Special Considerations for Special Populations: Congenital Heart Disease

Valerie Bosco, FNP, EdD
Alison Knauth Meadows, MD, PhD
University of California San Francisco
Adult Congenital Heart Program

Outline

• The exploding population
• Pre-transplant considerations
• Outcomes
• Cases
• Conclusions

Congenital Heart Disease: Spectrum of Disease

• CHD occurs in 8:1000 live births
• CHD represents a spectrum

simple complex

Congenital Heart Disease
Congenital Heart Disease

- 1938: First PDA ligation
- 1944: First aortic coarctation repair
- 1945: First Blalock-Taussig shunt
- 1950’s: Introduction of cardiopulmonary bypass - facilitated repair of TOF, CAVC, D-TGA, VSDs
- 1970’s: Fontan procedure introduced
- 1980’s: Neonatal surgery began - allowed repair of D-TGA with arterial switch procedure and palliation for HLHS

Congenital Heart Disease

- Advances in catheter based interventions
- Advances in imaging
  - 2D and 3D echocardiography
  - Cardiac MRI
  - Cardiac CT
- Advances in intensive care management
  - Balloon pump
  - ECMO
  - Ventricular assist devices
- Advances in medical management

Adults with Congenital Heart Disease

- More are surviving to adulthood
  - 85% are predicted to survive to adulthood
  - 85% increase since 1985
  - >1,000,000 ACHD patients in the USA
  - Adults with CHD > children with CHD

Adults with Congenital Heart Disease

- Sicker patients are surviving to adulthood
  - More than 1/3 have moderate to severe disease
  - Fewer than 30% are getting specialty care
  - Many young adults will reach end stage without good options
Options for ACHD Patients

Are there surgical or transcatheter options to treat residual hemodynamic burden?

- yes
  - Perform appropriate intervention
- no
  - Medical management

Consider transplant evaluation

Pre-transplant considerations in ACHD

- Multiple surgeries
  - Antibodies
  - Scar
  - Chest wall collaterals
  - Access issues
- Anatomic issues
  - Heterotaxy
  - Great vessel anatomy distortion
  - Venous anomalies

Pre-transplant considerations in ACHD

- Long standing heart failure (low output, congestion)
  - Renal failure
  - Liver failure
  - PLE
- Pulmonary arterial hypertension
- Chronic cyanosis
  - bleeding risk
  - thrombosis risk
Case #1: TS
NOT a transplant candidate

- 24 yo female
- Heterotaxy, dextrocardia, (P,L,L) CAVC unbalanced to the RV, LV hypoplasia, pulmonary atresia, transposed great vessels, bilateral SVCs without a bridging vein, left sided IVC

ARRGGHHHHH!!!!!!!
Case #1: TS
NOT a transplant candidate

- History of atrial flutter controlled on sotalol
- Bilateral iliac venous occlusions
- Now with profound symptoms
  - Resting and exertional cyanosis (rest 78%, with brief exercise 50%)
  - Severe headaches
  - Disfiguring AV malformation in left arm

Case #1: TS
NOT a transplant candidate

- Risk factors
  - Single ventricle
  - Chronic cyanosis
  - Limited venous access
  - Heterotaxy and dextrocardia
  - Multiple previous surgeries

Case #2: DB
Transplant candidate now, but can’t wait

- 23 yo female
- Shone’s syndrome including
  - Parachute MV with MS
  - Subvalvar and valvar aortic stenosis
  - Coarctation of the aorta
  - Severe LV diastolic dysfunction

Case #2: DB
Transplant candidate now, but can’t wait

- Status post surgical aortic valvuloplasty
- Status post percutaneous aortic valvuloplasty
- Status post CoA repair and subAS resection
- Status post AVR w/ St Jude’s mechanical valve
- Left with hemodynamic burdens
  - Mild MS
  - Severe LV diastolic dysfunction with LVEDp 30 and rising PVR
- NYHA Class III-IV
Case #3: LH
Good transplant candidate

- 38 yo Hispanic male
- Tetralogy of Fallot
- Status post repair (8 years of age)
- Developed severe cardiomyopathy (LVEF 7-25%) - ? alcohol toxicity
- Status post ICD for syncope and NSVT
- NYHA Class III, CHF management, multiple hospital admissions
- Considered for transplant in 2003

Case #3: LH
Good transplant candidate

- Admitted in cardiogenic shock (2005)
- 8/2005: To OR, but aborted transplant
- 9/2005: Successful heart transplant
- Follow-up at 1 and 5 years with no evidence of rejection, clinically well
- Co-morbidities
  - Diabetes
  - Osteoporosis
  - Hepatic congestion
  - Skin lesions

Case #3: LH
Good transplant candidate

- Predictors of favorable outcomes
  - Single sternotomy
  - Low PRA
  - Relatively acute onset of heart failure
  - Compliant with good family support

Summary

- Adults with CHD represent a unique and complex population of transplant candidates
- Many ACHD are not good candidates
- Transplant should be considered early in ACHD patients
  - To determine early whether transplant is an option in the future
  - To determine whether other interventions will help or hurt candidacy
  - To avoid detrimental effects of congestion, low output, and PAH on future candidacy