ECHOCARDIOGRAPHIC APPROACH TO CONGENITAL HEART DISEASE: THE POSTOPERATIVE ADULT

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OUTLINE

WHY ACHD?

WHAT GENERAL PRINCIPLES APPLY?

SPECIFIC LESIONS?
PREVALENCE OF CHD BY AGE

>1,000,000 Adults with congenital heart disease

Marelli et al. Circulation 2007
GENERAL GOALS OF CONGENITAL CARDIAC SURGERY

Create physiologically normal circulation

Decrease hypoxia

Increase pulmonary blood flow

“Separate” the pulmonary and systemic circulations

Allow for growth
GENERAL APPROACH

As with unrepaired congenital heart disease

Start with standard views to get a “lay of the land”

Follow blood flow through the heart as best possible

Use clues, like pacemaker leads

Evaluate residual stenoses, regurgitation, septal defects
VENTRICULAR SEPTAL DEFECT
SURGICAL TREATMENT

PALLIATION AND REPAIR

PALLIATION WITH PULMONARY ARTERY BAND

PATCH CLOSURE

A congenital defect that occurs between the right and left ventricles of the heart. A VSD allows blood to seep from the left to right side of the heart. The defect can be surgically corrected by sewing a patch over the hole.
ECHOCARDIOGRAPHIC EVALUATION

LEFT HEART
Residual VSD
Left ventricular size, ventricular function
Aortic valve function
    Right coronary cusp prolapse or thickening

RIGHT HEART
Right ventricular size and function
Right ventricular and pulmonary artery pressures
Evaluate for pulmonary stenosis if prior banding
ATRIOVENTRICULAR SEPTAL DEFECT
ATRIOVENTRICULAR SEPTAL DEFECT REPAIR

Surgical repair of ASD, VSD, cleft in mitral and/or tricuspid valve

May have residual mitral regurgitation

Abnormal left ventricular outflow tract
EVALUATION OF REPAIRED AVCD

Residual VSD or ASD

Residual “mitral” regurgitation
EVALUATION OF REPAIRED AVCD

Left ventricular outflow tract obstruction

Especially with annular rings or valve replacement
COARCTATION OF THE AORTA
COARCTATION OF THE AORTA

SURGICAL CORRECTION

- End–end
- Interposition graft
- Patch repair

- Subclavian flap repair
  - turndown

- Subclavian flap turnup

- Subclavian flap repair
  + end–end

- Subclavian reimplantation

- Subclavian reimplantation
  + end–end anastomosis
Arterial hypertension
May occur even with optimal repair

Blood pressure in left arm may be inaccurate

Recoarctation or residual coarctation

Aneurysms

COARCTATION OF THE AORTA
LONG TERM COMPLICATIONS
ECHOCARDIOGRAPHIC EVALUATION

Aortic size
Proximal and distal velocity through repair

Abdominal Doppler
TETRALOGY OF FALLOT
TETRALOGY OF FALLOT
ANATOMY

RIGHT VENTRICULAR OUTFLOW TRACT OBSTRUCTION

VENTRICULAR SEPTAL DEFECT

OVERRIDING AORTA

RIGHT VENTRICULAR HYPERTROPHY
TETRALOGY OF FALLOT
ANATOMY

RIGHT VENTRICULAR OUTFLOW TRACT OBSTRUCTION

VENTRICULAR SEPTAL DEFECT

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RIGHT VENTRICULAR HYPERTROPHY
“Make me a patent ductus arteriosus”
TETRALOGY OF FALLOT
BLALOCK TAUSSIG THOMAS
SHUNT

“Make me a patent ductus arteriosus”

Increases pulmonary blood flow → Decreases cyanosis
TETRALOGY OF FALLOT

SURGICAL CORRECTION

- Enlarge RVOT
- Closure of VSD
- Placement of transannular patch
TETRALOGY OF FALLOT

SURGICAL CORRECTION

Not cyanotic

Normal pulmonary blood flow

Pulmonary regurgitation

Right ventricular enlargement
TETRALOGY OF FALLOT

LONG TERM COMPLICATIONS

Pulmonary insufficiency and RV dysfunction
  Transannular patch associated with significant PI
Arrhythmias
  Supraventricular and ventricular
Sudden death
Exercise intolerance
  Decreased compared with age matched controls
TRANSTHORACIC ECHOCARDIOGRAM

Right heart size and function
Pulmonary regurgitation
Pulmonary stenosis
Left ventricular size and function
Aortic size and aortic valve function
ECHOCARDIOGRAM

PULMONARY STENOSIS AND REGURGITATION
ECHOCARDIOGRAM

RIGHT VENTRICULAR ENLARGEMENT
TRANSPOSITION OF THE GREAT ARTERIES
WHAT IS TRANSPOSITION OF THE GREAT ARTERIES?

AORTA FROM THE RIGHT VENTRICLE

PULMONARY ARTERY FROM THE LEFT VENTRICLE
PHYSIOLOGY

PULM ART

AORTA

PULM VEIN

RA-LV

SVC/IVC
PHYSIOLOGY

AORTA

PULM VEIN

PULM ART

ASD

VSD

PDA

SVC/IVC
Atrial level switch (1954)
SVC and IVC flow directed to left ventricle
Pulmonary vein flow directed right ventricle
Pericardial/prosthetic baffles vs. atrial tissue primary distinction between Mustard and Senning

Gaca et al. Radiology 2008
TRANSTHORACIC ECHOCARDIOGRAM

RV/LV size and function
AV valve function
Baffle obstruction or leak
Subpulmonic stenosis
Aortic size
ECHOCARDIOGRAM

PULMONARY VENOUS CHANNEL
ECHOCARDIOGRAM

SYSTEMIC VENOUS CHANNEL
GREG

36 year old man with congenital heart disease, in for routine follow-up
Transposition of the great arteries
   Initial palliation with a balloon septostomy
   Mustard repair at age 1 month
On no medications
Sinus bradycardia with junctional escape but normal heart rate variability
GREG’S ECHO
GREG’S MOST RECENT VISIT

Initially referred for evaluation of pulmonary stenosis

Subjectively normal exercise capacity

Rode his bike from home to his annual ACHD clinic evaluations
34 year old man with TGA and Mustard repair
Bradycardia requiring pacemaker implantation
Morning headaches and effort intolerance for 2 years
NYHA Class II, on beta blocker, ACE I, furosemide, digoxin
Sinus node dysfunction with atrial pacemaker
PROBLEM: SYSTEMIC RIGHT VENTRICLE
PROBLEM: BAFFLE OBSTRUCTION

[Image with arrows indicating Azygos run off]
TRANSTHORACIC ECHOCARDIOGRAM

LV/RV function
Branch pulmonary stenosis
Aortic dimension and neoaortic valve function

Brickner et al. NEJM 2000
PROBLEM: DILATED AORTAS AND PULMONARY STENOSIS AND ABNORMAL CORONARIES
FONTAN REPAIR
ATRIO OR CAVO-PULMONARY SHUNT
“MODIFIED” FONTAN - “NEW SCHOOL”
FONTAN PROCEDURE
ECHOCARDIOGRAM

MARKEDLY ENLARGED RIGHT ATRIUM
ECHOCARDIOGRAM

STAGNANT RIGHT ATRIAL FLOW
ECHOCARDIOGRAM

INTRATRIAL CONDUIT
CONCLUSION

Adult congenital heart disease is increasingly common

Echocardiography is a mainstay of evaluation

Knowing what to expect is optimal

Referral to an ACHD center is recommended