# Quality of Life in Adults with CHD

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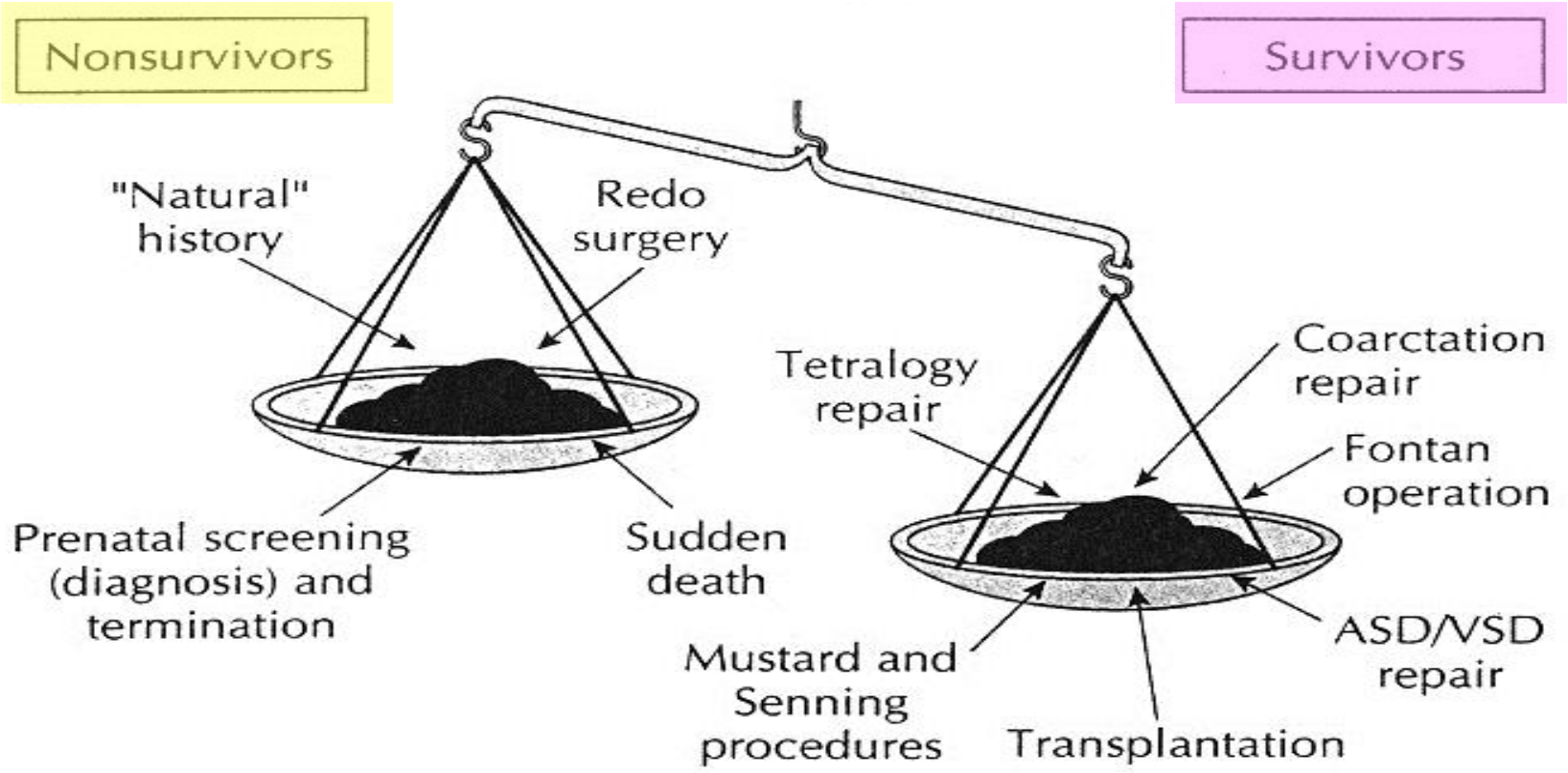
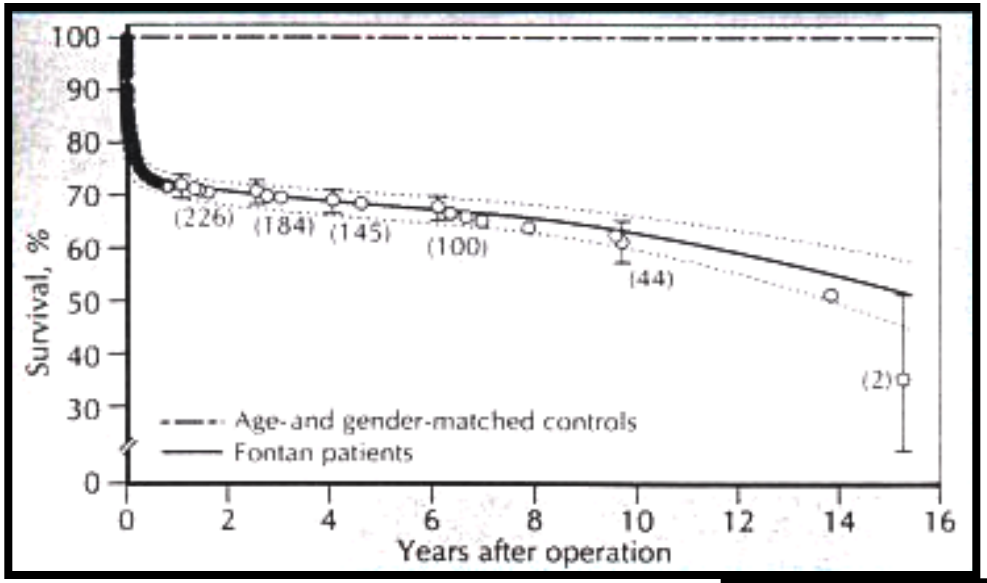
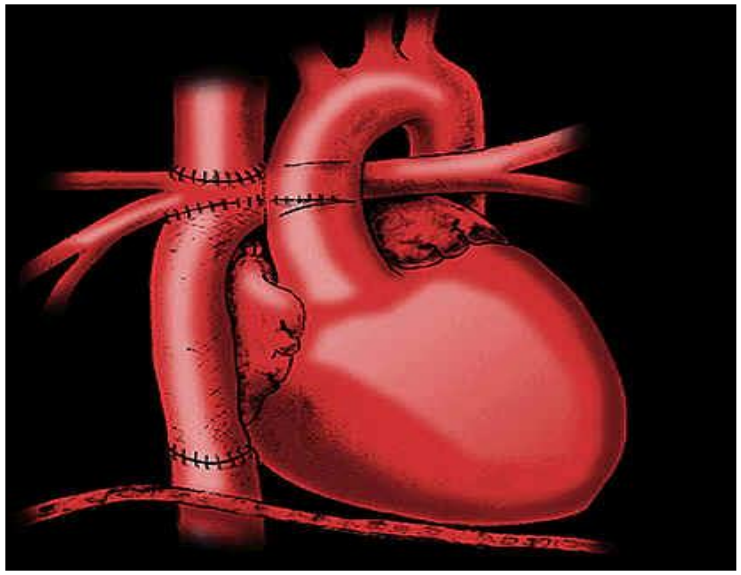
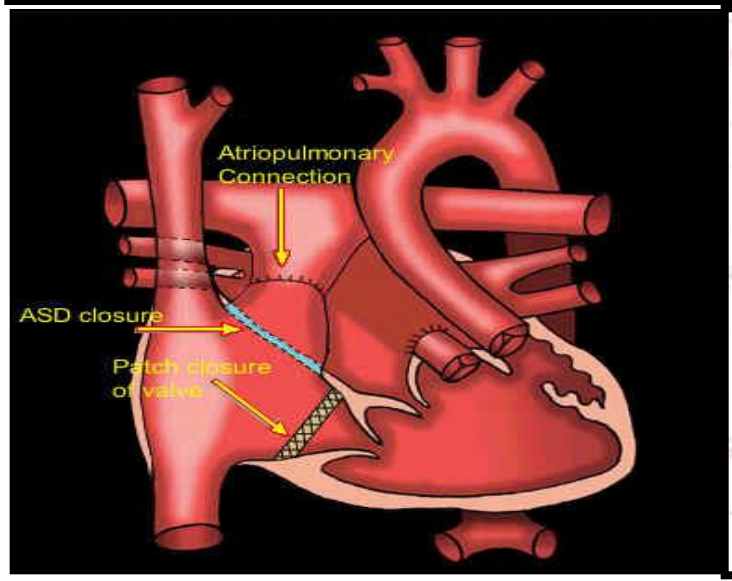
# Factors affecting balance of population

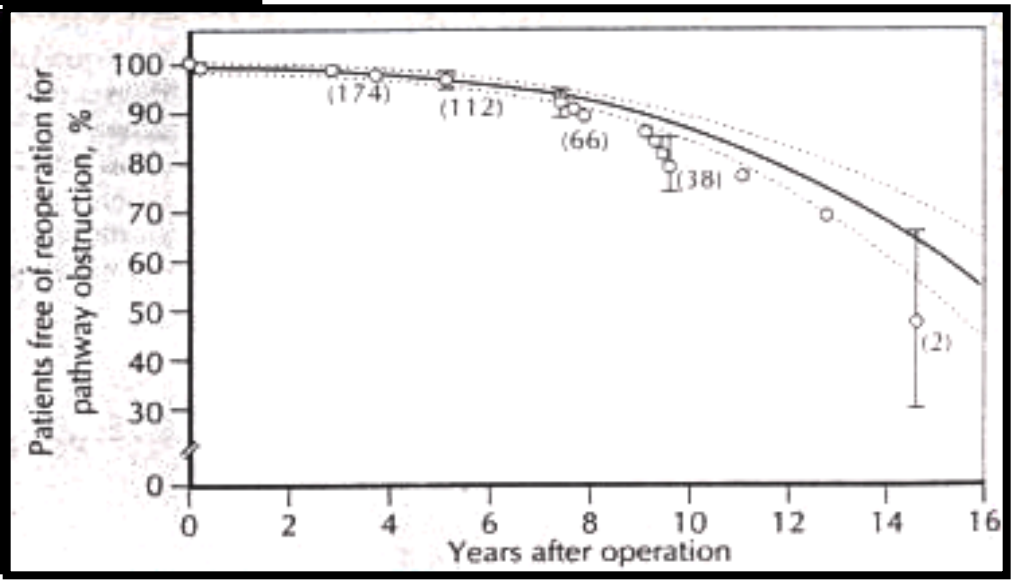


# Fontan op in ‘Functional Single Ventricle’

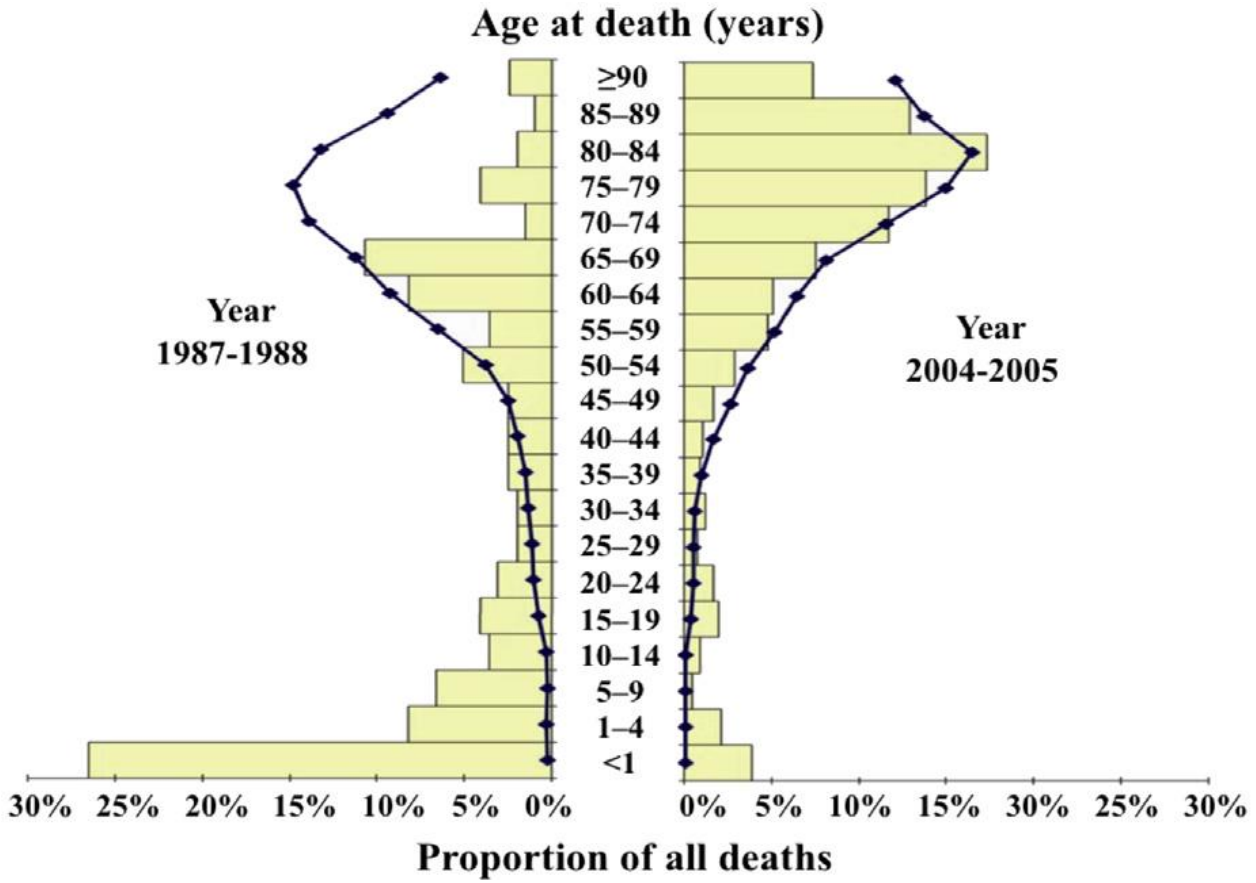




# Prevalence of CHD




Distribution of age at death in patients with congenital heart disease in 1987-1988 and 2004-2005. Histogram bars depict the proportion of all deaths (x-axis) according to age at death (y-axis) in our cohort of patients with congenital heart disease in the first (1987-1988; **left graph**) and final (2004-2005; **right graph**) years of observation. **Bold black curves with diamonds** represent the corresponding age at death distribution in the general Quebec population during the same periods of observation. Reproduced from Khairy et al.[7]

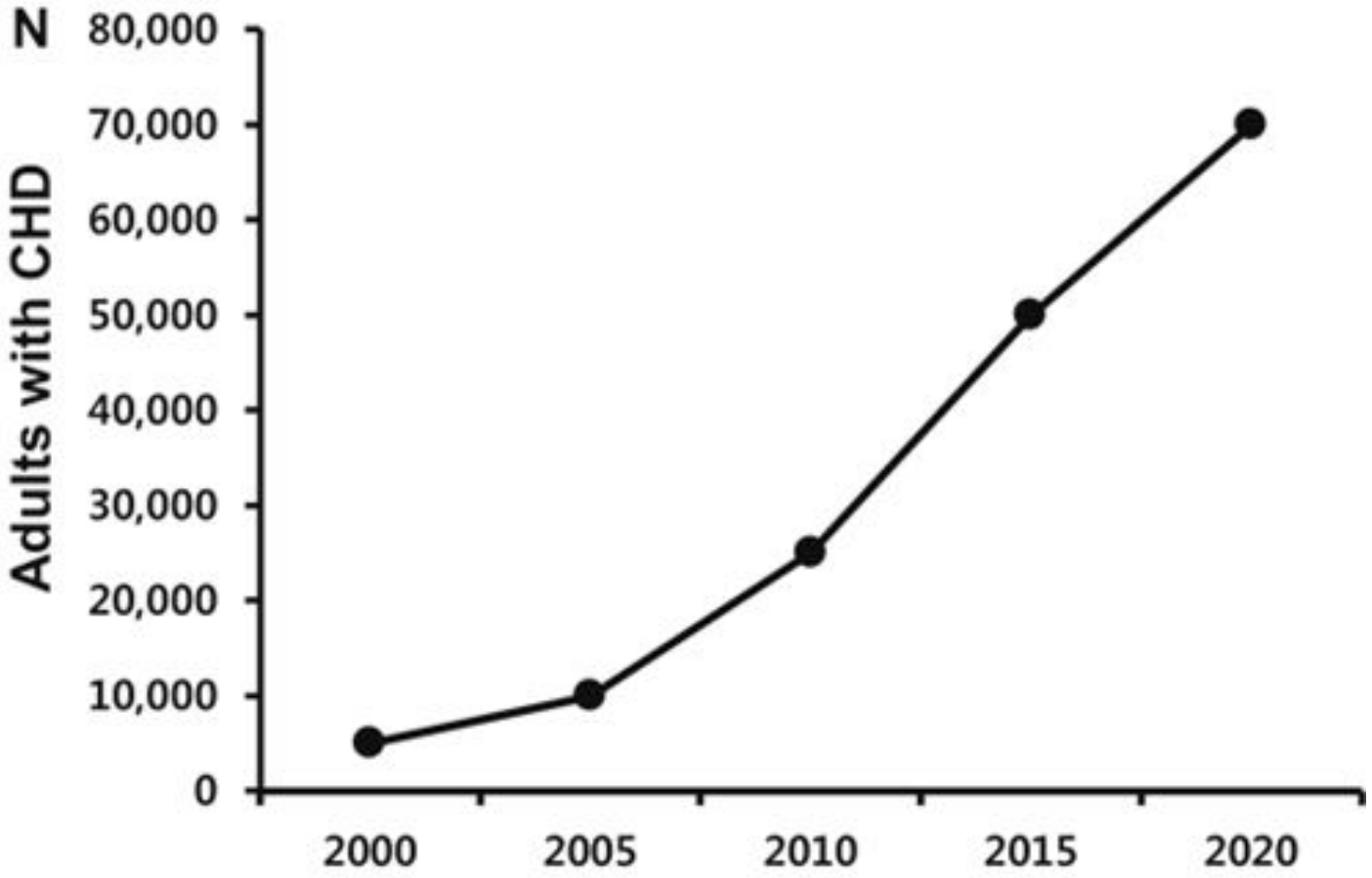
*(Avila P, et al. CJC, 2014;30:S410~S419)*

# Congenital Heart Disease Population

- At birth : 6 ~ 10 per 1000 live birth
- Natural Survivor at puberty (Presurgical era)
  : 5 ~ 15 %
- Natural & Unnatural GUCH Survivor
  (Surgical era) : over 85 %

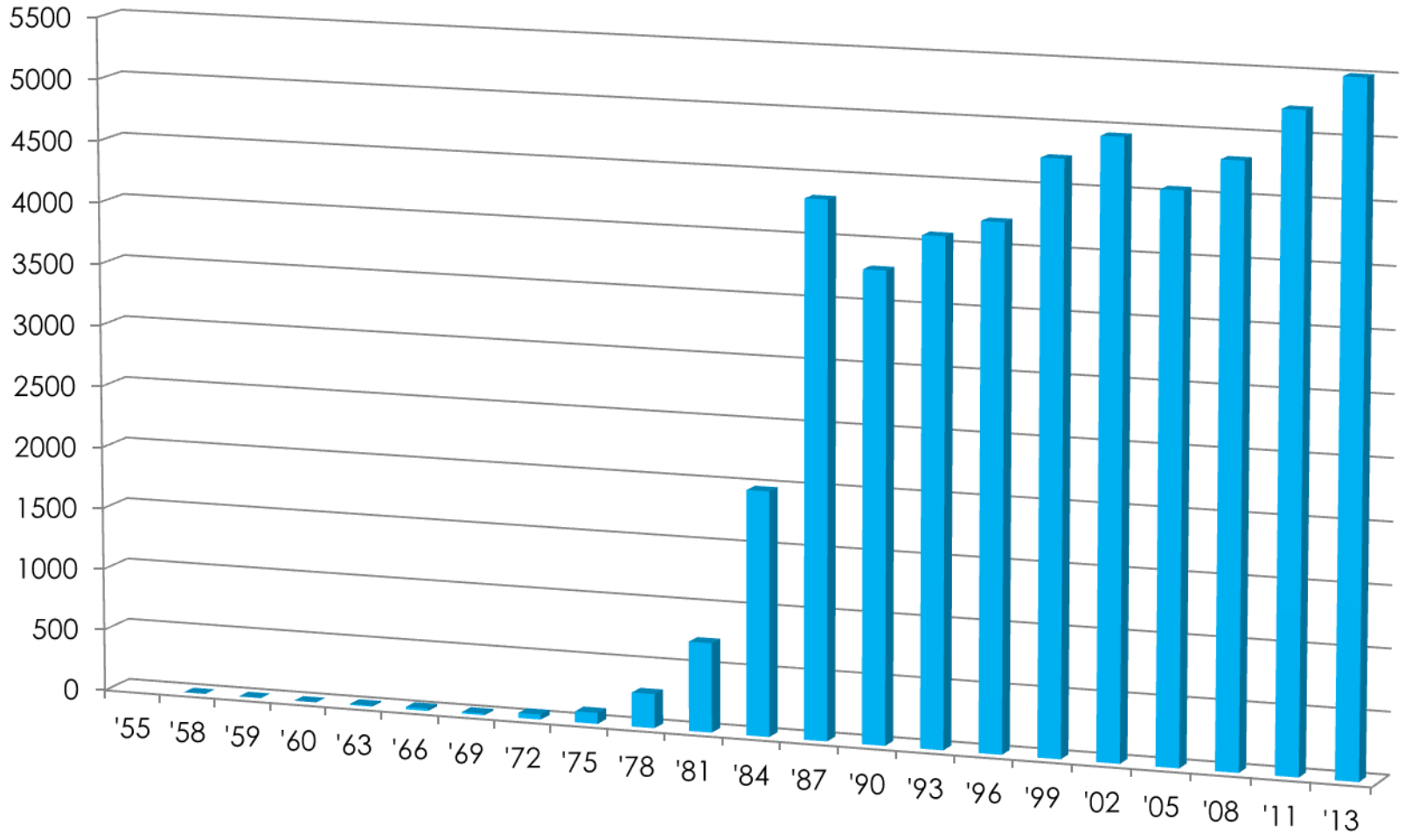
USA : may be over million in the early 2000s

# Congenital Heart Disease Population



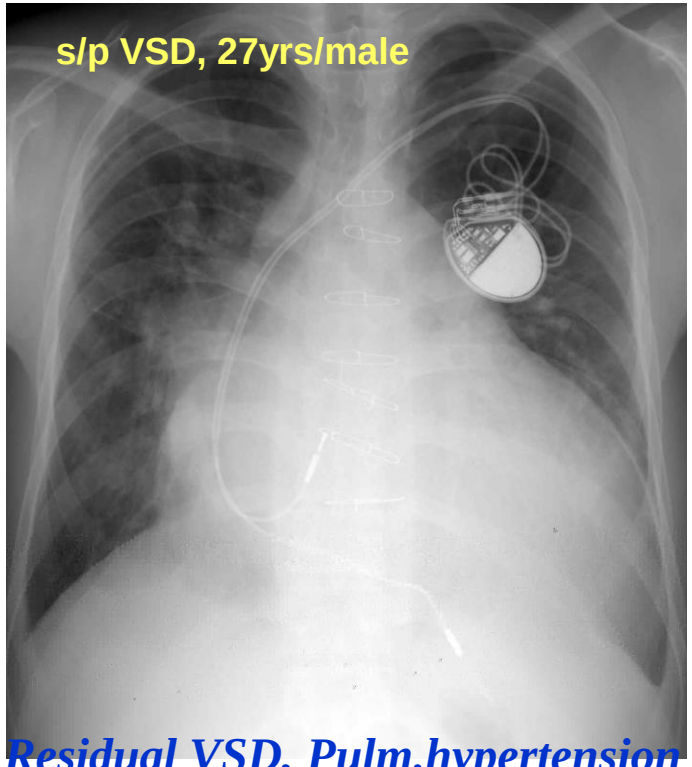

Kim GB Korean J Pediatr 2014;57(6):257-263

# Cardiac surgery for CHD in Korea



# Four Major Cardiologic Concerns in GUCH clinic

- Arrhythmia
- Infective endocarditis
- Myocardial failure
- Pulmonary vascular disease




***Residual VSD, Pulm.hypertension***
***Myocardial failure***

# Additional Issues

Adults with CHD face special challenges:

- Co-morbidities
- Neurocongintive deficit
- Inter-personal relationships
  - Negative body image
  - Social isolation
  - Parental overprotection
- Assuming responsibility for their own health care
- Pregnancy and contraception
- Vocational counseling
- Un-insurability

**Mark Gentle** with Sarah Richards
April 11 at 11:56am · Edited ·

#CHDAWARENESS We were a little apprehensive to post this but I feel it's important. Carter took his bandages off this morning. He cried for 45 minutes when he saw his scars. He said that he's afraid people will think he is ugly. This was hard to hear as a parent. We told him that his scars are beautiful and make him look like a superhero. How many likes can Carter get for his bravery?I Feel free to share. #CartersSong #HeartHeroes #CHD

# Holistic care of adults with CHD

- The emotional health of adolescents and adults with CHD should be a priority in the overall care of this population *[The 32nd Bethesda Conference, JACC 2001;37:1161~98].*

- The specialist service for grown-ups with CHD must provide support for the many psychosocial problems in this population *[ESC committee, Eur Heart J 2003;24(11):1035~84].*

# Health-related Quality of life (HRQOL)

"Health is not only the absence of disease and infirmity, but the presence of physical, mental and social well-being"

*[World Health Organization, 1948]*

"The degree of overall life satisfaction that is positively or negatively influenced by individuals' perception of certain aspects of life importance to them, including matters both related and unrelated to health"

*[Moons et al, 2005]*

# Evaluation of HRQOL

- Multiple dimensions
  - Physical functioning
  - Emotional functioning
  - Social functioning
  - School or occupational functioning
  - Disease-related and treatment-related aspects
- Determinants of an individual's QOL; family, Job/education, friends, health and leisure time
- QOL changes overtime

# What We Already Knew

- From EMBASE, PubMed, The Cochrane Library, PsycINFO DB
- keyword - QOL, adults CHD (2011~2015)
- Total 32 papers - systemic literature review papers - children papers = 22 papers
- Sample size: < 100 patients in 19 studies
  330 – 1109 in 3 studies

# What We Already Knew

- **Six deferent tools were used to assess QOL**
  - EuroQOL
  - SF-36
  - WHOQOL-BREF
  - a combination of a liner analog scale for QOL
  - Satisfaction with life scale
  - A combination of the satisfaction with life scale and the TAAQOL-CHD
- **Disease group**
  - Patients who were operated for Simple L to R shunt
  - Patients with TGA
  - Patients Dx with TOF
  - Patients with single ventricle physiology
  - Patients with a variety of CHD lesions

# What We Already Knew

- **QOL of patients compare to control**
  - 14 out of the 22 studies compared QOL in patients with CHD to a control
  - 8 studies found that QOL is worse in patients with CHD as compared to a control
  - 3 studies concluded that QOL is similar
  - 3 studies even found a better QOL in patients with CHD compared to a control

## Studies examining health-related QOL among adults with CHD

| Study authors | Country of study | Sample (n) | Age range (y) | QOL assessment | Comparison group | Results |
|---|---|---|---|---|---|---|
| Fekkes et al[36] | Netherlands | 148 | 15-30 | SF-36 Health Survey; QOL-CHD; TAAQOL | National norms | Adults with CHD and social impediments had health status and health-related QOL similar to general population; those without social impediments had equal or better health-related QOL |
| Ternestedt et al[45] | Sweden | 26 | ≥25 | Interview | Tetralogy of Fallot vs ASD | Patients with tetralogy of Fallot had better QOL than patients with atrial septal defect |
| Saliba et al[39] | France | 67 | 17-49 | Duke health profile | Reference norms | Although QOL scores among adults with univentricular heart were generally comparable to the general population, the following groups had impairment in one or more QOL domains: cyanotic patients, those with lower educational level, orthopedic problems, psychosocial problems, those aged >23 years, and those with mitral atresia |
| Lane et al[42] | United Kingdom | 276 | 16-85 | SF-36 | Reference norms | Among subgroups of adults with CHD, patients with inoperable conditions, cyanotic conditions, and surgically cured patients had worse QOL than surgically palliated or surgically corrected patients; all patient subgroups had worse physical functioning and general health perception than the general population |
| Kamphuis et al[41] | Netherlands | 78 | 18-32 | SF-36; TAAQOL | Reference norms | Adults with CHD had poorer health-related QOL than general population; there were poor correlations between QOL and physical status |
| Rietveld et al[44] | Netherlands | 82 | 17-77 | SF-36 | Compared within sample | Adults with many negative self-statements reported worse psychosocial adjustment and QOL than adults with few or moderate negative self-statements (regardless of cardiac disease severity); QOL was predicted by trait anxiety, whether the patient had a job, and negative self-statements |
| Gersony et al[40] | United States | ≤1681 | ? | Single-item question: "How would you describe your present health?" | National normative data | Adults with CHD generally reported perceived health status similar to general population |
| Simko and McGinnis[43] | United States | 124 | | SIP | Matched healthy control subjects | Adults with CHD reported worse QOL across all SIP scores |

*QOL-CHD*, QOL Congenital Disease Questionnaire; *TAAQOL*, Netherlands Organization for Applied Scientific Research Academic Medical Center Adult QOL Questionnaire; *SIP*, Sickness Impact Profile; *SF-36*, Medical Outcomes Trust 36-Item Short-Form Health Survey; *ASD*, atrial septal defect.

# What We Already Knew

## Determinants of QOL



**Table 1** Overview of potential determinants of quality of life in patients with congenital heart disease

| | Overgaard et al. (2011) [10] | Silva et al. (2011) [11] | Teixeira et al. (2011) [12] | Vigl et al. (2011) [13] | Cotts et al. (2012) [14] | Müller et al. (2012) [15] | Opic et al. (2012) [16] | Schoormans et al. (2012) [17•] |
|---|---|---|---|---|---|---|---|---|
| Demographic characteristics | | | | | | | | |
| Older age | | ↗ | = | ↘ | = | | | |
| Female sex | | = | = | ↘ | | | | |
| Higher academic performance/ education | | | ↗ | ↗ | | | | |
| Higher employment rate | | | | ↗ | | | | |
| More daily activities | | | | | | ↗ | | |
| Clinical characteristics | | | | | | | | |
| Cardiac surgery | | ↘ | ↘ | | | | | |
| Medication | | = | ↘ | | | | | |
| Better functional class | ↗ | | | | | | | |
| More severe heart defects | | = | = | ↘ | | | | |
| More severe residua | | ↘ | = | | | | | |
| CHD subtype | | | = | | | | | |
| Cyanosis | | = | | | | | | |
| Implantable cardioverter defibrillator | | | | | | | ↘ | |
| Physical limitations | | | ↘ | | | | | |
| Better exercise capacity | | | | | | ↗ | | |
| Psychosocial characteristics | | | | | | | | |
| Personal resources | | = | | | | | | |
| Family environment | | = | | | | | | |
| Better social support | | ↗ | ↗ | | | | | |
| Type D (distressed) personality | | | | | | | | ↘ |

↗ Determinant of better quality of life; ↘ Determinant of worse quality of life; = Unrelated to quality of life

# What We Already Knew

- **Determinants of QOL**

| Variables | Religion | | |
|---|---|---|---|
| | Yes | No | *t* |
| Psychological resilience | 93.45 ± 15.75 | 86.02 ± 14.30 | 2.195* |
| Physical health QoL | 60.09 ± 12.74 | 52.64 ± 11.58 | 2.719** |
| Psychological QoL | 63.48 ± 11.90 | 58.26 ± 13.66 | 1.777 |
| Social relationships QoL | 65.27 ± 15.91 | 59.17 ± 15.40 | 1.721 |
| Environment QoL | 67.73 ± 12.38 | 60.79 ± 14.09 | 2.279* |

* $p < 0.05$.
** $p < 0.01$.

*Bang et al., Int J Cardiol 170 (2013) 49–53*

# Emotional Adjustment

- Common research question :

*" Are adults with CHD more likely to have psychological difficulties than peer or normative group?"*

## Studies examining emotional functioning among adults with CHD

| Study authors | Country of study | Sample (n) | Age range (y) | Psychological assessment | Comparison group | Results |
|---|---|---|---|---|---|---|
| Utens et al[12] | Netherlands | 288 | 18-35 | 3 scales from DPQ: hostility, self-esteem, neuroticism | Reference norms (healthy men and women) | Patients with CHD had more desirable levels of hostility, self-esteem, and neuroticism; no difference in scores between cardiac diagnostic groups |
| Utens et al[14] | Netherlands | 166 | 19-25 | Young adult self-report | Dutch normative reference group | Patients with CHD had worse scores on 2 of 9 subscales; no relationship between cardiac diagnosis and psychological problems |
| Popelova et al[16] | Czech Republic | 32 | 19-64 | Zung's questionnaire | Instrument cutoff scores indicative of depression | 34% Received scores indicative of depression; depression associated with older age, worse functional status, and less education |
| Cox et al[15] | United Kingdom | 87 (with CHD) | 17-73 | GHQ30; HADS | Orthopedic outpatients | Adults with CHD had lower levels of psychopathology, as assessed by the GHQ30, than orthopedic outpatients |
| Van Rijen et al[13] | Netherlands | 362 | 20-46 | 3 DPQ scales: hostility, self-esteem, neuroticism | Reference norms (healthy men and women) | Patients with CHD had more desirable levels of hostility, self-esteem, and neuroticism |
| Brandhagen et al[17] | United States | 168 | 24-42 | SCL-90–R; dependency subscale of MMPI | Reference norms | Adults with CHD had higher levels of psychological distress on multiple scales of SCL-90–R; severity of cardiac defect was generally unrelated to psychological distress |
| Horner et al[18] | United States | 29 | 26-56 | Clinical interview | Psychiatric diagnostic criteria | 79% Met criteria for at least one psychiatric diagnoses |
| Bromberg et al[19] | United States | 22 | 19-60 | BSI | Psychiatric diagnostic criteria | 35% Of patients assumed to be "well adjusted" met criteria for psychiatric diagnosis; positive association between medical severity and depression |

*DPQ*, Dutch Personality Questionnaire; *GHQ30*, General Health Questionnaire 30; *HADS*, Hospital Anxiety and Depression Scale; *SCL-90–R*, Symptom Checklist-90–Revised; *BSI*, Brief Symptom Inventory; *MMPI*, Minnesota Multiphasic Personality Inventory.

# Risk Factors Related to Psychological Difficulties

- Younger patients more than older patients *[van Rijen EH, 2004; van Rijen EH, 2005]*
- Woman more than man *[van Rijen EH, 2004; van Rijen EH, 2005]*
- low maximal exercise capacity, physician-imposed restrictions and negative personal experiences with scaring *[Popelova et al, 2001]*
- Not related to Cardiac disease severity *[Brandhagen, 1991; Utens EM, 1994; Utents, 1998; Van Rijen, 2004]*

# Critics of Emotional Functioning Research

- Interview data suggested more psychological disturbance than self-report data
- Generalizations regarding international differences in the emotional functioning may be premature
- The role of disease severity is unclear

# Risk Factors for Poor QOL

- Greater social impediments
- Negative self-statement
- Anxiety
- Lack of employment
- Cyanosis
- low functional status
- No consistent relationship between medical severity or complexity of CHD and QOL

*[DeMaso, 1990; Krol, 2001; Uzark, 2003; Macran, 2006; Bans JS, 2013]*

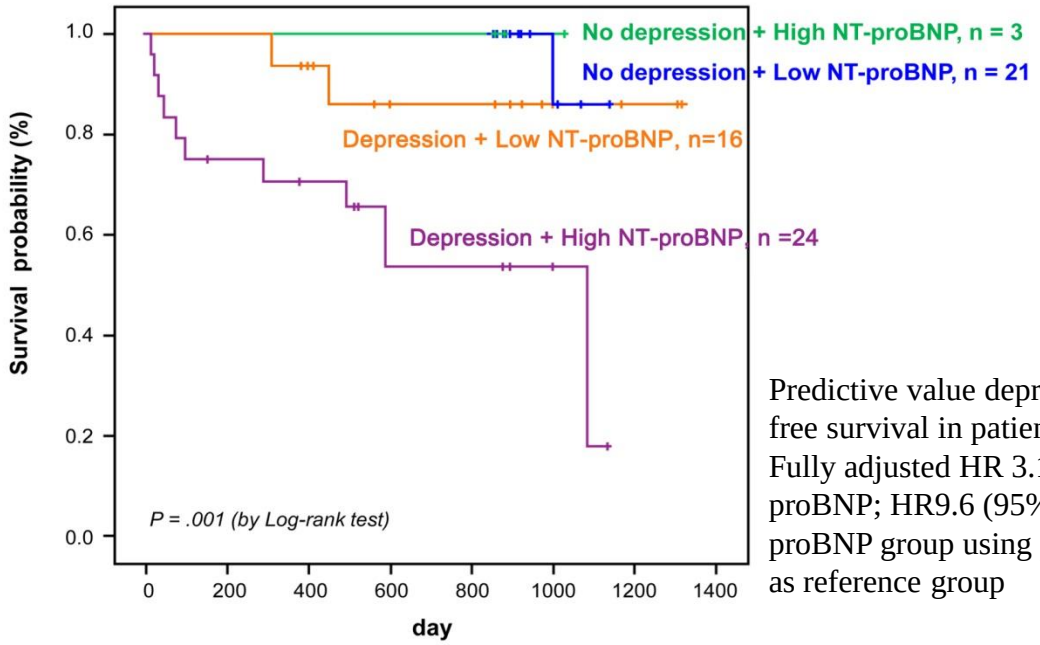
# Limitation of HRQOL research

- Sample bias
  - Those who receive follow-up care
  - Those who receive their care at a facility with a program of research
  - Not excluded based on cognitive functioning
- No exist disease-specific QOL measurement
- No longitudinal study(cross - sectional study)

# Avenues for Future Research

- **A combination with biomarker;** is needed to predict outcome such as

  - Clinical and prognostic value of depressive symptoms and NT-proBNP in patients with Eisenmenger Syndrome. *(Moon JR, et al. Psychosomatic Medicine, 2015; in press)*




Predictive value depressive symptoms and NT-proBNP for cardiac event-free survival in patients with Eisenmenger syndrome.
Fully adjusted HR 3.19 (95% CI 1.29 to 9.21, n=16) for depressed/lowNT-proBNP; HR9.6 (95% CI 2.09 to 29.54, n=24) for depressed/high NT-proBNP group using patients with no depression and low NT-proBNP levels as reference group

# Avenues for Future Research

- **A combination with biomarker;** is needed to predict outcome such as

  - Association between NT-proBNP and QOL in adults CHD *( Younge JO, et al. Cardiology in the Young, 2015;25:288-294)*

Table 3. Association between NT-proBNP and SF-36 domains in ConHD subgroups.

| | Simple | | | | Complex | | | |
|---|---|---|---|---|---|---|---|---|
| | Univariable analysis | | Multivariable** analysis | | Univariable analysis | | Multivariable** analysis | |
| | β | p | β | p | β | p | β | p |
| SF-36 subdomains | | | | | | | | |
| Physical functioning | 0.038 | 0.678 | 0.096 | 0.307 | −0.072 | 0.064 | −0.076 | 0.046* |
| Role physical functioning | 0.127 | 0.380 | 0.179 | 0.226 | 0.017 | 0.826 | 0.034 | 0.660 |
| Bodily pain | 0.038 | 0.762 | 0.095 | 0.452 | 0.064 | 0.133 | 0.060 | 0.158 |
| Social functioning | 0.016 | 0.875 | 0.069 | 0.497 | −0.006 | 0.864 | −0.004 | 0.901 |
| Mental health | −0.02 | 0.984 | 0.046 | 0.549 | 0.034 | 0.381 | 0.032 | 0.415 |
| Role emotional functioning | 0.090 | 0.511 | 0.159 | 0.261 | −0.030 | 0.672 | −0.017 | 0.813 |
| Vitality | 0.20 | 0.867 | 0.098 | 0.381 | 0.036 | 0.435 | 0.039 | 0.412 |
| General health | 0.033 | 0.791 | 0.073 | 0.569 | −0.42 | 0.335 | −0.041 | 0.358 |

ConHD = congenital heart disease; NT-proBNP = N-terminal pro-brain natriuretic peptide; SF-36 = 36-item Short-Form Health Survey.
*Statistical significance with $p \leq 0.05$
**Adjusted for age and gender

# Avenues for Future Research

- **Longitudinal research:** is needed to
  - Examine changed in QOL over time
  - Assess the direction of effects between QOL and possible determinants and concomitants

# Avenues for Future Research

- **Multicenter, international studies** should be initiated using **one common, well-defined method to measure QOL** in order to
  - Be able to make valid comparisons
  - Gain insight in whether the reported discordant results are due to mythological differences or whether they represent real differences, for example between countries or regions
  - Examine which factors impact on differences in QOL
  - Ensure the generalizability of the findings

# Avenues for Future Research

- **Intervention studies** should be conducted for the purpose of improving modifiable determinants of QOL

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# Assessment of Patterns of Patient-Reported Outcomes in Adults with Congenital Heart disease — International Study (APPROACH-IS): Rationale, design, and methods ☆,☆☆

Silke Apers [a], Adrienne H. Kovacs [b], Koen Luyckx [c], Luis Alday [d], Malin Berghammer [e], Werner Budts [f], Edward Callus [g], Maryanne Caruana [h], Shanthi Chidambarathanu [i], Stephen C. Cook [j], Mikael Dellborg [e,k], Junko Enomoto [l], Katrine Eriksen [m], Susan M. Fernandes [n], Jamie L. Jackson [o], Bengt Johansson [p], Paul Khairy [q], Shelby Kutty [r], Samuel Menahem [s], Gwen Rempel [t], Maayke A. Sluman [u], Alexandra Soufi [v], Corina Thomet [w], Gruschen Veldtman [x], Jou-Kou Wang [y], Kamila White [z], Philip Moons [a,e,aa,*], on behalf of the APPROACH-IS consortium and the International Society for Adult Congenital Heart Disease (ISACHD)

[a] KU Leuven — University of Leuven, Department of Public Health and Primary Care, B-3000 Leuven, Belgium
[b] Toronto Congenital Cardiac Centre for Adults, Peter Munk Cardiac Centre, University Health Network, Toron
[c] KU Leuven — University of Leuven, School Psychology and Child and Adolescent Development, B-3000 Leuv
[d] Division of Cardiology, Hospital de Niños, Córdoba, Argentina
[e] The Sahlgrenska Academy at University of Gothenburg, Institute of Medicine, Gothenburg, Sweden

Fig. 1. Geographic distribution of the APPROACH-IS participating centers.

**Table 1**
Variables measured in APPROACH-IS.

| | Source | Measurements | Required translations | Validity | Reliability | Responsiveness | Interpretation |
|---|---|---|---|---|---|---|---|
| *Socio-demographic variables* | | | | | | | |
| - Age<br>- Sex<br>- Marital status<br>- Presence of children<br>- Student status<br>- Educational level<br>- Employment status<br>- Patient-reported New York Heart Association assessment<br>- Religion | Patient self-report | Survey developed by research team | Chinese, Dutch, French, German, Hindi, Italian, Japanese, Norwegian, Spanish, Swedish, and Tamil | NA | NA | NA | NA |
| *Medical history* | | | | | | | |
| - Congenital heart disease diagnosis<br>- Complexity of heart defect<br>- Cardiac surgeries/interventions<br>- History of congestive heart failure/ arrhythmia/other medical condition<br>- Cardiac device implantation<br>- Cognitive impairment<br>- Frequency of follow-up<br>- Cardiac admission (total number since age 18)<br>- Chart-documented mood or anxiety disorder or another psychiatric diagnosis | Chart review | Survey developed by research team | Chinese, Dutch, French, German, Hindi, Italian, Japanese, Norwegian, Spanish, Swedish, and Tamil | NA | NA | NA | NA |
| *Primary outcomes* | | | | | | | |
| Perceived health status | Patient self-report | 12-item Short-form Health Survey version 2 (SF-12) | NA | Confirmed in medical populations [40] | Confirmed in medical populations [40] | Confirmed in medical populations [40] | Scores from 0 to 100 on eight health domains; higher scores = better perceived health |
| | | EuroQol-5D 3 level version (EQ-5D) | NA | Construct validity confirmed in cardiovascular patient populations [55] | Confirmed in cardiovascular patient populations [55] | Confirmed in cardiovascular patient populations [55] | Scores from 1 (no problems) to 3 (extreme problems) on five dimensions; score from 0 (worst imaginable health |

*(continued on next page)*

**Table 1** *(continued)*

| | Source | Measurements | Required translations | Validity | Reliability | Responsiveness | Interpretation |
|---|---|---|---|---|---|---|---|
| | | | | | | | state) to 100 (best imaginable health state) |
| Psychological functioning | Patient self-report | Hospital Anxiety and Depression Scale (HADS) | NA | Confirmed in medical populations [56,57] | Confirmed in medical populations [56,57] | NR | Subscale scores range from 0 to 21; higher scores = more symptoms |
| Health behaviors | Patient self-report | Health-Behavior Scale-Congenital Heart Disease (HBS-CHD) | Chinese, French, German, Hindi, Italian, Japanese, Norwegian, Spanish, Swedish, and Tamil | Content validity and validity based on relationship with other variables confirmed in adolescents with congenital heart disease [44] | Stability not yet confirmed [44] | Confirmed in adolescents with congenital heart disease [44] | Substance use risk score (0–100); dental hygiene risk score (0–100); physical exercise score (0–∞); total health risk score (0–100) |
| Quality of life | Patient self-report | Linear Analog Scale (LAS) | Chinese, French, German, Hindi, Italian, Japanese, Norwegian, Spanish, Swedish, and Tamil | Test content and validity based on relationship with other variables confirmed in adults with congenital heart disease [11] | Test–retest reliability confirmed in adults with congenital heart disease [11] | Confirmed in adults with congenital heart disease [11] | Scores from 0 (worst imaginable quality of life) to 100 (best imaginable quality of life) |
| | | Satisfaction with Life Scale (SWLS) | Tamil | Test content and validity based on relationship with other variables confirmed in adults with congenital heart disease [11] | Internal consistency and test–retest reliability confirmed in adults with congenital heart disease [11] | Confirmed in adults with congenital heart disease [11] | Total score from 5 (extremely dissatisfied) to 35 (extremely satisfied) |
| *Secondary outcomes* | | | | | | | |
| Sense of coherence | Patient self-report | 13 item Orientation to Life Questionnaire (SOC-13) | Tamil | Structural validity confirmed in adolescents with congenital heart disease [58]; Face, consensual, construct, criterion, and predictive validity confirmed in different populations [59] | Internal consistency and test–retest reliability confirmed in different populations [59] | Confirmed in different populations [59] | Total score from 13 to 91; higher values = stronger sense of coherence |
| Illness perceptions | Patient self-report | Brief Illness Perception Questionnaire (Brief IPQ) | Tamil | Concurrent validity confirmed in adults with renal disease, diabetes, and asthma; predictive validity confirmed in adults with myocardial infarction; discriminant validity confirmed in adults with diabetes, asthma, colds, myocardial infarction, and chest pain [53] | Test–retest reliability confirmed in adults with renal disease [53] | NR | Scores from 0 to 10 on eight dimensions; higher scores = more threatening view of the illness |

NA: not applicable; NR: not reported.


Thank you for attention.