

# Adult Congenital Heart Disease: The Scope of the Problem

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# Congenital Heart Disease

- ▶ Recognized for centuries
- ▶ Most common of all major birth defects
- ▶ 5-9/1,000 live births
  - ▶ Higher if BAV and PFO included
- ▶ Wide variety of lesions
- ▶ Natural and 'unnatural' histories

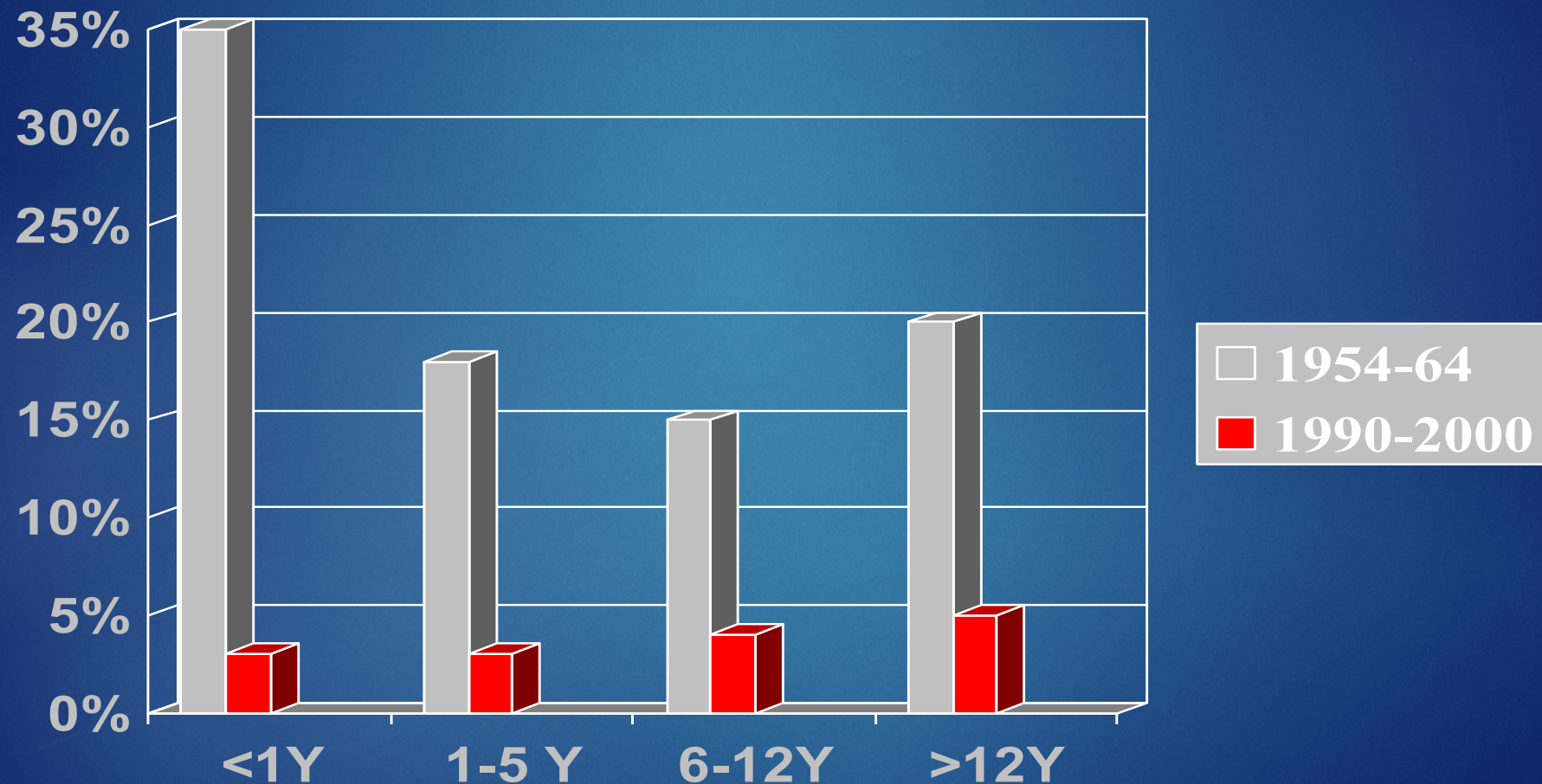
# Growth and Impact of CHD Surgery

- ▶ 1938 – Ligation of a patent ductus arteriosus (Gross & Hubbard)
- ▶ 1944 – First Blalock-Taussig shunt (Baltimore)
- ▶ 1945 – Resection of an aortic coarctation (Crafoord and Gross)
- ▶ 1953 – Gibbon closes ASD successfully using heart-lung machine
- ▶ 1954 – Repair of Tetralogy of Fallot (Lillihei, U Minn)
- ▶ 1957, 63 – Senning and Mustard atrial switches for D-TGA
- ▶ 1968 – Fontan procedure (Bordeaux)
- ▶ 1975 – Arterial switch for D-TGA (Jatene, Brazil)
- ▶ 1981 – Norwood procedure for HLHS (Boston)

# CHD Pioneers



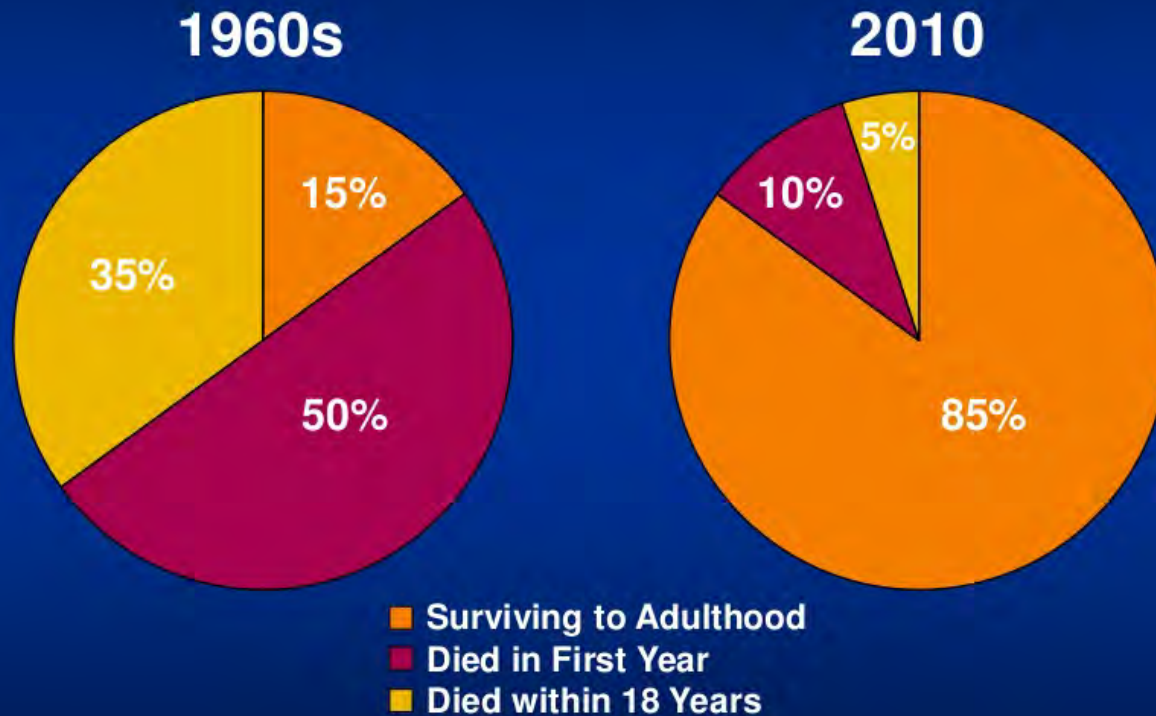
# CHD Surgical Mortality According To Age And Era



Slide Courtesy of Robert Sade, MD

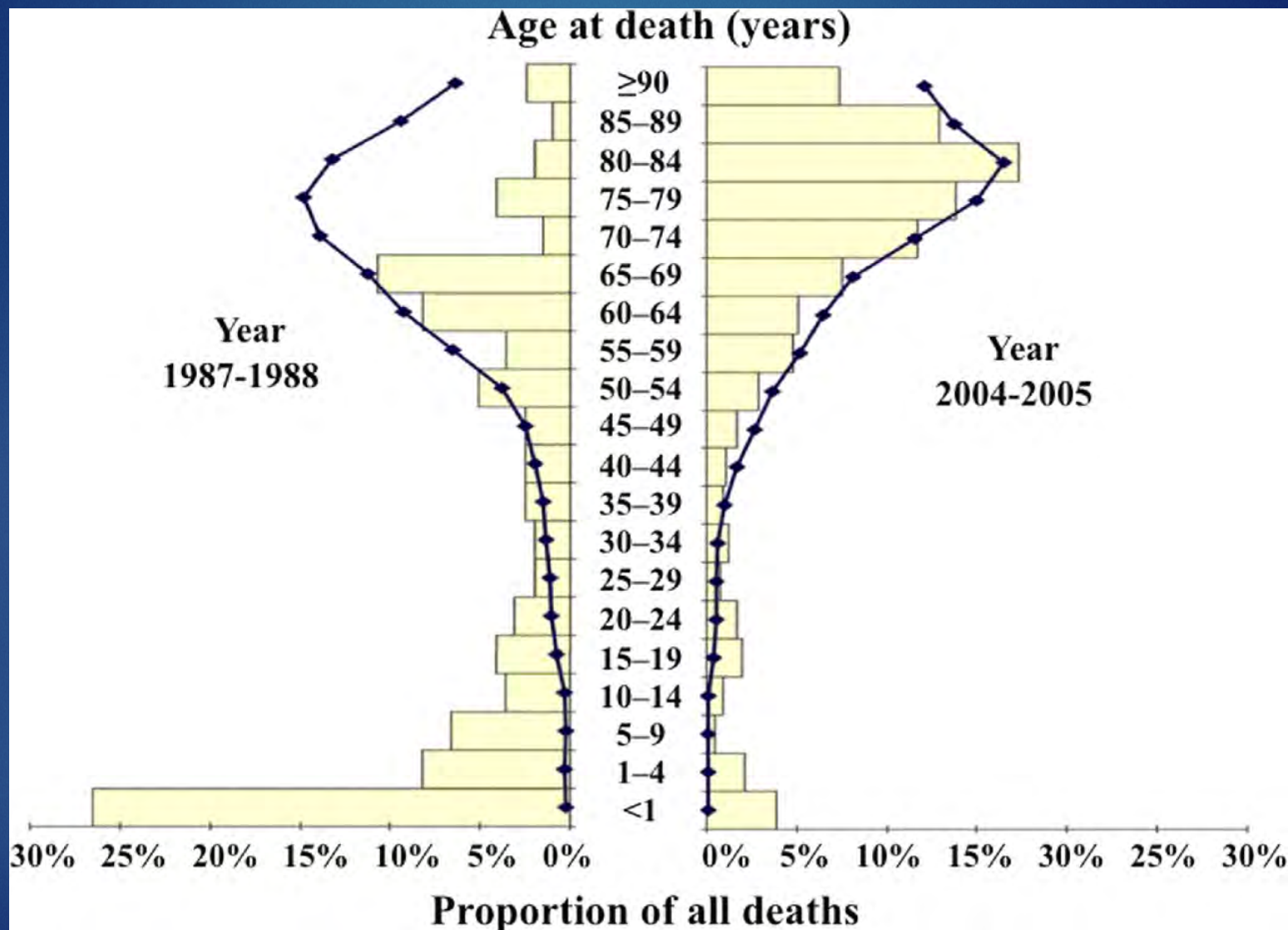
# Changing Times

Improving Natural History of Children with CHD  
= More Adults with CHD

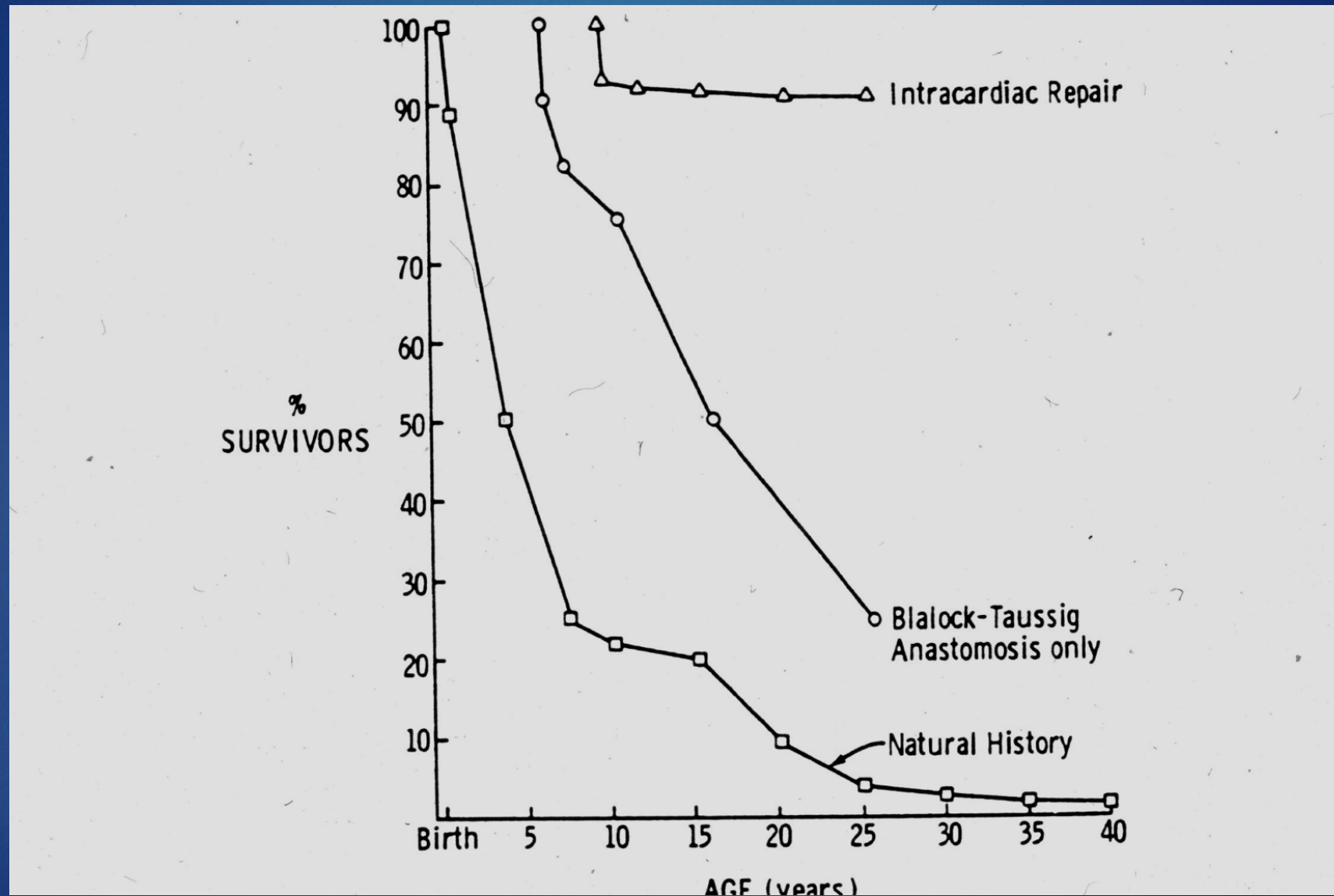


Vander Velde et al. *Eur Jour Epi.* 2005;20:549–57; Wames et al. *Circulation.* 2008;118:e714–e833.

# Age at Death in Patients With CHD



# TOF: Palliation and Repair



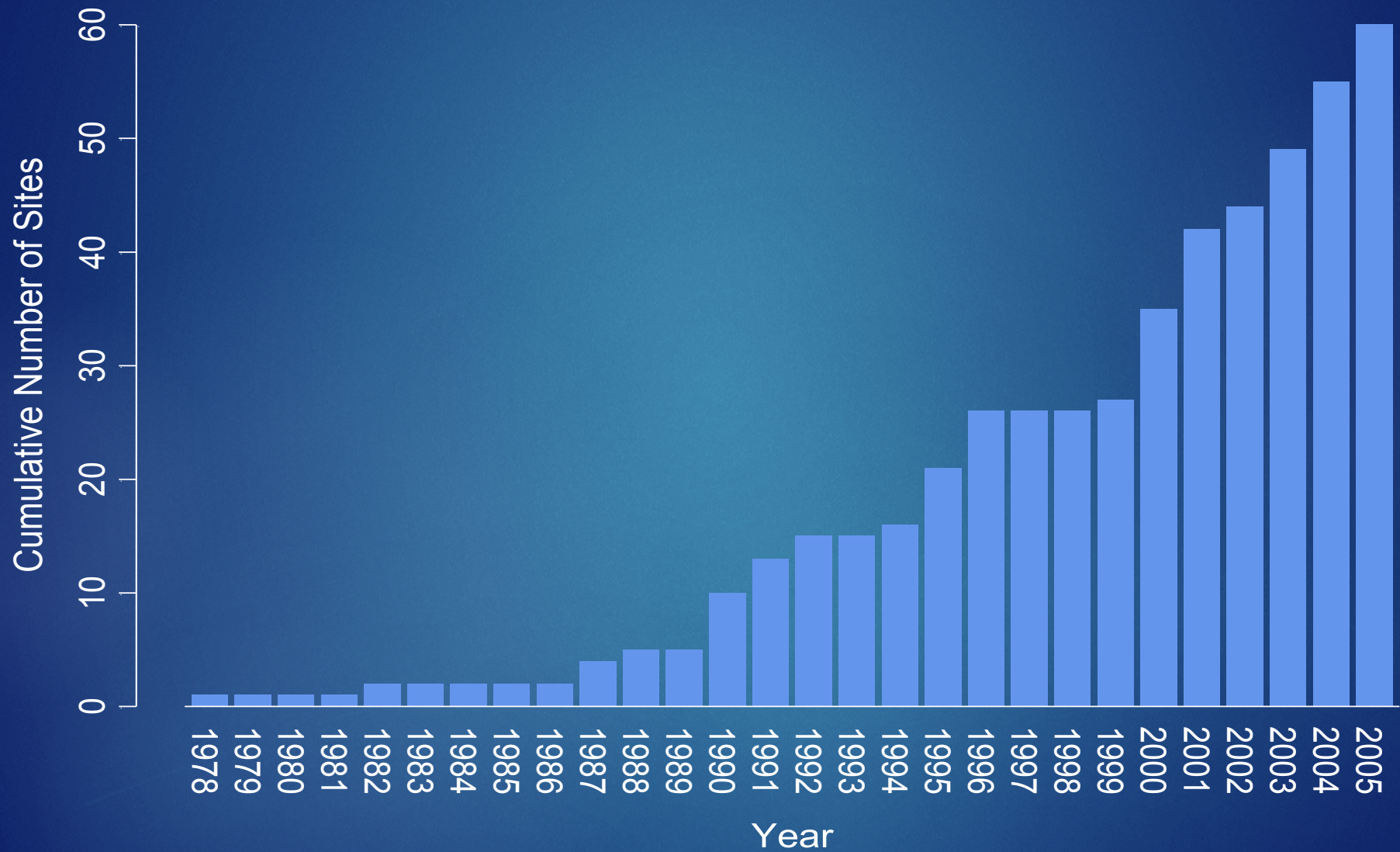
# Adult Congenital Heart Disease (ACHD)

- ▶ >39,000 infants born annually with CHD
- ▶ 90% survive to adulthood
- ▶ 20-25,000 operations for CHD annually
- ▶ Currently over 1,500,000 adults in US with CHD

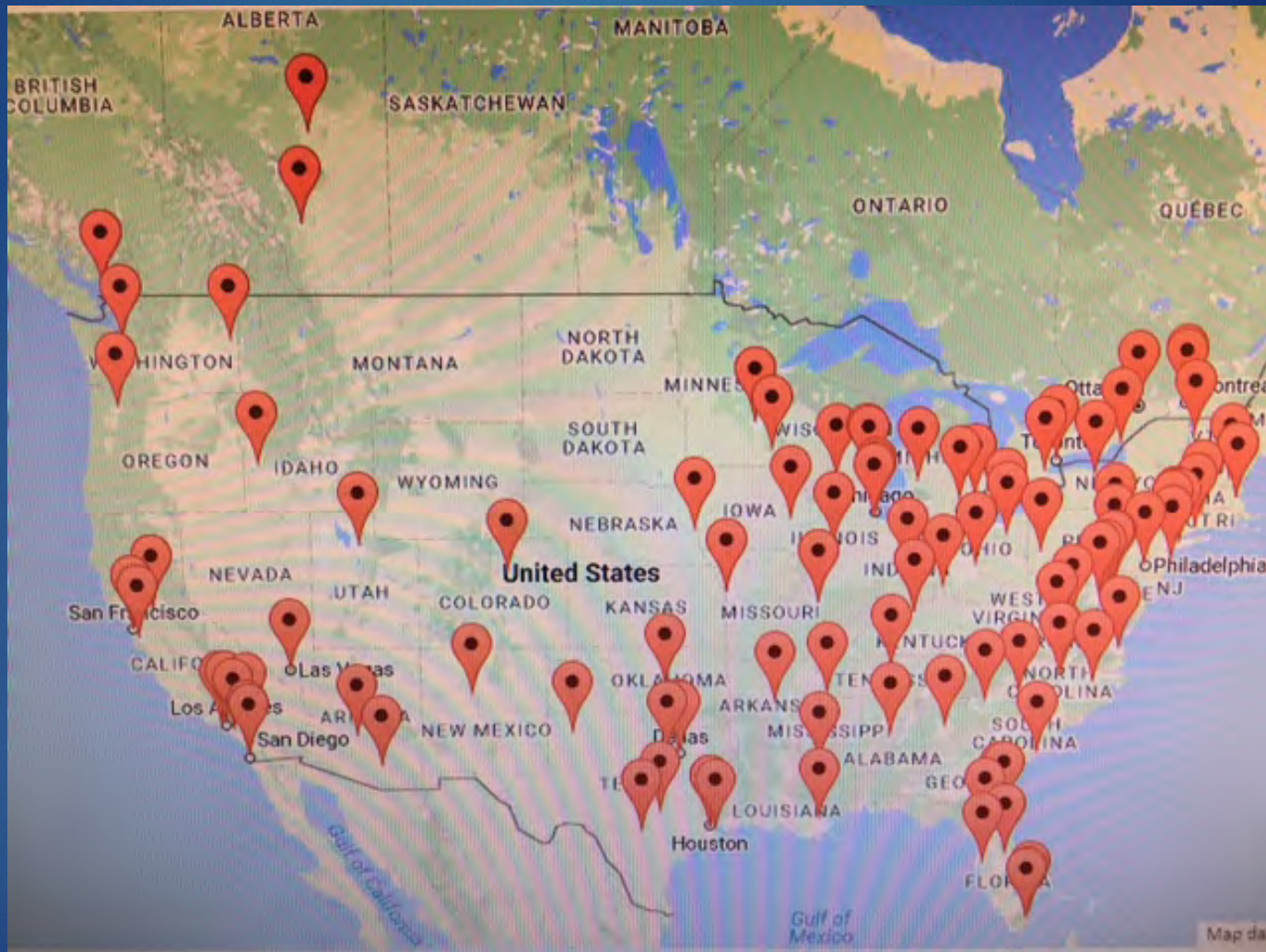
# Birth of a (sub) Subspecialty

- In the Beginning...
  - Boston Children's/ Mass General 1974
  - UCSF and UCLA 1980
  - Mayo 1988
  - Penn State Hershey 1991
- Currently 110 self proclaimed programs in US
- Little formal training available for ACHD physicians

# Program Establishment



# ACHD Programs in US and Canada, 2017





# Adult Congenital Heart Association

- 1998 at Boston Children's
- Now focuses on education, research and advocacy
- ACHA program accreditation



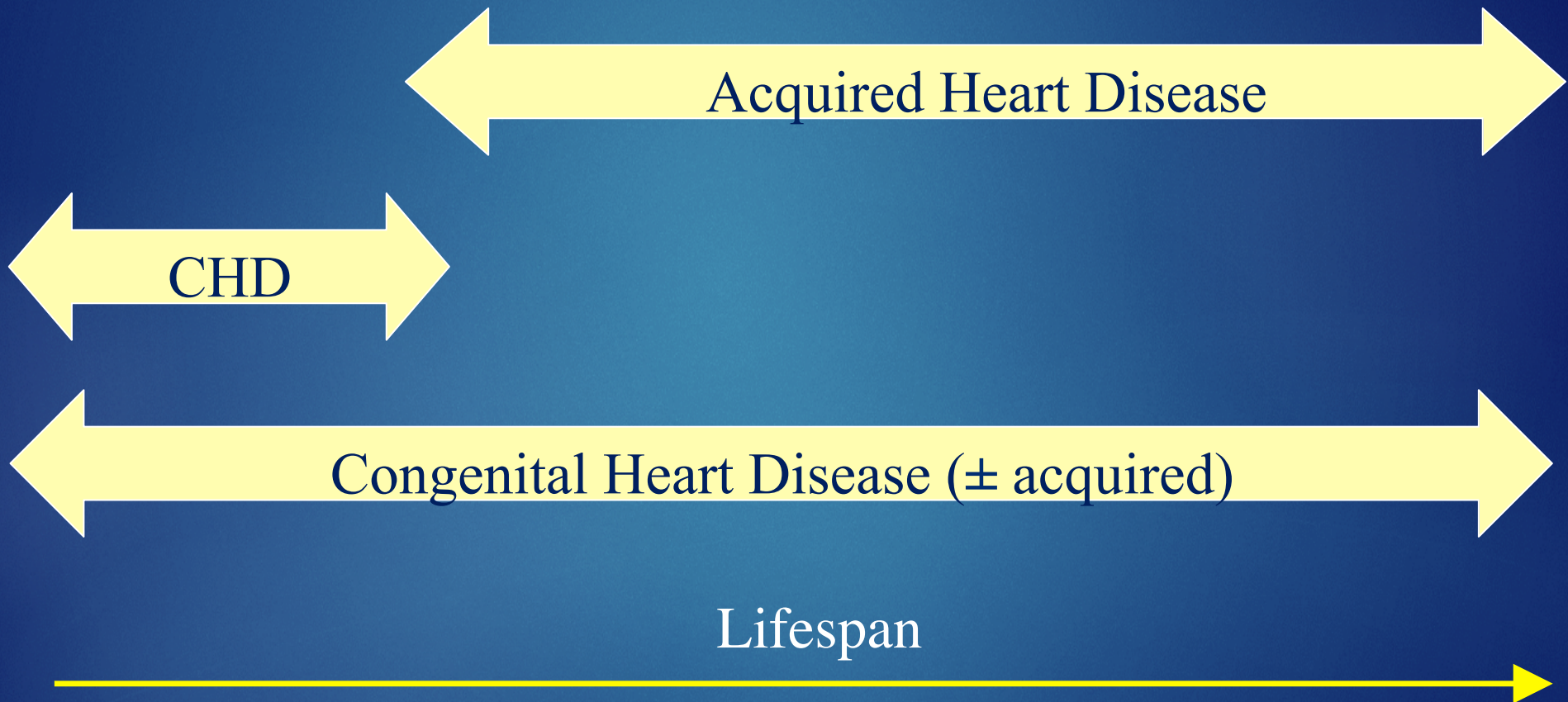
# Goals in ACHD

- ▶ Sufficient, trained providers
- ▶ Sufficient number of Regional Centers
  - ▶ Research
  - ▶ Clinical care
- ▶ Web of referral relationships
- ▶ Improved evidence base for practice

# Clinical Challenges in ACHD

- ▶ Relatively small numbers of patients/lesion
- ▶ High percentage of patients 'lost' to follow up
- ▶ Large burden of complex clinical care
  - ▶ History, lesion- and operation-specific
- ▶ High demands of imaging - anatomic, physiologic
- ▶ Imposition of acquired on congenital heart disease
- ▶ Consequences of aging

# ACHD: Changing the Conceptual Framework



# Societal Challenges in ACHD

- ▶ Transition of care
- ▶ Location of care
- ▶ Working relationship between Medicine and Pediatrics
- ▶ Societal recognition (coding, insurance, disability)
- ▶ Reluctance to refer
- ▶ Insurance access and adequacy
- ▶ Neurocognitive issues

# Timing of Transition

- Difficult to have a ‘one size fits all’ approach
  - Based on pt’s developmental stage
- Gradual process
  - Emphasize need for autonomy
- Start to educate teens
  - Diagnosis, meds, residual problems, symptoms to be worried about

# Location of Care

- Adult Only Hospital?
- Free-standing Children's Hospital?
- "Mixed" hospital?

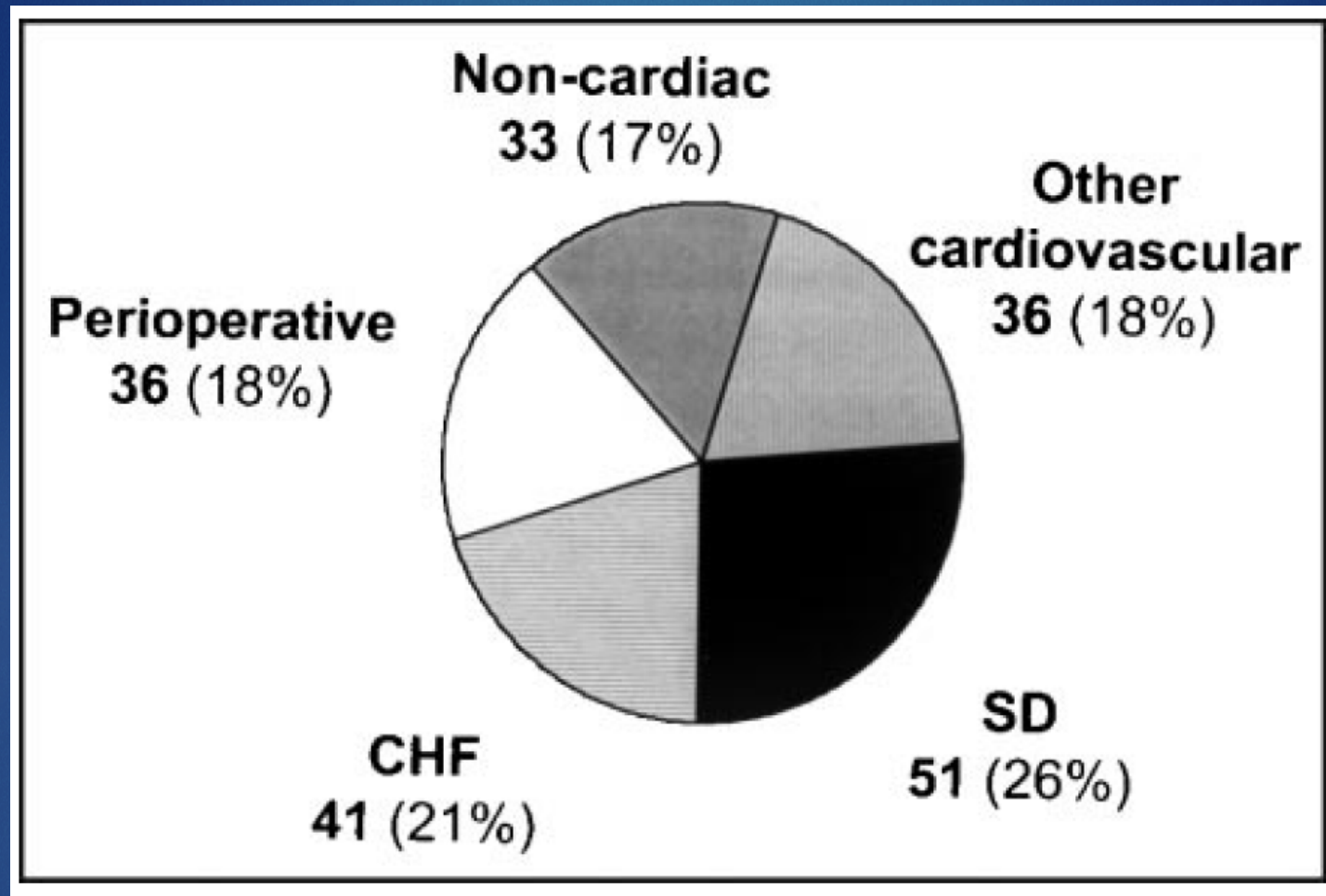
# Residuae and Sequelae of CHD Surgery

- ▶ Ventricular Dysfunction
- ▶ Vascular Problems
- ▶ Valve Disease
- ▶ Non-cardiovascular
- ▶ Prosthetic materials
- ▶ Electrophysiologic
  - ▶ Myocardial scars
  - ▶ Disorders of impulse formation and conduction
- ▶ Endocardial and myocardial effects

CHD Surgery is corrective, not curative!

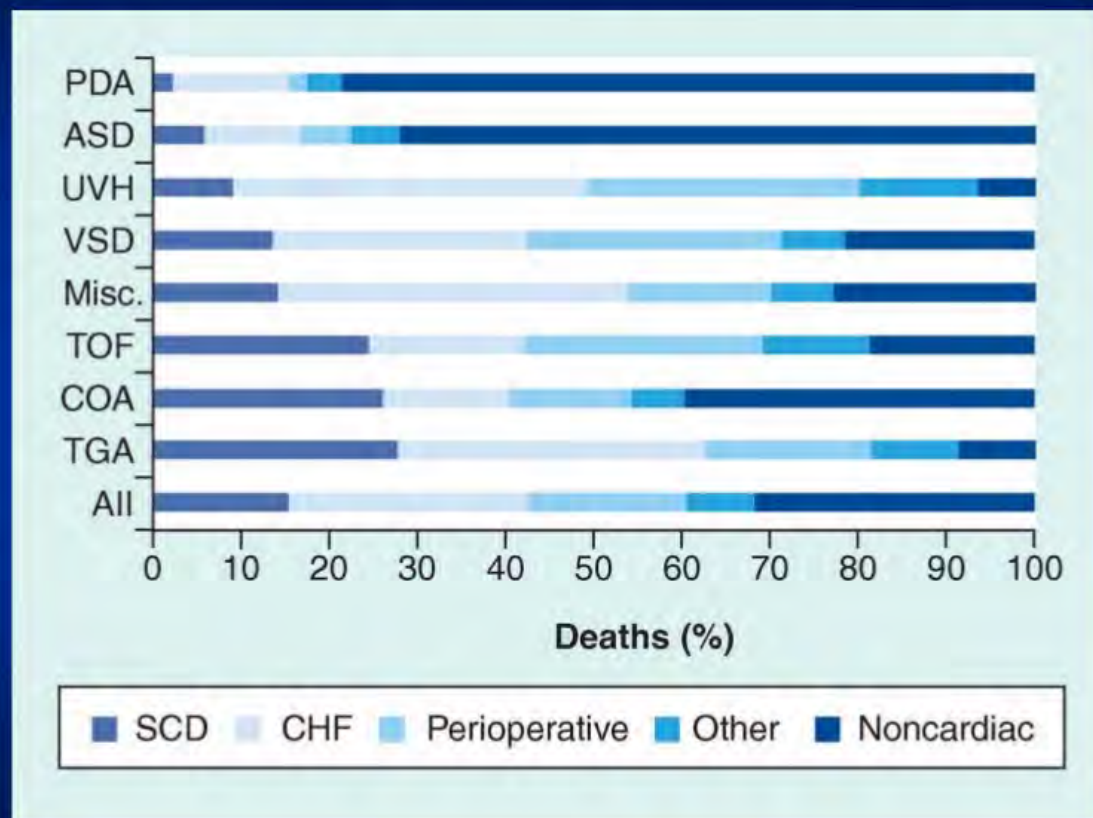
Perloff, Circ, 1991

# Causes of Death in ACHD



# SCD in ACHD

## Sudden Death Also Important Cause of CHD Mortality



# Death in ACHD

- ▶ Avoidable in 21%
  - ▶ 341 deaths in 4500 British pts
  - ▶ >50% of deaths in Class I or II patients
  - ▶ Errors in arrhythmia management
  - ▶ Lack of knowledge concerning pt.'s disease
  - ▶ Poor operative technique or planning
  - ▶ Poor postoperative care
  - ▶ Cyanotic and Fontan patients

Somerville, 1995

# Guidelines... Finally!

## PRACTICE GUIDELINE: FULL TEXT

### ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease

A Report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines on the Management of Adults With Congenital Heart Disease)

*Developed in Collaboration With the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons*

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# Recommendations for Care

- ▶ Every ACHD patient should be seen by an ACHD specialist at least once.
- ▶ Simpler lesions can be treated locally:
  - ▶ closed PDA
  - ▶ repaired secundum ASD
  - ▶ isolated small or repaired VSD
  - ▶ bicuspid AV without root involvement
  - ▶ mild pulmonic stenosis

# ACHD Guidelines

## ACC/AHA Guidelines for Monitoring ACHD

### Recommended visits to an ACHD center:

|                     |                                                                                      |                                                       |
|---------------------|--------------------------------------------------------------------------------------|-------------------------------------------------------|
| Simple              |    | At least once to determine needs for future follow-up |
| Moderate complexity |   | Every 12 to 24 months                                 |
| Great complexity    |  | Every 6 to 12 months                                  |

# Who Should Provide Care

- Pediatric Cardiologists??
- Adult Cardiologists??
- Ideally, an ACHD physician!

# But the Problem Is...

- Too few ACHD docs
- Too few ACHD fellowship programs
- Lots of current ACHD docs nearing retirement age....

# CHC PACC

(Program for Adult Congenital Cardiology)



# Where We are Headed...

- ▶ ACHA Accredited Comprehensive Care Center
- ▶ Fontan FIT Program
- ▶ Education Programs
  - ▶ Nursing, Inpt. providers,
  - ▶ Residents, cardiology fellows
  - ▶ PATIENTS!





Thanks

