

# End of Life Care

## Adult Congenital Heart Disease

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# Past to Present

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- Increasing number of ACHD
- More complex CHD patients surviving...
- Remaining at longterm risk of complication
- Premature death

# How Long to Live...

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*What if death comes soon?*

→ Can we plan the end of life?

→ **Communication for EOL**

*Tobler D et al. Curr Opin Support Palliat Care 2011; 5:291–296*

*Bowater SE et al. Curr Opin Support Palliat Care 2013; 7:8–13*

*Rosenthal TM, et al. Congenit Heart Dis. 2016;11:189-98*

# Medical Problem

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- Hemodynamic / Shunts
- Ventricular dysfunction, Ventricular failure
- Valve stenosis or regurgitation
- Need for further surgery or intervention
- Pulmonary hypertension
- Arrhythmia, Conduction abnormalitis
- Cyanosis, Thromboembolic disease
- Management of pregnancy

# Psychosocial Problem

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- Self-esteem
- Employment
- Insurance
- Socialization
- Sports and exercise recommendation
- Flying or driving recommendation
- Contraceptive advices
- Genetic counselling

# How Long to Live...

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## → Poor knowledge

How long can I live from now?

Can I have a job for life?

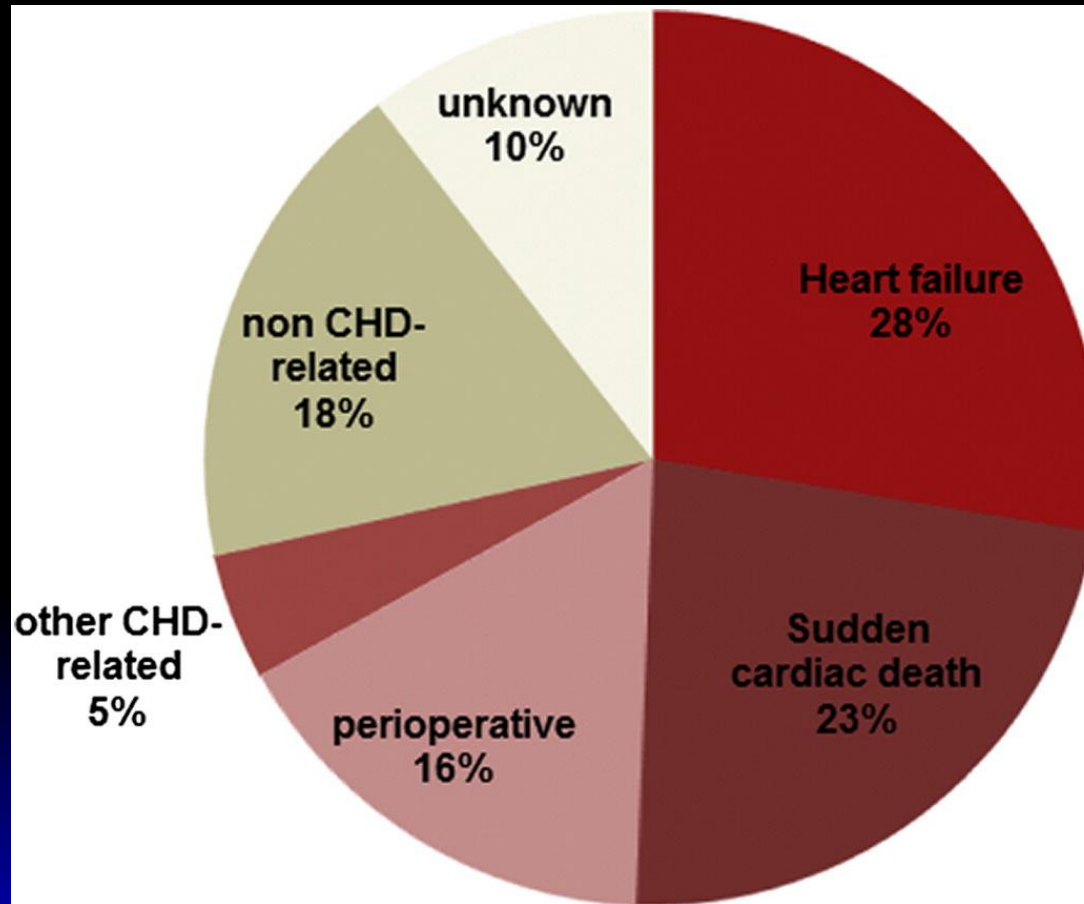
What kind of work I can do?

How much money for my CHD?

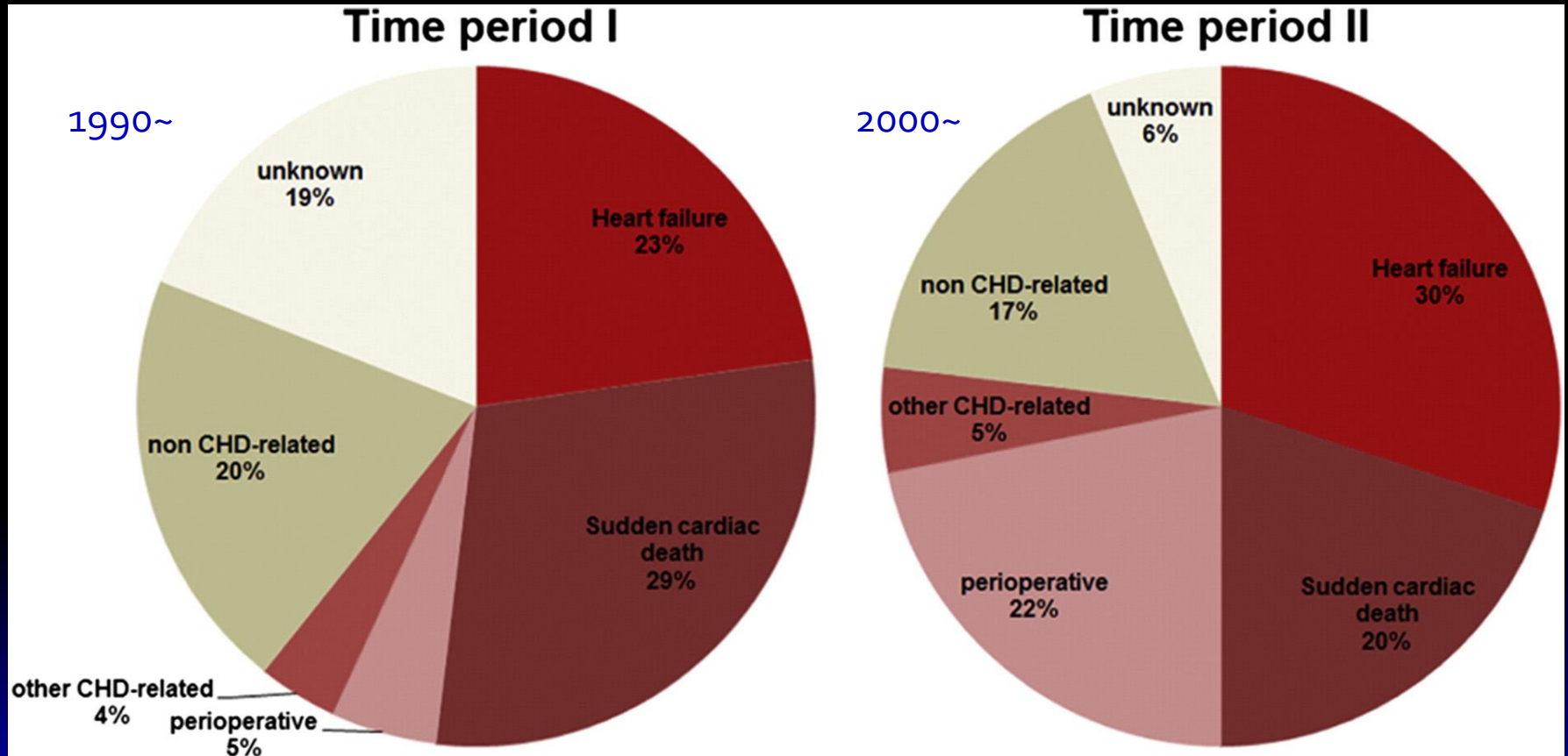
Can I marry someone?

Can the other person love me?

# Cause of Death...

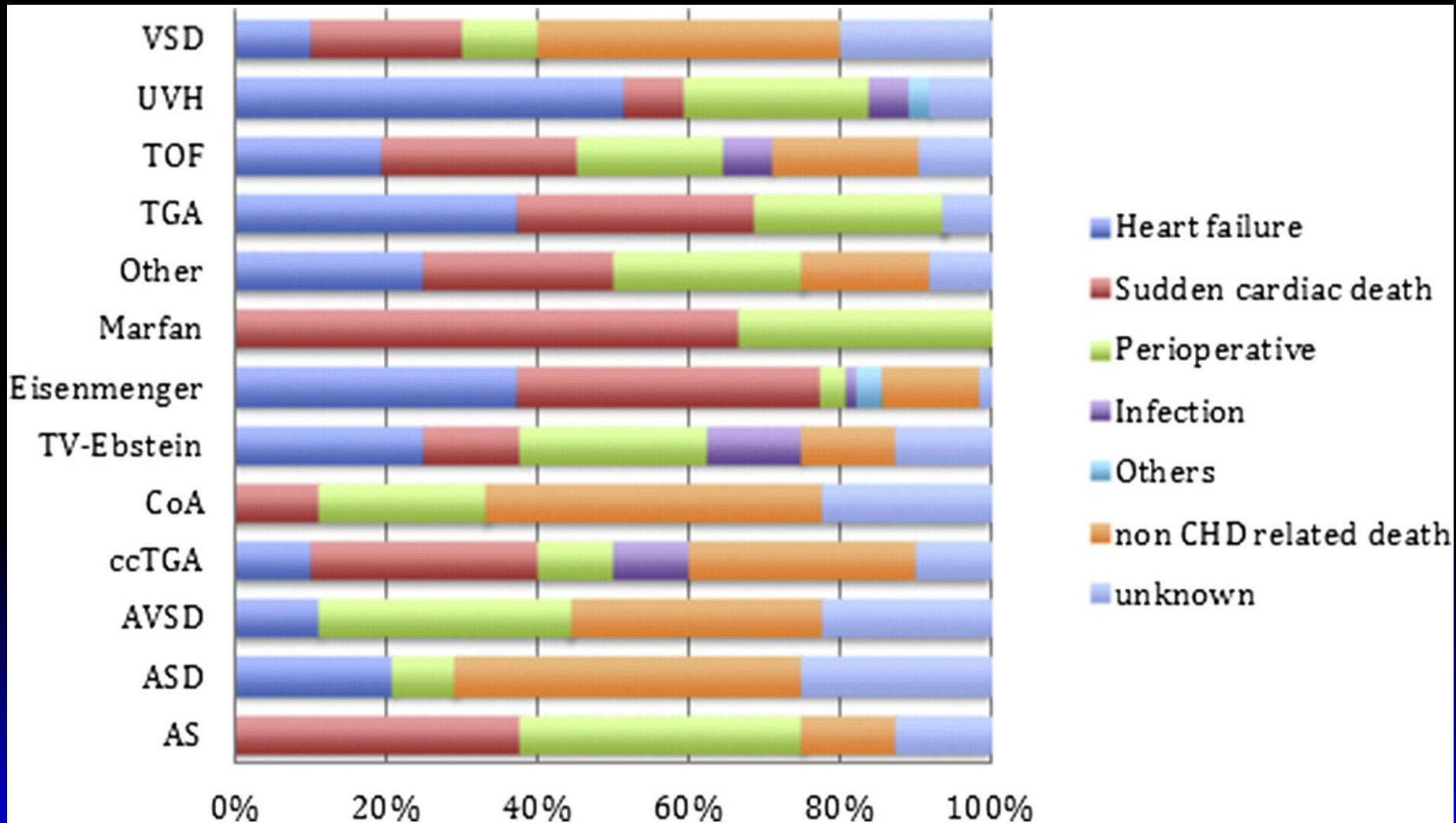


# Cause of Death...

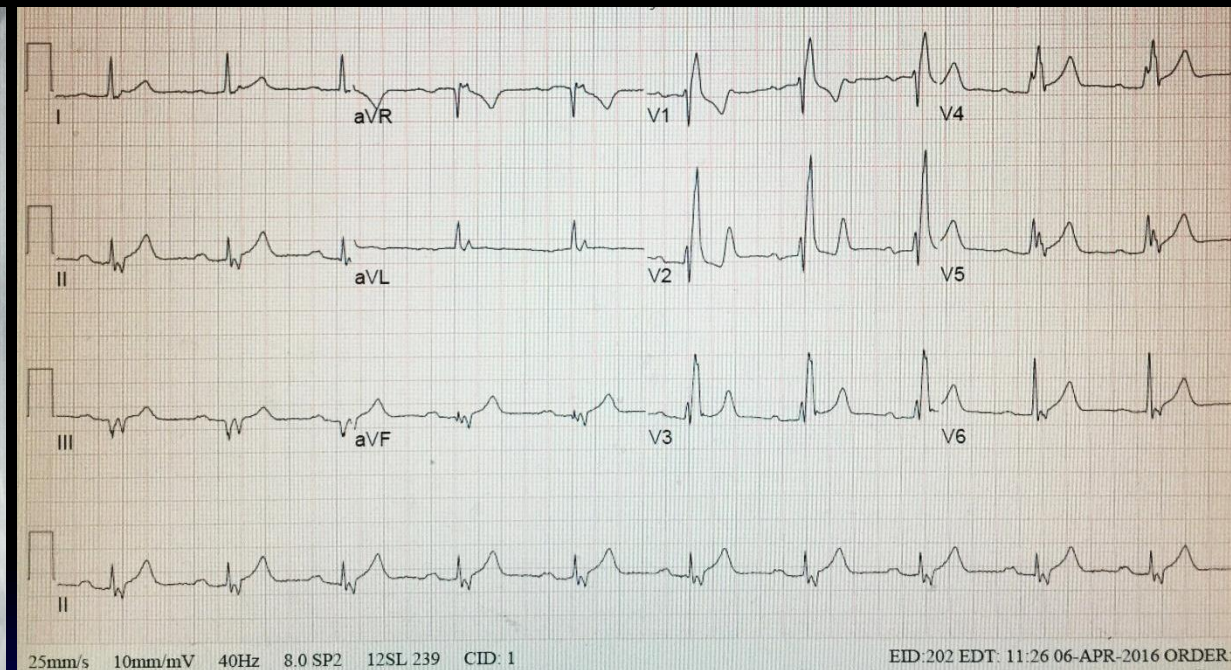
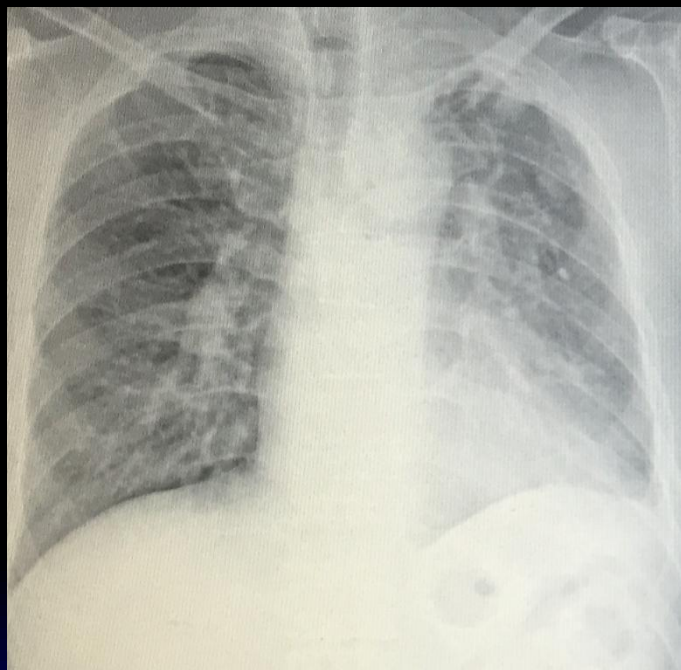




# Cause of Death...



# Case1. M/56 yr - S/P TOF repair



# What is the worst difficulty for lifetime?

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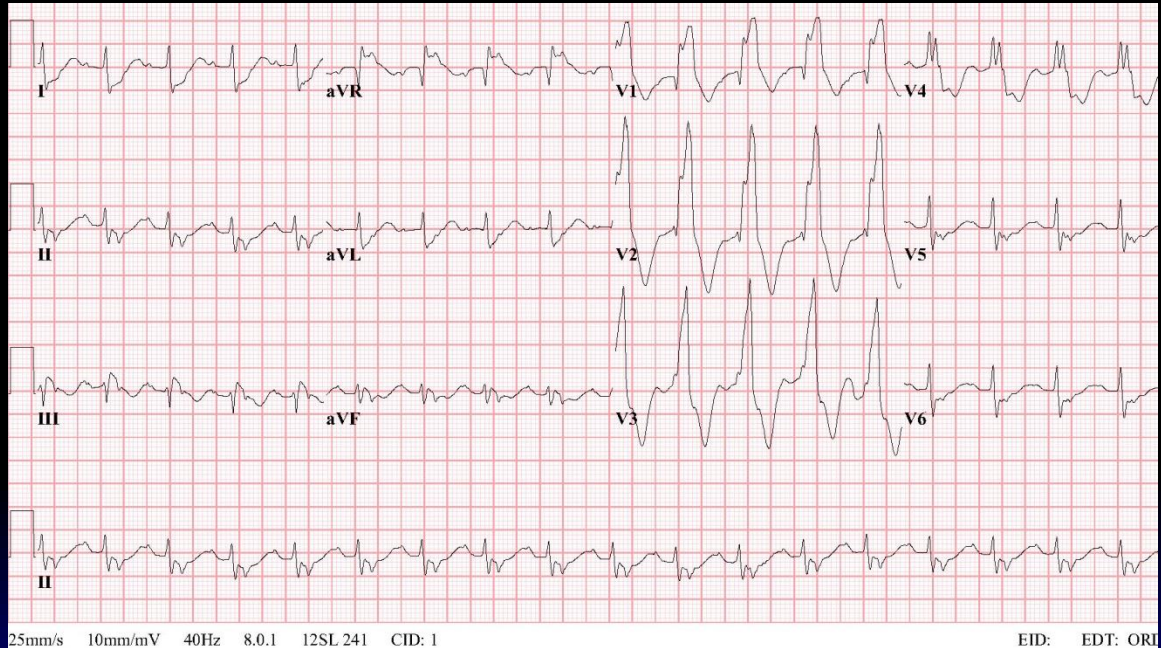


# Case2. F/48 yr - S/P VSD repair Pulmonary hypertension

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- 9 year-old: diagnosed as large VSD - VSD patch closure, but large residual shunt d/t detachment of patch : pulmonary HTN
- 35 year-old (2002.6.12) : Re-op for patch closure of VSD, partial closure of PFO
- 43 year-old (2010.6.12)
  - echoCG: mod TR 113mmHg, mild PR 68mHg, moderate distension of Rt. heart (especially RA)
  - On bosentan, sildenafil due to severe pulmonary HTN
- 47 year-old (2014.8) : O<sub>2</sub> support start
- 48 year-old (2015.4)
  - Aggravated Rt. side heart failure (dyspnea, generalized edema, ascites, atrial flutter)
  - Referred for further treatment (for Remodulin infusion)
- On admission: lower leg edema, ascites, O<sub>2</sub> 3-4L/min

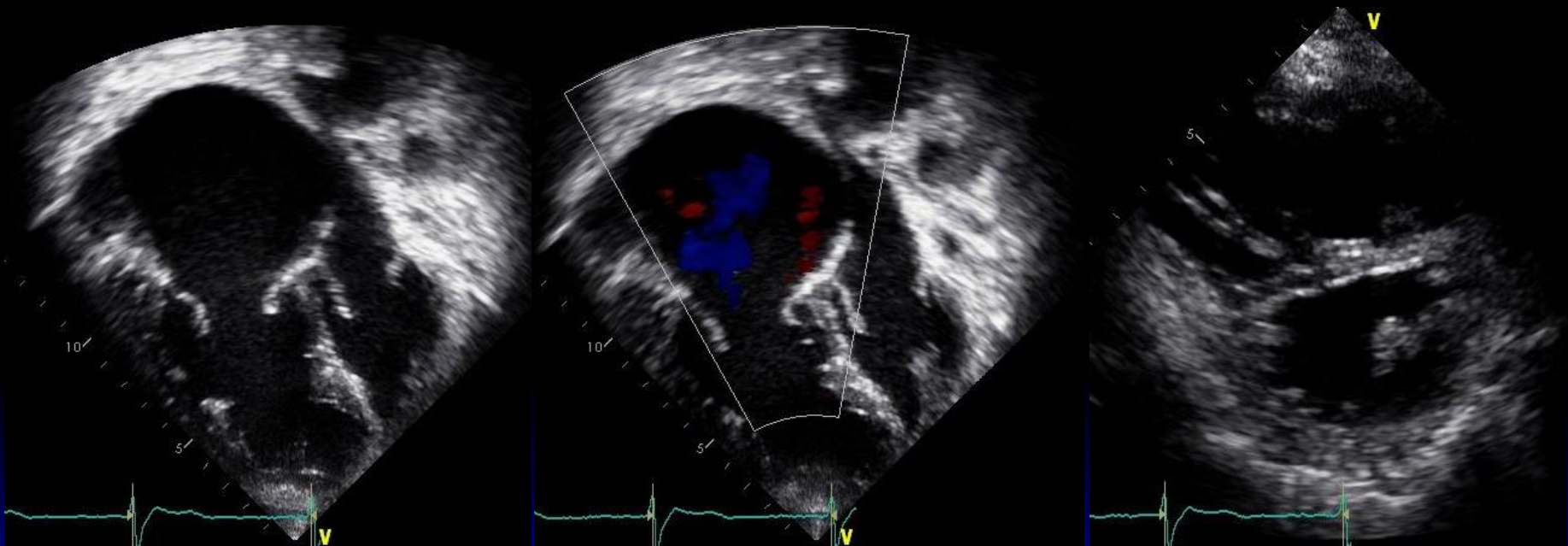
# Case2. F/48 yr - S/P VSD repair Pulmonary hypertension



Courtesy to Dr. GB Kim in SNUH

# Case2. F/48 yr - S/P VSD repair Pulmonary hypertension

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*Courtesy to Dr. GB Kim in SNUH*

# Case2. F/48 yr - S/P VSD repair Pulmonary hypertension

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- HD#2: remodulin S.C C.I start at MD 2ng/kg/min(0.008ml/hr)
- HD#8: sudden BP drop, drowsy mentality, low SpO<sub>2</sub>, ICU transfer, NO 60 ppm, FiO<sub>2</sub> 0.7, SIMV mode, SpO<sub>2</sub>: 70~90%
- HD #11: when CVP <25 : BP drop - difficulty for enough CRRT Removal, RBC Tf , Albumin, FFP Replacement. When CVP > 25 : Pulmonary edema- Desaturation continued
  - consider ECMO : may not be reversible, inappropriate
  - **discuss EOL**
- HD#16: SBP 60~70 mmHg, SpO<sub>2</sub> 60~70% even with IV inotropics - HR down
  - **discuss EOL → Decision for no cardiac compression**
- HD#17: expired

# Life prolonging intervention

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- Only definitive treatment for many ACHD
  - > Heart of Heart-Lung transplantation
- Transplantation for ACHD is Complicated
  - multiple previous operation
  - increased sensitization rates
  - complex venous anatomy
- Higher mortality after transplantation



# Palliative care

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- VAD : ventricular assist devices
  - bridge to transplantation
  - support the end stage heart failure
  - destination therapy in patients unsuitable for transplantation
  - new pump developed for Fontan circulation
- May the palliative care be the answer...?
- Any conflict...? an unwelcome idea...?

*Bowater SE et al. Curr Opin Support Palliat Care 2013; 7:8–13*

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# Discussion for End of Life

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- Death from heart failure : 26% of ACHD (51yr)
- Sudden cardiac death : 19% of deaths (39.1yr)

*Then, When is the right time to discuss EOL...?*

*--→ We can no longer wait for heart failure to develop before discussing EOL issues*

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# Discussion for End of Life

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- Early and proactive EOL discussion
  - beneficial in oncology / acquired HF
  - necessary for ACHD patients
- Less aggressive palliative care at the end
  - to manage symptom and other distress
  - even not to withdraw all care
  - usually unsuccessful resuscitation
- Earlier hospice referral

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# Effective Communication for EOL

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- Uncertainty
  - We cannot know exactly what will happen
- Identify the possibility of earlier or later death
  - several months to a year or two years
  - some live longer and others live shorter
- Resuscitation issue
  - When your heart or breathing stops...
  - When they would not prefer to be kept alive with life support

# Detail Communication for EOL

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- Actual location of EOL care  
Home > ICU >
- Actual location of death  
Home > ICU > special room in hospital

# Decision for EOL

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- What Patient thinks or believes :
  - able to change their mind after making important medical decision
  - doctors know the kind of care
  - wishes of EOL care will be respected by doctors, hospital, surrogate

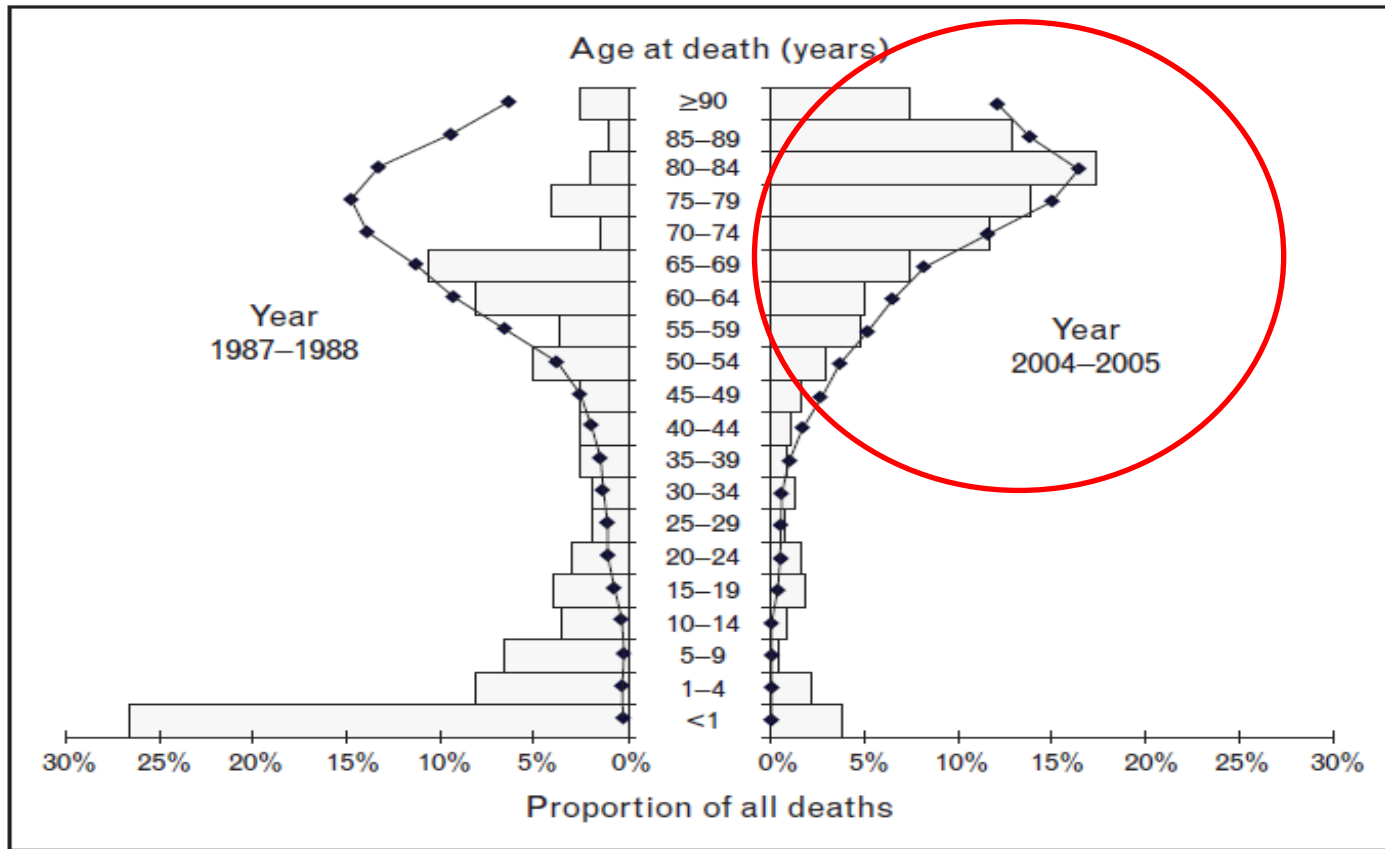
# Preferences – death and dying

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- Patient preferences regarding their own death and dying
  - honest answers from doctor
  - understanding treatment choices
  - visits from family and friends
  - having said everything they want to say
  - free from pain
  - physically comfortable, spiritually at peace
  - not being a burden to loved ones
  - knowing how to say goodbye
  - fulfilling personal goals / pleasures, having sense of own value
  - able to stay in their own home
  - dying a natural death / off life-expanding machines

# Distribution of age at death in CHD

Distribution of age at death in patients with congenital heart disease in 1987–1988 and 2004–2005





# Unique needs of Young

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- ACHD population - Young! But lifetime illness
- Reluctance to discuss EOL and palliative care
- Younger patients tend to receive :
  - more aggressive treatment plan
  - more option for the better care plan

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# Real Communication for EOL

**Table 1.** Recommendations to Facilitate Advance Care Planning and EOL Discussions.

- Normalize EOL discussions by holding them with all patients.
- Plan and practice language to conduct conversations about dying.
- Consider the cognitive and developmental ability as well as the cultural background of an individual patient when talking about EOL.
- As desired by the patient, include family members in planning discussions.
- When patients ask about prognoses and life expectancy, provide a general range of expected length of life for patients with a similar cardiac condition. Acknowledge prognostic uncertainty by explaining that life expectancy for adults with CHD can be difficult to predict.
- For patients with CHD of moderate or great complexity, consider using the term “a life-shortening medical condition.”
- Allow the topic of dying to come up early in conversations with patients and their families. Acknowledge that death occurs. Invite (but do not impose) a discussion of EOL issues early in the disease course with patients irrespective of defect complexity or life expectancy.
- Before initiating a conversation about dying, it is important to first ask the patient their preferences for information and participating in decision making.
- Consider scheduling a specific visit to talk about patient and family goals for their care, including preferences and options for EOL care.
- Encourage all adult patients to complete advance directives, including the appointment of a surrogate/proxy decision maker.
- Document discussions about preferences for EOL care and advance directives in inpatient and outpatient notes in a manner that “flags” them or makes them easily found. Make advance directives and documents naming substitute decision makers known to all members of the health care team.
- Identify the emotions experienced by patients as well as your own sadness or disappointment.
- Plan a way to acknowledge your own reactions to patients’ dying and to bring closure to the relationship.

Abbreviations: CHD, congenital heart disease; EOL, end of life.

# Real Communication for EOL

Table 2. The Ask-Tell-Ask Cycle.

Purpose	Examples
<p><b>Ask</b> Ask what patients currently understand about their CHD and what they would like to know.</p>	<ul style="list-style-type: none"> <li>• Tell me what you understand about your congenital heart disease, and what you expect?</li> <li>• What have other doctors or nurses told you about what to expect down the road?</li> <li>• What would you like to know about your health?</li> <li>• How would you like decisions to be made about your health care?</li> <li>• What have you told your family or other doctors you want when the time comes that your heart or breathing stops?</li> <li>• What worries do you have?</li> </ul>
<p><b>Tell</b> Provide information that is requested by the patients or is important to communicate to them</p>	<ul style="list-style-type: none"> <li>• You asked about how long I think you will live. Based upon how your health is doing now, we would expect that you have several months/a few years/a few decades. Of course, this kind of thing is not easy to predict and some patients live shorter and some live longer than we expect.</li> <li>• When your heart or breathing stops we can either try to revive you or allow you to die naturally.</li> <li>• I am afraid we have reached a phase in your illness in which you will near the end of life.</li> <li>• We are at a turning point in your heart disease and there are choices about which road to take.</li> <li>• I try to talk with all patients about what they would like to happen when they become very ill or near death. These might not be easy things to discuss, but it is very important that we know your preferences for EOL care and who you want to make your decisions about your health if you become unable to do so. Talking about this now will help your family if ever they need to make decisions on your behalf.</li> </ul>
<p><b>Ask</b> Confirm an understanding of what was said and provide an opportunity for patients to ask follow-up questions</p>	<ul style="list-style-type: none"> <li>• It is important that I explain things clearly to you. Please tell me what you understood.</li> <li>• What questions do you have?</li> </ul>

# How to Care End of Life

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- EOL plan : routine part for ACHD clinic
- Re-evaluate at every significant change
- Treatment for prior to death
- Well comfort – allowing natural death
- CPR – trying to revive, anticipated outcome
- Maintain contact – coping with emotions

# Goal of End of Life Care

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The ultimate goal is to provide a quality of dying experience with minimal distress for patients, their family members, and the care team.

# What is the better thought for lifetime?

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# Thank You So Much!

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*Quality of Life*  
*Quality of Dying*