State of the Art: Congenital Heart Disease 2002

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The following is a summary from the Power Point slides of Dr. Hsu's presentation

New Advances in Pediatric Cardiology

Management of the infant with congenital heart disease
- Fetal enchocardiography
- Infant surgery
- Heart transplantation
- Hypoplastic left heart syndrome

Interventional Cardiology

Fetal Ultrasound

Cardiac development
- 6 weeks gestation: heart chambers, great arteries formed.
- 14 weeks: major cardiac defects can be detected by ultrasound.
- 22-24 weeks: most common time for abnormalities to be identified.

Cardiac screening
- 4 chamber view
- 2 great arteries

Fetal Echocardiography

Ventricular hypoplasia
- Tricuspid atresia
- Hypoplastic left heart syndrome
- Single ventricle

Atrioventricular canal defect, VSD
Transposition of the great arteries
Pulmonary or aortic valve stenosis
Tetrology of Fallot
Coarctation of the aorta, ASD cannot be ruled out
Ventricular function
- Congenital defects
- Arrhythmias
  - SVT
  - Heart block

Follow-up studies may detect progression of defect
- Vavlvar stenosis - atresia
- Progressive ventricular hypoplasia

Motality associated with congenital heart disease in the United States

Boneva *et al*. Circulation 2001
National Center for Health Statistics of the CDC from 1979-1997
Mortality from heart defects declined 39% over the 18 years period at all ages
2.5 to 1.5 per 100,000 population
2.7% per year

Factors influencing improved infant survival

Echocardiographic diagnosis
Diagnostic catheterization 5%
Prostaglandin E
Surgical techniques
Post-operative care
Fetal echocardiography?

Impact of fetal diagnosis

Improved surgical outcome after fetal diagnosis of hypoplastic left heart syndrome (*Tworetzky et al, circulation 2001*)
Patient population: 88 patients

Prenatal	33
Postnatal	55

Surgiacal Interventions in HLHS

Single ventricle palliation: 3 stages
- Systemic venous return directly to lungs bypassing the heart, RV supplies blood flow to the body.
- Infancy: single RV with BT shunt (Norwood)
- 8 months: SVC to pulmonary artery connection
- 3 yrs: IVC to pulmonary artery connection

Heart transplantation

Outcome: Staged palliation for HLHS

Norwood Stage 1	30-45%
SVC-PA connection	4%
IVC-PA connection	4%
Overall 5 year survival	50%

Mortality on Heart Transplant Waiting List

16% children>6 months (n=497)
25% infants <6 months (n=639)

Decision Analysis of HLHS Treatment Strategies

A comparison of treatment strategies for HLHS using decision analysis
(*Jenkins et al. J Am Coll Card 2001)*
Pooled data from 5 centers: surgical mortality for staged surgery not increased if performed within one month of age

Treatment Strategies

Name	Description of surgery
Staged surgery	Stage 1 and 2 performed
Stage 1, the list	Stage 1 performed, then listed, receives organ if available
List, wait 1 month	If organ not found within 1 month, Stage 1 then Stage 2
List, wait 2 months	As above for 2 months
List, wait 3 months	As above for 2 months

List, wait until transplant - Pt waits until organ is available

Increasing the Donor Pool: ABO Incompatible Transplant

Hearts allocated by blood type compatibility
- O: Universal donor- recipient O, A or B
- Type O recipients more likely to die while waiting
- A,B hearts not used if recipient not available

Fetus synthesizes IgM (agglutinins)
Neonate: Anti-A and Bare IgG of maternal origin
Production of anti-A and B agglutinins begins at 3-6 months
Anti-A and B titers maximal levels by 5-10 yrs
Hypothesis: Neonates would be tolerant of an ABO incompatible donor due to the absence of IgG anti-A and anti-B agglutinins
Nost common mismatch: Recipient O, Donor A,B

ABO Incompatible Heart Transplants: Hospital for Sick Children: NEJM 2001

10 ABO incompatible transplants performed between 1996 and 2000
Survival 80%, comparable to ABO compatible
No hyperacute rejection
Mild humoral rejection noted at autopsy in one infant w/antibodies
2 infants developed antibodies to donor antigens but no evidence of damage to graft
Mortality on waiting list declined from 58% (1990-96) to 7% (1996-2000)

Post-transplant Management

Routine immunosupression
Normal rejection surveillance: clinical and endomyocardial biopsy
ABO antibody titer surveillance - titers = or > 1.4 consider plasmpheresis and increased immunosupression
Coronary angiography performed 6 and 12 months from DOT, then annually

Patient DS: blood type O

HLHS, poor RV function, tricuspid insufficiency and LV thrombus
Intubated on prostaglandins
Heart transplant at 17 days of age: Donor type B
Discharged post-op day #14
One episode of clinical rejection, treated with increased immunosuppression
All anti B antibody titers negative to date

New Advances in Interventional Cardiology

Therapeutic Catheter Interventions
- Alterative to surgery
- Adjunct to surgery

Implantable Devices
- Open: Stent
- Close: PDA, ASD, VSD

Intravascular Stent

Stenosis not amenable to balloon dilation

Post-surgical
- Branch pulmonary arteries
- RV-PA conduit
- Re-Coarctation
- Venous baffles

Native
- Coarctation
- Peripheral pulmonary stenosis

PDA Closure Devices

Gianturco Coil
- Ductus < 2 mm
- Hourglass appearance

Clinical trial: Amplatzer Ductal Occluder
- Larger PDA > 2 mm
- Any anatomic subtype

Amplatzer PDA Occluder: C*hildren's Hospital of NY (4/00 - 1/02)*

62 pts: I mos to 71 yrs
6 pts excluded: PDA too small, coil closure- 5 pts
Irreversible pulmonary hypertension- 1 pt
56 successful implantations
Ductus size: 3.5 mm (1.8-10 mm)
Closure rate: 53/66 (94.6%) closed at discharge, 55/56 (98/2%) closed on last follow-up

ASD Closure Devices

Two devices are FDA approved
Cardioseal
- Lower profile
- Stainless steel/Dacron

Amplatzer Septal Occluder
- Easily retrievable
- Optimal for larger defects

Clinical Applications of Transcatheter ASD Closure

Secundum ASD
- Isolated
- Multiple
- Standard of care shifting

Patent Foramen Ovale with paradoxical embolus
Fontan Fenestration

Future Directions in Pediatric Cardiology

Biomechanical devices
- Valves
- Growth potential

Evidence based medicine
- NHLB/NIH funded Pediatric Heart Disease Clinical Research Network
- 7 Centers in US and Canada
- Encourage multicenter trials in children with heart disease
- Clinical outcomes: medical, catheter-based and surgical therapies

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