M. Drakouli\textsuperscript{1}, E. Patiraki\textsuperscript{2}, M. Giannakopoulou\textsuperscript{2}, K. Petsios\textsuperscript{1}, I. Voutoufianaki\textsuperscript{1}, V. Matziou\textsuperscript{2}

\textsuperscript{1} Pediatric Cardiosurgical Intensive Care Unit, "Onassis" Cardiac Center, Athens, Greece
\textsuperscript{2} National and Kapodistrian University, Faculty of Nursing, Athens, Greece
Congenital heart defect (CHD) refers to a problem regarding the heart’s structure and function, due to abnormal heart development, during endometrial life. Congenital means present at birth.

Worldwide, CHD are estimated to be 1-2% of total births.

- The term Congenital heart disease (CHD) describes a number of different problems affecting the heart. It is the most common type of birth defect.
- Congenital heart diseases account for more deaths in the first year of life than any other birth defects.
- Congenital heart diseases are often divided into two types: cyanotic (blue skin color caused by a lack of oxygen) and non-cyanotic.

**Cyanotic:**
- Tetralogy of Fallot
- Transposition of the great vessels
- Tricuspid atresia
- Total anomalous pulmonary venous return
- Hypoplastic left heart
- Pulmonary atresia
- Ebstein’s anomaly
- Truncus arteriosus

**Non-cyanotic:**
- Ventricular septal defect (VSD)
- Atrial septal defect (ASD)
- Patent ductus arteriosus (PDA)
- Aortic stenosis
- Pulmonary valve stenosis
- Coarctation of the aorta
- Atrioventricular canal
- Atrioventricular canal (endocardial cushion defect)
Over the past decades, there is a significantly increased interest on quality-of-life (QoL) issues for children and adolescents with congenital heart disease (CHD).

In specific, an increasing number of studies in the QoL topic have emerge in the literature elaborating even further the notions of mortality and morbidity by taking under consideration data for reported physical functioning along with psychosocial well-being.
Assessment of Quality of Life in Children and Adolescents with CHD and the factors that determine it, through a systematic approach.
Material-Methods

- **Critical literature review**

- **Research of the literature in electronic databases:**
  - (PubMed, Web of Science, Scopus, Cinahl) with the following key-words:
  - "congenital heart disease", "quality of life", "children", "adolescents"

- **The inclusion criteria were:**
  - Original research studies
  - Publication date after 2000
  - Publication language-English
  - Included pediatric patients aged 0-18 years old
  - The Quality of Life in Children and Adolescents with CHD was described with clarity
Studies were found in databases PubMed, Cinahl, Scopus

436

Excluded for phenomenal consistency 72

Studied 104 articles

Available in another language 8

Excluded for vague content 4

Number of included studies 20
Results
Results

• Methodological quality of the studies varied greatly, while most studies suggested a moderate quality.

• Both generic and disease-specific instruments exist to measure pediatric QoL in children with CHDs.

• Disease-specific instruments are believed to be:
  - More comprehensive for a specific disease,
  - More sensitive to change in condition over time, and
  - A better discriminator of differences between sub-groups within a disease category.

  - PedsQL Cardiac Module - Uzark et al, 2003
  - CHAT - Kendall et al, 2001
  - ConQol - Macran et al, 2006
  - CHD-TAAQOL - Kamphuis et al, 2004
  - PCQLI - Marino et al, 2008
Main characteristics & findings in studies

<table>
<thead>
<tr>
<th>Authors</th>
<th>Aim</th>
<th>Sample</th>
<th>Type of study</th>
<th>Main Outcomes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kwon et al, 2011</td>
<td>Children &amp; adolescents with repaired ToF report QoL</td>
<td>20 children &amp; adolescents with repaired ToF (8-18 years old)</td>
<td>Prospective Study</td>
<td>Self-reported QoL appears similar to healthy peers. For both children and parents, QoL positively correlated with the child's exercise capacity.</td>
</tr>
<tr>
<td>Tahirovie et al, 2011</td>
<td>QoL in children after cardiac surgery for CHD</td>
<td>114 children &amp; adolescents after surgery for CHD (2-18 years old)</td>
<td>Cross-sectional Study</td>
<td>Children &amp; adolescents after cardiac surgery for CHD by self and parent assessment, have a lower QoL than healthy peers.</td>
</tr>
<tr>
<td>Sears et al, 2011</td>
<td>QoL in pediatric patients with implantable cardioverter defibrillators</td>
<td>60 children and adolescents (8-18 years old) &amp; their parents</td>
<td>Prospective Study</td>
<td>The pediatric sample reported lower psychosocial and physical QoL scores than healthy children's normative scores.</td>
</tr>
<tr>
<td>Strieper et al, 2010</td>
<td>This study evaluates whether a successful catheter ablation improved QoL scores in pediatric patients</td>
<td>27 children and adolescents with SVT (5-18 years old)</td>
<td>Prospective Study</td>
<td>Successful elimination of SVT substrate results in improved pediatric QoL scores as reported by patients. The greatest reported improvement post-ablation was in physical functioning.</td>
</tr>
<tr>
<td>Uzark et al, 2008</td>
<td>QoL in children with CHD as perceived by children and parents</td>
<td>475 children and adolescents (2-18 years old) and their parents</td>
<td>Cross-sectional Study</td>
<td>Children with more severe cardiovascular disease reported lower mean scores for physical functioning. Overall, children with CHD perceived lower QoL than healthy peers, across all age groups.</td>
</tr>
<tr>
<td>Pilla et al, 2008</td>
<td>HRQoL and right ventricular function in the midterm follow-up assessment after ToF repair</td>
<td>35 children (3-7 years old) with ToF repair</td>
<td>Cross-sectional study</td>
<td>The right ventricle function is not related to QoL after surgical repair of ToF. A trend for worse results was observed in the physical domain.</td>
</tr>
<tr>
<td>Landolt et al, 2008</td>
<td>HRQoL in children and adolescents after open-heart surgery</td>
<td>110 children and adolescents after surgery for CHD (7-16 years old)</td>
<td>Prospective Study</td>
<td>HRQoL is impaired in children with CHD after open-heart surgery. Medical variables, such as, duration of cardiopulmonary bypass, length of hospitalization, need for current cardiac medication and the quality of family relationships are important determinants.</td>
</tr>
</tbody>
</table>

**Disease Limitations**

- **Related to medication**
  - Refusing to take medication
  - Anxiety about side effects
  - Denial visiting the doctor or hospital, according to the medical schedule

- **Related to disease symptoms**
  - Feeling weakness, fatigue and pain, daily
  - Disease aggravation symptoms

- **Related to physical activity**
  - Reduced or significantly lower physical activity
  - Creative engagement
  - Alternative types of sports
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<tr>
<td>Brosig et al, 2007</td>
<td>Psychosocial outcomes for preschool children and families after surgery for complex CHD</td>
<td>13 children with TGA &amp; 13 children with HLHS (3-6 years old)</td>
<td>Proxy reports only</td>
<td>Cross-sectional Study</td>
</tr>
<tr>
<td>Hovels-Gurich et al, 2007</td>
<td>Long-term behavior and QoL after corrective cardiac surgery in infancy for ToF or VSD</td>
<td>20 children with ToF &amp; 20 children with VSD (5-12 years old) &amp; their parents</td>
<td>Cross-sectional Study</td>
<td>Children with preoperative hypoxemia in infancy due to cyanotic cardiac defects are not at significantly higher risk for behavioral problems and reduced QoL than those with acyanotic heart defects. Internalizing &amp; externalizing problems and reduced self-esteem were associated with reduced endurance capacity.</td>
</tr>
<tr>
<td>Goldbeck et al, 2006</td>
<td>The impact of severity of disease and social disadvantage on QoL in families with CHD</td>
<td>132 children and adolescents with CHD &amp; their parents</td>
<td>Prospective Study</td>
<td>A significant interactive effect indicated a cumulative negative impact of the severity of the disease and social disadvantage on the QoL of the patients.</td>
</tr>
<tr>
<td>Goldbeck et al, 2005</td>
<td>QoL in families of children with CHD</td>
<td>69 children and adolescents with CHD &amp; their parents</td>
<td>Cross-sectional Study</td>
<td>Parents with low own QoL agree significantly more with their children than parents with high QoL. Parent-child agreement on the children's QoL is limited and reflects complementary subjective viewpoints.</td>
</tr>
<tr>
<td>Culbert et al, 2003</td>
<td>QoL of children after repair of TGA</td>
<td>306 children and adolescents after surgery for TGA (11-15 years old)</td>
<td>Cross-sectional Study</td>
<td>QoL and health status as perceived by children after TGA repair is excellent when compared with healthy peers (except self-esteem) and is better after arterial switch operation than atrial repair.</td>
</tr>
</tbody>
</table>

**Stress and Emotional Disorders**

- **Family's (parents & child) stress related to:**
  - Disease symptoms
  - Disease prognosis
  - Disease complications
  - Potential danger for re-operation

- **Anxiety about death because of:**
  - Re-operation
  - Admission and long-lasting hospitalization for disease complications
  - Invasive diagnostic procedures
    (catheterization, transesophageal echo, etc)

- **Sleeping disorders**

- **Emotional disorders**
  - Aggression, anger, embarrassment, sadness, depression

- **Perceived physical appearance**
  - Low self-esteem, lack of confidence, internalizing symptoms
**Main characteristics & findings in studies**

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<tr>
<td>Guerra et al, 2012</td>
<td>QoL 4 years after complex heart surgery in infancy</td>
<td>130 parents with children with complex CHD (4 years old)</td>
<td>Prospective Cohort Study</td>
<td>Parents reported lower QoL than normative population, after complex heart surgery. A positive correlation was found between age at surgery, postoperative low cardiac output, socioeconomic status (SES) and QoL.</td>
</tr>
<tr>
<td>Limbers et al, 2012</td>
<td>Factors associated with perceived cognitive problems in children and adolescents with CHD (8-18 years old)</td>
<td>246 children &amp; adolescents &amp; their parents</td>
<td>Prospective Study</td>
<td>Cognitive problems and lower school progress seem to be positively correlated with low SES and greater disease severity.</td>
</tr>
<tr>
<td>Luyckx et al, 2012</td>
<td>Individual and Contextual determinants of QoL in adolescents with CHD</td>
<td>429 adolescents with CHD (14-18 years old)</td>
<td>Prospective Study</td>
<td>Perceived health status, sense of coherence and parental support, found to positively predict QoL.</td>
</tr>
<tr>
<td>Shearer et al, 2012</td>
<td>QoL in adolescents with CHD</td>
<td>10 adolescents with CHD (13-17 years old)</td>
<td>Descriptive Interpretive Study</td>
<td>Most adolescents viewed themselves as normal. They focusing on physical activity limitations and their need to fit in.</td>
</tr>
<tr>
<td>Cohen et al, 2007</td>
<td>QoL, Depressed Mood and Self-Esteem in adolescents with CHD</td>
<td>90 adolescents with CHD (12-18 years old)</td>
<td>Cross-sectional Study</td>
<td>Adolescents with severe heart disease reported higher levels of depressed mood and lower self-esteem than did adolescents with moderate/mild CHD and age-matched healthy controls.</td>
</tr>
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</table>

**Communication problems**

- **Within the family**
  - Disagreement/argument between parents and children
  - Feeling of being “overprotected”  
    - Guerra et al, 2012

- **Difficulty of making friendships with peers**
  - Internalizing symptoms
  - Feeling loneliness
  - Aggressive behavior to others
    - Limbers et al, 2012

- **Denying to cooperate with doctors and nurses, over medical procedures**

- **Disability of being socialized at school and community (social functioning)**
  - Shearer et al, 2012
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<tr>
<td>Costello et al, 2012</td>
<td>QoL of pediatric cardiac patients who previously required ECMO</td>
<td>41 children and adolescents &amp; their parents (5-18 years old)</td>
<td>Cross-sectional</td>
<td>Children &amp; adolescents with after survival with ECMO, seemed to have lower QoL than healthy population. Their parents reported problems in speech, writing, attention.</td>
</tr>
<tr>
<td>Uzark et al, 2012</td>
<td>Clinical utility of HRQoL assessment in pediatric cardiology outpatient practice</td>
<td>176 children &amp; adolescents with CHD (8-18 years old)</td>
<td>Descriptive</td>
<td>Children and adolescents with CHD reported problems in school progress 22.3% (school functioning problems). Cardiologists reported initiating interventions in 30.1% of patients.</td>
</tr>
<tr>
<td>Spijkerboer et al, 2006</td>
<td>Health-related QoL in children and adolescents after invasive treatment for CHD</td>
<td>113 children &amp; adolescents after surgery for CHD (8-15 years old)</td>
<td>Prospective</td>
<td>The total sample of CHD children (8-15 years old) obtained significantly lower mean scores on motor, cognitive and emotional functioning than reference peers, reflecting an experience of poorer functioning.</td>
</tr>
</tbody>
</table>

**Cognitive Disorders**

- **Cognitive disorders**
  - Difficulty solving math problems and developing critical mind
  - Clarity of mind
  - Speech disorders
  - Difficulty using language both verbally and written

- **Reduced school performance**:
  - Missing school due to medical procedures
  - Attention disorders - Difficulty concentrating to class

Costello et al, 2012

Uzark et al, 2012

experience of poorer functioning.
Determinants of CHD in QoL

A) Related to disease and medical treatment

- **Restoration surgery**
  - Number of surgeries
    - Full correction or palliative surgery
    - Age of full correction - surgery

- **Post-operative complications**
  - Direct (short influence in QoL)
  - Permanent disorders (long-term influence in QoL)
    - Neurological disorders (ECMO)
    - Placement of pacemaker/defibrillator (Heart rate disorders)

- **Treatment Limitations**
  - Regular, scheduled, postoperative check-up
  - Daily intake drugs
  - Limited physical activity

- **Compliance with treatment**
  - Symptoms

- **Re-operation**
Determinants of CHD in QoL

B) Social Factors

- **Family**
  - Family consistency
    - Domestic conflicts
    - Fraternal relations
    - Family preparation and education
      - Parent education about the nature of the disease and the direct identification signs of increasing (cyanosis, tachypnea, faint, tachycardia)
    - Disorders in Parents - QoL
      - Supportive family environment

- **School**
  - Prevention and early identification of learning and cognitive problems from preschool age (play-therapy, speech-therapy)
  - Participation in team sports, inclusion of physical activity in daily life

- **Peers**
  - Participation in activities
  - Encouragement to develop friendships
Conclusions

• QoL in children and adolescents with CHD is comparable to QoL of normative/healthy population, when the disease induced limitations are controlled and compliance with treatment is satisfactory.

• Severe symptoms are mainly affecting physical activity. Disease complications that can lead to painful diagnostic tests or re-operation, significantly seem to have a direct effect on their QoL.

• Children’s socialization, family consistency, early identification of learning, cognitive and emotional problems as well as participation in physical activities are acknowledged as important determinants of QoL improvement.

• Programmes providing psychosocial support for children with CHD and their caregivers should be family-focused and take under consideration risk factors (both medical and psychosocial).

• Creating individualized care plan at home and regular follow-up, in a consistent and continuous cooperation with parents, is an important nursing responsibility that decisively affects QOL of these children.
HRQoL in children with congenital heart diseases should be assessed according to age, growth level, severity and acceptability of the disease, as well as personality features.

Identification of significant impairments in QoL can impact clinical outcomes in children with CHD.