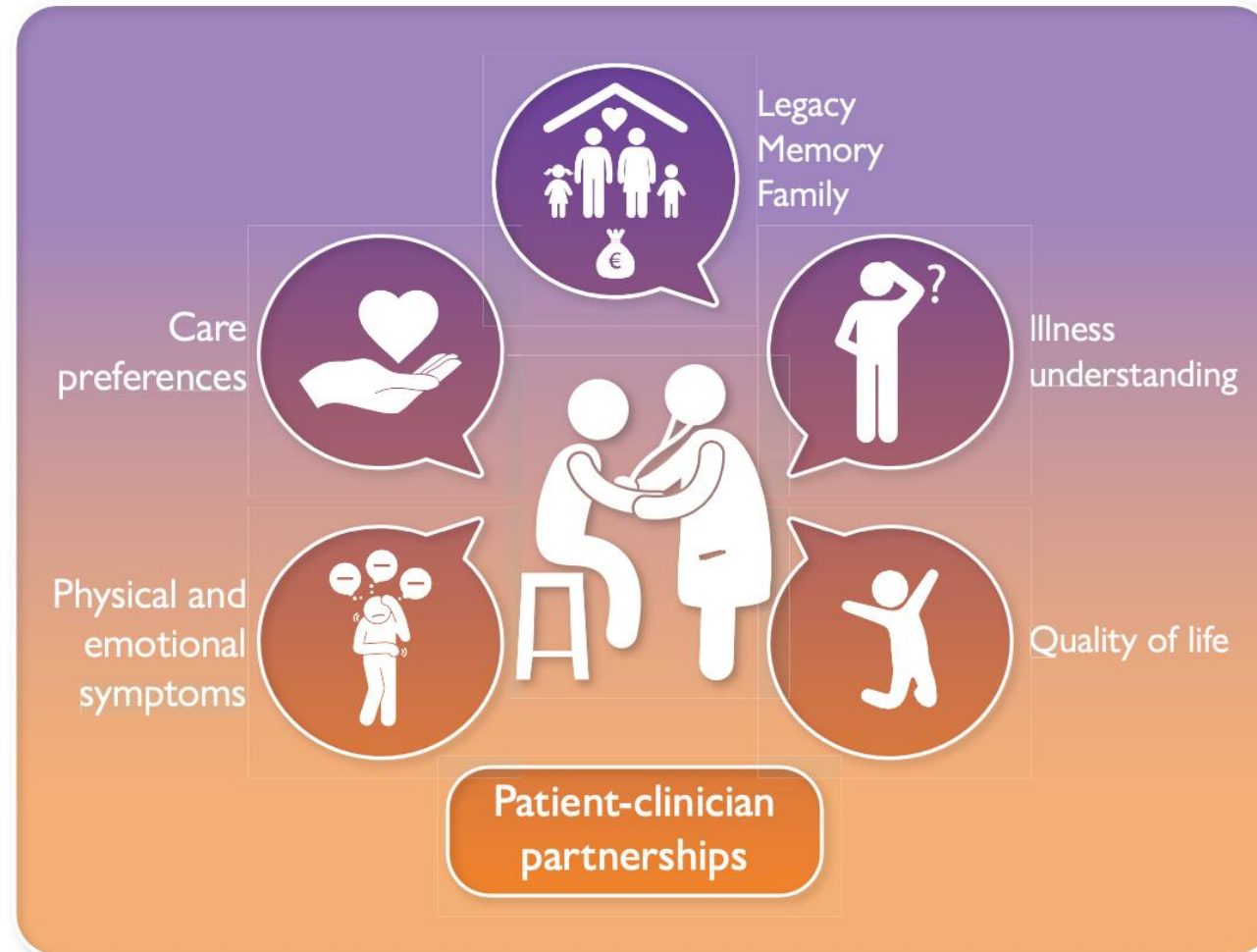
A view from the end: what the last year of life can teach us about palliative care on the adult congenital heart disease journey

Jill M. Steiner ^{1*} and James N. Kirkpatrick²

¹Division of Cardiology, Department of Medicine & Cambia Palliative Care Center of Excellence, University of Washington, 1959 NE Pacific St., HSB C502, Box 356422 Seattle, WA 98195, USA; and ²Division of Cardiology, Department of Medicine & Department of Bioethics and Humanities, University of Washington, 1959 NE Pacific St., Seattle, WA 98195, USA



ACHD palliative care needs



Needed future research

Meeting ACHD-specific goals of care

How and when to employ palliative care

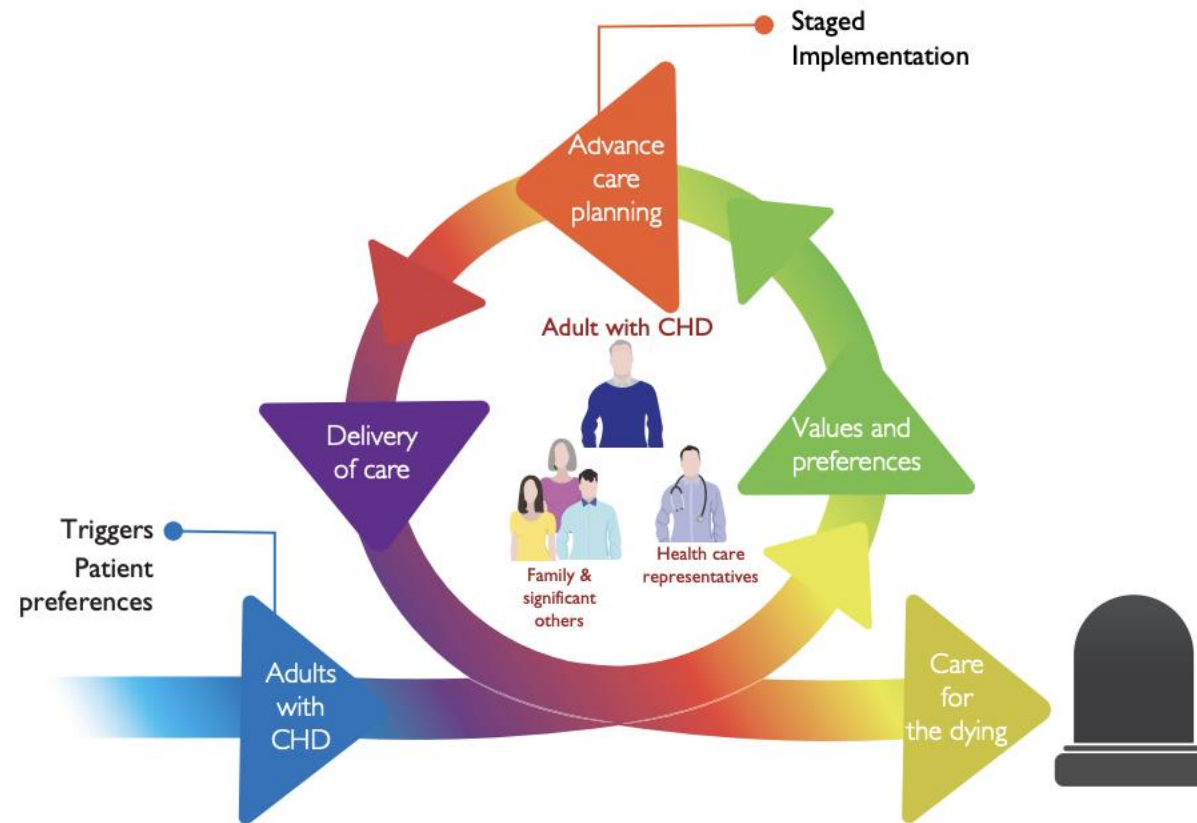
End-of-life trajectory

Changing end-of-life care needs

Identity, self-efficacy and resilience

Healthcare system response and support

Where does Palliative Care Intersect?



Amyloidosis - A Growing Population

- Cardiac amyloid - more common in older population, more common in men
- AL Amyloid annual incidence of 10 per million
- wtATTR - may account for 10% of the HEpEF population (and HFpEF numbers are quickly going to surpass HFrEF)



Amyloidosis

AL AMYLOIDOSIS



ATTR AMYLOIDOSIS



Common Cardiac Issues

- Heart Failure
- Arrhythmias
 - Atrial
 - Ventricular
- Heart Block
- Increased LA thrombus

Management of Heart Failure

- Volume management is the key
 - Loop diuretics and mineralocorticoid receptor antagonists (Spironolactone)
 - Cautious use of thiazide diuretics - Metolazone
- Usually poorly tolerate renin-angiotensin-aldosterone system inhibitors, as even low doses can precipitate significant hypotension
- β -blockers are also poorly tolerated
- SGLT2 inhibitors may offer benefits in selected patients, although data specific to cardiac amyloidosis remains limited.

Management of Arrhythmia

- Anticoagulation with a DOAC recommended
- Rate and rhythm control
 - Don't tolerate B blockers
 - Digoxin tends to bind to amyloid fibrils, increasing the risk of toxicity.
 - Calcium channel blockers have not been shown to provide significant benefit and may exacerbate hypotension.
 - Amiodarone is the drug of choice (half life 25 to 100 days)

Non cardiac issues

- Orthostatic Hypotension
 - Frequently encountered due to autonomic dysfunction
 - peripheral vasoconstrictors like midodrine may be used to support blood pressure
- GI symptoms
 - Very common and challenging to treat

GI symptoms

- Gastroparesis
 - D2 receptor antagonists
 - Metoclopramide
 - Motilin receptors
 - Erythromycin
- Constipation
 - PEG
- Diarrhoea
 - Loperamide
 - Octreotide

Questions/Discussion

Wrap Up

- Please fill out the feedback survey following the session! Link has been added into the chat.
- A recording of this session will be e-mailed to registrants within the next week.
- Please join us for the next session in this series on **Interaction of heart failure and lung disease** on **August 20, 2025 from 12-1pm ET**

Thank You



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