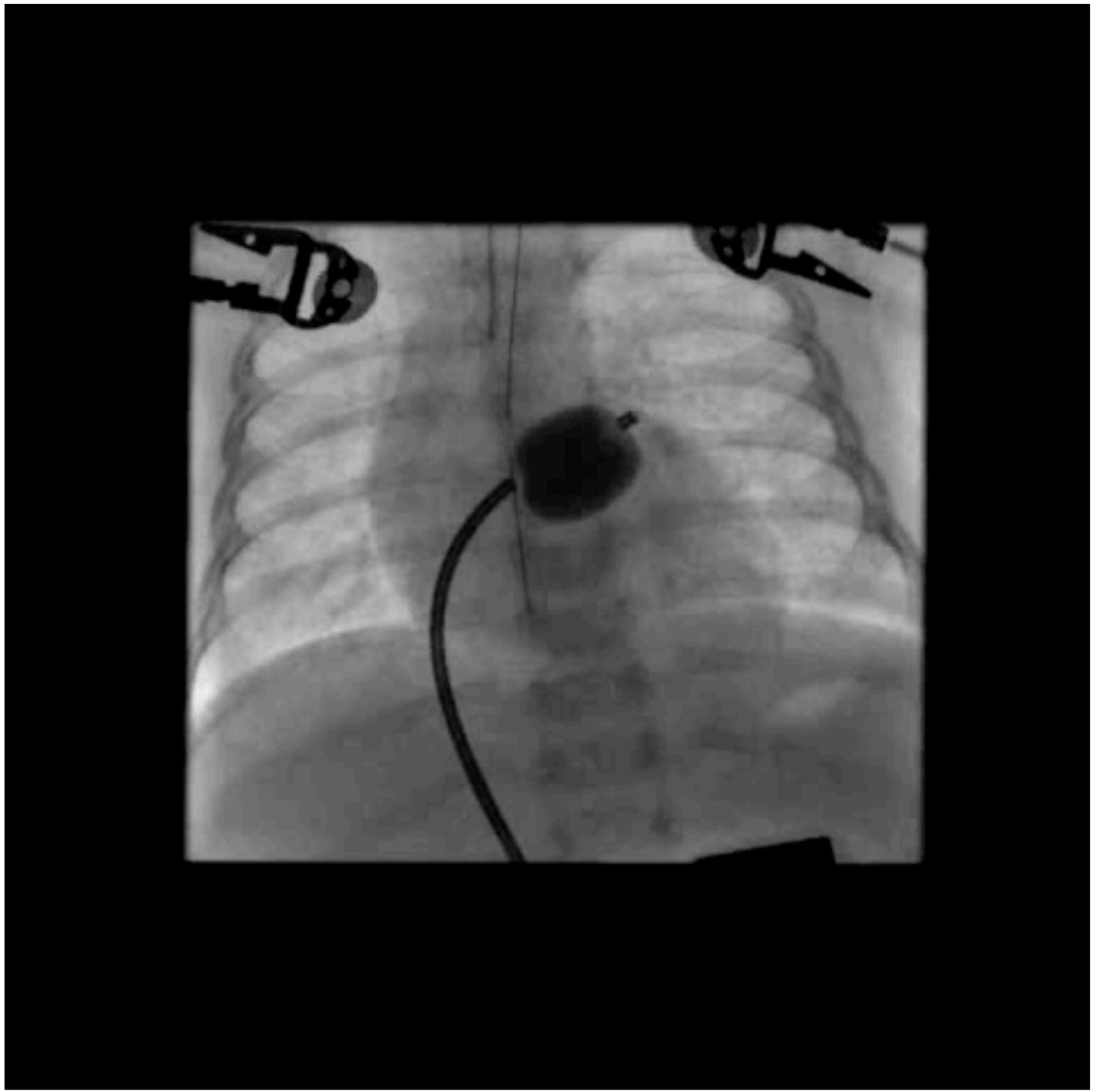
Myths and Misconceptions in Pediatric Cardiology

May 16, 2015

No disclosures

Myth #1

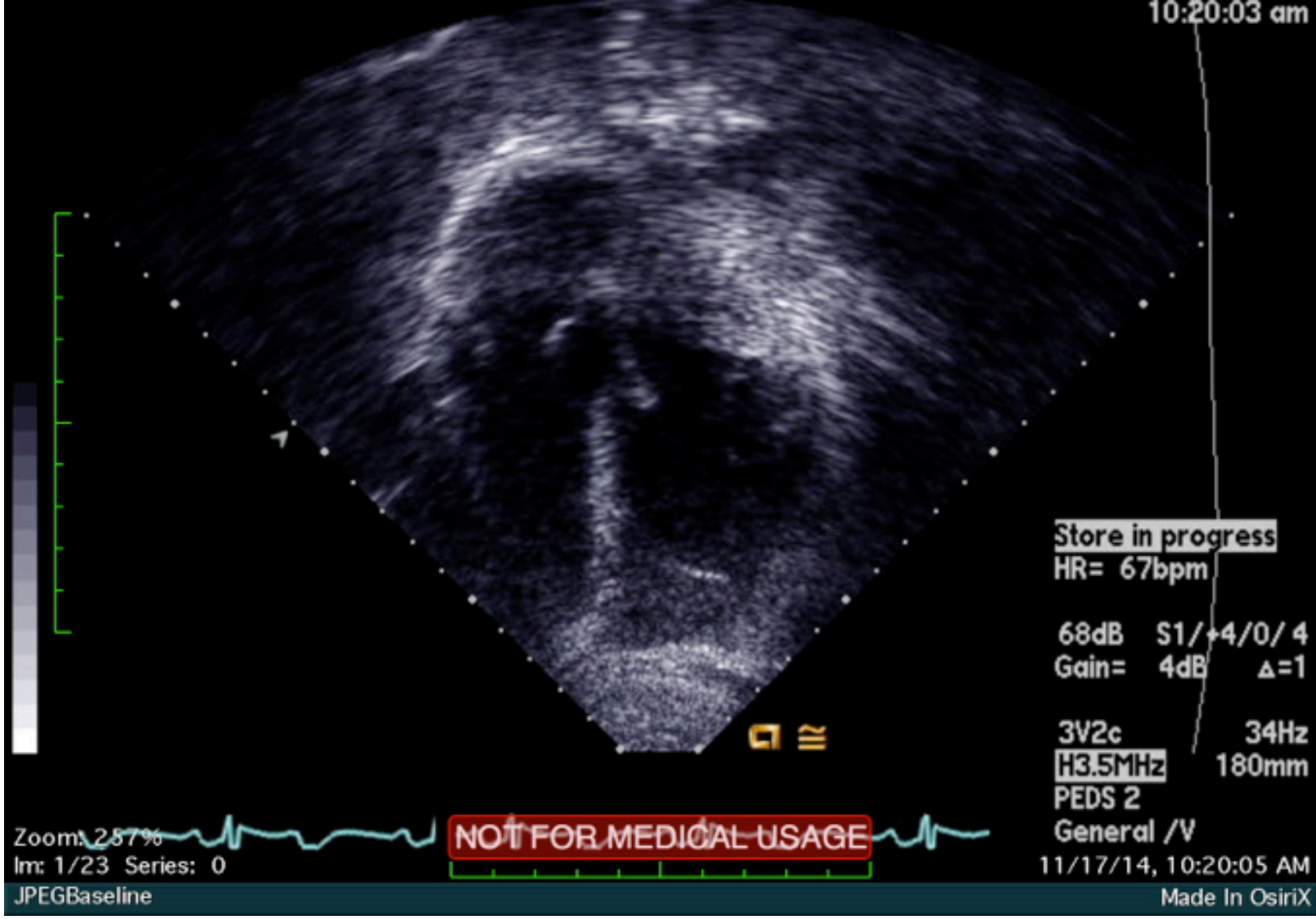
- Early balloon atrial septostomies were performed using Foley catheters



Myth #2

- The first seven patients who underwent the arterial switch operation for D-TGA died

- Patients with L-TGA can survive into their 80s without surgical correction



Myth #3

Myths….or reality?

A. Early balloon atrial septostomies were performed using a foley catheter

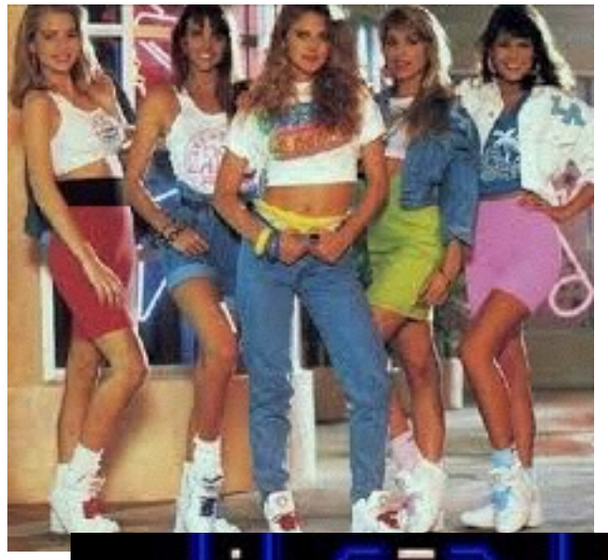
B. The first 7 patients who underwent arterial switch procedure for D-TGA died in the peri-operative period

C. Patients with L-TGA can live normal full lives- without an operation

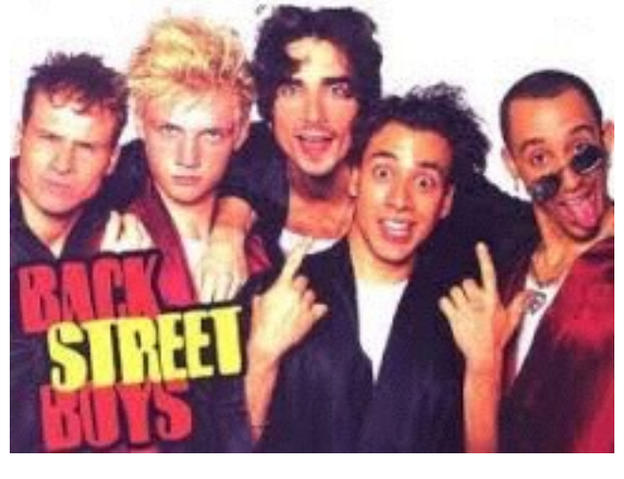
Journey through Cardiology and CV surgery

Evolution of Pediatric CV Surgery

Evolution of Pediatric CV Surgery



Evolution of Pediatric CV Surgery



Evolution of Pediatric CV Surgery

Myths?

- Patients born with HLHS have a poor prognosis
- Physical activity in patients with congenital heart disease is dangerous
- Patients post surgical repair of congenital heart disease should not get pregnant
- Patients post surgical repair of congenital heart disease have morbidities and poor neurodevelopmental outcomes

What is Hypoplastic Left Heart Syndrome?

- Inadequate development of the left side of the heart
  - Mitral valve atresia or stenosis- Primary cause?
  - Aortic valve atresia or stenosis
  - Hypoplasia of the aorta
    - Ascending, transverse, coarctation
  - Hypoplasia of the left ventricle

Statistics on HLHS

- 5-7% of all Congenital heart disease
- Lethal if untreated
- Extracardiac anomalies uncommon- 10-15%
- Chromosomal abnormalities uncommon- 10-15%

What is the big deal with HLHS?

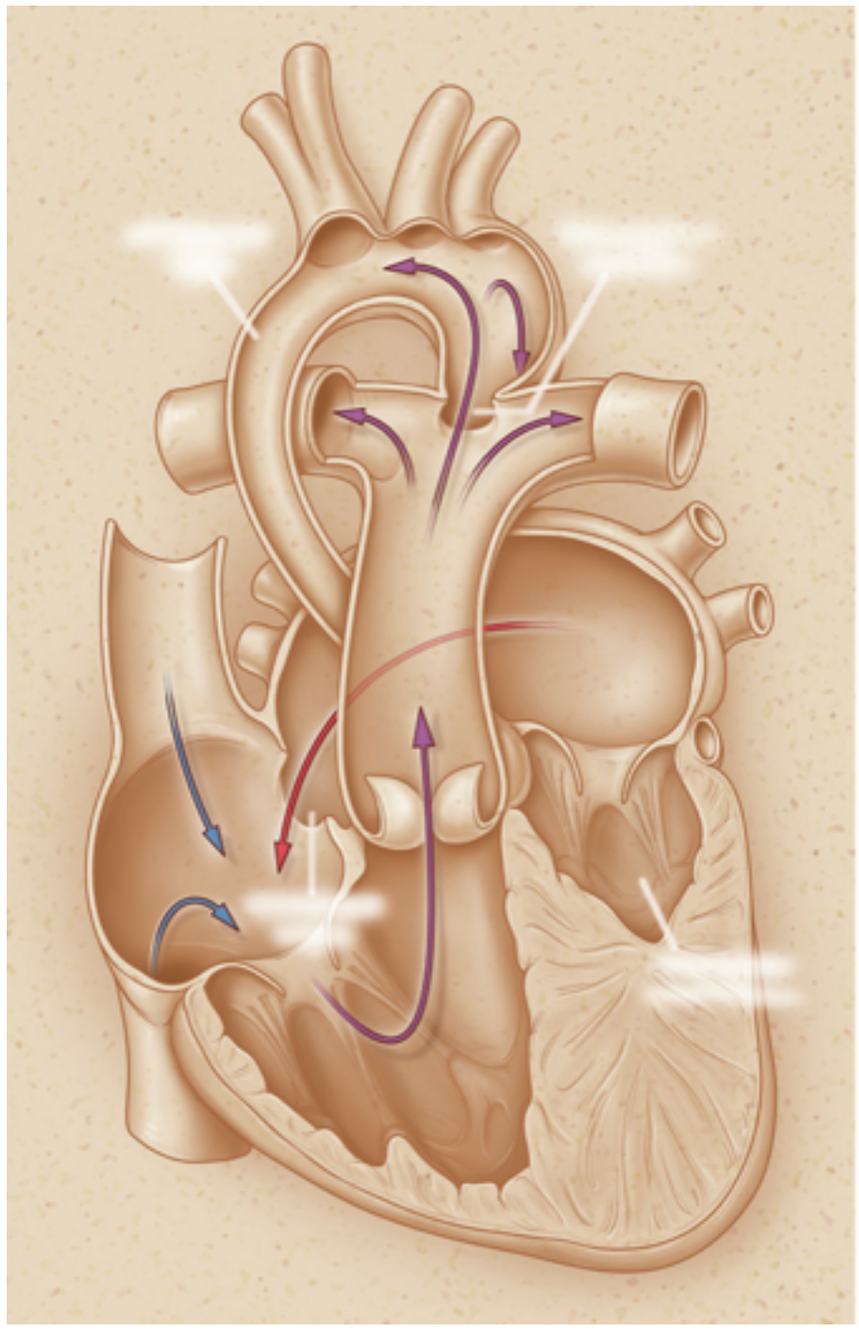
- HLHS treatment/management/outcomes is the Benchmark for Cardiology and CV surgery programs
    - "Benchmark" surgery for STS database
        - **Defines your performance as a center**
- "If you do this well, you can do everything well"
- Lessons learned in the operative and preoperative management of HLHS can be applied to every other surgery

Why HLHS?

- Most benchmarking typically uses discharge mortality and morbidity data
- Long term survival requires
  - Navigation of multiple surgeries, re-do operations, catherizations
  - Requires advanced imaging
  - Requires sophisticated cardiology/surgical decision making
- Requires a large cohesive team

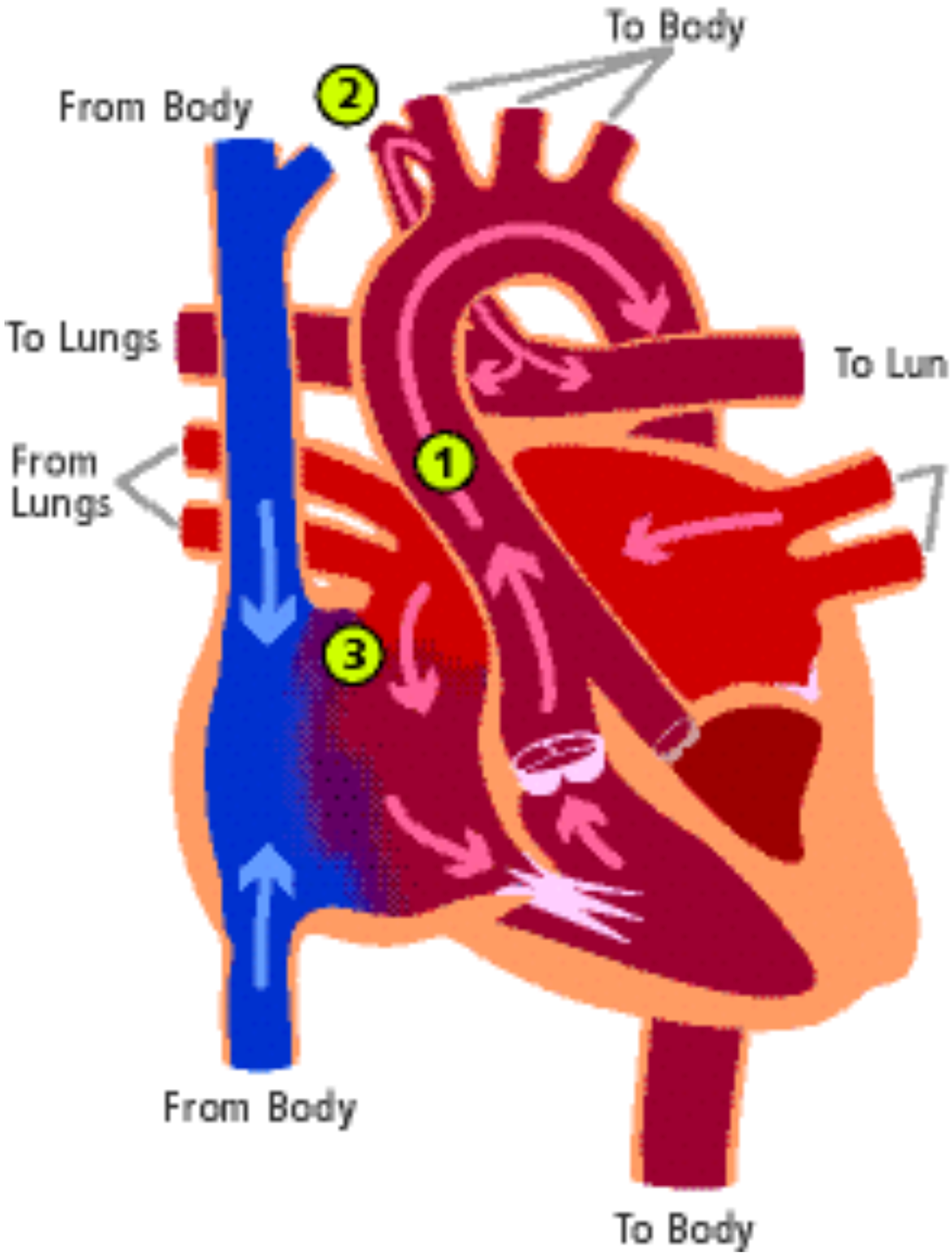
HLHS

- The heart is structurally normally related
- Follow the blood flow
- Patients are started on PGE to maintain ductal patency



Norwood procedure

- Often done ages 3-7 days of life
- Pulmonary artery anastomosed to the small ascending aorta
- Aortic arch is augmented
- Shunt provides pulmonary blood flow
- Removal of the atrial septum allows blood to reach the right side of the heart



Norwood results

- Early 1990s- Early survival 50%
- 1990s- 70% survival to discharge
- 2010s- 80-84% survival to discharge
- Highest performing centers- >90%

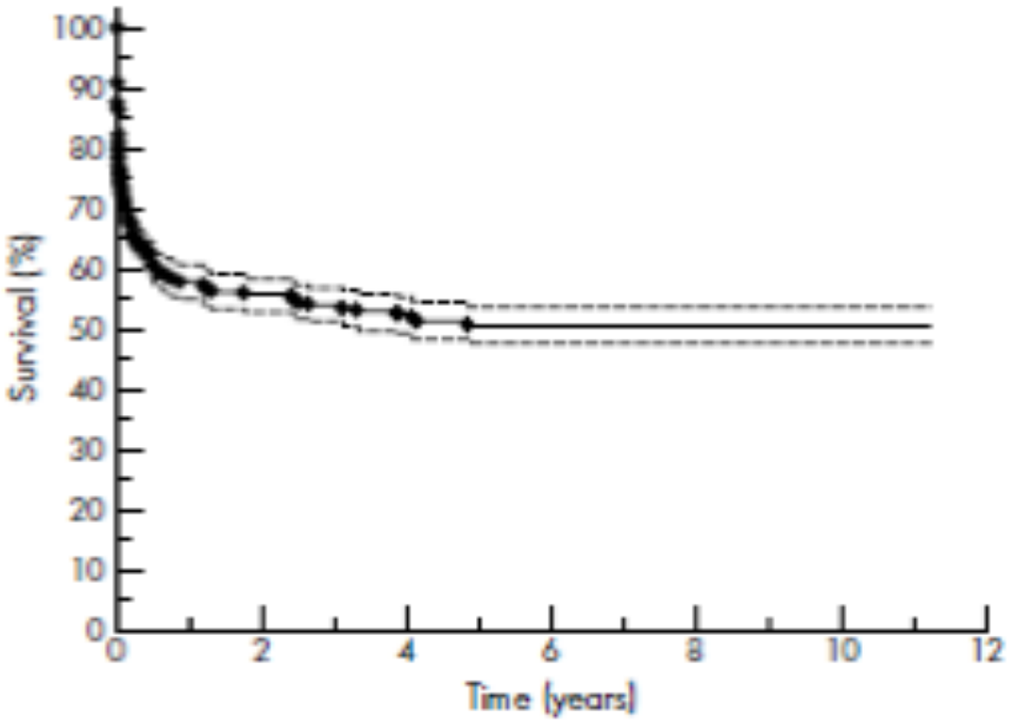
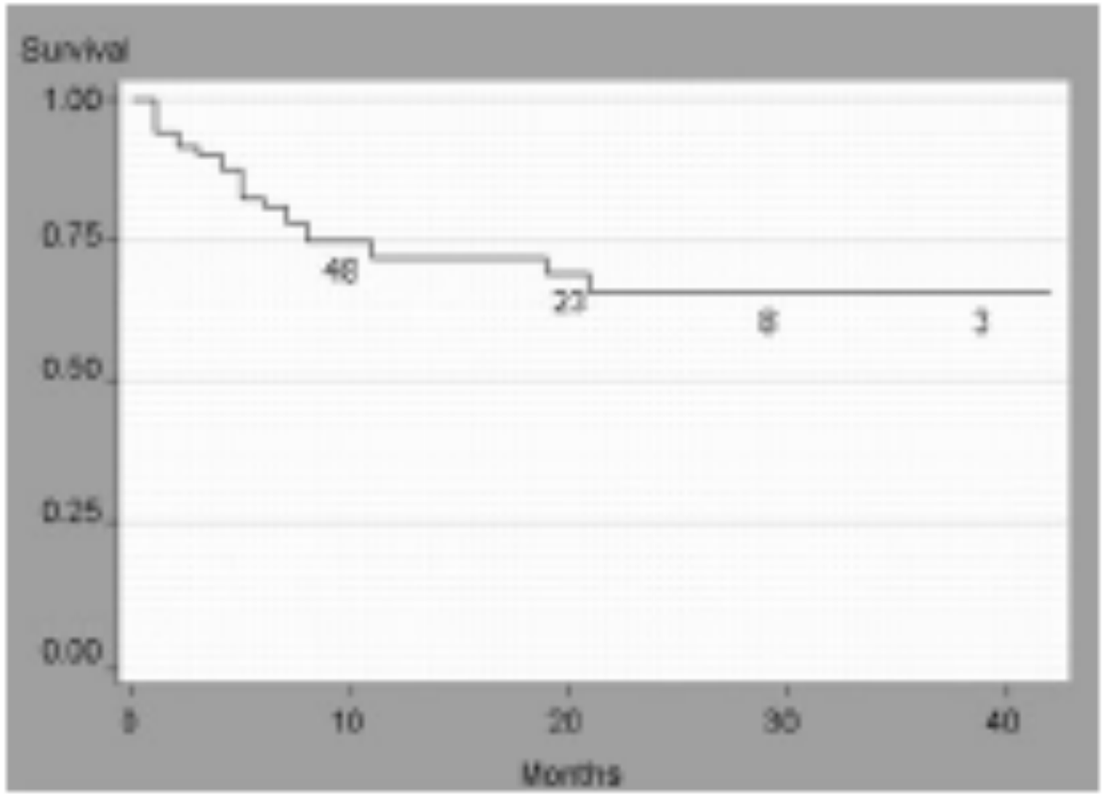
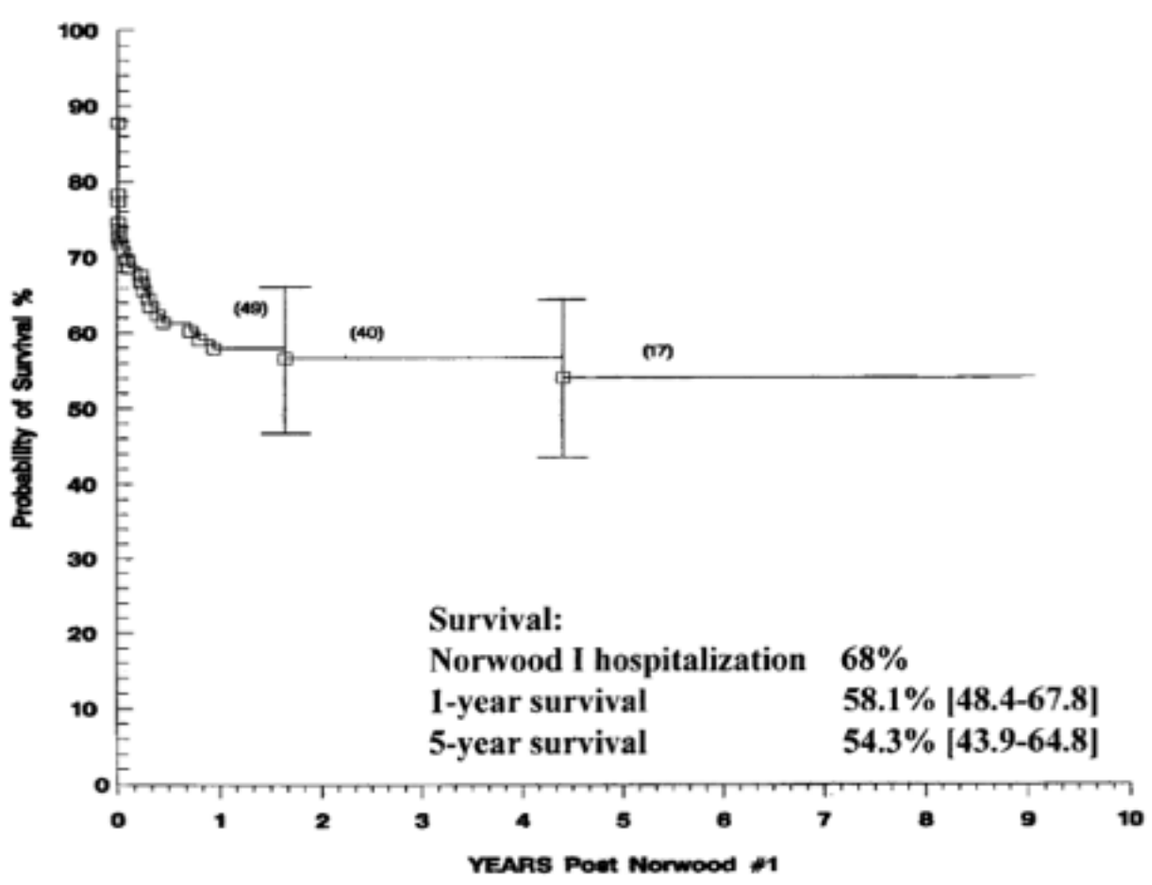


Figure 1 Actuarial survival after staged surgical management of the hypoplastic left heart syndrome. Data are mean





Our institutional data- 10 years

- >90 consecutive newborns
- Operation on median DOL #6 (1-42)
- 21% high risk!!!
- 34% female
- Median BWt 3.17 Kg (1900-4500g)

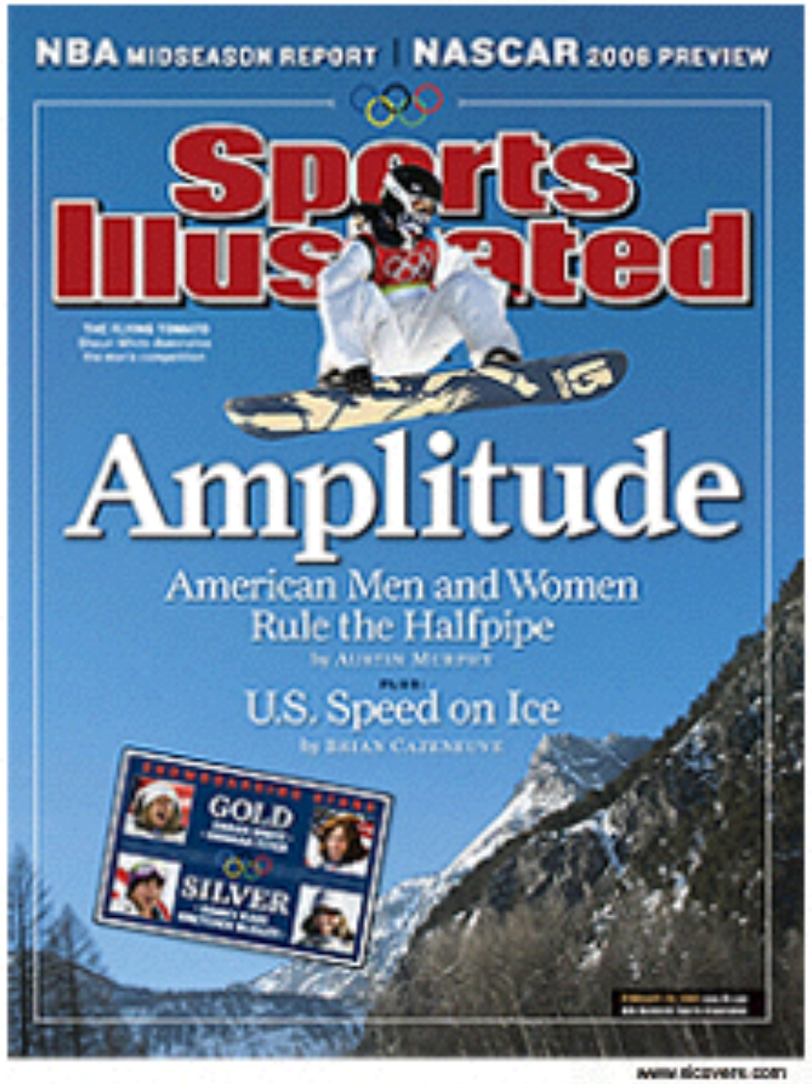
Our institution
10 year experience

- Hospital discharge survival- 89%
  - Standard risk- 91%
  - High risk survival- 74%
    - Over last 5 years- 92%
- ECMO- 17%
  - Poor prognosis- 53%
  - More common in 5-10 years range

HLHS

- 90% of patients surviving to discharge
- Hospital discharge survival should be above the STS “benchmark” of 83-84%
- 10 year survival is now the expectation- 60-80%
- Outcomes are very good even for the most difficult of surgeries


Rolling Stone
SHAUN WHITE
Big Air, Big Hair & the Killer Inside
ROCK GOD, INTERRUPTED
Billy Corgan's Missing Years
2010'S BIGGEST ALBUMS
THE LOST 'EXILE' SESSIONS
THE COP'S DIRTY WAR AGAINST OBAMA


NBA MIDSEASON REPORT | NASCAR 2006 PREVIEW
Sports Illustrated
Amplitude
American Men and Women Rule the Halfpipe
by Austin Murphy
plus:
U.S. Speed on Ice
by Brian Cazeneuve
GOLD
SILVER

Is exercise dangerous or unhealthy in patients with CHD?

AHA Scientific Statement

Promotion of Physical Activity for Children and Adults With Congenital Heart Disease

A Scientific Statement From the American Heart Association

Patricia E. Longmuir, PhD; Julie A. Brothers, MD; Sarah D. de Ferranti, MD, MPH;
Laura L. Hayman, PhD, RN, FAHA; George F. Van Hare, MD; G. Paul Matherne, MD, MBA, FAHA;
Christopher K. Davis, MD, PhD; Elizabeth A. Joy, MD, MPH, FACSM;
Brian W. McCrindle, MD, MPH, FAHA, Chair; on behalf of the American Heart Association
Atherosclerosis, Hypertension and Obesity in Youth Committee of the Council on Cardiovascular
Disease in the Young

Circulation. 2013;127:2147-2159

Promotion of Physical Activity for Children and Adults with CHD

- Published by AHA
- Title uses “Promotion” instead of “Restriction”
- “Counseling of pts with CHD should emphasize the importance of daily physical activity and decreasing sedentary behavior”
- Limited research on patients with CHD and exercise
    - Limited data on whether restriction is necessary
    - Patients with CHD tend to be sedentary
    - Patients with CHD have lower levels of physical activity, higher incidence of obesity compared to healthy children
- “Counseling to encourage daily participation in physical activity should be a core component of every patient encounter”

Exercise in CHD

- Moderate intensity exercise
  - Physical activity incorporates all types of physical movement, not solely “exercise”
    - Health benefits of moderate exercise are within reach of nearly all patients
- Only rare patients need to be restricted from activity with family and friends
- **For most, physical activity can be unlimited and should be strongly promoted**

Current guidelines

- Healthy adults
  - Muscle strengthening 2 or more days each week
  - 75 minutes of vigorous or 150 minutes of moderate activity each week
- Children 60 minutes or more of daily physical activity

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BETHESDA CONFERENCE REPORT

36th Bethesda Conference: Eligibility Recommendations for Competitive Athletes With Cardiovascular Abnormalities

Barry J. Maron, MD, FACC, *Conference Co-Chair*
Douglas P. Zipes, MD, MACC, *Conference Co-Chair*

This Conference, sponsored by the American College of Cardiology Foundation, was held at Galerie 1, New Orleans Marriott, New Orleans, Louisiana, on November 6, 2004. Please refer to the appendix of each Task Force report for author disclosure information.

Benefits of physical exercise

Benefits of physical exercise

- Physiological improvements in skeletal muscle function
- Vascular function
- Immune system function
- Obesity prevention
- Complex psychological, cognitive and social function

Physical activity promotion

- Daily physical activity is a cornerstone of CV health promotion

- We are doing a disservice to our patients if we are not encouraging them to exercise regularly

- Providers feel ill prepared to promote healthy lifestyles

    - May be lacking in skills, knowledge, resources, time needed to implement physical activity promotion

Physical activity promotion

- "Effective behavior change counseling enables the individual to identify personally relevant goals and use intrinsic motivation to develop and implement action plans that are meaningful and appropriate"

- The clinician can bring to the fore the patient's intrinsic motivation for physical activity

- Patient centered

- Motivational interviewing

Motivational interviewing

- RULE (**R**esist, **U**nderstand, **L**isten, **E**mpower)
  - **R**esist the “righting reflex”
    - Natural tendency of providers to overtly support the “good” or desired behavior
      - Intended to be helpful, these comments turn the patient’s focus toward justification of the negative behavior (defensiveness)

Motivational interviewing

- RULE (**R**esist, **U**nderstand, **L**isten, **E**mpower)
  - **U**nderstand and explore the patient's own motivation for change and perception of the current situation
  - Why do you want to be more active?
  - If you get tired during exercise, what type of exercise are you doing?

Motivational interviewing

- RULE (**R**esist, **U**nderstand, **L**isten, **E**mpower)
  - **L**isten with empathy, and validate the patient's feelings attend to non-verbal stimuli
  - Perhaps you can't exercise after work- would you have time before work? After school?

Motivational interviewing

- RULE (**R**esist, **U**nderstand, **L**isten, **E**mpower)
  - **E**mpower the patient encouraging hope and optimism that they can achieve their goal
  - I know you want to get fit, but maybe we haven't had the best plan for you yet, together we will figure out a plan to get you exercising

Most common causes of sudden death

- Hypertrophic cardiomyopathy- 35-40%
- Coronary artery anomalies- 25%
- LVH- 10%
- Dilated Cardiomyopathy- 3-5%
- Congenital heart disease- 5% (**Rare with exercise**)
- Long QT syndrome- 3%
- Myocarditis- 3%
- Arrhythmogenic right ventricular dysplasia- 2%
- Commotio Cordis- < 1%

Which patients DO we limit?

- Ventricular dysfunction- Significantly decreased ejection fraction
    - Risk of ventricular tachyarrhythmia
    - Restricted to recreational sports, no competitive sports
- Aortic dilation
    - Risk of aneurysm or dissection
    - Moderate intensity exercise is safe
- Hypoxia
    - Largely self limit
- Anticoagulation
    - Avoid sports where body contact is a routine aspect of the sport
- Implanted devices
    - Avoid sports with contact

Counseling patients to promote physical activity

- Ask about exercise and encourage all families at every visit

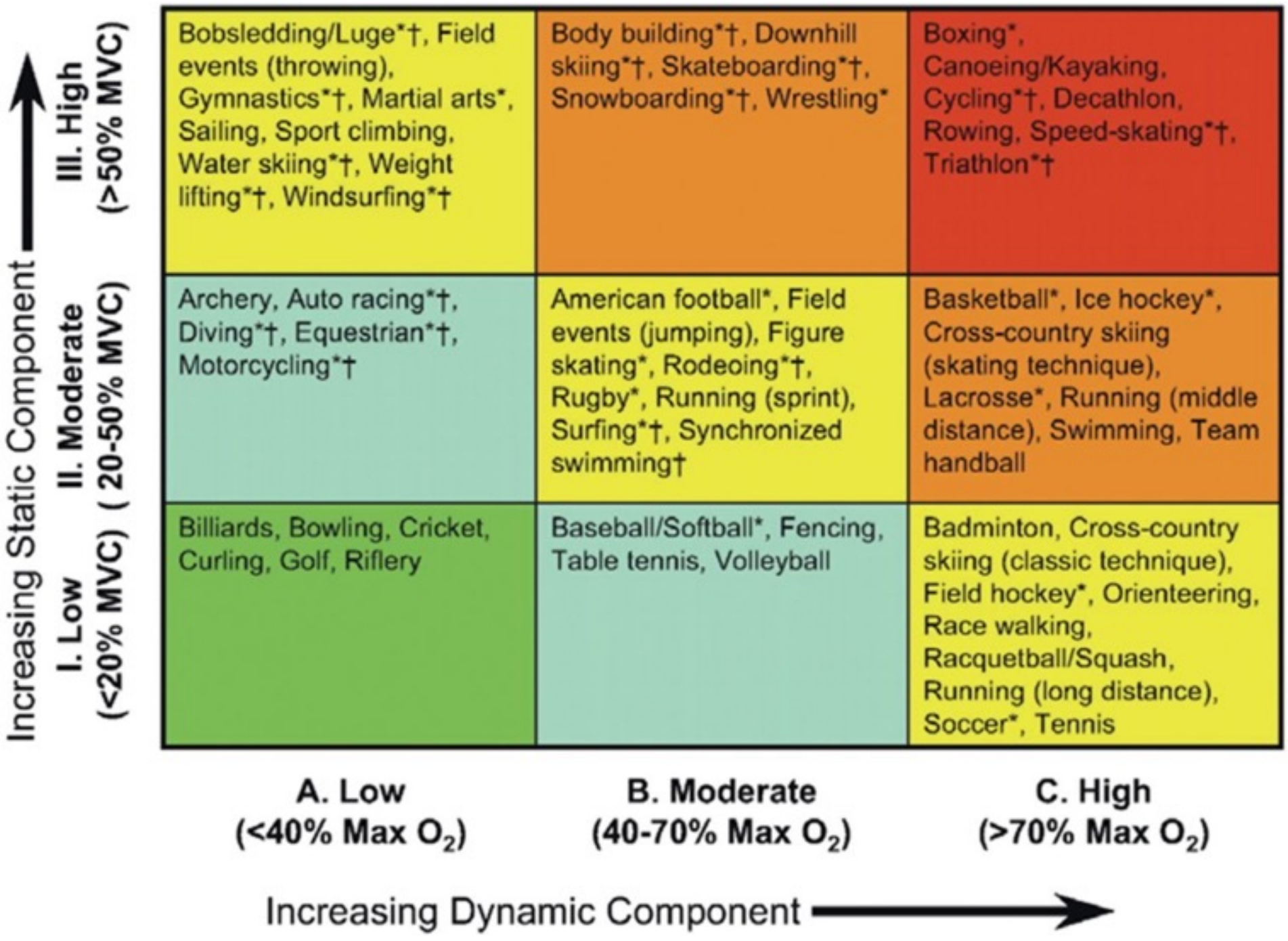
- Inform families that there are health risks associated with prolonged sedentary periods, even if patient meets the exercise goals

- Encourage patients to identify realistic and measurable goals when counseling them to change their physical activity behavior

- Counsel adults and children to block time for exercise during their weeks

- If heart is successfully repaired, without sequelae, full activity without restriction

Classification of sports by degree of peak dynamic and static components achieved during competition.



Patricia E. Longmuir et al. Circulation. 2013;127:2147-2159

So what now?

- Survival now into the 20s-30s for all lesions

- But is it morbidity free survival?

Neurodevelopmental disabilities

- Most common sequelae after congenital heart disease

- Most damaging sequelae after congenital heart disease

- Very limited data on this subject

ND disabilities

- Children undergoing cardiac surgery in infancy
  - Reasoning
  - Learning
  - Executive function
  - Inattention
  - Impulsive behavior
  - Language skills
  - Social skills

ND disabilities

- In childhood
  - Poor school performance
    - More likely to require remedial services
    - More likely to require OT, PT or ST
    - Strained interpersonal relationships
    - Behavior problems
- As Adults
  - Less educational achievements
  - Employability
  - Insurability
  - Quality of life

Neurodevelopmental outcomes following surgery for CHD

Neurodevelopmental Outcomes After Cardiac Surgery in Infancy

J. William Gaynor, MD[a], Christian Stopp, MS[b], David Wypij, PhD[b], Dean B. Andropoulos, MD[c], Joseph Atallah, MD, CM, SM, FRCPC[d], Andrew M. Atz, MD[e], John Beca, MD[f], Mary T. Donofrio, MD[g], Kim Duncan, MD[h], Nancy S. Ghanayem, MD[i], Caren S. Goldberg, MD[j], Hedwig Hövels-Gürich, MD[k], Fukiko Ichida, MD[l], Jeffrey P. Jacobs, MD[m], Robert Justo, MD[n], Beatrice Latal, MD[o], Jennifer S. Li, MD[p], William T. Mahle, MD[q], Patrick S. McQuillen, MD[r], Shaji C. Menon, MD[s], Victoria L. Pemberton, RNC, MS, CCRC[t], Nancy A. Pike, RN, PhD[u], Christian Pizarro, MD[v], Lara S. Shekerdemian, MD[w], Anne Synnes, MDCM[x], Ismee Williams, MD[y], David C. Bellinger, PhD[b], Jane W. Newburger, MD, MPH[b], for the International Cardiac Collaborative on Neurodevelopment (ICCON) Investigators

- Pediatrics; Volume 135, Number 5, May, 2015

ND outcomes after cardiac surgery in infancy

- Analyzed individual participant data from studies of children evaluated with the Bayley Scales of Infant Development

- Heart surgery 1996-2009

- Outcomes

  - Psychomotor Development Index (PDI)

  - Mental Development Index (MDI)

ND outcomes after surgery in infancy

- 1770 pts
- 22 institutions
- Assessed at age 14.5 +/- 3.7 months

Results

- Patients with CHD when compared to age matched peers
  - PDI- 77.6 +/- 18.8
  - MDI- 88.2 +/- 16.7
- Percentage of patients > 1 SD below the mean
  - PDI 63%
  - MDI 35%
- Percentage of patients > 2 SD below the mean
  - PDI 37%
  - MDI 15%

Risk Factors for lower PDI or MDI

- Risk factors for lower PDI or MDI
  - Lower birth weight
  - White race
  - Presence of genetic/extracardiac anomaly (all $p<0.01$)
  - Male sex
  - Less maternal education
- No improvement over time!

More on data

- Perhaps patient risk factors that are not related to the surgery are more important determinants of ND outcome than operative management strategies

- Fetal brain development is abnormal in patients with CHD

  - MRI studies

    - Delayed cortical development and folding

    - White matter injury present in 1 in 5 infants before CV surgery

“Lack of improvement in ND outcomes over the long study period despite contemporaneous improvements in survival, surgical strategies, preoperative care, thus may be due to greater effects of innate patient factors and brain maturation which outweigh the impact of modifiable management strategies.”

What are the risk factors for ND delays?

Published in final edited form as:
J Thorac Cardiovasc Surg. 2007 May ; 133(5): 1344–1353.e3. doi:10.1016/j.jtcvs.2006.10.087.

Patient characteristics are important determinants of neurodevelopmental outcome at one year of age after neonatal and infant cardiac surgery

J. William Gaynor, MD[a], **Gil Wernovsky, MD**[b], **Gail P. Jarvik, MD, PhD**[c], **Judy Bernbaum, MD**[d], **Marsha Gerdes, PhD**[e], **Elaine Zackai, MD**[f], **Alex S. Nord, BA**[c], **Robert R. Clancy, MD**[g], **Susan C. Nicolson, MD**[h], and **Thomas L. Spray, MD**[a]

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Risk factors

- 247 infants
- All types of CHD
- Looked at Psychomotor developmental index (PDI) and Mental developmental index (MDI)

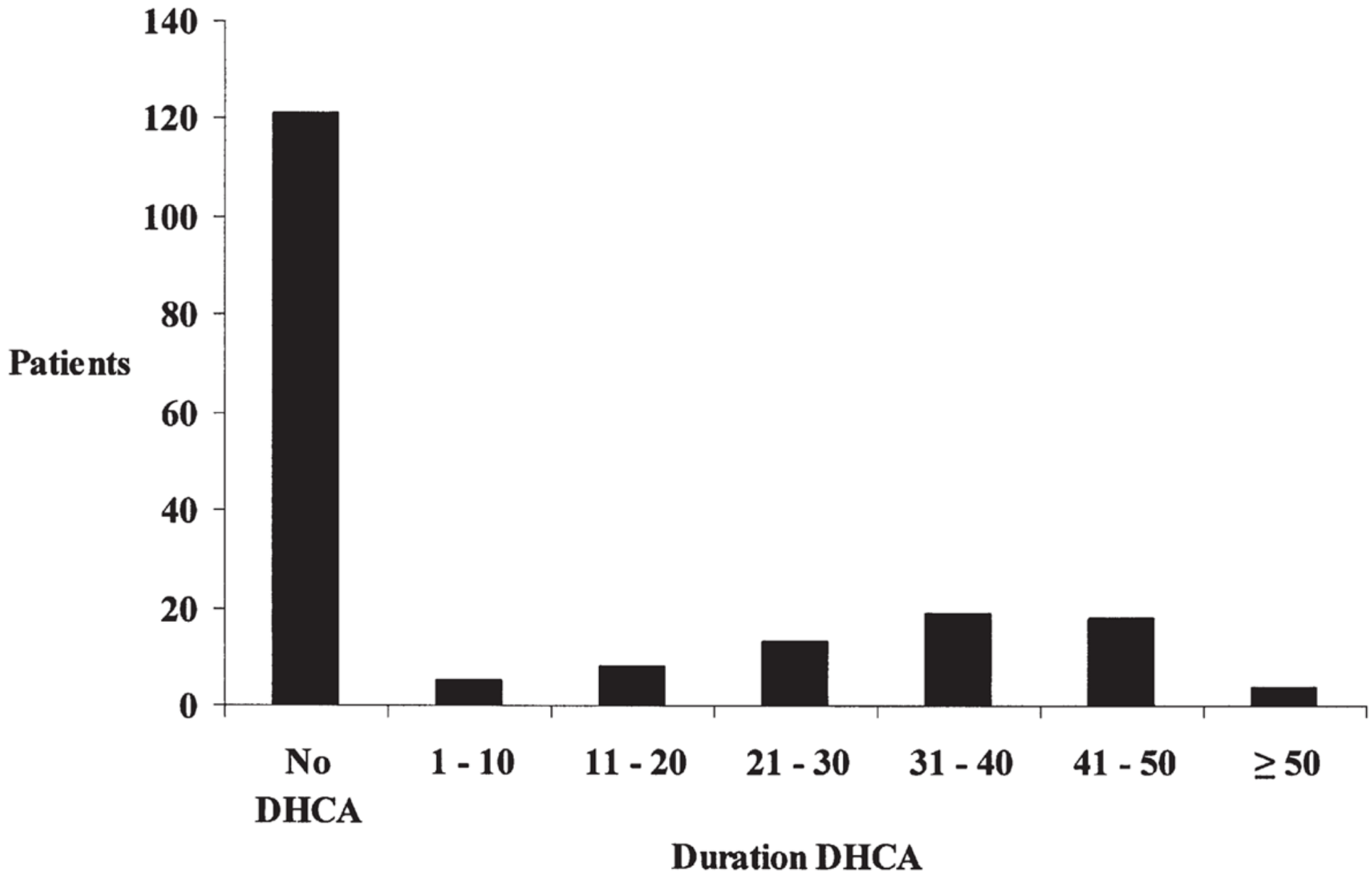

Figure 1.
Bar graph showing use and duration of DHCA. The *x-axis* is DHCA duration in 10-minute increments. The *y-axis* is number of patients. *DHCA*, deep hypothermic circulatory arrest.

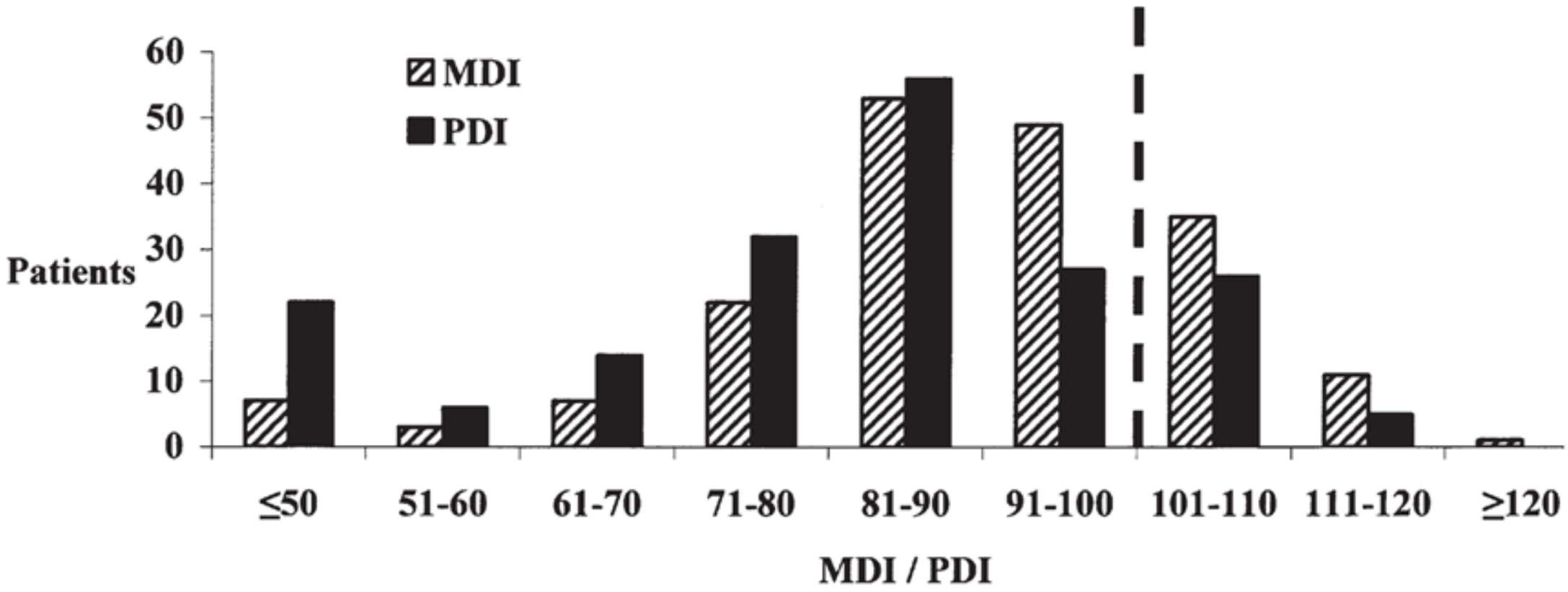

Figure 2.
Bar graph showing distribution of MDI and PDI scores. The *x-axis* is MDI/PDI score in 10-point increments. The *y-axis* is number of patients. In the general population, MDI and PDI scores are normally distributed with a mean of 100 and a standard deviation of 15. In this cohort, the distribution is shifted to the left, indicating worse performance. The PDI is more severely affected than the MDI. *MDI*, Mental Developmental Index; *PDI*, Psychomotor Developmental Index.

Multivariate analysis

- For entire cohort
  - MDI 90.6 +/- 14.9
  - PDI 81.7 +/- 17.2
- Non-genetic syndrome
  - MDI 93.7 +/- 13.6
  - PDI 85.1 +/- 14.6

- Patient related factors- Birth weight, ethnicity, presence of a genetic syndrome are all important risk factors
- Procedure related factors- lowest NP temperature, length of hospital stay

Conclusions

"Overall, patient-specific factors (gender, ethnicity, birth weight, birth head circumference, Apgar score at 1 minute, Apgar score at 5 minutes, genetic syndrome, *APOE* genotype) explained more of the variability in the MDI (13.0% vs 5.3%) and the PDI (20.7% vs 7.6%) than did intraoperative factors (weight at surgery, cooling time, DHCA time, CPB time, lowest NP temperature, hematocrit). "

Published in final edited form as:
Pediatr Cardiol. 2014 February ; 35(2): 344–352. doi:10.1007/s00246-013-0781-6.

Academic Proficiency in Children Following Early Congenital Heart Disease Surgery

Sarah B. Mulkey, MD[1,2], **Christopher J. Swearingen, PhD**[1], **Maria S. Melguizo, MS**[1], **Rachel N. Reeves**[3], **Jacob A. Rowell**[2], **Neal Gibson, PhD**[4], **Greg Holland, PhD**[4], **Adnan T. Bhutta, MBBS**[1], and **Jeffrey R. Kaiser, MD, MA**[5]

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[3]Arkansas Children's Hospital Research Institute, Little Rock, AR

[4]Arkansas Research Center, Arkansas Department of Education, Conway, AR

[5]Departments of Pediatrics and Obstetrics and Gynecology, Baylor College of Medicine, Houston, TX

Results

- CHD patients scored 13% lower on proficiency tests than non-CHD peers in both literacy and mathematics

- 8 fold increase in receiving special education compared to non-CHD patients

ND Conclusions

- Patients who have had surgery for CHD are at risk of multiple learning delays and psychosocial disorders

- Predetermined risk factors seem to play a larger role in the determination of delay in PDI and MDI than does surgical factors (although surgical factors still play a role)

- Early intervention may mitigate some delays

Pregnancy in patients with Congenital Heart disease

Pregnancy in CHD

- Generally safe
- High risk patient groups
    - Single ventricle
    - Severe Pulmonary hypertension
        - NYHA class
        - CHF symptoms
- Increased risk groups
    - Arrhythmias
        - TOF
- Cardiovascular management in pregnancy

Circulation. 2014 Jul 15;130(3):273-82

Summary

- Survival/outcomes in all CV surgeries are now very good
- Promote exercise in your CHD patients
- Monitoring neurodevelopmental outcomes is important in this patient population
- Any questions, please call!

Myths….or reality?

A. Early balloon atrial septostomies were performed using a foley catheter

B. The first 7 patients who underwent arterial switch procedure for D-TGA died in the peri-operative period

C. Patients with L-TGA can live normal full lives- without an operation