Ductal-Dependent Forms of Congenital Heart Disease: Clinical Significance of Prenatal Diagnosis

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**Ductus Dependant Lesions**

- **Types**
  - **Right side obstruction**
    - Pulmonary atresia (IVS, VSD)
      - Functional Atresia
    - Tricuspid atresia
    - Severe TOF
  - **Left side obstruction:**
    - Aortic arch (CoA, IAA)
    - LVOT obstruction
    - HLHS
  - **Parallel Circulation:**
    - TGA
    - DORV
    - Isolated ventricular inversion
  - **Complex CHD with obstructed systemic or PBF**
**Postnatal Dx** 250 pts  
**Prenatal Dx** 68 pts  
**Clinical condition**  
- Metabolic acidosis  
- Multiorgan failure  
- Longer hospital stay  
- Preoperative mortality  
  - 15 / 250  
  - 0 / 68  
- Postoperative mortality  
  - 20 / 235  
  - 0 / 68  

**Conclusions:**  
- Prenatal diagnosis reduces mortality and morbidity in TGA

*Circulation. 1999;99:916-918*
HLHS

- 88 patients:
  - 33 prenatal Dx (11 terminated)
  - 55 Postnatal Dx

- Surgery:
  - 14 / 22
  - 38 / 55

- Survival till second stage: 75%
  - Prenatal Dx: 100%
  - Postnatal Dx: 65%

- Lower preoperative morbidities

- Conclusions: Prenatal diagnosis of HLHS was associated with improved preoperative clinical status and with improved survival after first-stage palliation

Circulation. 2001;103:1269-1273

Improved Surgical Outcome After Fetal Diagnosis of Hypoplastic Left Heart Syndrome

Wayne T. Tworetzky, MD; Jeff D. McElhinney, MD; V. Mohan Reddy, MD; Michael M. H recycle, MD; Frank L. Hauser, MD; Norman H. Silverman, MD

Background: Hypoplastic left heart syndrome (HLHS) is frequently diagnosed prenatally, but this has not been shown to improve surgical outcomes.

Methods and Results: We reviewed patients with HLHS between July 1992 and March 1999 to determine the influence of prenatal diagnosis on preoperative clinical status, outcomes of stage 1 surgery, and parental decisions regarding care. Of 88 patients, 33 were diagnosed prenatally and 55 after birth. Of 22 prenatally diagnosed patients, 22 were live-born, and pregnancy was terminated in 11. Of 22 prenatally diagnosed patients who were live-born, 14 underwent surgery, and parents elect to forego treatment in 8. Of 55 patients diagnosed postnatally, 38 underwent surgery, and 17 did not because of parental decisions or clinical considerations. Prenatally diagnosed patients were less likely to undergo surgery than postnatally diagnosed infants (P=0.008). Among live-born infants, there was a similar rate of death or NEC. Among patients who underwent surgery, survival was similar (59.52%). All patients who had a prenatal diagnosis and underwent surgery survived, whereas only 25 of 38 postnatally diagnosed patients survived (P=0.009).

Conclusions—Prenatal diagnosis of HLHS was associated with improved survival after first-stage palliation in comparison with patients diagnosed after birth. (Circulation, 2001;103:1269-1273.)

Key Words: Prenatal diagnosis; Hypoplastic left heart syndrome; Infant diseases

Hypoplastic left heart syndrome (HLHS) can be easily recognized on prenatal ultrasound and is proportionately one of the most common serious cardiac defects diagnosed prenatally. The standard “amnion fluid index” used for screening of congenital heart disease demonstrates either a small left heart or an enlarged left ventricle from isolated diaphragmatic defects. Prenatal diagnosis of HLHS, however, has not been demonstrated to improve survival outcomes.

In HLHS, the left heart complex is underdeveloped and unable to support the systemic circulation. Neonates with HLHS may be asymptomatic but become severely ill as the ductus arteriosus closes. Without treatment, this defect is almost invariably lethal. Current management strategies for infants with HLHS includes staged palliative surgery, monitoring with serial echocardiographic evaluation (Fontan operation) or stage conversion. Despite advances in surgical technique and postoperative care for infants with HLHS, the first stage of surgical palliation continues to have significant morbidity compared with other neonatal cardiac operations.

Prenatal diagnosis of HLHS allows time for discussion between parents and physicians to plan and prepare them for possible outcomes. Without the benefit of prenatal diagnosis, most infants with HLHS are born outside of tertiary care centers, which delays diagnosis and appropriate care. The delay in diagnosis, in turn, may lead to systemic hypotension, shock, and multiglandular damage, which can diminish chances for surgical success and lead to long-term sequelae.

Precise smaller studies have failed to demonstrate a positive impact of prenatal diagnosis on the surgical outcomes of patients with HLHS. Furthermore, the different influences of prenatal and postnatal diagnosis on parental decisions or preoperative clinical status have not been fully understood. The aim of this present study was to examine the impact of prenatal and postnatal diagnosis on (1) the preoperative clinical status of these infants and (2) outcomes.
Clues for Diagnosis

- 4 Chambers abnormalities
  - Symmetrical or Asymmetrical
  - Chambers size
  - Abnormal MV or TV
4 Chambers View

Asymmetrical 4ch

Symmetrical 4ch
Small RV
4 Chambers View
4 Chambers View
LVOT and RVOT
Aortic arch
Ductal arch
3 Vessels View

P
A
svc
d
C

SHA22
KACC 2000
3 Vessels View
Axial Arch View
Axial Arch View
Conclusions  Prenatal detection of reversed flow in the aortic arch or ductus arteriosus is associated with complex congenital heart disease with major diminution of forward flow to the corresponding great vessels.
Arch Color Flow Doppler
Axial Arch View
Arch Color Flow Doppler
Arch Color Flow Doppler
Influence of ultrasonographers training on prenatal diagnosis of congenital heart diseases: a 12-year population-based study

- Sonographers
  - Trained 19
  - Not trained 21
- CHD detection
  - 75% vs 38%
  - P < 0.001

Prenat Diagn 2008; 28: 1016–1022
Take Home Messages

- Ductus Dependant Lesions should not be missed
- Prenatal Diagnosis: Life Saving
- Training & Education
- Important Clues:
  - 4 Chambers Symmetry
  - Outflow tract Parallel
  - Vessel size in 3 vessels view
  - Arch Color Flow Doppler
Mark Your Calendar 29-31 March

Echocardiography Workshop on Congenital Heart Disease

TEE Imaging from Knowledge to Clinical Practice
“First Hands-on TEE Workshop”

main topics
- Live Transmission from Operating room & Cath Laboratory
- “Hands-on” Mannequin Simulator training for TEE procedure
- Wet Lab for Bovine Hearts
- Computer Based TEE Training
- 3D TEE During Procedures
- Read with expert sessions
- Emphasis on TEE modality

target audience
This course is designed for pediatric and adult cardiologists, Anesthesiologists, ER Physicians, fellows in training, and Cardiac Sonographers. Participants should have a good working knowledge of echocardiography.

venue
Cardiac Sciences Auditorium, King Abdulaziz Cardiac Center
King Abdulaziz Medical City for National Guard Health Affairs
Riyadh, Saudi Arabia

Early Registration: 01 March 2011

National Guard Health Affairs
King Saud Bin Abdulaziz University for Health Sciences
and the Postgraduate Training Center - Riyadh

International Guest Speaker:
Dr. Richard Humes, MD, FAAP, FACC
Chief, Cardiology - The Children’s Hospital of Michigan
Detroit Medical Center (DMC)

29th - 31st March 2011
24th - 26th Rabi II 1432 H

TEE Simulator Practice
Special Thanks

- Dr Merna Atiyah
- Obstetric Team
- Techs: Natalia & Carmelita
- Nurses

Thank you for your attention