Beyond the Heart: Tackling the Social, Emotional, and Educational Challenges of Kids with Fontan Hearts

May 17, 2022

Single Ventricle Program at Lucile Packard Children's Hospital Stanford

Lucile Packard Children's Hospital Stanford

FONTAN OUTCOMES NETWORK

Case Presentation

- 15-year-old male with DORV w/ mitral stenosis s/p Y-graft Fontan at age 3.
- Cath at age 5: Fontan pressure 16 mmHg. LPCWp 10-11mmHg; RPCWp 9mmHg
- Cardiac MRI: TR (regurgitant fraction 15%), and decreased RV function (EF 25%).
- Echo: normal function, mild TR
- On ACEi, ASA
- Age 12 (clinic visit): C/O trouble sleeping, exercise intolerance, fatigue, and circumoral cyanosis with exertion.
- Endorsed anxiety, depression and was also really struggling with school.
- Difficulty finding a consistent therapist to treat anxiety and depression.

- ND testing (age 12): Met criteria for ADHD, Executive Function Deficit, and Major Depressive Disorder.
- Severely decreased exercise capacity with O2 sats 87-92%. Cath recommended.
- Sleep study c/w mild obstructive sleep apnea. Started oxygen, and it helped improve sleep
- Referred to Endocrine for obesity and glucose intolerance
- Repeat Cath: Fontan pressure 18, TPG 3-4, PCW 14-16, PVR 1.2-1.6, coiled VV collaterals (sats improved).
- Recommended Sildenafil/Tadalafil but not started due to insurance issues.

- 2 years later (age 14): diagnosed with PLE in setting of diarrhea, low albumin, and elevated stool alpha 1 antitrypsin.
- Repeat Cardiac MRI showed mild to moderate TR (regurgitant fraction 22-28%), and mildly depressed RV systolic function (EF 47%).
- Repeat Cath: Fontan pressures 18, LVEDP 13-14, and PVR 4.8 WU.
- PH team and Heart Failure team referral.
- Started on budesonide, Lasix, Aldactone, Sildenafil (stopped recently due to HA).
- Currently:
  - Ongoing school support from SVP education liaison
  - Ongoing support from SVP MHC to address issues surrounding anxiety and depression.
  - Ongoing follow up with Heart Failure Team at LPCH.

MENTAL HEALTH IN SVHD

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AHA SCIENTIFIC STATEMENT

Psychological Health, Well-Being, and the Mind-Heart-Body Connection: A Scientific Statement From the American Heart Association

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Table 1. Effect Estimates for Associations of Negative Psychological Factors With Cardiovascular Events and Conditions

Negative psychological factors	Parameter/ end point	Effect estimates (95% CI)
Depression	Incident MI	RR, 1.30 (1.22–1.40)[42]
	Incident CHD	RR, 1.30 (1.18–1.44)[42]
	Stroke	RR, 1.45 (1.31–1.61)[45]
	Obesity	RR, 1.37 (1.17–1.48)[49]
	Hypertension	RR, 1.42 (1.09–1.86)[51]
	Diabetes	RR, 1.32 (1.18–1.47)[52]
Anxiety	CVD mortality	RR, 1.41 (1.13–1.76)[39]
	Incident CHD	RR, 1.41 (1.23–1.61)[39]
	Coronary artery spasm	RR, 5.20 (4.72–5.40)[40]
	Incident stroke	RR, 1.71 (1.18–2.50)[39]
	Heart failure	RR, 1.35 (1.11–1.64)[39]
Work-related stress	Incident CVD events	RR, 1.4 (1.2–1.8)[18]
Any-cause stress	Incident CHD/CHD mortality	RR, 1.27 (1.12–1.45)[19]
PTSD	Incident CHD	RR, 1.61 (1.46–1.77)[22]
Social isolation and loneliness	Incident CVD events	RR, 1.5 (1.2–1.9)[18]
Pessimism	CHD mortality	OR, 2.17 (1.21–3.89)[50] (highest vs lowest quartile)
Anger and hostility	Incident CHD	HR, 1.19 (1.05–1.35)[33]
	Recurrent CHD	HR, 1.24 (1.08–1.42)[33]

CHD indicates coronary heart disease; CVD, cardiovascular disease; HR, hazard ratio; MI, myocardial infarction; OR, odds ratio; PTSD, posttraumatic stress disorder; and RR, risk ratio.

MENTAL HEALTH IS HEART HEALTH

American Heart Association, 2021

Psychological Health, Well-Being, and the Mind-Heart-Body Connection

ANXIETY

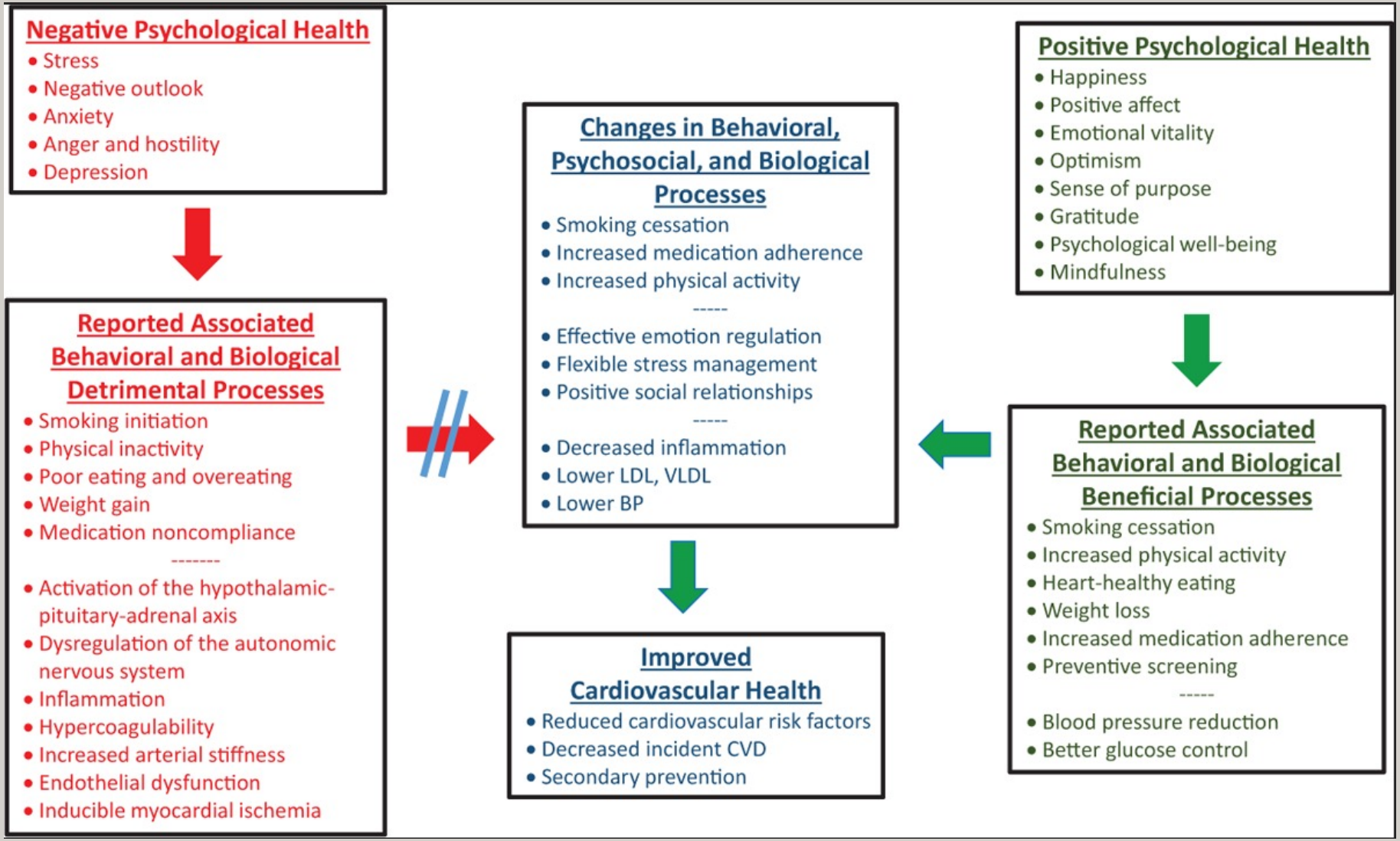
CVD Mortality Risk = Increase by 40%

SOCIAL ISOLATION

CVD Adverse Events Risk = Increase by 50%

NON-ADHERENCE

Strong associations with treatment/medication non-adherence


Negative Psychological Health
• Stress
• Negative outlook
• Anxiety
• Anger and hostility
• Depression
Reported Associated Behavioral and Biological Detrimental Processes
• Smoking initiation
• Physical inactivity
• Poor eating and overeating
• Weight gain
• Medication noncompliance

• Activation of the hypothalamic-pituitary-adrenal axis
• Dysregulation of the autonomic nervous system
• Inflammation
• Hypercoagulability
• Increased arterial stiffness
• Endothelial dysfunction
• Inducible myocardial ischemia
Changes in Behavioral, Psychosocial, and Biological Processes
• Smoking cessation
• Increased medication adherence
• Increased physical activity

• Effective emotion regulation
• Flexible stress management
• Positive social relationships

• Decreased inflammation
• Lower LDL, VLDL
• Lower BP
Improved Cardiovascular Health
• Reduced cardiovascular risk factors
• Decreased incident CVD
• Secondary prevention
Positive Psychological Health
• Happiness
• Positive affect
• Emotional vitality
• Optimism
• Sense of purpose
• Gratitude
• Psychological well-being
• Mindfulness
Reported Associated Behavioral and Biological Beneficial Processes
• Smoking cessation
• Increased physical activity
• Heart-healthy eating
• Weight loss
• Increased medication adherence
• Preventive screening

• Blood pressure reduction
• Better glucose control

Cardiology in the Young

cambridge.org/cty

Review

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Psychological functioning in paediatric patients with single ventricle heart disease: a systematic review

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Abstract

Background: Patients with single ventricle heart disease are living into adulthood due to medical and surgical advancements but have significant physical comorbidities and an increased risk for psychological comorbidities compared to healthy subjects or those with other CHD diagnoses. This study aimed to systematically review psychological functioning in paediatric single ventricle heart disease. *Methods:* Literature was searched using PubMed, Embase, PsycInfo, CINAHL Complete and Scopus. Peer-reviewed articles that included patients ages 0–25 years with single ventricle heart disease, and quantitative measures of psychological outcomes were included. Meta-analysis using a fixed-effect model was conducted for internalising and externalising t-scores, utilised by the Achenbach Child Behavior Checklist. *Results:* Twenty-nine records met the criteria for inclusion. 13/24 studies demonstrated increased risk for internalising disorders, such as anxiety/depression; 16/22 studies demonstrated risk for externalising

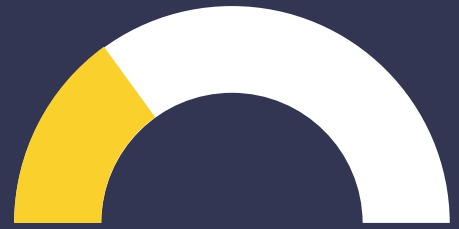
MENTAL HEALTH IN SVHD

24

STUDIES ON INTERNALIZING RISK (ANXIETY, DEPRESSION)

22

STUDIES ON EXTERNALIZING RISK (BEHAVIOR, ADHD)



30% OF STUDIES CONDUCTED IN <5 YEARS

ANXIETY, DEPRESSION, ADHD

MENTAL HEALTH IN SVHD

65% OF YOUTH WITH SVHD WITH MENTAL HEALTH DX

5x

FIVEFOLD INCREASED RISK OF ANXIETY DX

NEARLY 6X INCREASED RISK FOR ADHD DX

DeMaso DR, Calderon J, Taylor GA, Holland JE, Stopp C, White MT, Bellinger DC, Rivkin MJ, Wypij D, Newburger JW. Psychiatric Disorders in Adolescents With Single Ventricle Congenital Heart Disease. Pediatrics. 2017 Mar;139(3):e20162241. doi: 10.1542/peds.2016-2241. Epub 2017 Feb 1. PMID: 28148729; PMCID: PMC5330395

MENTAL HEALTH IN SVHD

SVHD	DTGA	TOF
COMPARING TEENS W/ SVHD, DTGA AND TOF	ANXIETY, ADHD, DISRUPTIVE BEHAVIOR DX HIGHER IN SVHD	ANXIETY DX HIGHER IN SVHD

Cassidy AR, Bernstein JH, Bellinger DC, Newburger JW, DeMaso DR. Visual-spatial processing style is associated with psychopathology in ado?lescents with critical congenital heart disease. Clin Neuropsychol 2019; 33: 760–778.

WHERE TO GO FROM HERE?

RESEARCH

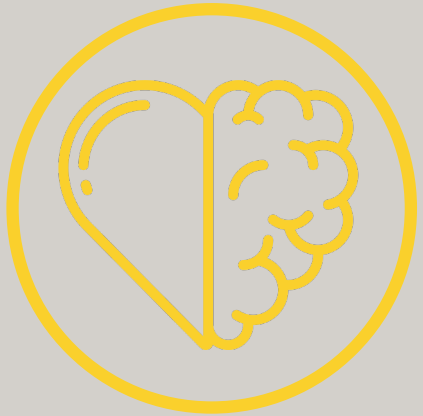
- Larger Samples
- Risk and Protective Factors
- SDOH

TRAINING

- Cardiology clinicians on the "front lines"

INTERVENTION

- Integrated mental health care
- Prevention efforts
- Treatment studies



INTEGRATE

- Mental health AS heart health
- Validate
- Value
- Advocate

Why neurodevelopmental surveillance of people with SVHD?

- Most average or better intelligence and strong verbal skills
- More likely to have behavioral, developmental, and/or learning challenges
- The following are specific areas to monitor:
  - Anxiety and/or Depression
  - Attention Deficit Hyperactivity Disorder (ADHD) –> 3-4x more common CHD
  - Executive Functioning Deficits - weak working memory, planning, organization, inhibition, cognitive flexibility
  - Visual-Spatial reasoning
  - Motor development

Mental health support in school

- General education support – most districts partner with community-based non-profits to provide free, individual counseling services on school sites. These services typically require a referral from a school staff member and parents to sign a waiver to allow participation.
- Special education support – Students with behavioral health issues that impact ability to learn and access the school curriculum are eligible to receive the mental health services needed to benefit from their education program.
  - ERMHS (Educationally Related Mental Health Services)– completed by the district.
  - Allowable services under ERMHS
    - Individual and/or group counseling
    - Parent counseling
    - Social work services
    - Psychological services
    - Behavior intervention services (including 1:1 aides)
    - Residential Placement

Mental health resources

- Private insurance – in or out of network
  - packet for families
  - Insurance exception / appeal
- Internal hospital clinicians
- School-based (ERMHS or general education)
- Private foundations (ie, Ollie Hinkle Heart Foundation)
  - www.theohhf.org -> Ollie's Branch; Free Mental Health Services
  - St. Louis, Kansas City, Chicago, SF Bay Area (in progress)
  - "heart warrior", parents, grandparents, siblings, pre-natal

Creative solutions to "bridge the gap":

- Can a social worker provide bridging services?
- Create relationships with adult clinic resources
- Mindfulness, meditation, yoga practice
- Workbooks for depression and anxiety

Beyond the Heart: Cardiologist role

- Our "Why": Thriving people, not functioning organ
- "I" to "We" : Team based care
  - More effective referrals & linkage
  - Feels better for MD & parent
- Heart Center- Program Development & Funds Flow
  - Collaborative program proposals, stepwise support, academic "nest
- Cardiologist : Trusted source, provide context & prioritization interdisciplinary teammates
- Check-in: "How are you doing?" ABP Roadmap Project
  - Longterm relationship
- " Flip the Script" : Our words matter, direct impact of positivity, be open to what may yet unfold
- Recognize potential immediate impact of evidence-based interventions "beyond the heart"