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Quality of Life in Congenital Heart Disease Survivors: Creating New Models of Intervention to Promote Resilience and Minimize Disability

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Advancing the Development of Pediatric Therapeutics (ADEPT 6)

Conflict of Interest Disclosures for Bradley S. Marino, MD, MPP, MSCE

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Stock Shareholder	**Nothing to disclose**
Other (identify)	**1. Creator of the Pediatric Cardiac Quality of Life Inventory** **2. National leadership roles with the AHA, AAP, CNOC, and PCHA**

Figure 1:

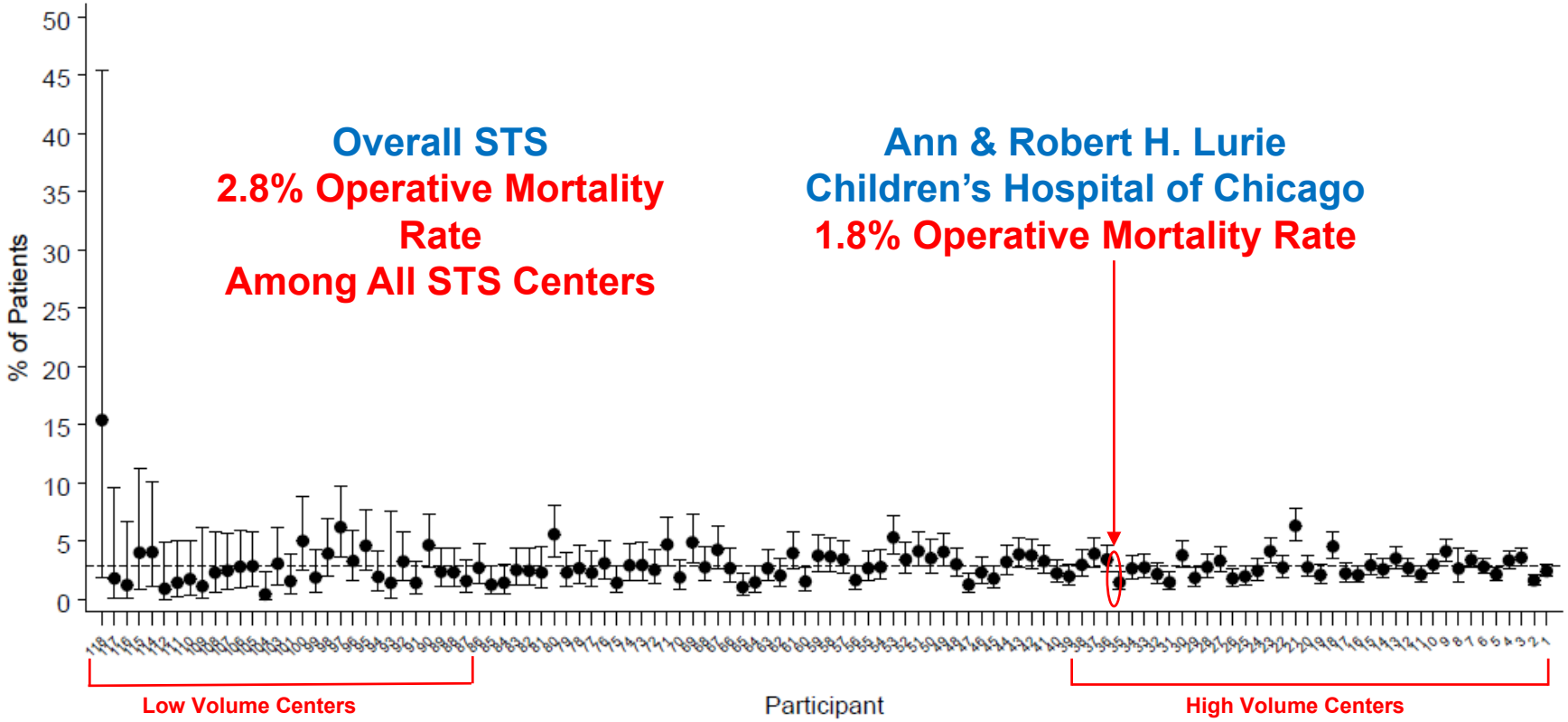
STS Congenital Heart Surgery Operative Mortality

STS Period Jan 2015 – Dec 2018

Duke Clinical Research Institute

All Patients

Operative Mortality + 95% CI



Dotted line on graph represents overall value for the STS: 2.84%

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Important Morbidities after Repair vs Palliation:

- Neurodevelopmental/Neurocognitive
- Psychosocial and Psychiatric
- Late surgery or catheter re-interventions
- Arrhythmias
- Chronic heart failure/transplantation
- Pulmonary hypertension
- Endocarditis
- Other important end organ dysfunction:
  - Chronic renal and liver insufficiency
  - Coagulopathy leading to thromboembolic complication

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Causes of CNS Sequelae Are Cumulative and Interactive

- Patient/Preoperative factors:
  - Genetic abnormalities, *in utero* factors, birth weight, gestational age, socioeconomic status, parental ability
  - Smaller brain volumes and brain maturity at birth
- Global morbidity and sequelae of heart disease
  - Chronic severe cyanosis
  - Malnutrition
  - Arrhythmias
  - Cardiac arrest

Causes of CNS Sequelae Are Cumulative and Interactive (continued)

- Sequelae of cardiac interventions, such as cardiac surgery or catheterization

- Pre- and post-operative hemodynamic instability
  - Impaired cerebrovascular pressure autoregulation and reactivity to CO2
  - ↑ vulnerability to ↓ BP and ↓ CI

Cumulative White Matter Injury

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An MRI Study of Neurological Injury Before and After Congenital Heart Surgery

William T. Mahle, MD; Federica Tavani, MD; Robert A. Zimmerman, MD; Susan C. Nicolson, MD; Kristin K. Galli, MD; J. William Gaynor, MD; Robert R. Clancy, MD; Lisa M. Montenegro, MD; Thomas L. Spray, MD; Rosetta M. Chiavacci, BSN; Gil Wernovsky, MD; C. Dean Kurth, MD

TABLE 4. Incidence of Abnormalities on Brain MRI and MRS

	Preoperative	Early Postoperative	Late Postoperative
Lactate	53%	NA	NA
Infarction			
Total	8%	23%	12%
New		19%	6%
PVL			
Total	17%	52%	0%
New		42%	
Hemorrhage			
Total	4%	43%	0%
New		38%	
Atrophy	0%	0%	12%

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Mahle WT, *Circulation* 2002

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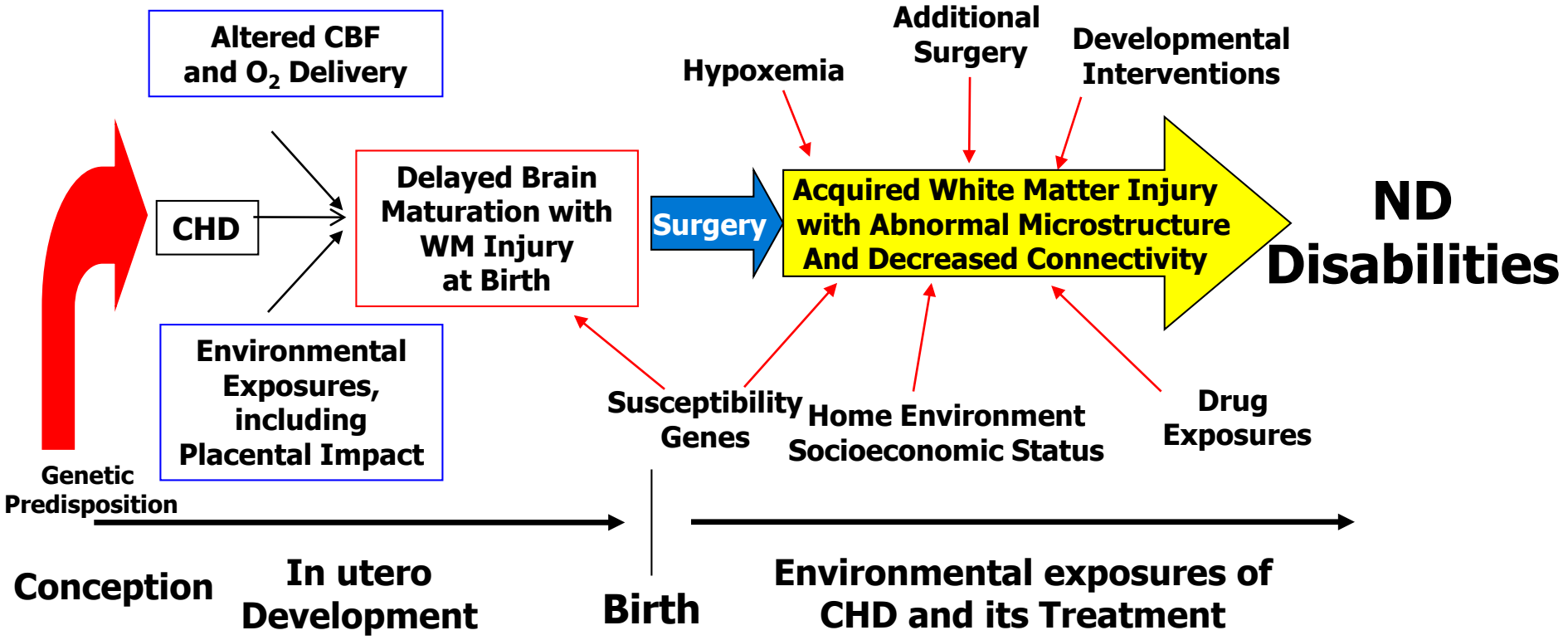
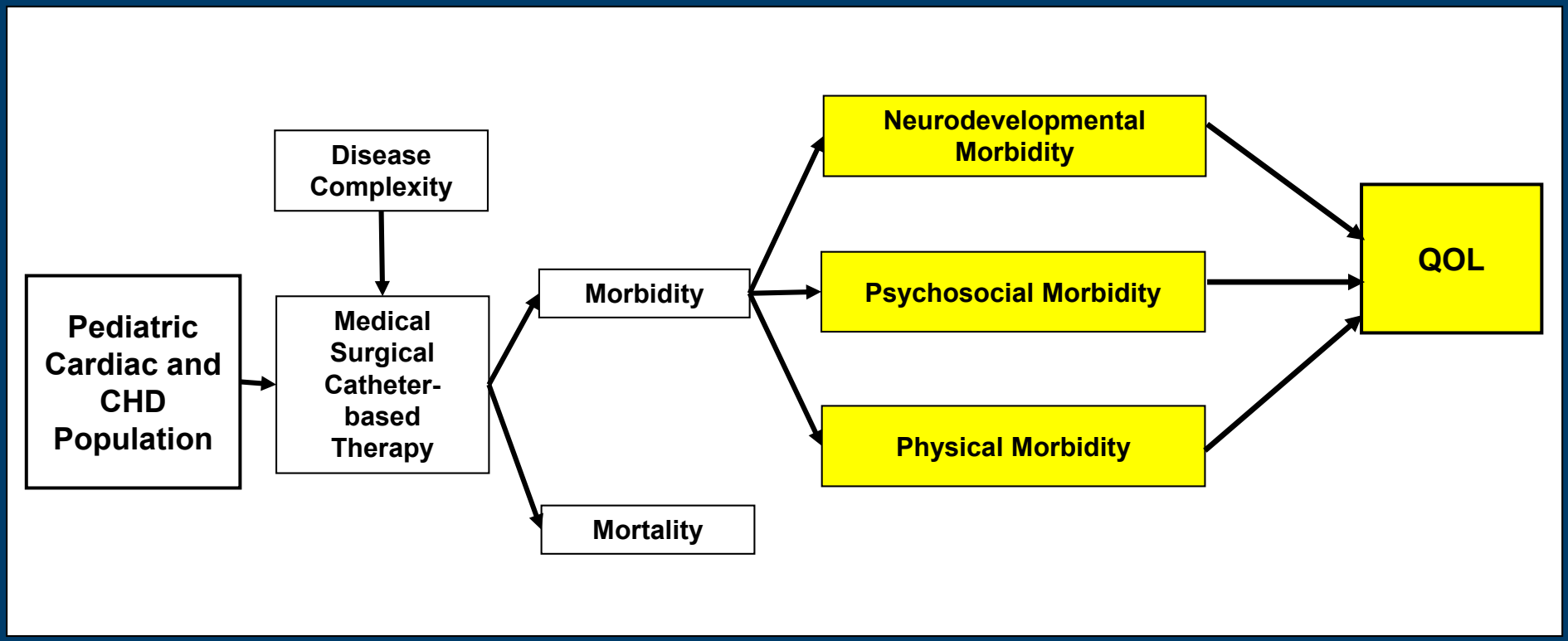
Model for Neurobehavioral Dysfunction in CHD Surgical Survivors



Adapted from J. William Gaynor

How Should We Measure Outcomes in Children with Heart Disease?

Health-related Quality of Life



ND Phenotype in CHD Survivors

- Mild cognitive impairment - normal to slightly lower IQ and Academic Achievement
- Impaired pragmatic language
- Decreased visual construction and perception
- Poor executive functioning
- Inattention and increased impulsivity (ADHD)
- Diminished fine and gross motor skills
- Psychosocial maladjustment (internalizing problems)

Marino et al, Circulation, 2012

ND Phenotype in CHD Survivors

- Individual deficits or delays are may be mild, but often occur across multiple domains
- "High prevalence – low severity" picture doesn't meet classic criteria of a "learning disability"
- Many of these children have difficulties in school, but may not qualify for special services

Learning Challenges Are Common in CHD

- Students with complex CHD have generally lower academic achievement than typical peers
  - **1/3-1/2** of students with complex CHD receive special education programming
  - **1 in 6** are placed in substantially-separate classrooms
  - **1 in 5** repeat a grade at least once
  - **1 in 4** receive occupational therapy, physical therapy, speech therapy, and/or psychotherapy

Bellinger et al., 2011; Bellinger, Watson, et al., 2015; Jaworski, White, DeMaso, Newburger, Bellinger, & Cassidy, 2017; Shillingford et al., 2008

Associations Among CHD, ASD, ADHD, and Learning Disabilities in US Children Aged 2-17 Years, 1997-2011

Variable	CHD N = 374	No CHD N = 158,243	OR
Autism/ASD	2.6%	0.6%	4.6 (1.9-11.0)
ADHD/ADD	10.3%	6.6%	1.6 (1.1-2.4)
Learning Disability	20.9%	7.6%	3.8 (2.9-5.2)

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Razzaghi H et al, *J Pediatr* 2015

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Psychosocial Phenotype in CHD Survivors

- Impaired social interaction and deficits in social cognition

  Bellinger et al, ***Cardiology Young*****, 2008**

- Impaired core communication skills – incidence of autism spectrum disorders

  Swillen et al, ***Am J Med Gen*****, 2015**

- Increased incidence of Psychiatric disorders

  Olsen et al, ***Circulation*****, 2011**

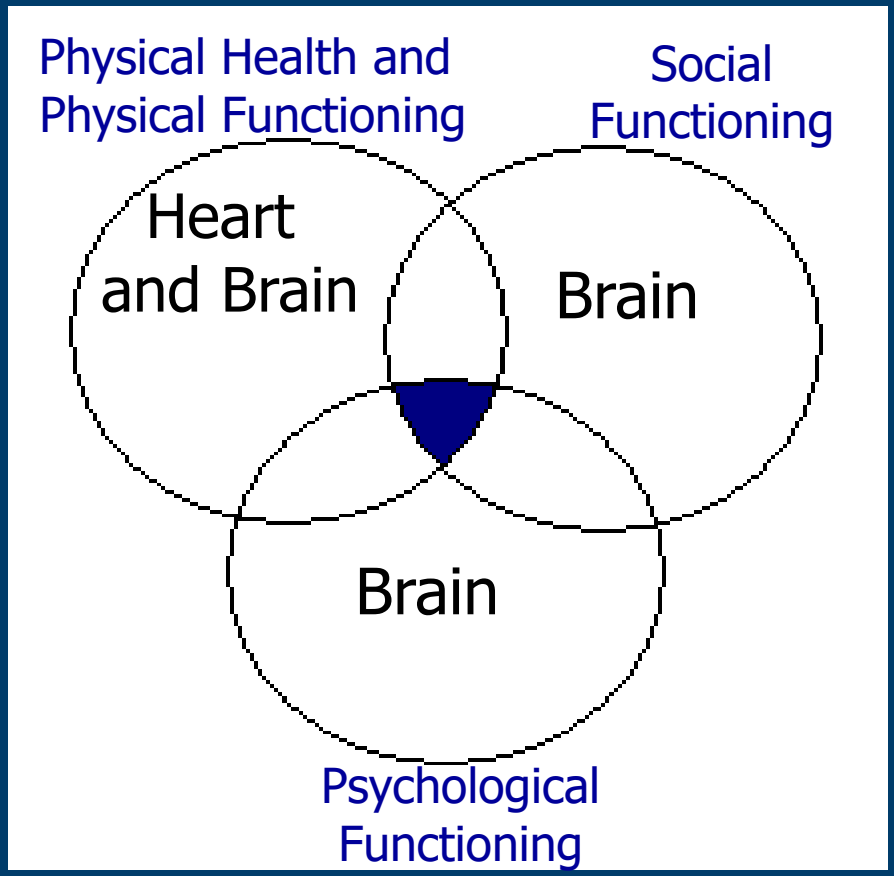
- Issues with behavioral/emotional functioning
  - Anxiety
  - Depression
  - Post-traumatic stress

  Marino et al, ***Circulation*****, 2012**

Definition of Quality of Life (QOL)

- Physical Health and Physical Functioning
- Psychological Functioning
- Social Functioning

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



Ware 1984, Aaronson 1988

Quantitation of HRQOL with a Cardiac-Specific QOL Measure

Validation of the Pediatric Cardiac Quality of Life Inventory

AUTHORS: Bradley S. Marino, MD, MPP, MSCE,[a,b] Ryan S. Tomlinson, BSE,[a] Gil Wernovsky, MD,[c,d] Dennis Drotar, PhD,[e] Jane W. Newburger, MD, MPH,[f,g] Lynn Mahony, MD,[h] Kathleen Mussatto, RN, PhD,[i] Elizabeth Tong, RN, MS,[j] Mitchell Cohen, MD,[k] Charlotte Andersen, RN, MS,[a] David Shera, ScD,[l] Philip R. Khoury, MS,[a] Jo Wray, PhD,[m] J. William Gaynor, MD,[n] Mark A. Helfaer, MD,[d] Anne E. Kazak, PhD,[o] and Judy A. Shea, PhD,[p] for the Pediatric Cardiac Quality of Life Inventory Testing Study Consortium

Divisions of [a]Cardiology, [b]Critical Care Medicine, and [e]Behavioral and Clinical Psychology, Department of Pediatrics, Cincinnati Children's Hospital Medical Center, Cincinnati, Ohio; Divisions of [c]Cardiology, [l]Epidemiology and Biostatistics, and [o]Psychology, Department of Pediatrics, [d]Division of Critical Care Medicine, Department of Anesthesiology and Critical Care, and

WHAT'S KNOWN ON THIS SUBJECT: Little is known regarding the impact of important clinical and patient factors (disease severity, medical care utilization, patient-parent consensus, and patient self-perception, competency, and behavior) on HRQOL in the pediatric cardiac population.

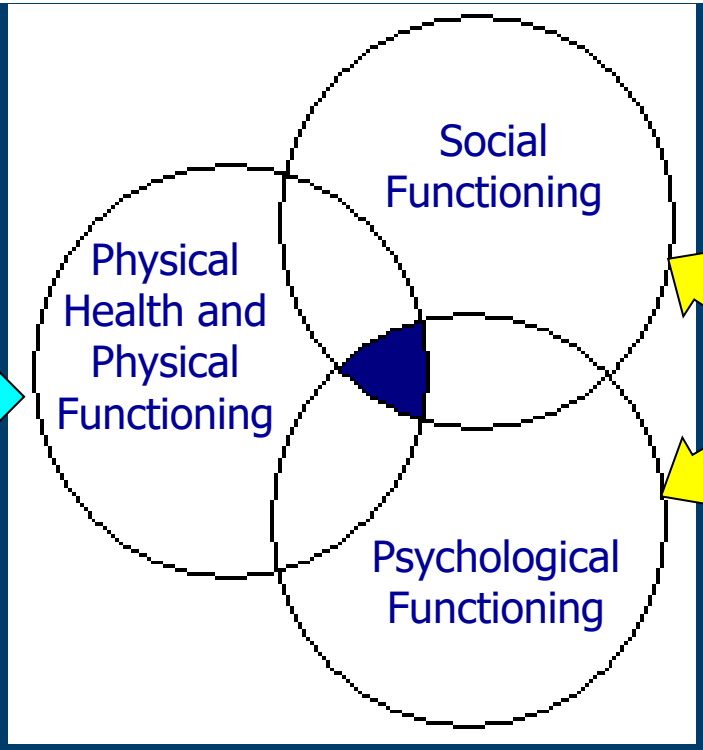
WHAT THIS STUDY ADDS: This large, multicenter study showed that lower HRQOL was associated with greater disease severity and medical care utilization, poorer patient self-perception and competency, and increased behavioral and emotional problems in the pediatric cardiac population. The PCQLI is valid and reliable.

Marino et al, *Pediatrics* 2010

PCQLI Subscales: Disease Impact and Psychosocial Impact

PCQLI Total Score = 100

Disease Impact (DI) Max Score=50

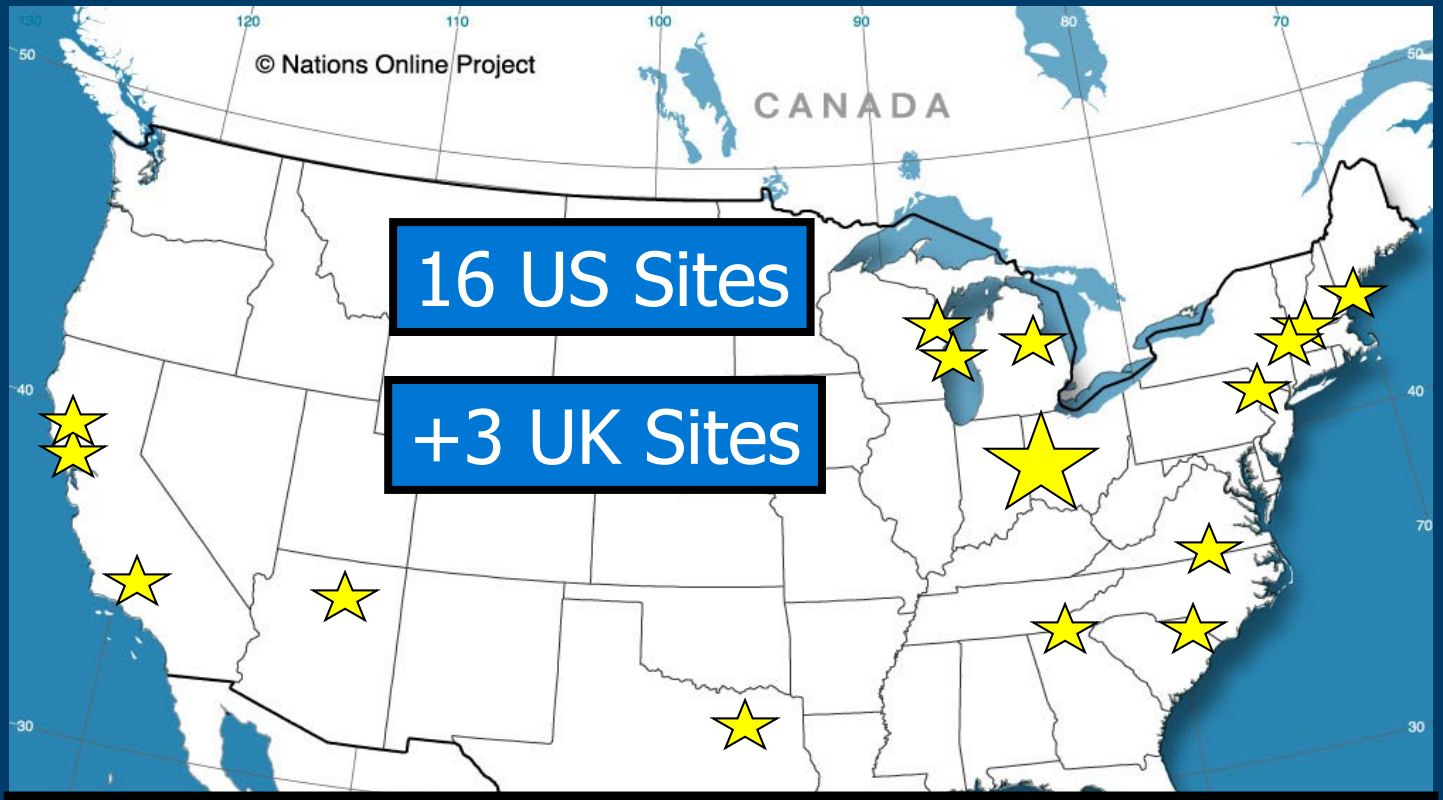


Psychosocial Impact (PI) Max Score=50

Marino et al, *Quality of Life Research* 2008

Higher Score = Higher Quality of Life

PCQLI Validation Study Sites: United States and United Kingdom



Total US and UK Enrollment
3,270 Patient-Parent Pairs = 6,540 Respondents

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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
- QOL in CHD survivors worsens with increasing disease complexity
  Marino et al, *Quality of Life Research* 2008
  Marino et al, *Pediatrics* 2010
  Wray et al, *Cardiology Young* 2012
- 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

Diagnosis	Patient PCQLI Score	Parent PCQLI Score
VSD w/o Intervention (n=78)	82	88
Bicuspid Ao Valve w/o Intervention (n=37)	82	84.9
Ao Stenosis w/o intervention (n = 38)	82	86.1
CoA w/o VSD (n=112)	80	82.9
VSD with Intervention (N = 90)	79	82
Ao Stenosis with intervention (n = 96)	76	77.3
TGA (n = 159)	76	79.2
CoA with VSD (n=52)	76	79.9
Pacemaker w/o CHD (n = 126)	76	70.8
TOF w/o PA (n = 160)	75	75.5
Pacemaker with CHD (n = 141)	67	64.6
TOF with PA (n = 41)	66	67.5
FONTAN (n =250)	66	65.1

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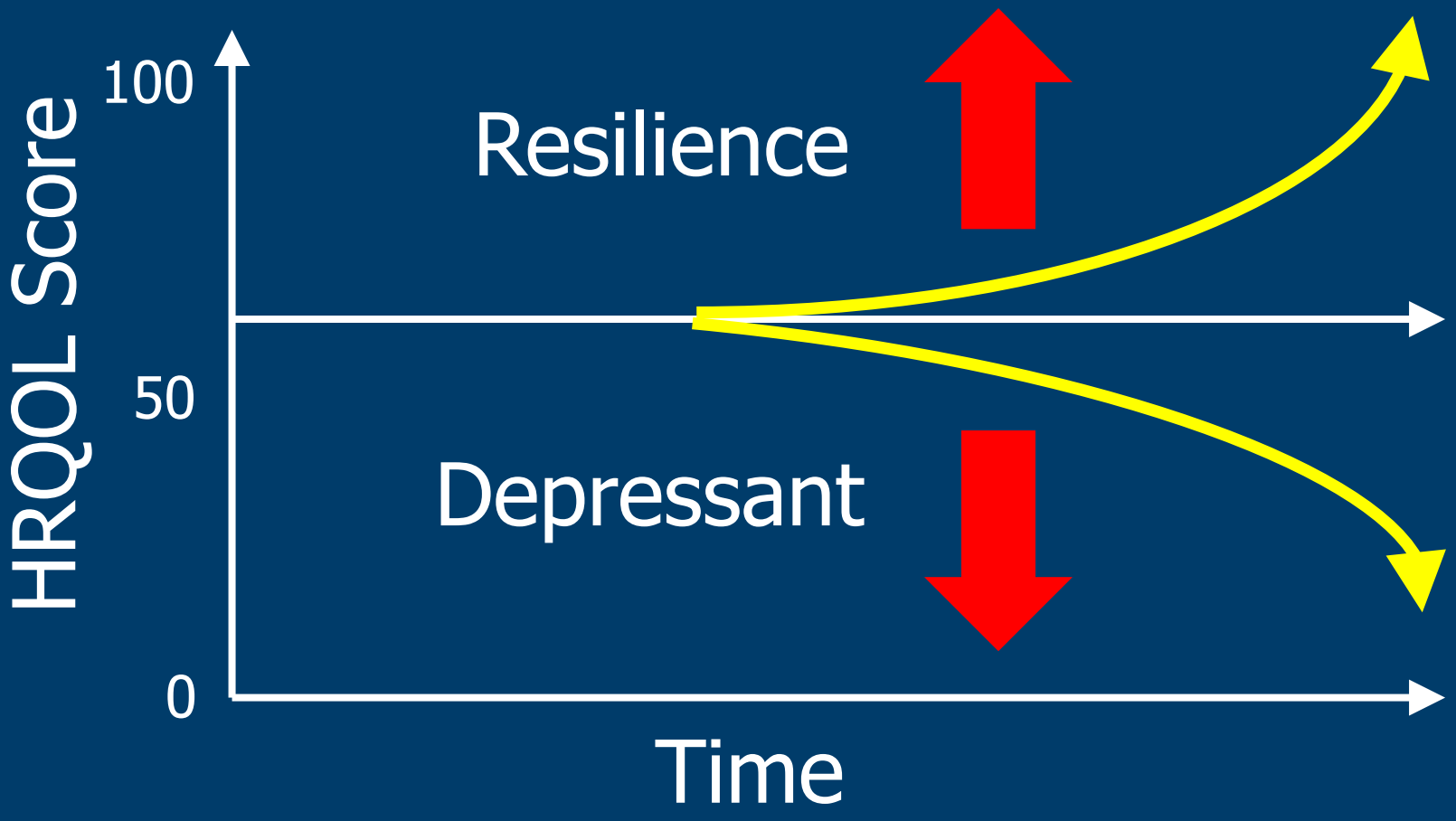
Variation in QOL within CHD Subgroups

PCQLI Median Total Score (Range)				
	Child	Parent of Child	Adolescent	Parent of Adolescent
AS (n=75)	86.2 (51.9, 100)	86.8 (54.1,100)	89.5 (65.1, 97.8)	85.2 (40.8, 99.3)
TOF (n=125)	75.6 (48.7, 100)	78.6 (43.5, 100)	79.6 (39.6, 99.3)	78.7 (33.5, 100)
Fontan (n=219)	64.4 (32.2, 99.1)	66.1 (30.7, 100)	70.5 (39.6, 100)	69.7 (26.0, 98.5)

Marino B, Moss and Adams' 2013

Patient Independent ARC of QOL: Resilience vs Depressant Factors



Marino B, Moss and Adams' 2013

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Surgical Complexity Minimally Impact Long-term Quality of Life

ORIGINAL RESEARCH

American Heart Association | American Stroke Association

Impact of Surgical Complexity on Health-Related Quality of Life in Congenital Heart Disease Surgical Survivors

Amy M. O'Connor, DO; Jo Wray, PhD, MSc; Ryan S. Tomlinson, MD; Amy Cassedy, PhD; Jeffrey P. Jacobs, MD; Kathy J. Jenkins, MD, MPH; Kate L. Brown, MRCP, MPH; Rodney C. G. Franklin, MBBS, MD; Lynn Mahony, MD; Kathleen Mussatto, RN, PhD; Jane W. Newburger, MD, MPH; Gil Wernovsky, MD; Richard F. Ittenbach, PhD; Dennis Drotar, PhD; Bradley S. Marino, MD, MPP, MSCE

Background—Surgical complexity and related morbidities may affect long-term patient quality of life (QOL). Aristotle Basic Complexity (ABC) score and Risk Adjustment in Congenital Heart Surgery (RACHS-1) category stratify the complexity of pediatric

Surgical Complexity assessed by RACHS-1 category and Aristotle Basic Complexity score only accounts for 2% of the variability in QOL score in CHD surgical survivors age 8-18 yrs

and RACHS-1 categories were associated with lower QOL scores ($P<0.005$), correlation with QOL scores was poor to fair ($r=-0.10$ to -0.29) for all groups. Ordinary least squares regression showed weak association with $R^2=0.06$ to $R^2=0.28$. After accounting for single-ventricle anatomy, number of doctor visits, and time since last hospitalization, surgical complexity scores added no additional explanation to the variance in QOL scores.

Conclusions—ABC scores and RACHS-1 categories are useful tools for morbidity and mortality predictions prior to cardiac surgery and quality of care initiatives but are minimally helpful in predicting a child's or adolescent's long-term QOL scores. Further studies are warranted to determine other predictors of QOL variation. **(*J Am Heart Assoc*. 2016;5:e001234 doi: 10.1161/JAHA.114.001234)**

Key Words: Aristotle Basic Complexity • congenital heart disease surgery • quality of life • Risk Adjustment in Congenital Heart Surgery

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O'Connor et al, *JAHA*, 2016

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The Impact of Surgical and Intensive Care Unit Factors on Long-Term Quality of Life in Congenital Heart Disease Surgical Survivors

Bradley S. Marino, MD, MPP, MSCE, Cassedy Amy, PhD, Kate Brown, MD, Mirjana Cvetkovic, MD, John M. Costello, MD, MPH, Rodney Franklin, MD, J. William Gaynor, MD, Simon Laker, RSCN, Katherine Levinson, MD, Helen MacGloin, MD, Lynn Mahony, MD, Annette McQuillan, RSCN, Kathy Mussatto, PhD, Deirdre O'Shea, MD, Jane Newburger, MD, MPH, Michelle Sykes, MD, Sarah A. Teele, MD, Gil Wernovsky, MD, Dennis Drotar, PhD, Jo Wray, PhD

Marino et al, *JACC* , 2015

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Surgical and ICU Predictors of QOL in the CHD Surgical Survivors

- 8 Cardiac Centers US and UK
- 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

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Marino et al, *JACC* (Abstract), 2015

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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 ($p<0.0002$)

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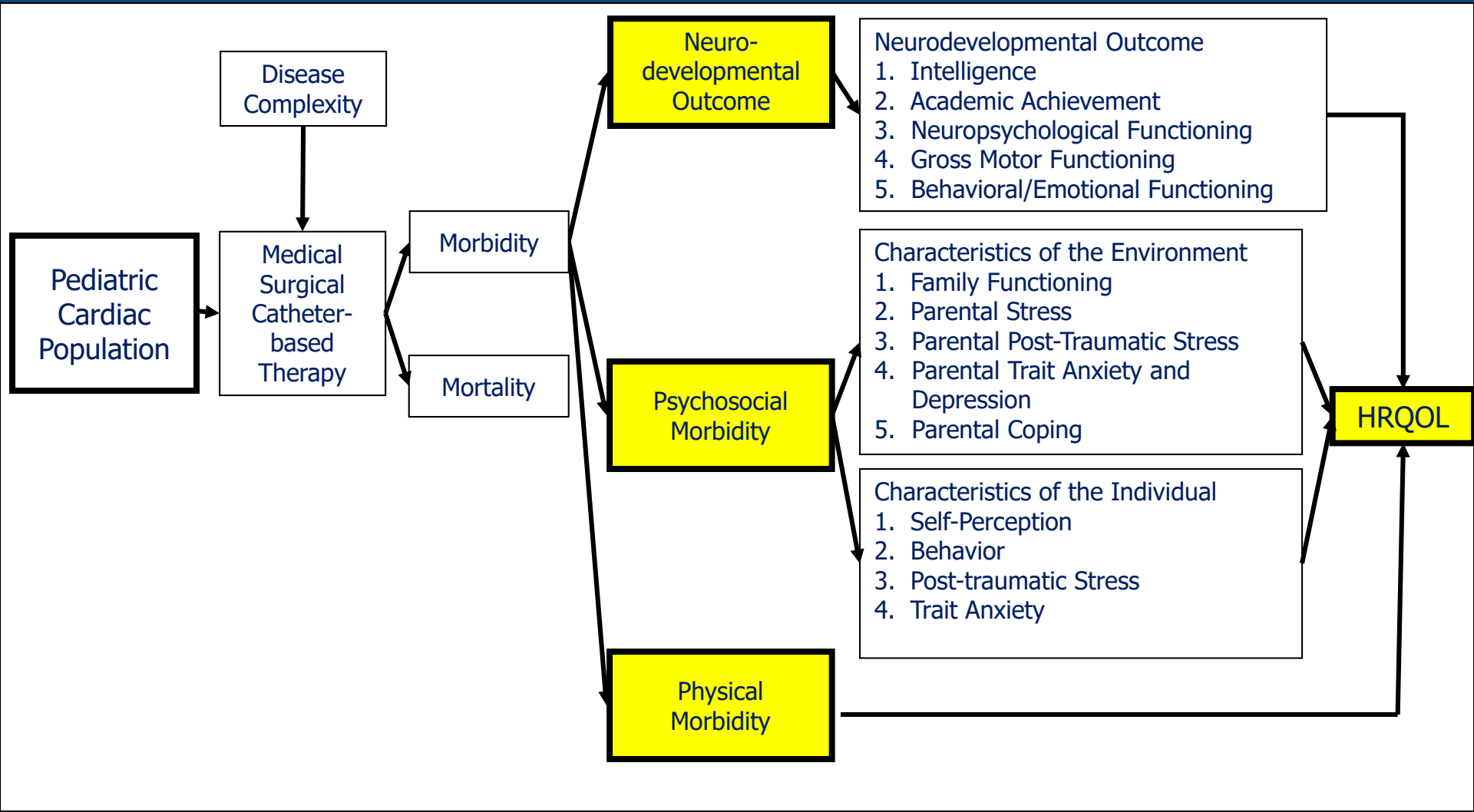
The R^2 values for models ranged from 0.27 - 0.32
Surgical and Intensive Care Unit Variables accounted for only 10-15% of the variation in HRQOL score

QOL Conceptual Model in Pediatric Heart Disease



A Model for Assessing the Impact of Neurodevelopmental and Psychosocial Morbidity Factors on QOL

- Multi-Site Cohort – PCQLI Validation Study (US data)
- Single Site Cohort - CCHMC
- Multi-site Psych Extension at 6 Centers in US and UK
  - Psychosocial Predictors of Psychosocial QOL Score – Multivariate Path Model (US and UK Data)
  - The Individual High Risk Group – Fontans (US and UK Data)

Lower PCQLI Total Score Correlates with Worse Self-Perception and Behavioral and Emotional Functioning [n=1,605]

Instrument	Domain	Respondent Group	Correlation Coefficient
SPPC/SPPA	Global Self Worth (Self-Perception)	Child	0.49
		Adolescent	0.40
Achenbach	Total Competence Scale (Activity, Social, School)	Child/Parent	0.31/0.33
		Adolescent/Parent	0.31/0.38
	Internalizing Problem (Anxious/Depressed, Withdrawn/Depressed, Somatic Complaints)	Child/Parent	-0.52/-0.51
		Adolescent/Parent	-0.51/-0.51

All comparisons significant <0.0001

Marino et al, *Pediatrics* 2010

Lower PCQLI Total Scores Correlate with Increased Risk for Affective Disorder, Anxiety Disorder, and ADHD [n=1,605]

Instrument	Domain	Respondent Group	Correlation Coefficient
Achenbach (DSM)	Affective Disorder	Child/Parent	-0.55/-0.47
		Adolescent/Parent	-0.49/-0.51
	Anxiety Disorder	Child/Parent	-0.38/-0.44
		Adolescent/Parent	-0.37/-0.39
	Somatic Problems	Child/Parent	-0.34/-0.34
		Adolescent/Parent	-0.36/-0.36
	ADHD	Child/Parent	-0.33/-0.33
		Adolescent/Parent	-0.25/-0.24

All comparisons significant <0.0001

Marino et al, *Pediatrics* 2010

Worse Neurodevelopmental Outcome is Associated with Worse PCQLI Total Scores and School Performance

ELSEVIER

The Journal of Pediatrics

Volume 173, June 2016, Pages 154-159

Original Article

Executive Functioning and School Performance among Pediatric Survivors of Complex Congenital Heart Disease

Portions of the study were presented at the scientific session and expo of the American College of Cardiology, April 2-5, 2011, New Orleans, LA.

Melissa Gerstle PhD [1, 2], Dean W. Beebe PhD [1, 2], Dennis Drotar PhD [1, 2], Amy Cassedy PhD [3, 4], Bradley S. Marino MD, MPP, MSCE [5, 6]

Cincinnati Children's ND Cohort

- 143 complex CHD survivors recruited at CCHMC 2008-2010
- Age: 8-16 (11.4±2.4) yrs
- 67% Male, 85% Caucasian
- CHD Groups s/p repair or palliation
  - Transposition of the great arteries (n=41)
  - Tetralogy of Fallot (n=51)
  - Fontan (n=51)

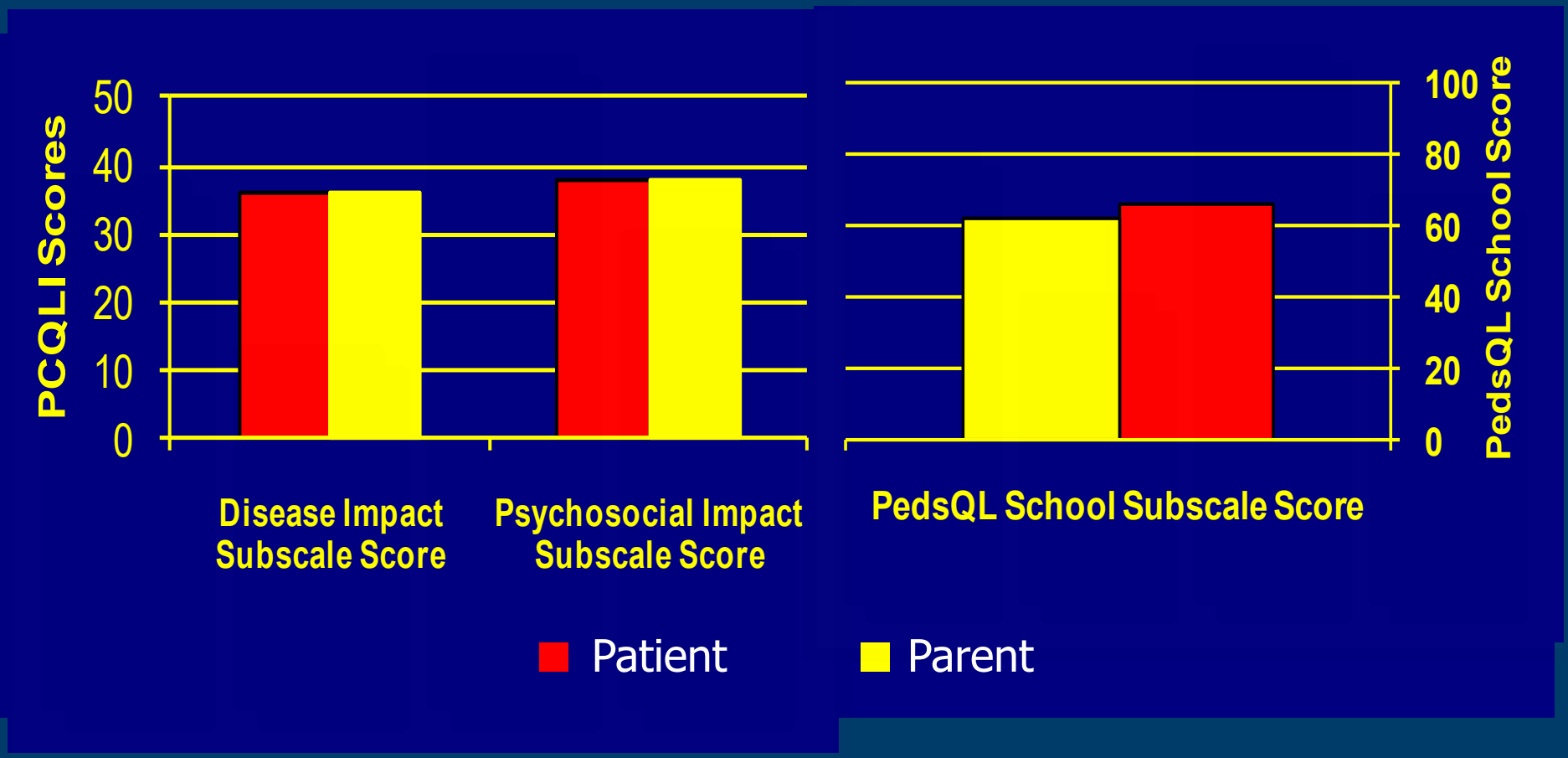
QOL, Behavioral and Emotional Functioning, and Gross Motor Skill Assessment

- QOL
  - PCQLI QOL scores
  - PedsQL School Functioning subscale score
- Behavioral and Emotional Functioning
  - Achenbach: Child Behavior Checklist
- Gross Motor Skill
  - Bruininks-Oseretsky Test 2

Neuropsychological Functioning

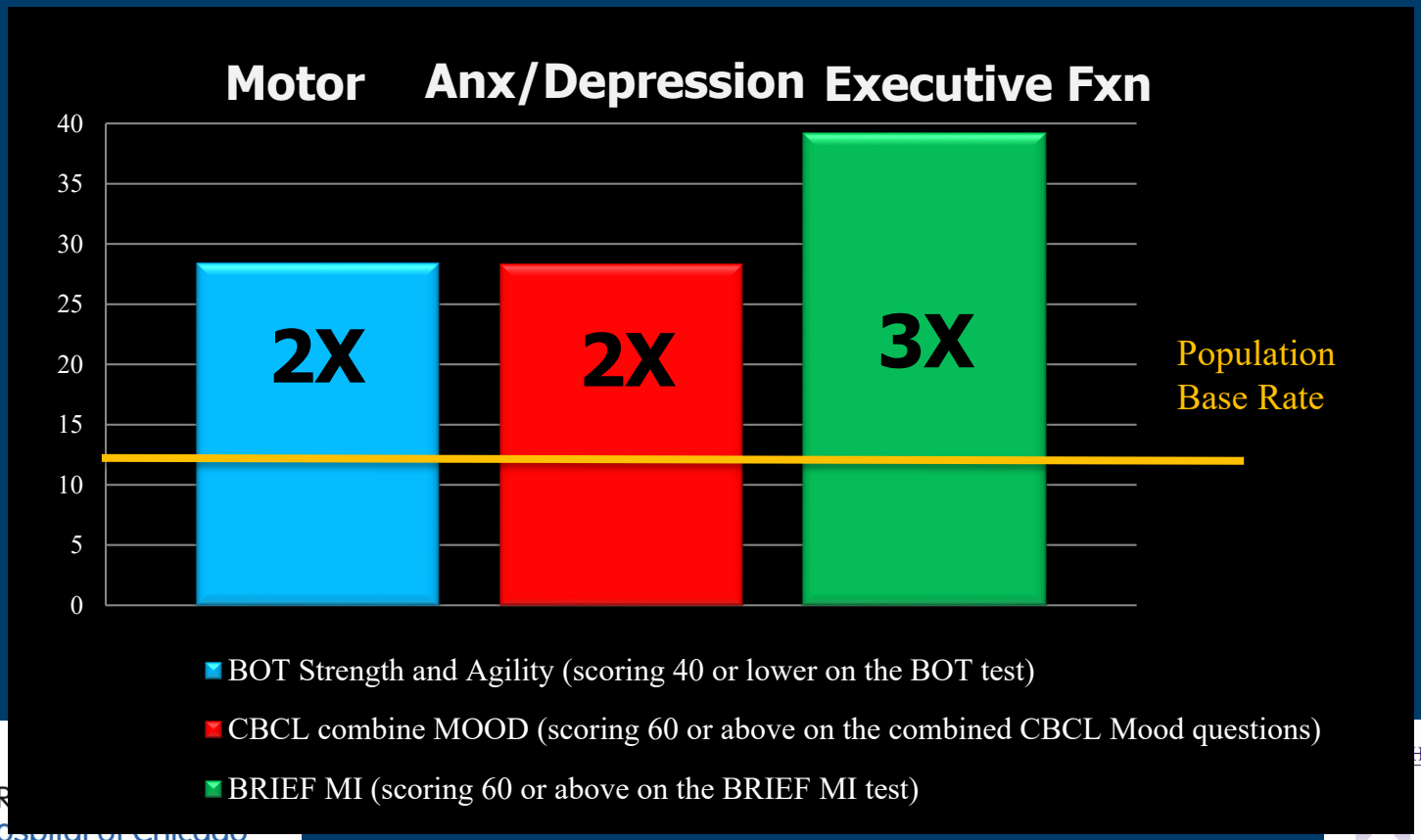
- **Intelligence**
  - Wechsler Intelligence Scale for Children-IV
- **Academic achievement**
  - Wechsler Individual Achievement Test-II
- **Language**
  - Clinical Evaluation of Language Fundamentals-IV
  - Woodcock Johnson-III
- **Visual construction and perception**
  - Beery-Buktencia Developmental Test of Visual-Motor Integration
- **Executive functioning**
  - Behavior Rating Inventory of Executive Functioning
  - Wisconsin Card Sorting Test of Executive Function
- **Attention**
  - Connors' Continuous Performance Test-II
- **Memory**
  - Wide Range Assessment of Memory and Learning- Screener
- **Fine motor skills**
  - Grooved Pegboard

ND Cohort
Patient and Parent PCQLI Subscale Scores and PedsQL School Functioning Subscale Score



Percentage of ND Patients "At Risk" for Problems with Motor, Mood, and Executive Functioning

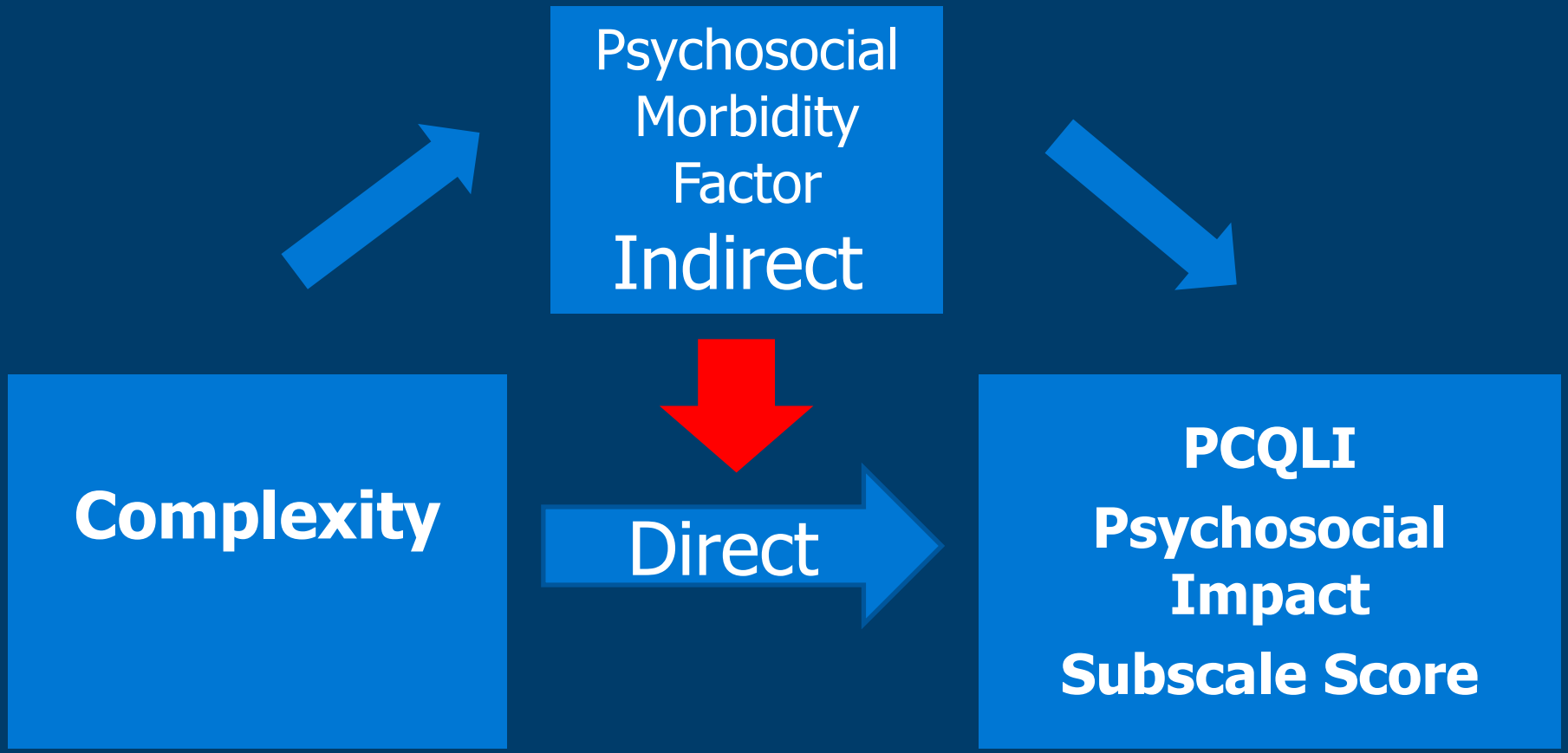
Percent of ND Patients Scoring ≥ 1 SD Worse than the National Mean



Multivariate Regression Models for PCQLI, PedsQL, and CBCL Patient and Parent-Proxy Scores

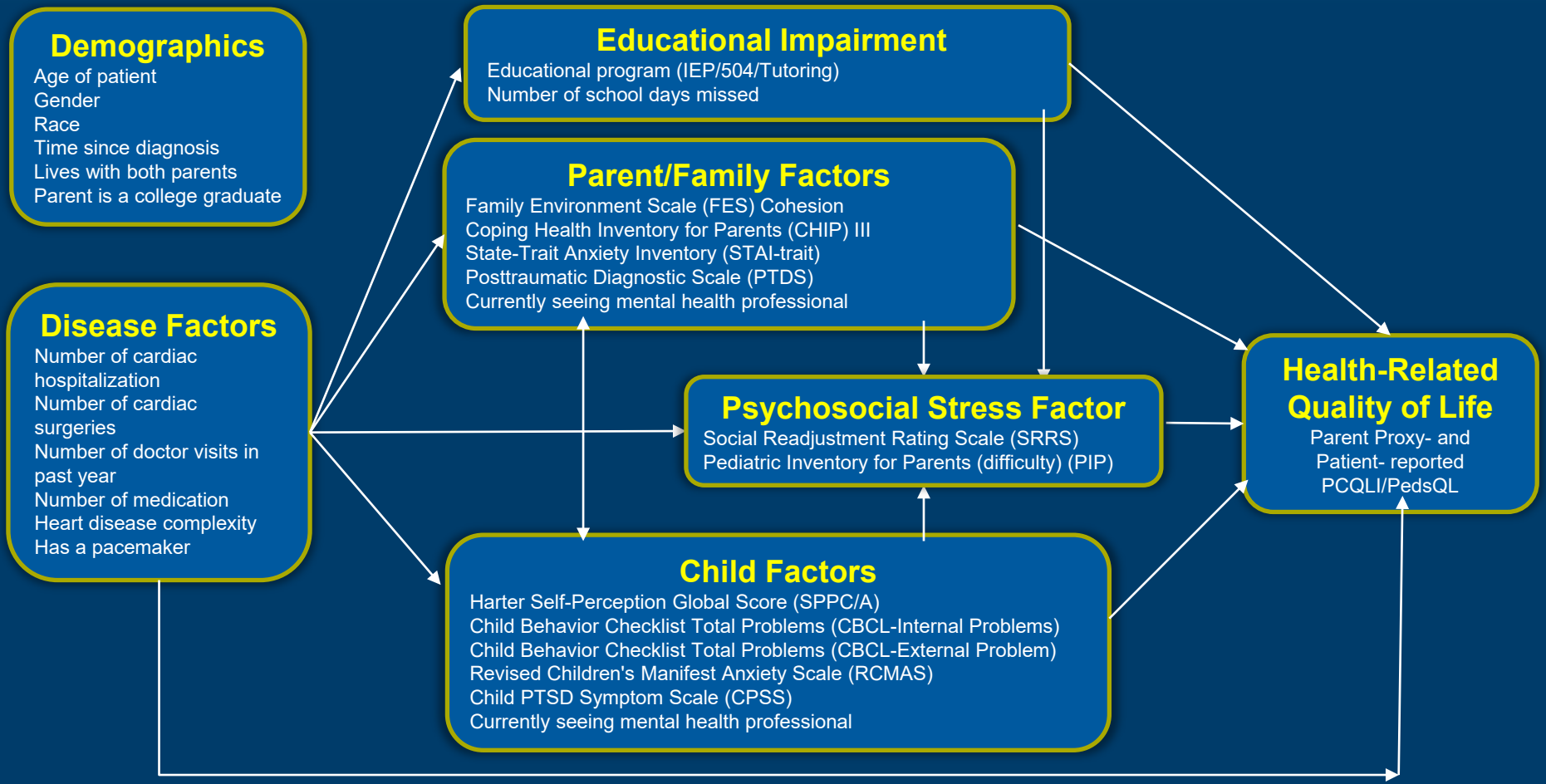
- **PCQLI Total Score**
  - Worse executive functioning (metacognition), gross motor ability, and mood were significantly associated and predicted an additional 47% of the variance (ΔR^2) ($p \leq 0.01$)
- **PedsQL School Functioning and CBCL School Competence Scores**
  - Worse executive functioning (metacognition) was significantly associated for and predicted an additional 37-54% of the variance (ΔR^2) ($p \leq 0.001$)
- Demographics and anatomy (age, gender, income, single ventricle) accounted for only 2-8% ($p < 0.05$) of the variance in QOL scores

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



Wallander & Varni Model of Risk Resilience and Adaptation

Wallander JW, Varni J, *J Ped Psych*, 1998



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Ernst, Marino et al, *Ped Cardiology,* 2018

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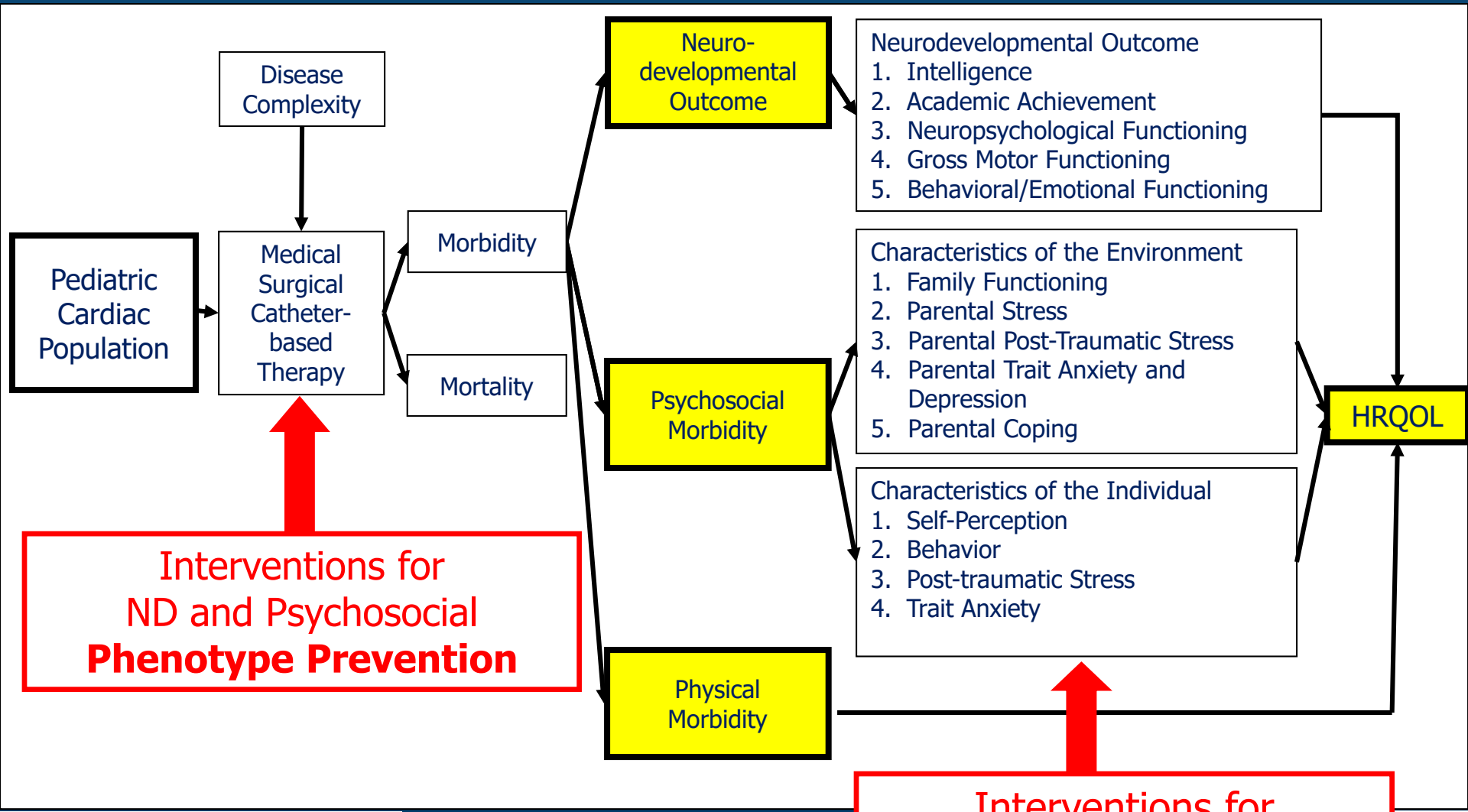
Multivariate Path Model for Impact of Psychosocial Variables on QOL

6 Centers US and UK (n=861)	OUTCOMES	
MODEL	**Patient PCQLI PI**	**Parent PCQLI PI**
	β (p-value)	β (p-value)
Child Factors		
Harter Self-Perception Profile for Children/Adolescents	-0.17 (<.0001)	----------
Child Behavior Checklist Internalizing Problems Score (CBCL)	-0.13 (<.0001)	-0.36 (<.0001)
Revised Children Manifest Anxiety Scale (RCMAS)	-0.34 (<.0001)	----------
Child Post-traumatic Symptom Scale (CPSS)	-0.12 (0.0003)	-0.11 (<.0001)
Parental Factors		
Parental Post-traumatic Stress Disorder (PTSD)	----------	-0.19 (<.0001)
Parent sought Mental Health Treatment	----------	0.07 (0.005)
Parental Stress		
Pediatric Inventory for Parents: Difficulty Score (PIP)	----------	-0.13 (<.0001)
Educational Impairment		
Patient had an Special Educational Program	-0.09 (0.0002)	----------
Number of missed school days in past year	----------	----------
Disease Impact		
Number of Hospitalizations	----------	-0.18 (<.0001)
Number of Surgeries	----------	----------
Number of MD Visits per year	----------	----------
Number of Medications	----------	-0.1 (<.0001)
Increasing Heart Dx Complexity	-0.15 (<.0001)	----------
Demographics		
Patient Age (AGE)	0.22 (<.0001)	----------
R^2	0.50	0.51

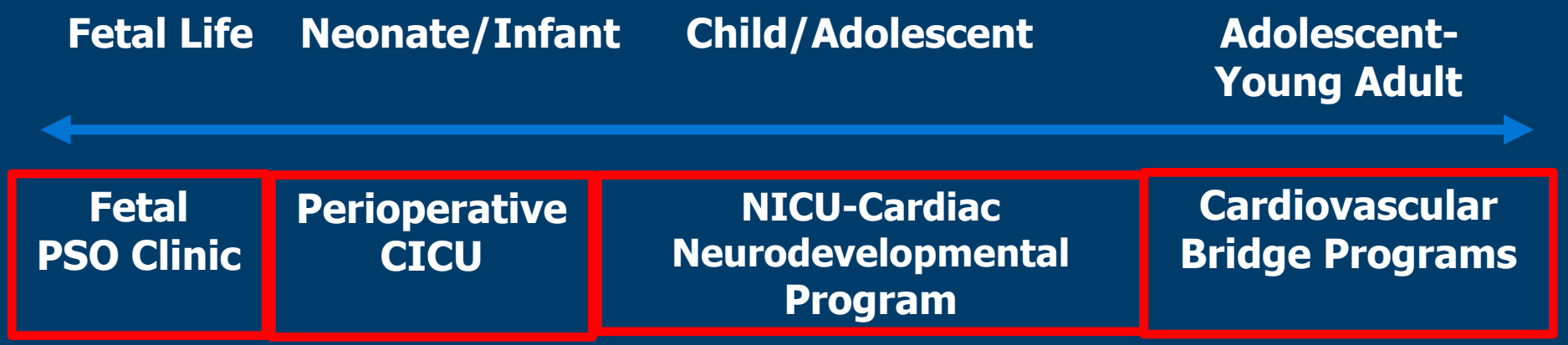
QOL Conceptual Model in Pediatric Heart Disease



"Nurturing" Psychosocial and Neurodevelopmental Resilience to Improve HRQOL

- Neurobehavioral and Psychotherapy in the child and adolescent to minimize impact of social cognition issues, psychiatric issues, autism spectrum and affective disorders, and ADHD
- Psychosocial support to have the child improve their self-perception
- Therapy in the patient and parent to:
  - Prevent and treat PTSD symptomatology prior to invasive procedures, in the ICU, and during follow-up
  - Prevent and treat Anxiety and depression
- Programs to reduce parental stress relative to raising a child with chronic disease

Cardiovascular Care AND Neurodevelopmental and Psychosocial Support Across the Lifespan



Comprehensive Cardiovascular Care AND Neurodevelopmental and Psychosocial Follow-up

AHA Scientific Statement

Neurodevelopmental Outcomes in Children With Congenital Heart Disease: Evaluation and Management

A Scientific Statement From the American Heart Association

This statement has been approved by the American Academy of Pediatrics.

Bradley S. Marino, MD, MPP, MSCE, FAHA, Co-Chair; Paul H. Lipkin, MD; Jane W. Newburger, MD, MPH, FAHA; Georgina Peacock, MD, MPH; Marsha Gerdes, PhD; J. William Gaynor, MD; Kathleen A. Mussatto, PhD, RN; Karen Uzark, PhD, CNP, FAHA; Caren S. Goldberg, MD, MS; Walter H. Johnson, Jr, MD; Jennifer Li, MD; Sabrina E. Smith, MD, PhD; David C. Bellinger, PhD; William T. Mahle, MD, FAHA, Co-Chair; on behalf of the American Heart Association Congenital Heart Defects Committee of the Council on Cardiovascular Disease in the Young, Council on Cardiovascular Nursing, and Stroke Council

Background—The goal of this statement was to review the available literature on surveillance, screening, evaluation, and management strategies and put forward a scientific statement that would comprehensively review the literature and create recommendations to optimize neurodevelopmental outcome in the pediatric congenital heart disease (CHD) population.

Methods and Results—A writing group appointed by the American Heart Association and American Academy of Pediatrics reviewed the available literature addressing developmental disorder and disability and developmental delay in the CHD population, with specific attention given to surveillance, screening, evaluation, and management strategies. MEDLINE and Google Scholar database searches from 1966 to 2011 were performed for English-language articles cross-referencing CHD with pertinent search terms. The reference lists of identified articles were also searched. The American College of Cardiology/American Heart Association classification of recommendations and levels of evidence for practice guidelines were used. A management algorithm was devised that stratified children with CHD on the basis of established risk factors. For those deemed to be at high risk for developmental disorder or disabilities or for developmental delay, formal, periodic developmental and medical evaluations are recommended. A CHD algorithm for surveillance, screening, evaluation, reevaluation, and management of developmental disorder or disability has been constructed to serve as a supplement to the 2006 American Academy of Pediatrics statement on developmental surveillance and screening. The proposed algorithm is designed to be carried out within the context of the medical home. This scientific statement is meant for medical providers within the medical home who care for patients with CHD.

Conclusions—Children with CHD are at increased risk of developmental disorder or disabilities or developmental delay. Periodic developmental surveillance, screening, evaluation, and reevaluation throughout childhood may enhance identification of significant deficits, allowing for appropriate therapies and education to enhance later academic, behavioral, psychosocial, and adaptive functioning. (*Circulation*. 2012;126:1143-1172.)

Marino et al, *Circulation*, 2012

NICU-Cardiac Neurodevelopmental Program

Supporting development throughout childhood and adolescence

Ann & Robert H. Lurie Children's Hospital of Chicago

Monitoring and Intervening on Development

Cardiac ND Follow-Up Clinic at Ann & Robert H. Lurie Children's Hospital of Chicago

- Offered to high risk NICU/Cardiac patients
- Regular developmental evaluations infancy through adolescence
- Psychology and Neuropsychology evaluation and management
- Physical, occupational, speech therapy
- Dieticians, social workers, nurses and APNs
- Care from Developmental Pediatricians and Special Educators

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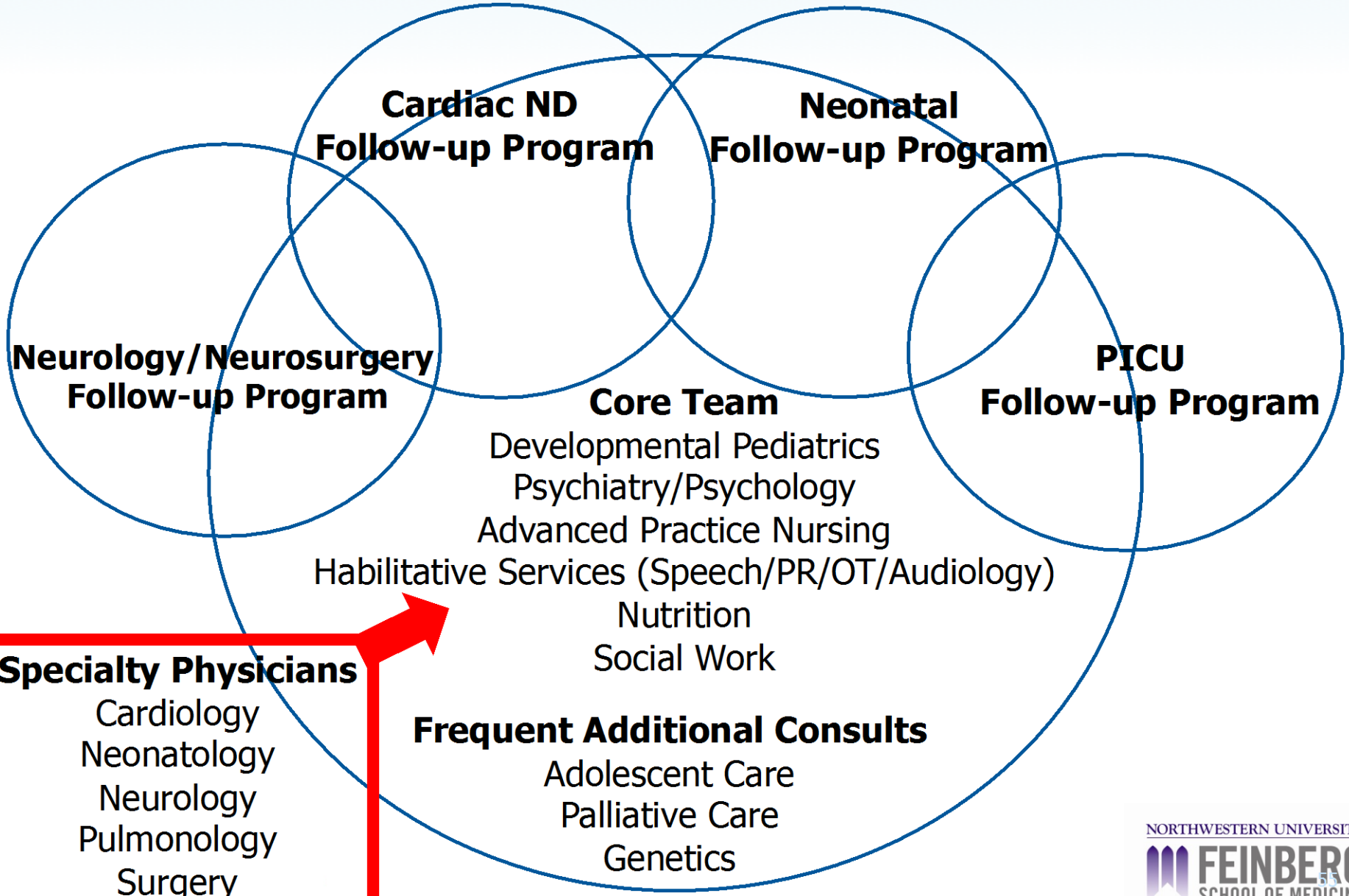
Goals of ND Assessment in Pediatric Heart Disease and CHD Survivors

- To diagnose developmental disability and developmental delay through Surveillance, Screening, Evaluation, and Management
- To put interventions in place to prevent or treat the ND and Psychosocial phenotype noted in the pediatric cardiac population to maximize long-term outcome
  - To maximize health-related QOL
  - To maximize educational attainment
  - To reduce the incidence of anxiety, depression, and stress in family members and family dysfunction
  - To maximize adult transition and ACHD outcome

Development of a Singular Developmental Medical Home for High-Risk Children



CNOC

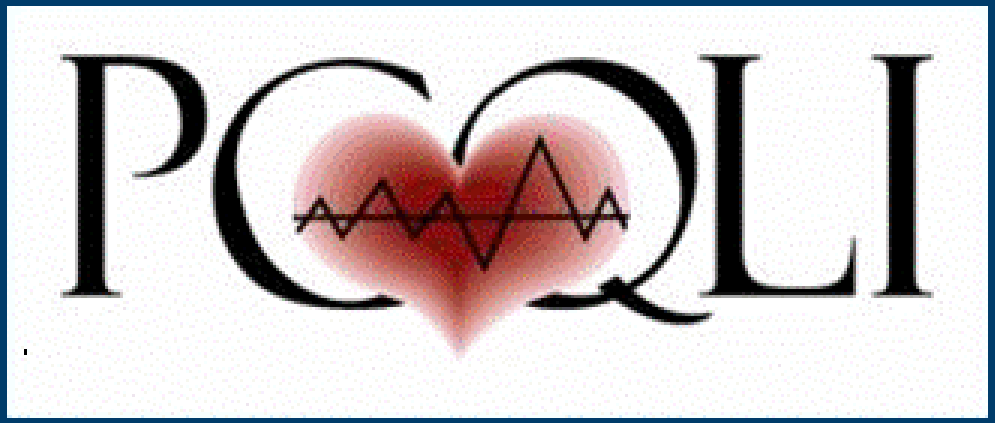
Cardiac Neurodevelopmental Outcome Collaborative

Summary

- QOL in CHD survivors is lower than heart-healthy children and worsens with increasing disease complexity
- There is wide variation in QOL among CHD survivors
- QOL in CHD survivors is strongly associated with neurodevelopmental and psychosocial deficits
  - Executive dysfunction
  - Worse self-perception
  - Internalizing Problems (Depression/Withdrawal)
  - Affective disorders
  - ADHD
  - Increased Post-traumatic Stress Symptomatology in patients and parents
  - Trait Anxiety in patients and parents
  - Parental Stress

Summary

- Interventions are needed to prevent and treat neurodevelopmental and psychosocial issues in complex CHD survivors and their families to improve the patients' long-term HRQOL
- High-risk cardiac patients per AHA/AAP Guidelines should be referred directly for formal developmental and medical evaluations to assess the neurodevelopmental AND psychosocial status of the patient and the parent to maximize HRQOL
- These interventions may be performed during the prenatal period, in the ICU, and by integrated multi-disciplinary Cardiac Neurodevelopmental Programs



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