Ultrasound Diagnosis & Prognosis of Common Cardiac Abnormalities



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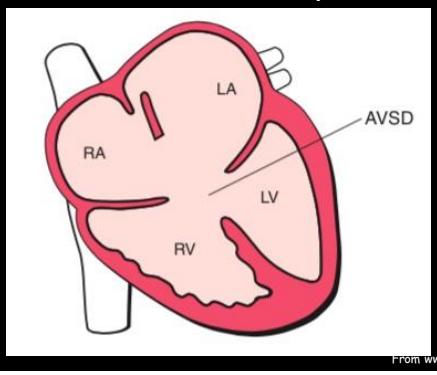
Common Cardiac Abnormalities

- Atrioventricular Septal Defect
- Hypoplastic Left Heart Syndrome
- Tetralogy of Fallot
- Transposition of Great Arteries

2

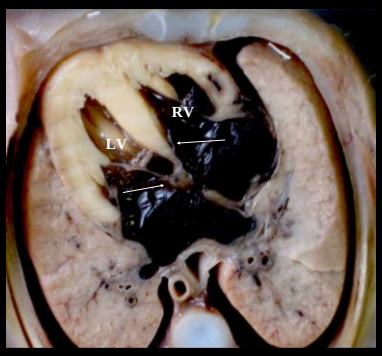
Atrioventricular Septal Defect



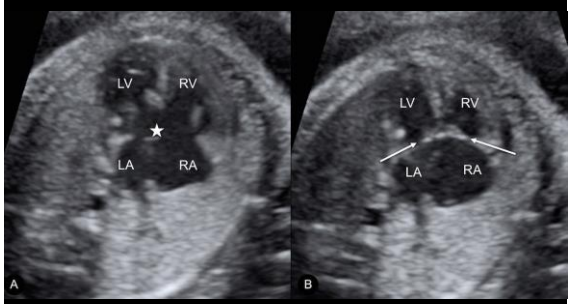
From www.PedHeart.com

Atrioventricular Valves

Normal



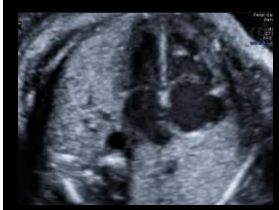
Atrioventricular Septal Defect



Diastole Systole

Atrioventricular Septal Defect

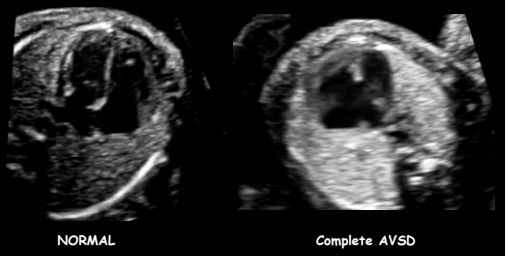
Four-chamber view




NORMAL Complete AVSD

Atrioventricular Septal Defect

Four-chamber view



NORMAL

Complete AVSD

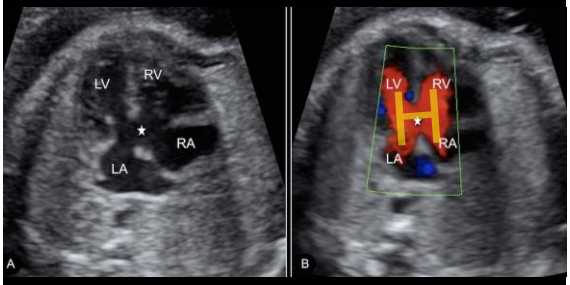
Atrioventricular Septal Defect

Ultrasound Findings:

- Apical 4-chamber view – most optimal
  Linear arrangement of valves

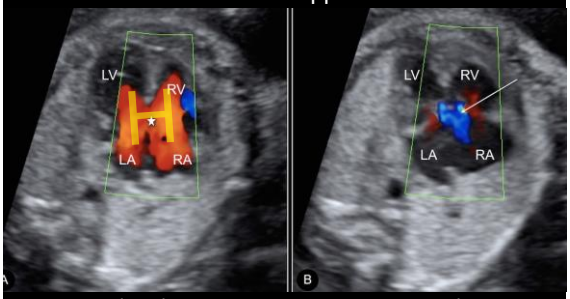
Atrioventricular Septal Defect

Color Doppler



Atrioventricular Septal Defect

Color Doppler

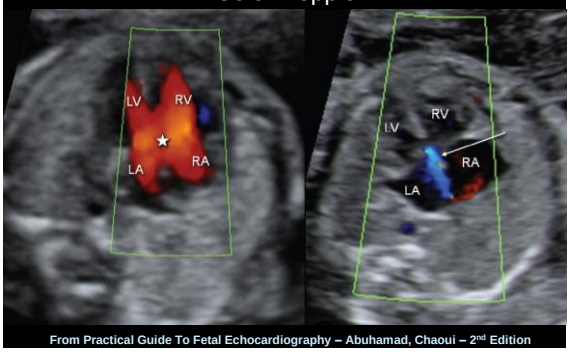


Diastole

Systole

Atrioventricular Septal Defect

Color Doppler



From Practical Guide To Fetal Echocardiography – Abuhamad, Chaoui – 2nd Edition

Atrioventricular Septal Defect

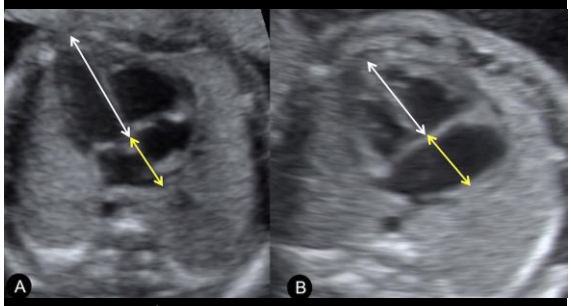
Color Doppler

Atrioventricular Septal Defect

Ultrasound Findings:

- Apical 4-chamber view – most optimal
- Color Doppler very helpful
  - Regurgitation & H sign

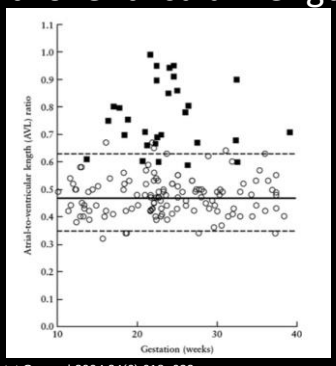
Atrioventricular Length



Normal AVSD

Ultrasound Obstet Gynecol 2004;24(6):618–622

Atrioventricular Length



Ultrasound Obstet Gynecol 2004;24(6):618–622

Atrioventricular Septal Defect

Ultrasound Findings:

- Apical 4-chamber view – most optimal
- Color Doppler very helpful
- Look at atrioventricular length ratio
  - Increased in AVSD

Atrioventricular Septal Defect

Short axis view

NORMAL Complete AVSD

Atrioventricular Septal Defect

Ultrasound Findings:

- Apical 4-chamber view – most optimal
- Color Doppler very helpful
- Look at atrioventricular length ratio
- Short-axis views are diagnostic
  - Single valve with multiple leaflets

Atrioventricular Septal Defect

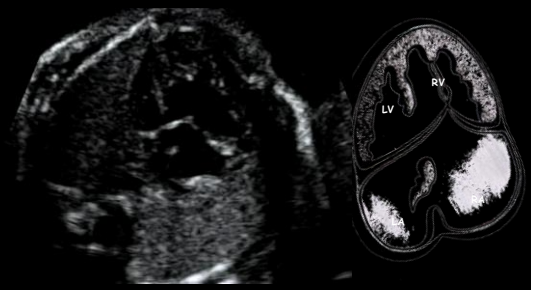
- Accounts for 4-5% of CHD in infants and occurs in 0.19 per 1000 live births.
- Most common form of CHD diagnosed in the fetus, accounting for 18% of fetal CHD
- Synonyms include endocardial cushion defect and atrioventricular canal defect

AVSD forms

⇩ ⇩

Balanced | Unbalanced

Atrioventricular Septal Defect



Unbalanced type

Atrioventricular Septal Defect

Partial

- Atrial septum primum defect
- Cleft in mitral valve
- Two distinct mitral & tricuspid valve annuli

Atrioventricular Septal Defect

Partial

AVSD and Down syndrome

➢AVSD accounts for 40% of congenital heart disease in children with Down syndrome

➢AVSD does NOT change the survival rate in patients with Down syndrome

1) Complete balanced AVSD

2) Partial AVSD

3) Complete Unbalanced AVSD

Risk for DS

Atrioventricular Septal Defect

Clues to Diagnosis

- Apical 4-chamber view is the most optimal
- Color Doppler very helpful (H sign / regurgitation)
- Short-axis views are diagnostic
- Note the atrioventricular length
- Partial & small defects are difficult to detect

Hypoplastic Left Heart Syndrome

Hypoplastic Left Heart Syndrome

Spectrum of Malformations

Severe hypoplasia of left ventricle
and left ventricular outflow tract

⬇

Obstruction to Systemic Cardiac Output

Hypoplastic Left Heart Syndrome

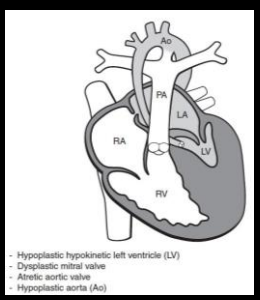
Classic Types:

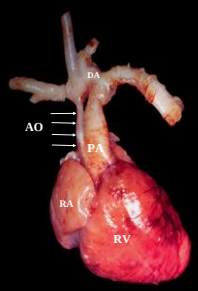
Mitral & aortic atresia

Aortic atresia (Patent MV)

Ultrasound Obstet Gynecol 2000;4:271

Hypoplastic Left Heart Syndrome





From: A Practical Guide to Fetal Echocardiography – Abuhamad, Chaoui

Hypoplastic Left Heart Syndrome

Variants:

- Critical AS with hypoplastic LV
- Severe coarctation of Aorta
- Severely unbalanced AVSD

Ultrasound Obstet Gynecol 2000;4:271

Hypoplastic Left Heart Syndrome

- Incidence: 0.1 – 0.25 / 1000 live births
- Karyotypic abnormalities in 5 %
- Extracardiac anomalies in up to 28%
- Preponderance in ♂ fetuses (7/10)
- Represents 5 % of all CHD

Ultrasound Obstet Gynecol 2000;4:271
Pediatrics 1988;82:698

Hypoplastic Left Heart Syndrome

Prenatal US follow-up

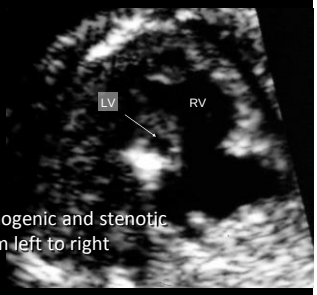
Fetal Growth Restriction due to

20 % reduction in combined cardiac output

Am J Epidemiol 1996;143:505

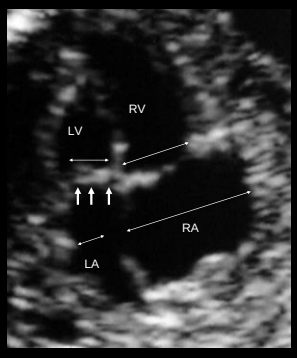
Hypoplastic Left Heart Syndrome

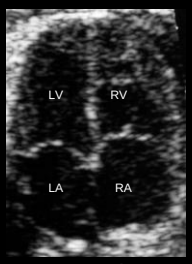
Four-Chamber View



- Diminutive, hypertrophic,
- hypokinetic LV
- RV apex forming
- Mitral valve dysplastic echogenic and stenotic
- Foramen ovale leaflet from left to right
- Color Doppler fills RV only

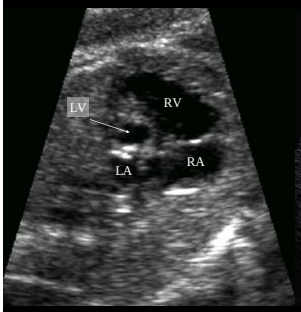
Hypoplastic Left Heart Syndrome



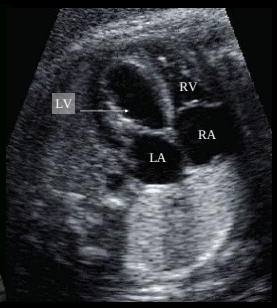


Normal

Hypoplastic Left Heart Syndrome



Aortic & mitral atresia



Critical aortic stenosis

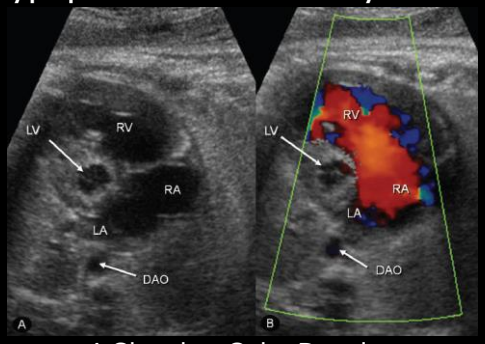
Hypoplastic Left Heart Syndrome

Four-Chamber View

Hypoplastic Left Heart Syndrome

Four-Chamber View

Hypoplastic Left Heart Syndrome



4-Chamber Color Doppler

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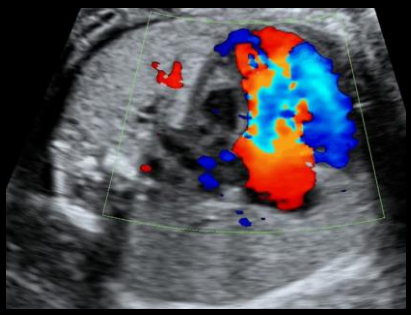
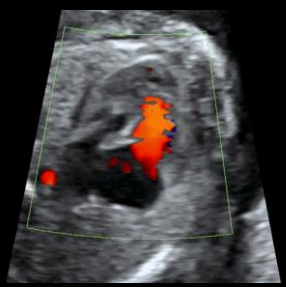
Hypoplastic Left Heart Syndrome



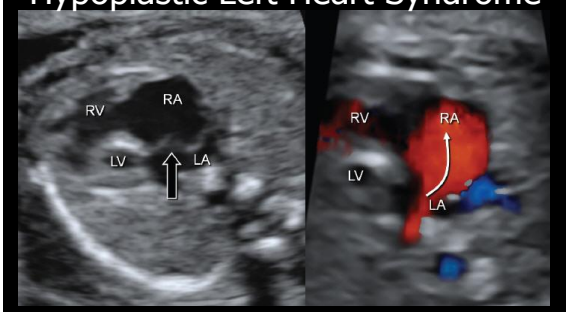
4-Chamber Color Doppler

Hypoplastic Left Heart Syndrome



4-Chamber Color Doppler

Hypoplastic Left Heart Syndrome

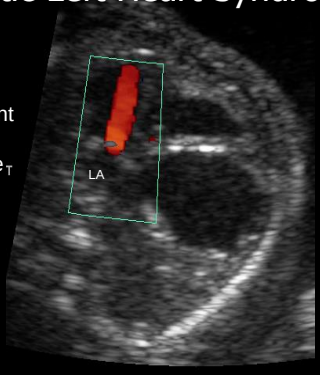


Color Doppler – Foramen Ovale

From: A Practical Guide to Fetal Echocardiography – Abuhamad, Chaoui

Hypoplastic Left Heart Syndrome

Left-to-right shunt
At
Foramen Ovale



Hypoplastic Left Heart Syndrome

Left Ventricular Outflow Tract

Hypoplastic Left Heart Syndrome

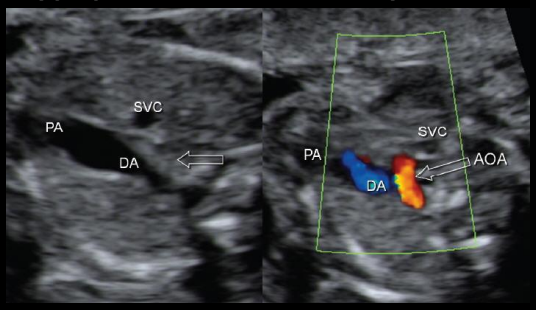
Right Ventricular Outflow Tract

Hypoplastic Left Heart Syndrome

Three-Vessel Trachea View

- Two vessels are seen
- Dilated PA, next to SVC
- Nonvisible or diminutive aorta
- Color Doppler shows reverse flow in aortic isthmus

Hypoplastic Left Heart Syndrome



3-Vessel Trachea View

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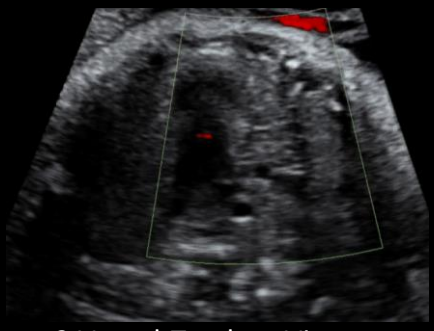
Hypoplastic Left Heart Syndrome



3 Vessel-Trachea View

Hypoplastic Left Heart Syndrome

3 Vessel-Trachea View

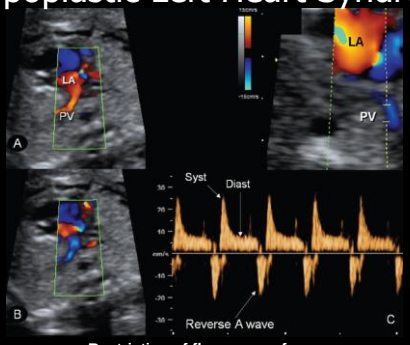
Hypoplastic Left Heart Syndrome

Prenatal Follow-up:

- Monthly
- Fetal growth (Fetal Growth Restriction)
- Size inter-atrial communication (PV Doppler)
- Function of tricuspid valve - RV

Semin Fetal Neonatal Med 2005;10(6):553

Hypoplastic Left Heart Syndrome



Restriction of flow across foramen

From: A Practical Guide to Fetal Echocardiography – Abuhamad, Chaoui

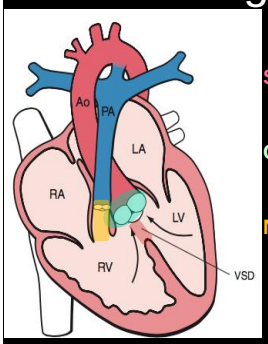
Hypoplastic Left Heart Syndrome

Poor Prognostic Factors:

- Low birth weight
- Prematurity
- Non-cardiac anomalies
- Obstruction to PV return (restricted atrial septum)
- Poor RV function

Ultrasound Obstet Gynecol 2000;4:271
Pediatrics 2007;119(1):109

Tetralogy of Fallot



Subaortic malaligned VSD

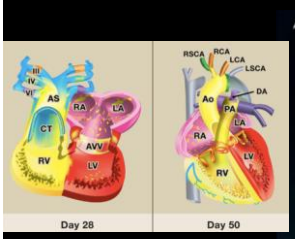
Overriding dilated aortic root

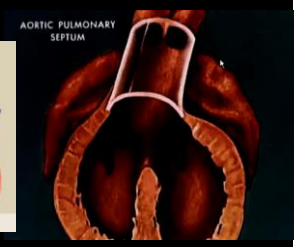
Narrow stenotic PA

RV hypertrophy is not present in fetus

From Practical Guide To Fetal Echocardiography – Abuhamad, Chaoui – 2nd Edition

Embryology of Conotruncus





Srivastava - Cell 2006
https://www.youtube.com/watch?v=5DlUk9IXUaI - Rushmer, Blandau

52

Tetralogy of Fallot

- Classic form (PS) (~ 80 %)
- Pulmonary atresia with VSD
- Absent pulmonary valve

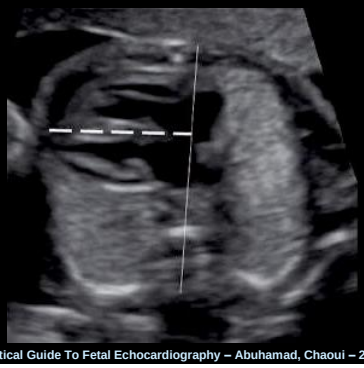
Tetralogy of Fallot

Ultrasound Findings:

- Four chamber view normal
- Left axis deviation

TOF – 4CV

TOF – Cardiac Axis



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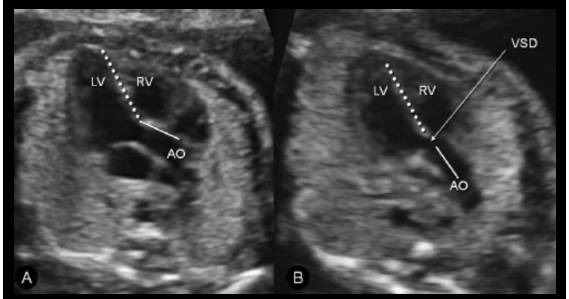
TOF – Cardiac Axis

TOF - LVOT



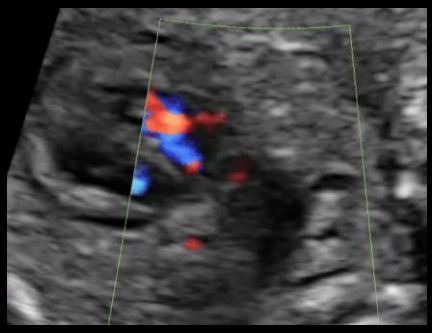
Normal

TOF

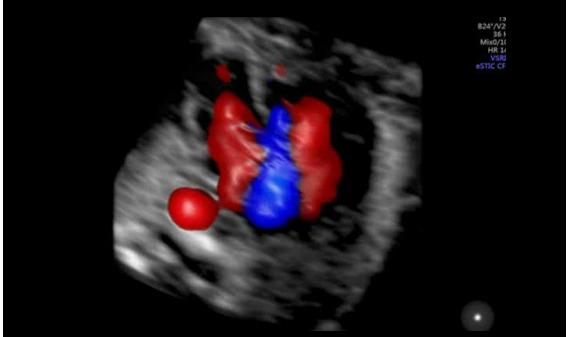
TOF - LVOT

TOF - LVOT

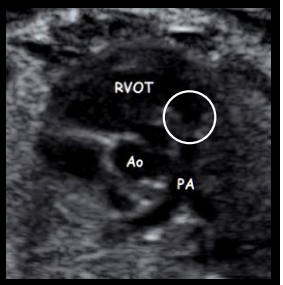
TOF - LVOT



TOF - LVOT



TOF - RVOT



Normal

TOF

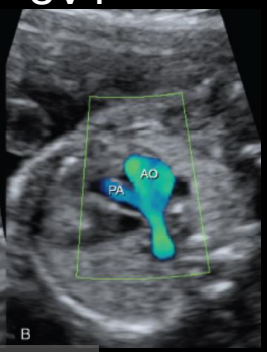
TOF - RVOT

TOF - RVOT

5-Chamber View

3-Vessel View

TOF – 3VT



3-Vessel Trachea View

Tetralogy of Fallot

Ultrasound Findings:

- Five chamber view abnormal
- Aortic dextroposition
- Dilated aortic root (3rd trimester)
- Perimembranous subaortic VSD
- Infundibular pulmonary stenosis

Tetralogy of Fallot

Common Associated Cardiac Anomalies

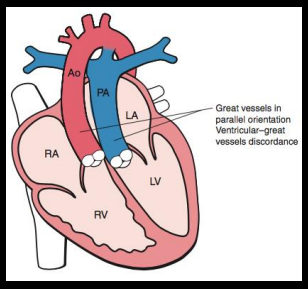
- Patent foramen ovale/ ASD in 85%
- Right sided Aortic arch in 25%
- Persistent LSVC in 11%

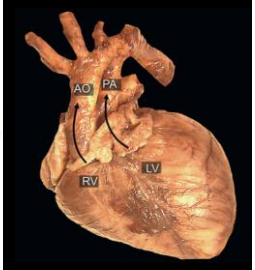
Tetralogy of Fallot

Associated Extracardiac Anomalies

- Chromosomal abnormalities in 30%
- Anomalies of anatomic organs, common
- Deletion 22q11 (DiGeorge) in 10-15%
  - Right Aortic arch
  - Thymic hypogenesis / agenesis
  - Extracardiac anomalies
  - Polyhydramnios

Transposition of Great Arteries

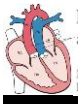




70

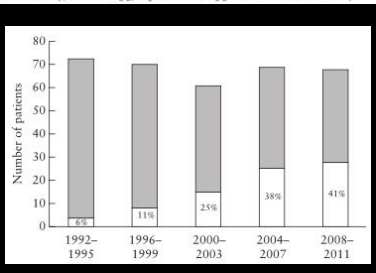
How often is TGA detected prenatally?

71



Prenatal diagnosis of transposition of the great arteries over a 20-year period: improved but imperfect

M. C. ESCOBAR-DIAZ*†‡, L. R. FREUD*†, A. BUENO*†, D. W. BROWN*†, K. G. FRIEDMAN*†, D. SCHIDLOW*†, S. EMANI§¶, P. J. DEL NIDO§¶ and W. TWORETZKY*†



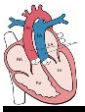
Overall rate of detection 23 %

Ultrasound Obstet Gynecol 2015; 45: 678

72

Does it matter to detect TGA prenatally?

73



Prenatal detection of transposition of the great arteries reduces mortality and morbidity

C. L. VAN VELZEN*, M. C. HAAK†, G. REIJNDERS*, M. E. B. RIJLAARSDAM‡, C. J. BAX*, E. PAJKRT§, J. HRUDA¶, F. GALINDO-GARRE**, C. M. BILARDO††, C. J. M. DE GROOT*, N. A. BLOM‡^ and S. A. CLUR^

1st year mortality overall 4.9 %

0% in prenatal diagnosis

11.4% without prenatal diagnosis

Table 2 Frequency and outcome of indicators of presurgical morbidity in 139 infants with transposition of the great arteries (TGA) between 2002 and 2012

Clinical characteristic	*Category of TGA*	*With prenatal diagnosis (n = 34)*	*Without prenatal diagnosis (n = 105)*	P
Lowest pH value presurgery	All	7.20 ± 0.15	7.24 ± 0.12	0.088
	Simple	7.19 ± 0.16	7.24 ± 0.14	0.113
Highest lactate level presurgery (mMol/L)	All	4.75 ± 2.45	4.97 ± 4.82	0.807
	Simple	4.82 ± 2.66	5.44 ± 5.46	0.589
Oxygen saturation at admission (mmHg)†	All	73.6 ± 15.9	67.0 ± 15.55	0.048*
	Simple	69.4 ± 15.6	63.3 ± 15.1	0.099
Renal†	All	1/23 (4.3)	17/89 (19.1)	0.039*
dysfunction†	Simple	1/18 (5.6)	15/69 (21.7)	0.021*
AST (units/L)‡	All	60.9 ± 23.9	204.5 ± 941.5	0.633
	Simple	59.3 ± 26.1	256.0 ± 1085.0	0.613
ALT (units/L)‡	All	38.7 ± 29.5	90.2 ± 489.6	0.730
	Simple	32.4 ± 18.3	112.6 ± 561.9	0.673
Closure of duct before PGE therapy†	All	—	21/89 (23.6)	0.000*
	Simple	—	13/66 (19.7)	0.002*
Inotropes presurgery	All	7/33 (21.2)	27/103 (26.2)	0.602
	Simple	7/25 (28.0)	22/77 (28.6)	0.712
Rashkind procedure	All	20/33 (60.6)	67/104 (64.4)	0.691
	Simple	17/25 (68.0)	57/77 (74.0)	0.557

Ultrasound Obstet Gynecol 2015; 45: 320–325

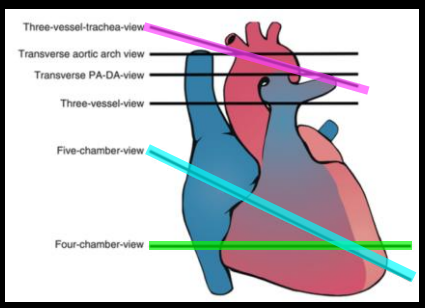
Transposition of Great Arteries

Ultrasound Views for Diagnosis

Four-chamber view
Left Ventricular outflow tract
Three-vessel trachea view
Spatial orientation of great arteries

75

Transverse Views



TGA: Four-Chamber View

77

TGA: Four-Chamber View

TGA with Mesocardia

TGA: Four-Chamber View

Four-chamber view is typically normal except for:

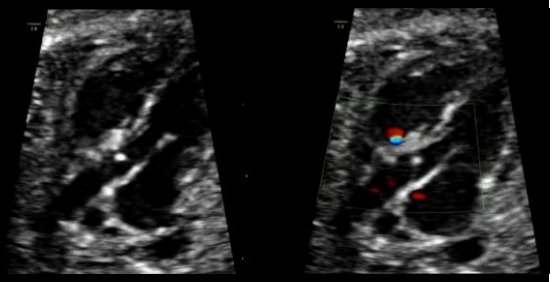
- Associated VSD
- Mesocardia
- Significant LV Outflow obstruction

79

TGA: Left Ventricular Outflow

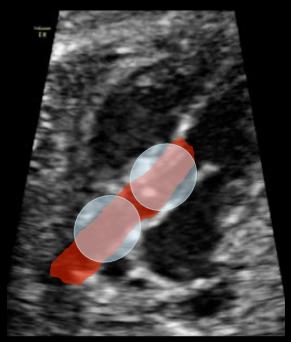
80

Left Ventricular Outflow



Normal

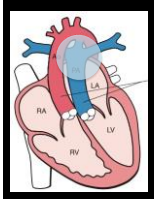
Left Ventricular Outflow



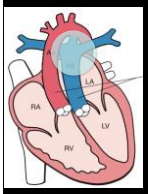
Left Ventricular Outflow

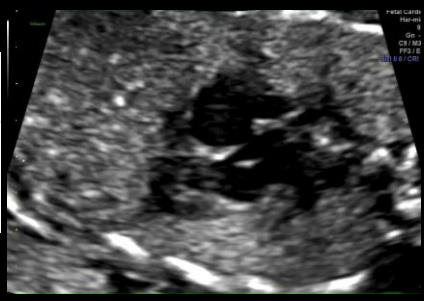
Normal

TGA: Left Ventricular Outflow

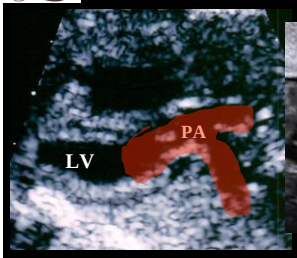


TGA: Left Ventricular Outflow





TGA: Left Ventricular Outflow



TGA: Left Ventricular Outflow

Left ventricular outflow view is always abnormal:

- No override
- LV outflow divides into 2 branches

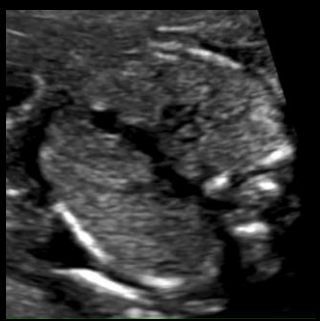
87

3VT View

13 wks

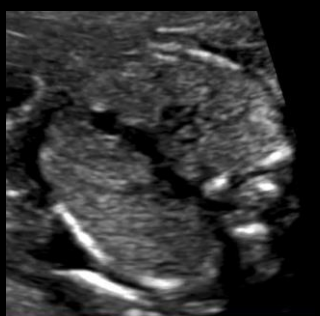
Normal

TGA: 3VT View



89

Transposition of Great Arteries

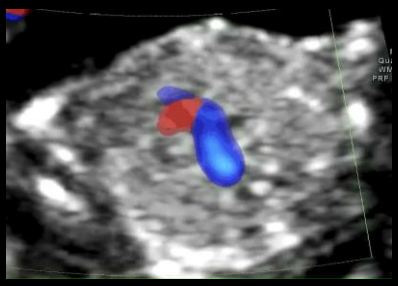


3VT – 2 vessels seen

Transposition of Great Arteries

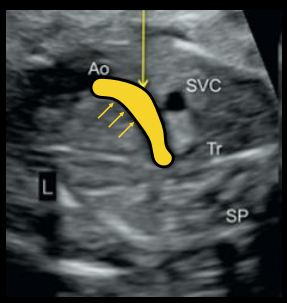
3VT – 2 vessels seen

Transposition of Great Arteries



3VT – 2 vessels seen

Transposition of Great Arteries



Convex-shape of Aorta

TGA: 3VT View

3VT view is very commonly abnormal:

- 2 vessels seen
- Aorta and SVC
- PA is posterior to Ao
- Ao has a convex appearance

95

TGA: Orientation of Great Arteries

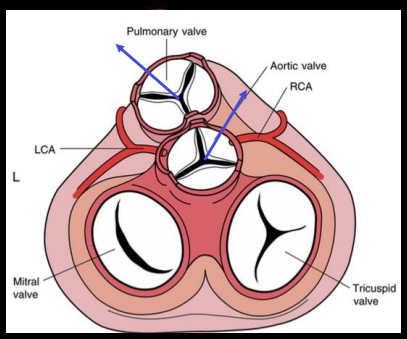
Aorta

- Points to right shoulder
- Posterior to PA
- Gives head & neck vessels
- Aortic arch (candy cane)

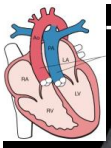
Pulmonary

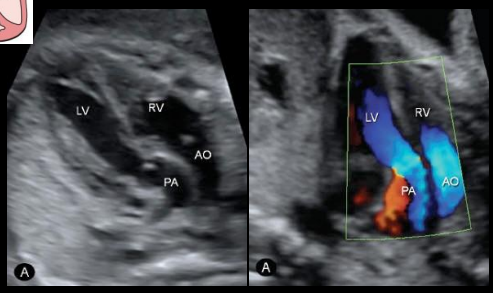
- Points to left shoulder
- Anterior to aorta
- Divides to right & left
- Ductal arch (hockey stick)

Orientation of Great Arteries



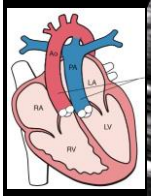
Transposition of Great Arteries



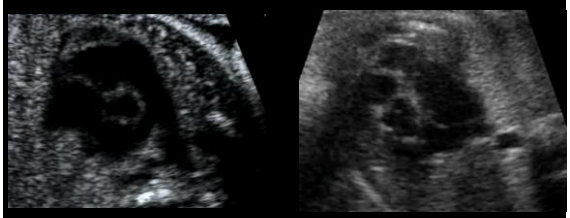


From Practical Guide To Fetal Echocardiography: Normal & Abnormal Hearts– Abuhamad, Chaoui – 3rd Edition -Oct 2015

Transposition of Great Arteries



Orientation of Great Arteries



Normal TGA

Common Cardiac Malformations

- Learn normal cardiac anatomy
- Understand anatomic features of cardiac malformations
- Know informative ultrasound planes for diagnosis
- Learn about associated cardiac and extracardiac malformations