Predictors of Long-Term Quality of Life in Children and Adolescents with Congenital Heart Disease

Kathy Mussatto, PhD, RN
Nurse Scientist, Co-Director of Research, Herma Heart Center
Children’s Hospital of Wisconsin

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I have no relevant financial relationships to disclose.

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Objectives
• Review conceptual model of quality of life (QOL)
• Describe factors that do and do not predict QOL
• Note key lessons learned from QOL research
• Suggest strategies to optimize QOL

Key Drivers of Health-related Quality of Life in Pediatric Heart Disease
Definition of Quality of Life (QOL)

- Physical Health and Functioning
- Social Functioning
- Psychological Functioning

QOL describes a child’s ability to function in situational contexts (family, school, peer) and derive personal satisfaction from doing so.

Psychosocial Factors Mediate the Relationship between Heart Disease Complexity and Lower QOL

Complexity of Disease and Treatment → Psychosocial Morbidity Indirect → QOL

Psychosocial Impact Subscale Score

PCQLI

Lesson Learned

Who should define QOL?

The person living with the outcome.

How Are YOU Doing?

Calculations

Expectations + Reality

Ideal Self + Actual Self

“Quality of life is enhanced when the distance between the individual’s attained and desired goals is less.”

(Bergner, 1989)
**Literature Search**

**Keywords:**
- Congenital heart defects
- Quality of life
- Neurodevelopment
- Psychological adaptation
- Child development

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**MedLine Results - All Languages**

# of Citations on QOL and Adaptation in CHD

<table>
<thead>
<tr>
<th>Year Range</th>
<th>Citations</th>
</tr>
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<tbody>
<tr>
<td>1960-1969</td>
<td>14</td>
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<tr>
<td>1970-1979</td>
<td>40</td>
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<tr>
<td>1980-1989</td>
<td>29</td>
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<tr>
<td>1990-1999</td>
<td>170</td>
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<tr>
<td>2000-2014</td>
<td>386</td>
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</table>

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**Quantification of HRQOL with a Cardiac-Specific QOL Measure**

Validation of the Pediatric Cardiac Quality of Life Inventory

- Marino et al, Pediatrics 2010

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**PCQU Validation Study Sites:**

United States and United Kingdom

- 16 US Sites
- +3 UK Sites

Total US and UK Enrollment

3,174 Patient-Parent Pairs = 6,348 Respondents
Pediatric Cardiac Quality of Life Inventory Research Consortium [19 Centers US and UK]

- QOL in CHD survivors is lower than heart-healthy children.
  - Mellion et al, J Pediatrics, 2014
  - Huyserbo et al, Ped Health Care, 2014

- QOL in CHD survivors worsens with increasing disease complexity.
  - Huysebo et al, Quality of Life Research, 2008
  - Marino et al, Pediatrics, 2010

- Increasing medical care utilization (number of surgeries, cardiac catheterizations, hospitalizations, and physician visits annually) is associated with lower QOL in CHD survivors.
  - Marino et al, Pediatrics, 2010
  - Wray et al, Cardiology Young, 2012

- There is wide variation of QOL score within cardiac sub-groups.
  - Marino et al, Pediatrics, 2010
  - Wray et al, Cardiology Young, 2012

Generic PedsQL QOL Scores:
CHD Survivors have Lower QOL Scores than Healthy Controls

Variation in QOL within CHD Subgroups

<table>
<thead>
<tr>
<th>PCQLI Median Total Score (Range)</th>
<th>Child</th>
<th>Parent of Child</th>
<th>Adolescent</th>
<th>Parent of Adolescent</th>
</tr>
</thead>
<tbody>
<tr>
<td>AS (n=75)</td>
<td>86.2</td>
<td>86.8</td>
<td>89.5</td>
<td>85.2</td>
</tr>
<tr>
<td>(51.9, 100)</td>
<td>(54.1, 100)</td>
<td>(65.1, 97.6)</td>
<td>(40.8, 99.3)</td>
<td></td>
</tr>
<tr>
<td>TOF (n=125)</td>
<td>75.6</td>
<td>78.6</td>
<td>78.7</td>
<td>78.7</td>
</tr>
<tr>
<td>(48.7, 100)</td>
<td>(43.5, 100)</td>
<td>(39.6, 99.3)</td>
<td>(23.5, 100)</td>
<td></td>
</tr>
<tr>
<td>Fontan (n=219)</td>
<td>64.4</td>
<td>66.1</td>
<td>70.5</td>
<td>69.7</td>
</tr>
<tr>
<td>(32.2, 99.1)</td>
<td>(30.7, 100)</td>
<td>(39.6, 100)</td>
<td>(26.0, 98.5)</td>
<td></td>
</tr>
</tbody>
</table>

Lesson Learned
Children and families are incredibly resilient.

They spend a lot more time thinking about what they can do than about what they can’t do.
Surgical and ICU Predictors of QOL in the CHD Surgical Survivors

- 8 Cardiac Centers US and UK
- Detailed Surgical and ICU data collection
- CHD Lesions (n=575)
  - Transposition of the Great Arteries (n=149, 26%)
  - Tetralogy of Fallot (n=169, 29%)
  - Single Ventricle s/p Fontan (n=257, 45%)
- Patients: Male (60%), Caucasian (83%), Age 11.7±2.9 yrs

Predictors of QOL in the CHD Surgical Survivors

- Independent predictors of Lower PCQLI Total score in patients and parent proxy-reporters
  - Greater number of surgeries (p<0.04)
  - Greater number of inotrope days (p=0.026)
  - Post-operative neurologic deficit persisting at DC (p=0.004)
  - Greater number of Total Cardiac Hospitalizations (p=0.035)
  - Shorter time from last hospitalization (p=0.004)
  - Greater number of MD visits in the prior 12 mo (p<0.0001)
  - Unmarried parents (p<0.04)
  - Non-College parental education (p<0.0007)
  - African-American race (p<0.0002)

The R² values for models ranged from 0.27 - 0.32

Surgical and Intensive Care Unit Variables accounted for only 10-15% of the variation in HRQOL score.
Lesson Learned

When you are a kid, there is no such thing as a “minor” heart procedure.

Depression in adolescents with CHD

- 231 adolescents with CHD
- Parental attitude
- Resilience
- NYHA functional class
- Academic achievement
- Age
- Not type of heart disease

Explained 62% of the variance in depression

JR Moon et al, Heart and Lung, 2009

Depression in adolescents with CHD

- 54 adolescents and adults with Fontan physiology -
- Lower physical health status
- More depressed
- Equivalent QOL, mental health and social support

Explained 55% of the variance in QOL

NA Pike et al, J Cardiovasc Nurs, 2012

The Importance of Self-Perceptions to Psychosocial Adjustment in Adolescents With Heart Disease

- Children’s Hospital of Wisconsin
  - n=92 adolescents, 11-18 years (mean 14.2 ± 2.1 years)
  - Examined self-perceptions as a determinant of psychosocial adjustment in CHD adolescent survivors
  - Self-perceptions and the Discrepancy between competence and importance explained most variance in both QOL and behavior problems ($R^2 = .24-.34$)

Disease Severity and Self-Perception of Health

<table>
<thead>
<tr>
<th>Clinical Severity</th>
<th>Self-Perception of Health</th>
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</thead>
<tbody>
<tr>
<td>Insignificant</td>
<td>Excellent</td>
</tr>
<tr>
<td>Mild</td>
<td>Very Good</td>
</tr>
<tr>
<td>Moderate</td>
<td>Good</td>
</tr>
<tr>
<td>Marked</td>
<td>Fair</td>
</tr>
<tr>
<td>Severe</td>
<td>Poor</td>
</tr>
</tbody>
</table>

$r = .17$

< 3% of the variance in self-perception of health was explained by disease severity
Predictors of QOL

- Self-Perception of Health
- Discrepancy Score
- Global Self-Worth
- Need for Medications

\[ R^2 = .34 \]

\[ R^2 = .17 \]

Did NOT Predict QOL:
- Gender
- Age
- Race
- Birth Order
- Family Structure
- Severity of Illness

Model \( R^2 = .54 \)

Lesson Learned

No child can ‘see’ himself directly. He only sees himself from the reflections of others.”

Dorothy Corkille Briggs, Your Child’s Self-Esteem, 1975

Reliability Within Specialty Groups

<table>
<thead>
<tr>
<th></th>
<th>Intensivist</th>
<th>Output Cardiologist</th>
<th>ICU RN</th>
<th>Outpt RN</th>
<th>APN</th>
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</thead>
<tbody>
<tr>
<td>Predicted QOL</td>
<td>79 ± 11</td>
<td>79 ± 11</td>
<td>83 ± 10</td>
<td>83 ± 9</td>
<td>86 ± 9</td>
</tr>
<tr>
<td>ICC</td>
<td>0.35</td>
<td>0.36</td>
<td>0.34</td>
<td>0.38</td>
<td>0.35</td>
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</table>

Agreement Between Specialty Groups

<table>
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<tr>
<th></th>
<th>ICU MD</th>
<th>ICU RN</th>
<th>Outpt MD</th>
<th>Outpt RN</th>
<th>APN</th>
</tr>
</thead>
<tbody>
<tr>
<td>ICC</td>
<td>0.19</td>
<td>0.10</td>
<td>0.20</td>
<td>0.14</td>
<td>0.22</td>
</tr>
</tbody>
</table>

Methods

- Clinicians: ≥ 3 years experience; ≥ 70% clinical time
  - Cardiac ICU MDs and RNs
  - Outpatient cardiologists and RNs
  - Cardiac advanced practice nurses
- Provided PCQLI patient clinical summaries
- Asked to predict QOL score
Conclusions

- Reliability within groups of clinicians to predict QOL as reported by individual children with heart disease & their parents is poor.
- Agreement between groups of clinicians to predict QOL is poor.
- Clinicians tend to underestimate QOL for patients who report good QOL, and overestimate QOL in those who report poor QOL.
- Clinicians need to be cognizant of these data when communicating with patients & their families about expected outcomes in the intensive care unit.

“Nurturing” Neurodevelopment and Psychosocial Resilience in Children and Adolescents to Improve QOL

- Early intervention to:
  - Minimize impact of executive dysfunction
  - Minimize impact of affective disorder
  - Minimize impact of ADHD
- Occupational therapy to improve gross motor function and physical activity
- Psychosocial support to improve self-perceptions, social competence, and autonomy
- Parental support to reduce stress and promote positive parenting

Summary

- Interventions needed to prevent and treat ND and psychosocial issues in complex CHD survivors and their families to improve the patients long-term QOL.
- These interventions should span the care continuum.
- High-risk CHD survivors should be referred directly to Cardiac ND Programs for formal developmental and medical evaluations to “Nurture” and improve their QOL.
- There is little data on neurodevelopmental outcomes in the ACHD population and research is needed to see the impact on QOL in this ever-growing population.

Challenge

Quality of Life should be one of the outcomes that we assess, follow, and potentially treat, as part of routine care for pediatric heart disease.
Contact Information

Kathy Mussatto, PhD, RN
Nurse Scientist
Co-Director of Cardiac Research
Herma Heart Center
Children's Hospital of Wisconsin
PO Box 1097, MS B550A
Milwaukee, WI 53201

414-266-2073
Fax 414-266-6248
kmussatto@chw.org