### **Adult Congenital Heart Disease**

Pankaj Madan, MD, MS, Medical Director South Texas Adult Congenital Heart Center Methodist Hospital, San Antonio

# Which statement is FALSE about congenital heart disease population?

- 1. More children than adults have congenital heart disease.
- 2. It is the fastest growing population of heart disorders in adults.
- 3. >90% of children born with congenital heart disease reach adulthood.
- 4. Adults with CHD require specialized care.

# What is TRUE about Adult congenital heart disease care?

- 1. Most patients are seen in specialized ACHD clinics
- 2. Majority of patients are seen by pediatric cardiologists.
- 3. Majority of the patients are seeing adult cardiologists
- 4. Majority of the patients are lost to follow-up

# What is true about ACHD subspecialty?

- 1. No specific training. Adult cardiologists are comfortable in taking care of ACHD patients.
- 2. No additional training required. Pediatric cardiologists are qualified to take care of ACHD patients.
- 3. Additional years of training beyond pediatric or adult cardiology required for expertise in ACHD care. There is additional board certification.

## **ACHD** population

- 40,000 infants born with CHD/ year
- THE most common birth defect
- Successful outcome is a moving target
  - Surviving initial surgical repair
  - Surviving to 1 year of age
  - Normal childhood
  - Normal adolescence

Survival <u>to</u> adulthood → Survival <u>through</u> adulthood

### **Improving survival of CHD patients**



Percent survival to 18 years old

## **ACHD: Population**

# Surviving to adulthood is now expected

### **Changing Mortality in Congenital Heart Disease**

Paul Khairy, MD, PHD,\* Raluca Ionescu-Ittu, MSC,†§ Andrew S. Mackie, MD, SM,† Michal Abrahamowicz, PHD,§ Louise Pilote, MD, MPH, PHD,‡§ Ariane J. Marelli, MD†

Montreal, Quebec, Canada



Khairy et al. JACC 2010

### **Congenital Heart Disease population**



### **Congenital Heart Disease population**





### **Congenital Heart Disease population**





# Patients reaching Adulthood with CHD



Gilboa et al. Circulation 2016;134:101–109

# Patients reaching Adulthood with CHD



Gilboa et al. Circulation 2016;134:101–109

### PROBLEM

# Probability of SCD free survival after surgical correction



### Age at death for adults with CHD



Oehslin et al. Am J Cardiol 2000; 86: 1111

### Mortality in Adult congenital heart disease



Verheugt et al. EHJ 2010; 31: 1220-9

### Most of the mortality is cardiac related



J Am Coll Cardiol 2007;50:1263–71)

# Sudden cardiac death and heart failure leading causes of cardiac mortality



### **Morbidity in ACHD**

One year hospitalization rate of patients with severe and other cardiac lesions compared with the adult population of Quebec (April 1999- March 2000)



Mackie AS. Am J Cardiol 2007; 99(6): 639-643

#### From: The Changing Demographics of Congenital Heart Disease Hospitalizations in the United States, 1998 Through 2010

JAMA. 2013;309(10):984-986. doi:10.1001/jama.2013.564



# Patients reaching Adulthood with CHD

140		
120	U	nce reaching adulthood *
100		
80		Survival is not as expected
60		
40	•	Heart Failure and
20		arrhythmias
	•	Morbidity is substantial

## Long term complications

### Arrhythmias

### **Heart Failure**

- Atrial
- Ventricular
- SCD

Adults with Congenital Heart Disease

Vascular lesions

**Residual Shunts** 

Valvular heart disease

## Long term complications

- **Adult Comorbidities**
- CAD, PVD
- DM
- OSA, COPD
- Renal and Hepatic insufficiency

vaivulai iicait

disease

#### 5-years risk of death For 40-years old patients



Diller et al. Circulation. 2015;132:2118-2125.

### Sudden Cardiac death in ACHD patients



Silka et al. JACC 1998: 32: 245

### Prevalence of Atrial arrhythmias in ACHD patients



Vander Velde et al. Eur J Epidemiol 2005; 20: 549-557

### **Exercise intolerance in ACHD population**



## **ACHD** population

• High risk group of young individuals

 Requires specialized and multidisciplinary care.

## Who is providing the care?

- Many being seen by Pediatric Cardiologists NOT trained in ACHD
- Many being seen by Adult Cardiologists NOT trained in ACHD
- Few being seen in ACHD clinics
- Several have been lost to follow-up

### ACHD Patients in USA vs Those in ACHD Clinics



Krasuski et al. Circulation. 2016;134:110–113

### **Understanding Loss of CHD Follow-Up**



The blue bars indicate patients who were not seen by a cardiologist within the indicated age range but were seen again by a cardiologist in an older age group (ie, transiently lost to follow-up).

Adapted from: Mackie A, et al. Circulation. 2009;120:302-309.

## **Reasons for being "Lost"**

### **Patient Obstacles:**

- Patient assumes "cure"
- Poor communication from parents or pediatrician
- Loss of previous health records
- Gradual symptom onset Lack of health insurance

### **Physician Obstacles:**

- Physician assumes "cure"
- Uninformed about specific potential problems
- No prior records available
- No reported symptoms
- Symptoms ascribed to more common causes

### **ACHD clinic: recurring themes**

• Patients not aware of their medical and surgical history

• They receive inappropriate medical or surgical treatment

• Patients are misinformed.

### **Group 1: Simple CHD**

Unrepaired conditions: Repaired or unrepaired
 – Isolated small ASD
 conditions

Patients should be seen at ACHD center at least once

VSD

32<sup>nd</sup> Bethesda Conference for care of ACHD

### **Group 2: Moderately complex CHD**

**Repaired or Unrepaired:** 

• Anomalous Left Coronary Artery from Pulmonary Artery (ALCAPA)

 Pulmonary valve regurgitation (moderate to severe)

0

Patients should be seen
periodically at an ACHD
center

• Infundibular right ventricular outflow obstruction (moderate to severe)

- letralogy of Fallot
- VSD with any valve problems

**32<sup>nd</sup> Bethesda Conference for care of ACHD** 

## Group 3: Greatly complex CHD

#### **Repaired or Unrepaired:**

- Congenitally Corrected Transposition of the Great Arteries (ccTGA or I-TGA)
- Double outlet ventricle

### All patients who have undergone the following procedures:

- Arterial switch procedure
- Blalock-Taussig shunt
- Patients should be seen
- : regularly at ACHD center.
- Inansposition of the Great Arteries (d-TGA)

Senning procedure

- Tricuspid atresia
- Truncus arteriosus/hemitruncus
- Other abnormalities of AV connections (i.e., crisscross heart, isomerism, heterotaxy syndromes)

All Patients with Eisenmenger syndrome

All Patients who are cyanotic ("blue")

32<sup>nd</sup> Bethesda Conference for care of ACHD

### When to refer

- If your patient had a surgery named after a physician.
- When more than one isolated structural defect is present.
- You can't figure out anatomy in 5 seconds
- Your patient has never been seen by an ACHD physician







Search	

Home > Newsroom, Events & Resources > Media Newsroom > Releases > News Release

News Release: ABMS Announces Certification in New Subspecialty Adult Congenital Heart Disease

NEWS RELEASE

ABMS Media Contact: Lori Boukas (312) 436-2626 Iboukas@abms.org

See complete list of specialty and subspecialty certificates

#### ABMS Announces Certification in New Subspecialty: Adult Congenital Heart Disease

CHICAGO - December 5, 2012 - The American Board of Medical Specialties (ABMS) announces the creation of physician certification in a new subspecialty: Adult Congenital Heart Disease (ACHD). The ABMS Board of Directors and ABMS Reserved Powers Board approved the subspecialty at its September 2012 meeting. The subspecialty will be offered by the American Board of Internal Medicine (ABIM) and will

## **Additional material**

### **Transplantation in ACHD**

- Complex anatomy with multiple prior surgeries
- Highly sensitized
- Increased pulmonary vascular resistance
- Concomitant organ dysfunction e.g. liver

### **Transplant outcomes in CHD**



### Not all CHD are created equal

