Prenatal Natural History of Left Ventricular Outflow Obstructive Lesions and Fetal Imaging

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Obstructive Lesions of Left Ventricular Outflow Tracts

- Fetal Development of Ventricular Outflow Tracts (OFT) and Obstructive Lesions
- Prenatal Natural History of Obstructive Lesions of OFT
- Fetal Cardiac Imaging of Evolution of Obstructive Lesions of OFT
The distal part of the right ventricle and outflow tract (OFT) originate from a “Secondary and Anterior Heart Fields”

Formation of the Ventricular Outflow Tracts

- The OFT has exclusively muscular walls and the lumen is lined throughout by a layer of endocardial jelly.
- The endocardial jelly concentrates itself into pairs of facing cushions spiraling round and approaching one another throughout the outflow tract.

Formation of the Ventricular Outflow Tracts

The arrangement of the distal outflow tract: Cells derived from the neural crest infiltrate the distal cushions, which fuse to divide the distal OFT into aorta and the pulmonary trunk.
Formation of the Ventricular Outflow Tracts

Formation of valve in proximal portion of OFT: the cushions divide into two layers that give rise to the valvar leaflets luminally and the walls of the supporting sinuses murally. **Myocardialisation**: Muscular tissue added to the OFT in its most proximal portion by the process of “Myocardialisation”.

Rotation of the OFT: Rotation of the myocardium at the base of the OFT leads to positioning of the great arteries with respect to each other at the ventriculo-arterial junction.
Natural History of Left Ventricular Outflow Obstructive Lesions in Fetuses

Left heart OFT obstructive lesions in the fetus can evolve through pregnancy.
Natural History of Left Ventricular Outflow Obstructive Lesions in Fetuses

The onset in earlier time in gestation and severity of OFT obstructive lesions affect the left heart growth and postnatal outcomes.
Natural History of Left Ventricular Outflow Obstructive Lesions in Fetuses

The onset in earlier time in gestation and severity of OFT obstructive lesions affect the left heart growth.

Aortic Stenosis

Aortic Atresia

Growth of the Left Ventricle in OFT obstructive lesion detected in Mid-trimester

Hornberger et al. Circulation 1995
Natural History of Left Ventricular Outflow Obstructive Lesions in Fetuses

The onset in earlier time in gestation and severity of OFT obstructive lesions affect the left heart growth.

Moderate Aortic Stenosis  Severe Aortic Stenosis

Growth of the Mitral Valve in OFT obstructive lesion detected in Mid-trimester

Hornberger et al. Circulation 1995
Natural History of Left Ventricular Outflow Obstructive Lesions in Fetuses

The onset in earlier time in gestation and severity of OFT obstructive lesions affect the left heart growth.

Aortic Stenosis

Growth of the Aorta in OFT obstructive lesion detected in Mid-trimester

Aortic Atresia

Hornberger et al. Circulation 1995
Natural History of Left Ventricular Outflow Obstructive Lesions in Fetuses

The onset in earlier time in gestation and severity of OFT obstructive lesions affect the left heart growth.

Change in LV/RV length ratio from Mid-trimester to Post-natal intervention

Hornberger et al. Circulation 1995
In midgestation fetuses with Aortic Stenosis and normal LV length, reversed flow in the arch and foramen ovale, monophasic mitral inflow, and LV dysfunction predict → HLHS.

Mäkikallio et al. Circulation. 2006
Natural History of Left Ventricular Outflow Obstructive Lesions in Fetuses

In Left Ventricle OFT obstructive lesion initial mid-trimester left heart dimensions and flow pattern predict the development or progression of left heart hypoplasia:

- Mitral valve and ascending aorta z scores,
- Left ventricular length,
- Reduced rates of growth of left heart structures,
- Mitral valve inflow pattern
- The foramen ovale flow from LA to RA, and retrograde transverse arch flow.
- LV dysfunction

Endocardial fibroelastosis (EFE): Severity of EFE corresponds to some indexes of left heart size, geometry, and function.

Absence of EFE or presence of grade I EFE favors a biventricular outcome postnatally.

McElhinney et al. Am J Cardiol 2010
Left Ventricular Outflow Tract Obstructive Lesions: Fetal Cardiac Imaging

Serial fetal cardiac evaluation:

- Measurements of left heart structures
- Left ventricular function
- Fetal flow dynamics

Left Ventricular Outflow Tract Obstructive Lesions: Fetal Cardiac Imaging

Progression of Aortic Stenosis to HLHS

Aortic Stenosis with Smaller LV in Early Mid-trimester

Aortic Stenosis with Globular LV Later in Mid-trimester
Left Ventricular Outflow Tract Obstructive Lesions: Fetal Cardiac Imaging

Progression of Aortic Stenosis to HLHS

Aortic Stenosis with dysfunctional and hypertrophied LV

Aortic Stenosis with Smaller LV and Grade II EFE in Late Mid-trimester
Left Ventricular Outflow Tract Obstructive Lesions: Fetal Cardiac Imaging

Progression of Aortic Stenosis to HLHS

HLHS with Grade II EFE of LV in Third Trimester

HLHS with Grade III EFE of LV in Third Trimester

HLHS with MA & AS in Third Trimester
Left Ventricular Outflow Tract Obstructive Lesions: Fetal Cardiac Imaging

Progression of Aortic Stenosis to HLHS

HLHS with Hypoplastic Ascending Aorta

HLHS with Retrograde Flow in Ascending Aorta and Arch
In fetuses with aortic stenosis following initial mid-trimester left heart dimensions and flow pattern predict biventricular outcome postnatally:

- Aortic Root and Ascending Aorta z scores within ± 2,
- The presence of forward flow in transverse arch and RA to LA foramen ovale flow,
- Absence of LV dysfunction
- Absence of or only grade I EFE.

Development of Fetal Aortic Stenosis in Mid-trimester

Aortic Stenosis with Dysplastic Valve, Normal LV Function

Aortic Stenosis with Turbulent Colorflow, Normal LV Size

Aortic Stenosis with Increased LVOT Pressure Gradient
Aortic Stenosis in a Neonate

- Aortic Stenosis with Dysplastic Valve
- Aortic Stenosis with Turbulent Colorflow
- Aortic Stenosis with Discrepant RV-LV

Postnatal Outcome of Left Ventricular Outflow Tract Obstructive Lesions in Fetuses
Left Ventricular Outflow Tract Obstructive Lesions: Fetal Cardiac Imaging

- Discern the natural history of left ventricular outflow tract obstructive lesions
- Helps in decision making for need for intervention
- May provide the patient selection criteria for intervention
- Provides the predictors of postnatal outcomes
Thank You
In neonates with critical aortic stenosis independent factors associated with greater survival benefit for the Norwood procedure versus biventricular repair:

- Younger age at diagnosis and entry,
- Lower z-score of the aortic valve and LV length,
- Higher grade of endocardial fibroelastosis,
- Larger ascending aorta.
In fetuses with aortic stenosis following initial mid-trimester left heart dimensions and flow pattern predict biventricular outcome postnatally.
Segmental Approach to CHD

Cardiac Development: Re-alignments

Manner J. Anat Rec 2000
Outflow tract development:
Contribution of second heart field
Truncus arteriosus septation

A

EGA 7½, CS 15

B

EGA 8, CS 18

C

LVOT
RV
LV
PA

D

EGA 7½, CS 16

E

RA
LA

F

LA
Formation of the Ventricular Outflow Tracts

The distal part of the right ventricle and outflow tract originate from a secondary heart field.