Prenatal Evaluation of the Fetus with Transposition of the Great Arteries

Children's Mercy Fetal Cardiology Education Series

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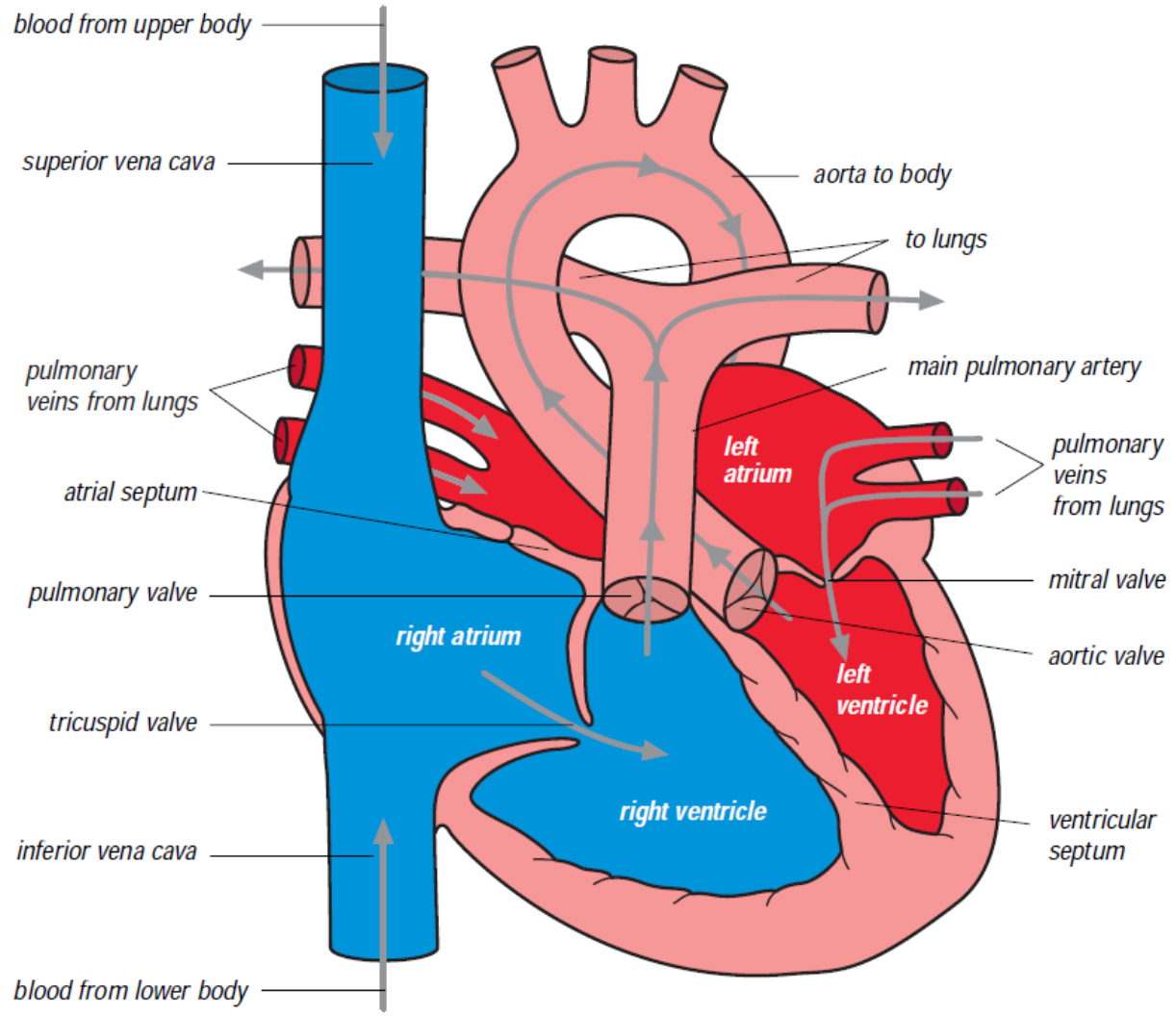
Disclosures

- No disclosures

Objectives

- Understand the basic anatomy of D-transposition of the great arteries (D-TGA)
- Define fetal imaging goals for D-TGA
- Recognize aspects of D-TGA anatomy on shared D-TGA fetal imaging cases
- Discuss prenatal and postnatal management and outcomes

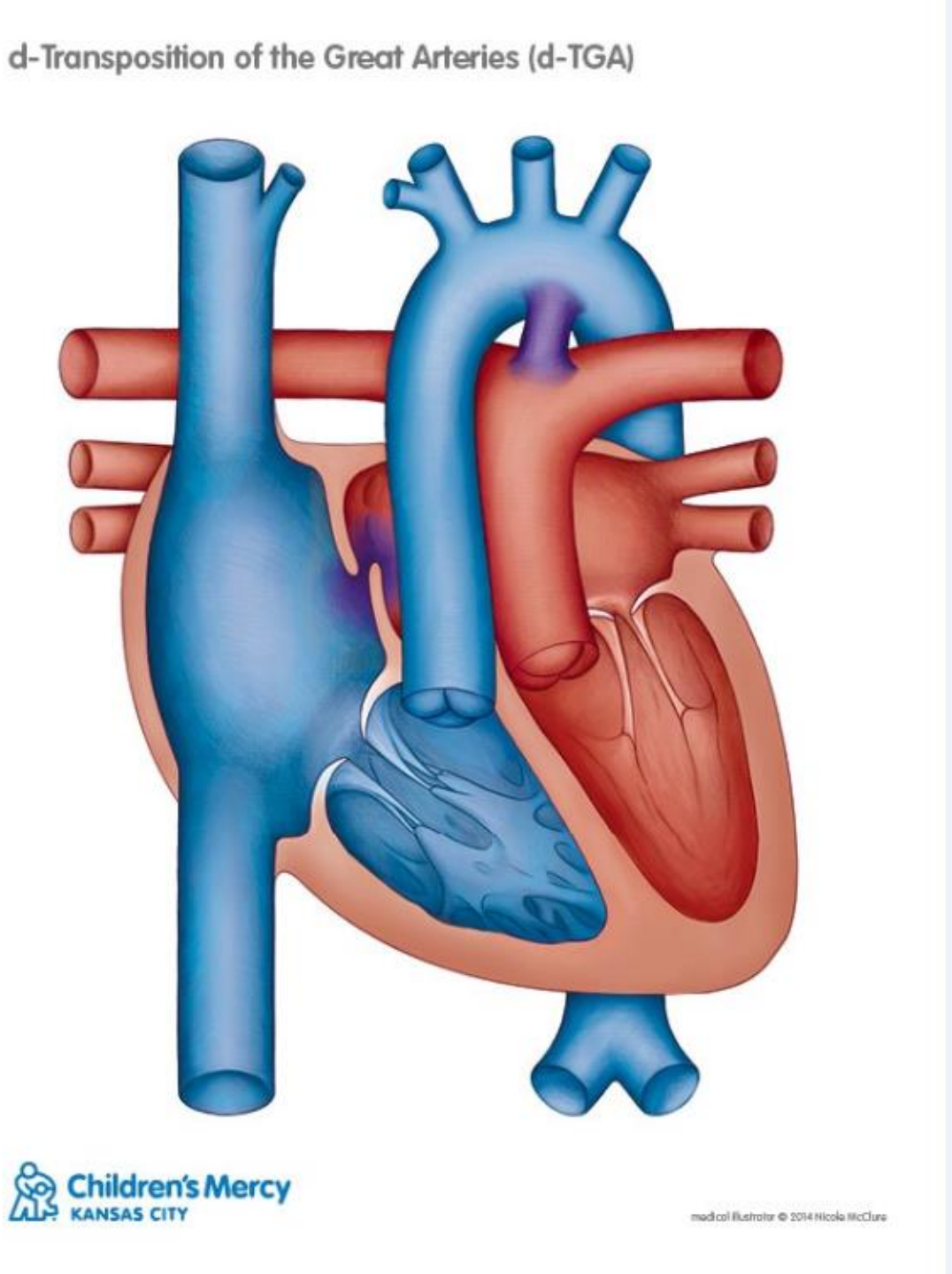
Normal Heart



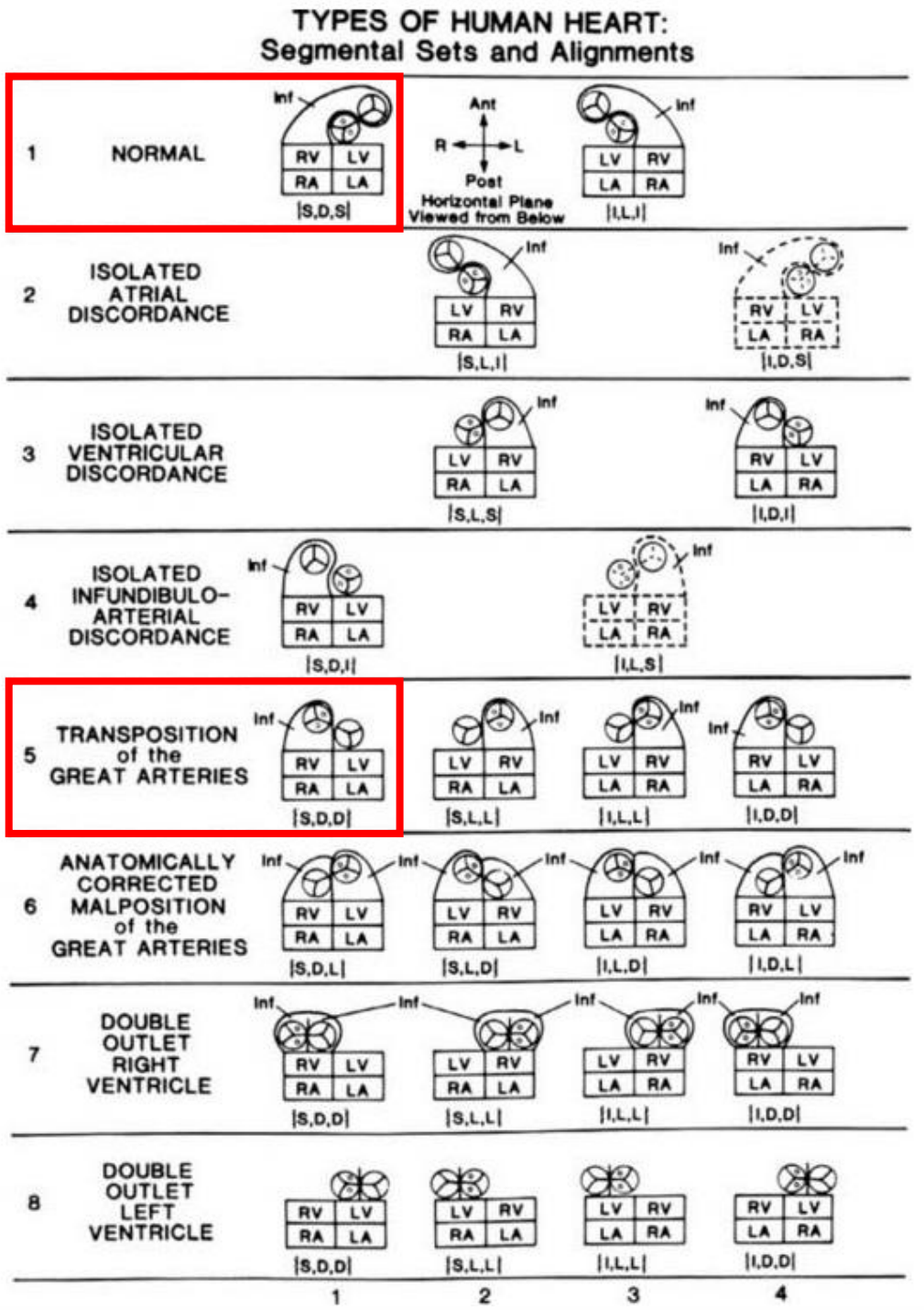
Anatomy of D-TGA

- Aorta arises from the RV and pulmonary artery from the LV (ventriculoarterial discordance)
- Differences from normal:
  - Aorta is anterior and rightward instead of posterior and rightward
  - Subaortic conus instead of subpulmonic conus
  - Fibrous continuity between mitral and pulmonary valves instead of mitral and aortic valves
- Atrioventricular concordance and normal D-looping of the ventricles, {S, D, D} segmental anatomy

Anatomy of D-TGA



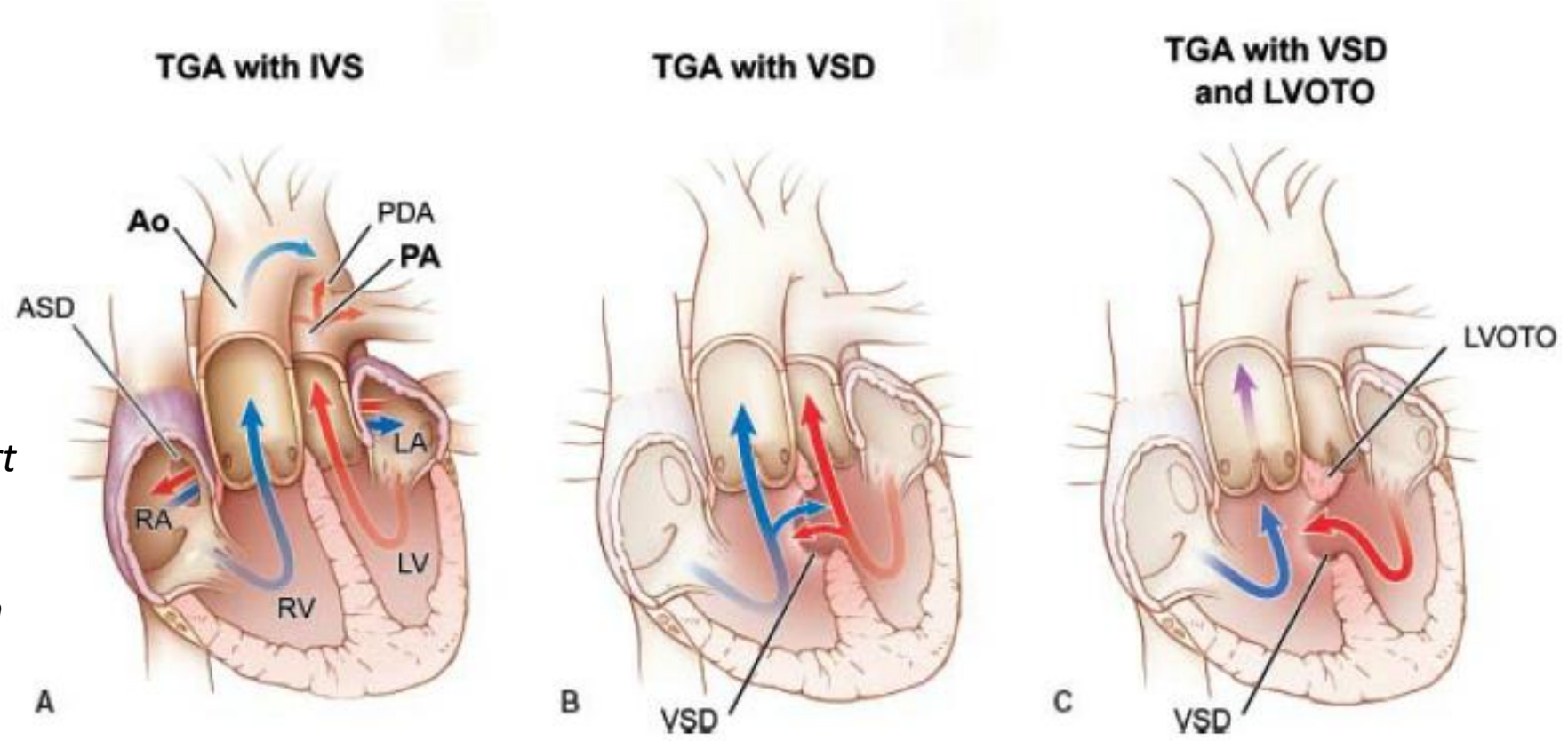
Anatomy of D-TGA



Foran RB, Belcourt C, Nanton MA, et al. Isolated infundibuloarterial inversion {S,D,I}: a newly recognized form of congenital heart disease. Am Heart J 1988;116:1337-50

Anatomy and Associated Lesions

- Approximately 50% of D-TGA fetuses have an intact ventricular septum



Moss and Adams' Heart Disease in Infants, Children, and Adolescents, 9th Edition

Frequency and Genetics

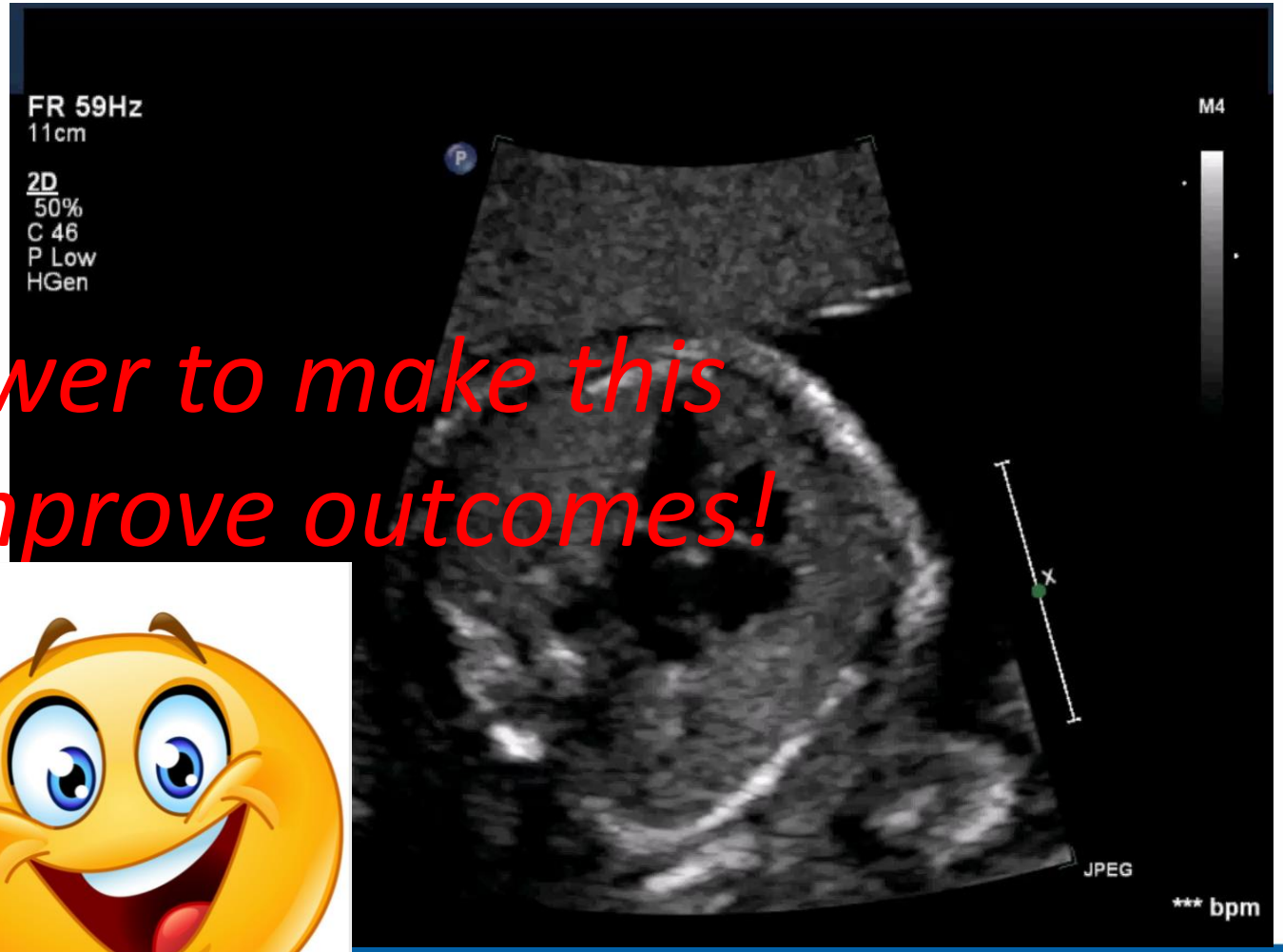
- Incidence: approximately 2.5 per 10,000 live births
- Common cause of cyanotic congenital heart disease
- Strong male predominance (60-70%)
- No known causative genetic abnormality
- Extracardiac differences are unusual

Prenatal Evaluation

- D-TGA is commonly missed on prenatal scanning!

Prenatal Evaluation

- 4C View will appear normal!



You have the power to make this diagnosis and improve outcomes!

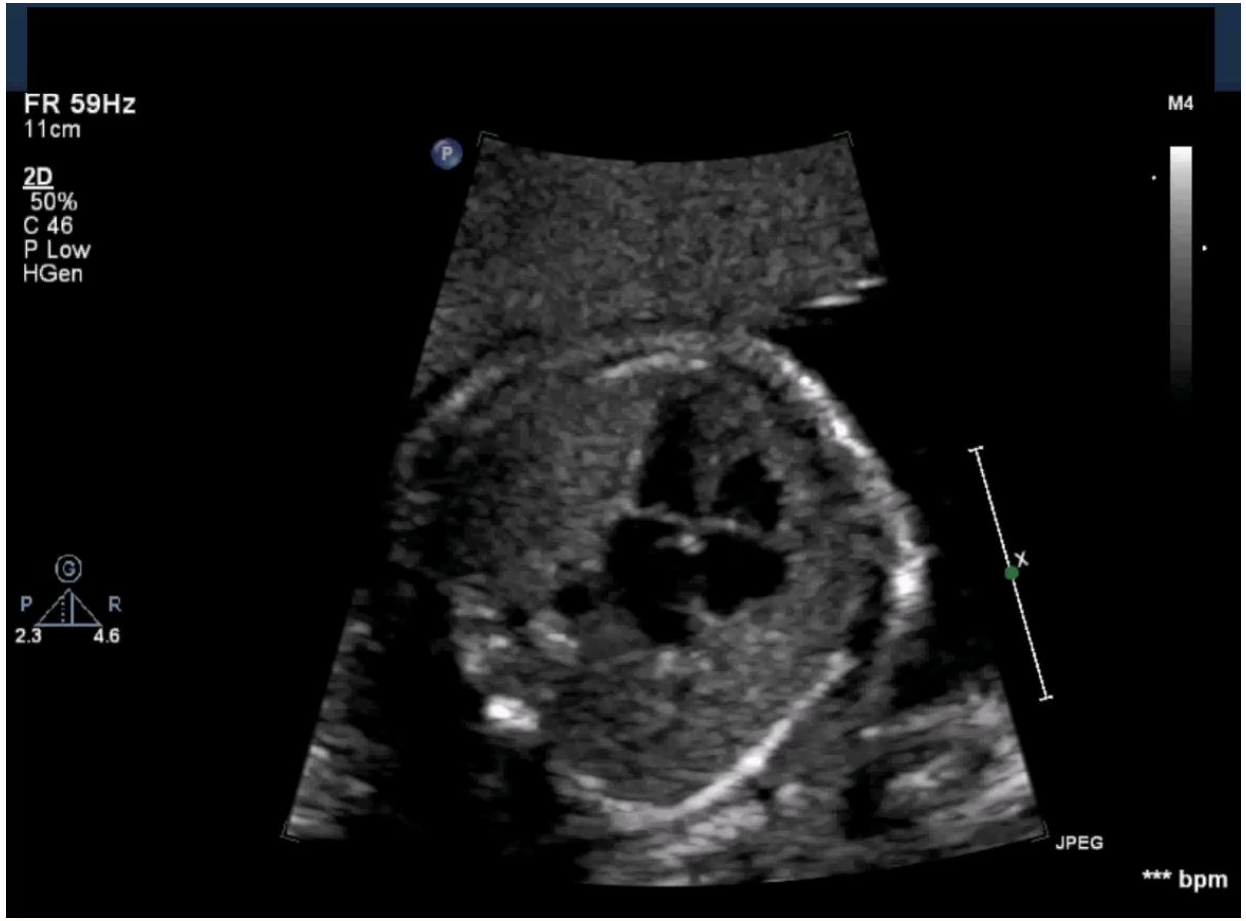
Prenatal Evaluation

- Diagnosis requires inspection of the great arteries
- Normally related great arteries will cross as you scan the heart moving cephalad
- D-TGA: great arteries arise in parallel and do not cross

Prenatal Evaluation

NORMAL

D-TGA



D-TGA Fetal Echo Features

Key Echocardiographic Features

- Presence or absence of great vessels crossing as they course just above the heart.
- Relationship of the great vessels to each other as they arise from the heart.
- Relative size of the great vessels—is the pulmonary artery larger than the aorta by approximately 25%, as is expected in transposition of the great arteries?
- Presence or absence of ventricular septal defects.
- Presence or absence of left or right outflow tract obstruction.
- Presence or absence of semilunar valve stenosis.
- Presence or absence of a patent ductus arteriosus with shunting from the pulmonary artery to the descending aorta.
- Ductus arteriosus is typically smaller than in normally related great vessels.
- Ductal and aortic arches appear as running parallel in their course in the superior mediastinum.
- Atrial septal defect and redundancy of the septum primum.

Rychik, Jack. Zhiyun, Tian. *Fetal Cardiovascular Imaging: A Disease-Based Approach.* Elsevier Saunders, 2012.

D-TGA Fetal Echo Features

- Two key structures predict postnatal stability
  - Foramen ovale
  - Ductus arteriosus
- Restriction will limit mixing of oxygenated and deoxygenated blood leading to severe hypoxemia and hemodynamic instability

Will the baby need a balloon atrial septostomy?

- BAS needed in 20-40% of neonates with D-TGA
- Done to improve mixing at the level of the atria and improve arterial oxygen saturation
- Challenging to accurately predict by prenatal data!

Will the baby need a balloon atrial septostomy?

FULL TEXT ARTICLE

Usefulness of the Prenatal Echocardiogram in Fetuses With Isolated Transposition of the Great Arteries to Predict the Need for Balloon Atrial Septostomy

Trisha V. Vigneswaran MRCPCH, Vita Zidere MD, Owen I. Miller MD, John M. Simpson MD and Gurleen K. Sharland BSc, MD

American Journal of Cardiology, The, 2017-05-01, Volume 119, Issue 9, Pages 1463-1467, Copyright © 2017 Elsevier Inc.

- Need for emergent BAS:
  - Foramen ovale/total septal length of <0.5
  - Fixed appearing septum primum flap

Will the baby need a balloon atrial septostomy?

FULL TEXT ARTICLE

Fetal Predictors of Urgent Balloon Atrial Septostomy in Neonates with Complete Transposition

Rajesh Punn MD and Norman H. Silverman MD, DSc (Med), FASE

Journal of the American Society of Echocardiography, 2011-04-01, Volume 24, Issue 4, Pages 425-430, Copyright © 2011 American Society of Echocardiography

- Need for emergent BAS:
  - Hypermobile septum with bidirectional flow
  - Reverse diastolic flow in the PDA

Will the baby need a balloon atrial septostomy?

- Need for emergent BAS:
  - Atrial septal excursion (ASE) ratio
  - Degree of redundancy of the atrial septum
  - >atrial redundancy = less likely to become restrictive after birth
  - No fetus with ratio >0.5 required urgent BAS

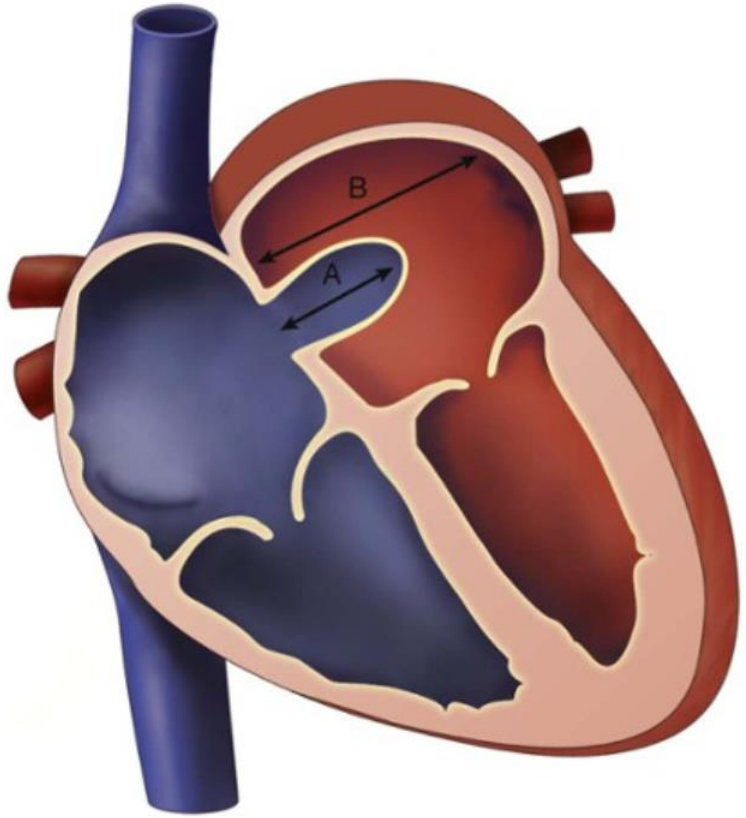
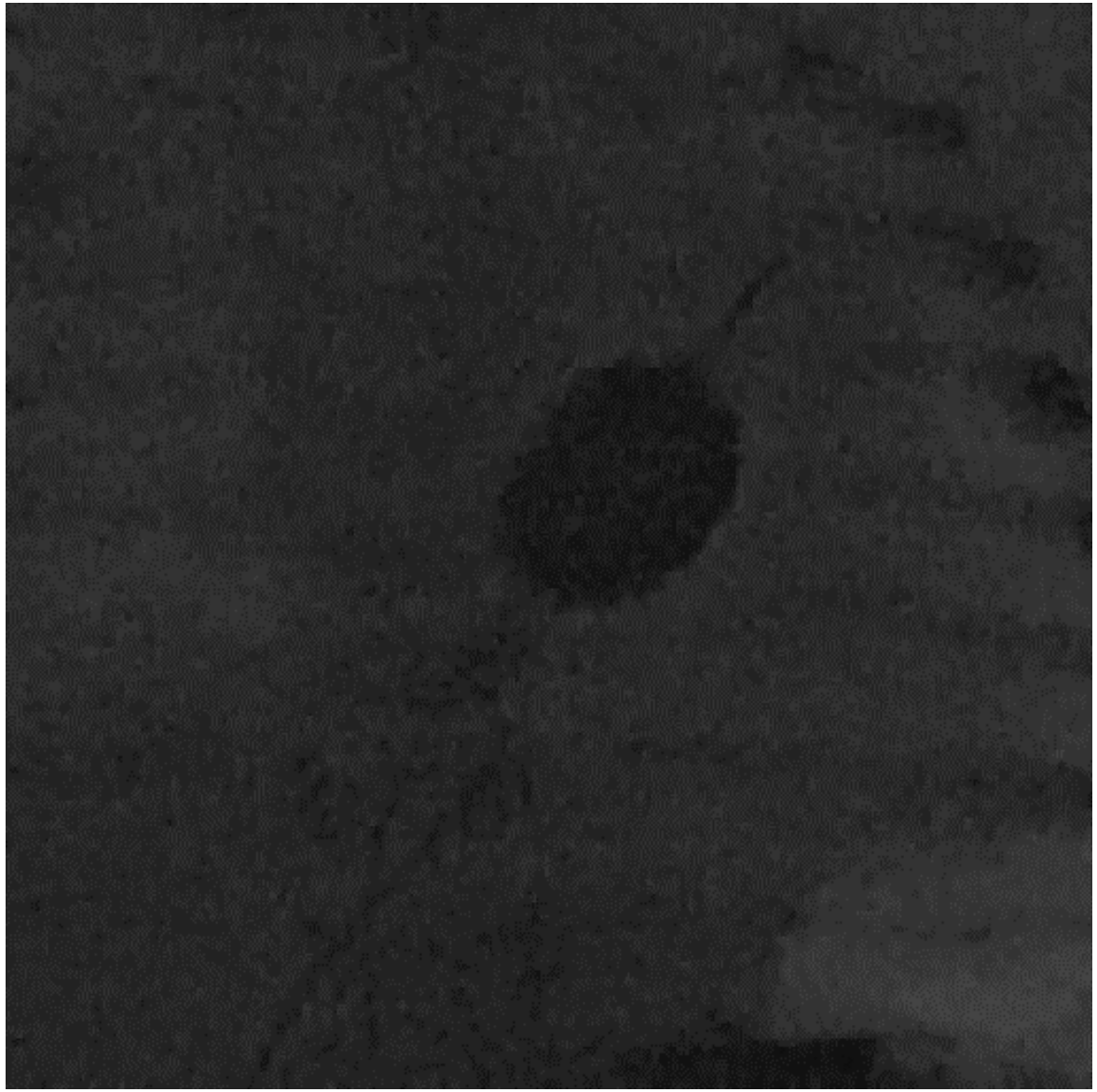


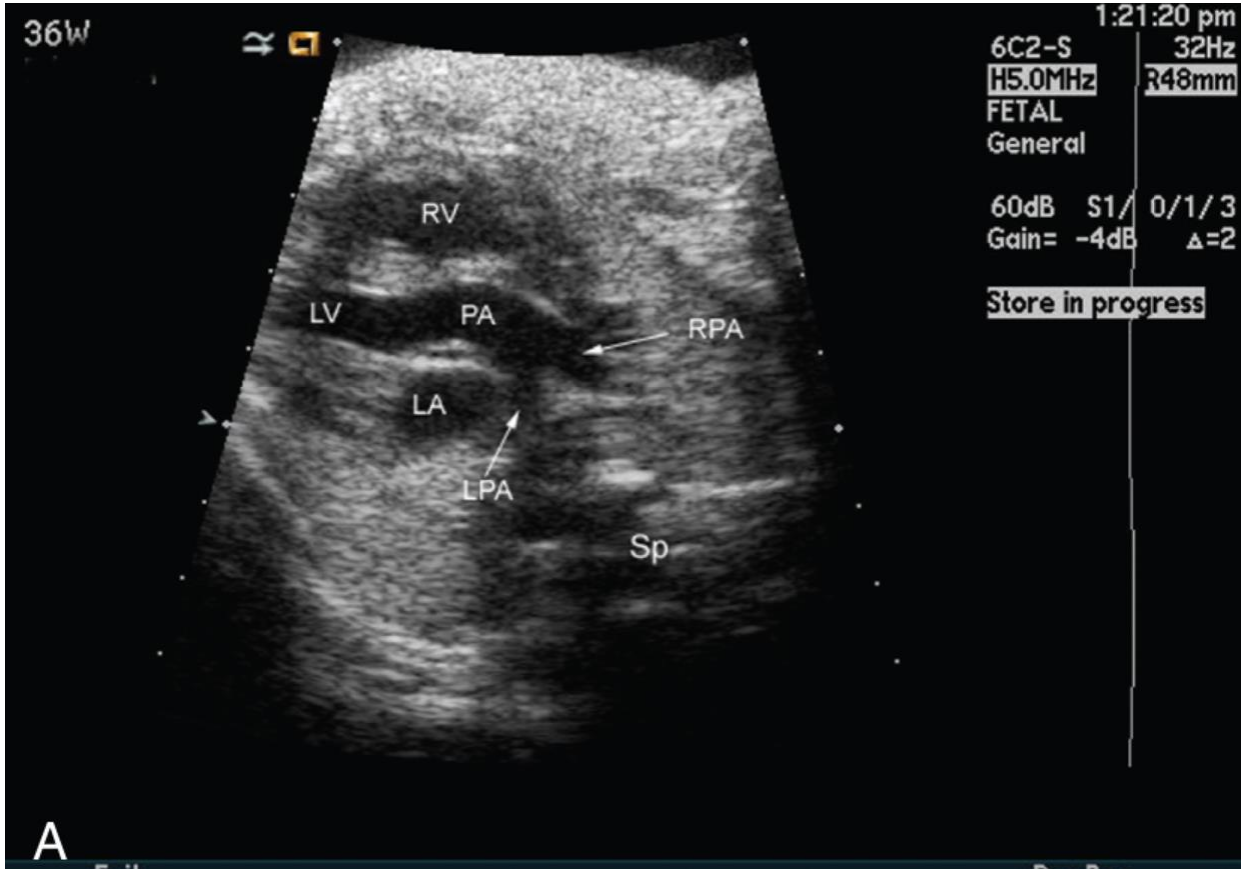
FIGURE 14-2 Calculation of the atrial septal excursion (ASE) ratio, an estimate of atrial …

Balloon atrial septostomy



D-TGA Fetal Echo Features

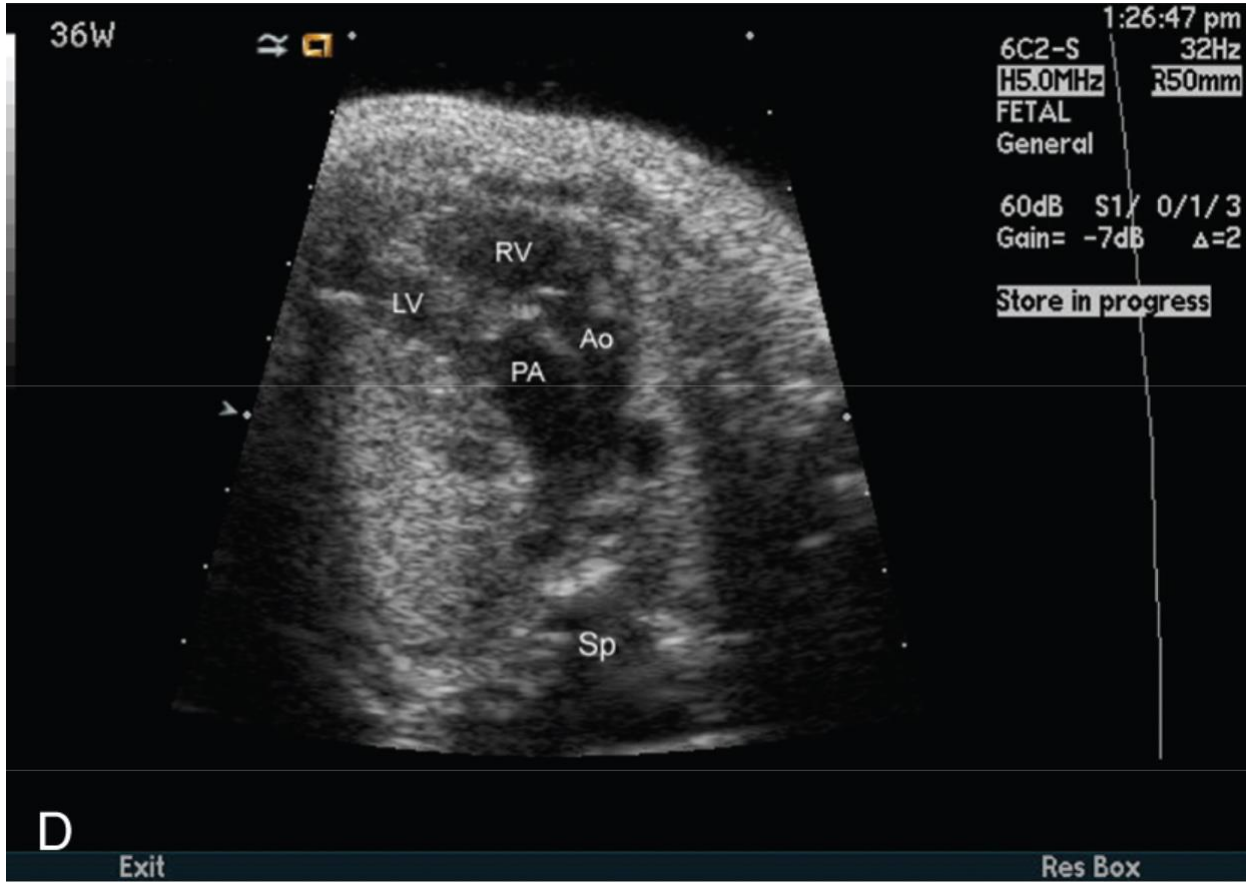
- Long axis of LV



Rychik, Jack. Zhiyun, Tian. *Fetal Cardiovascular Imaging: A Disease-Based Approach.* Elsevier Saunders, 2012.

D-TGA Fetal Echo Features

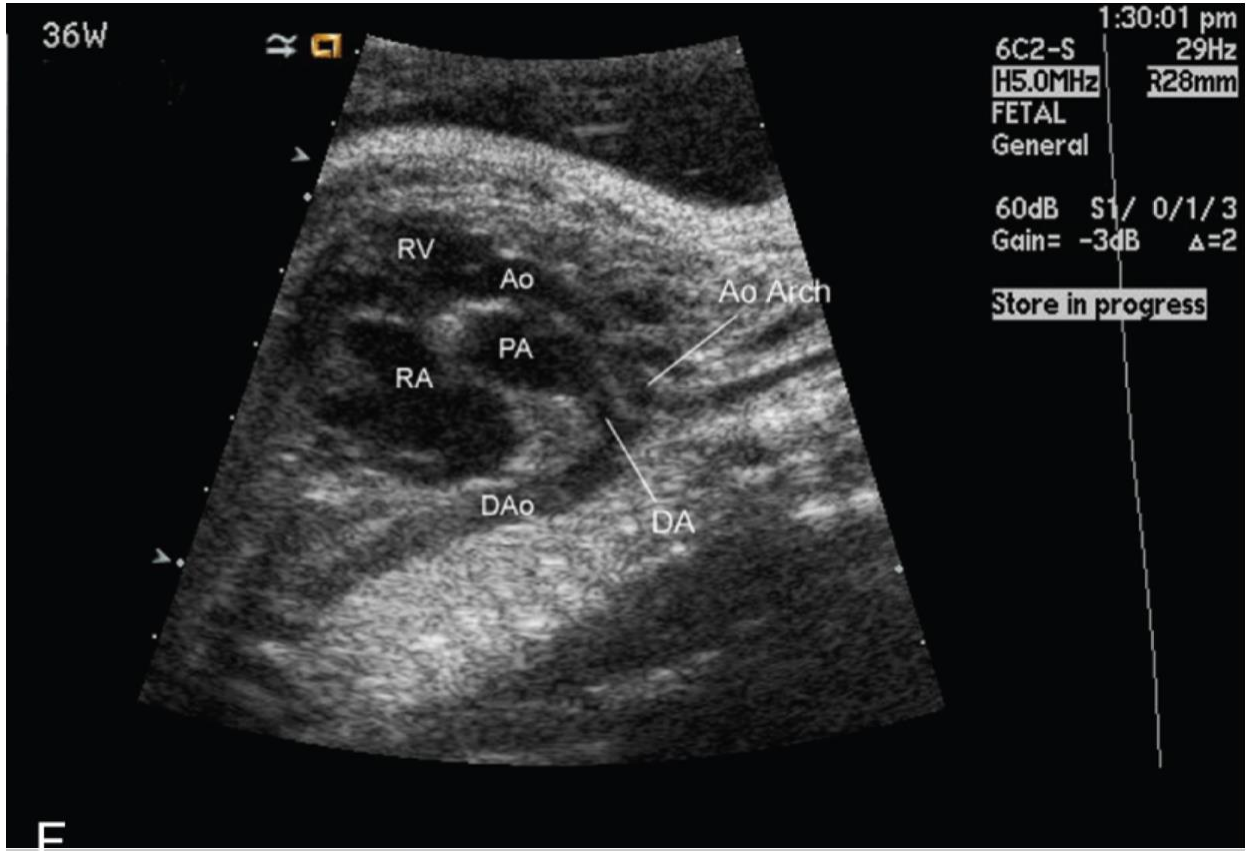
- Parallel great vessels



Rychik, Jack. Zhiyun, Tian. *Fetal Cardiovascular Imaging: A Disease-Based Approach.* Elsevier Saunders, 2012.

D-TGA Fetal Echo Features

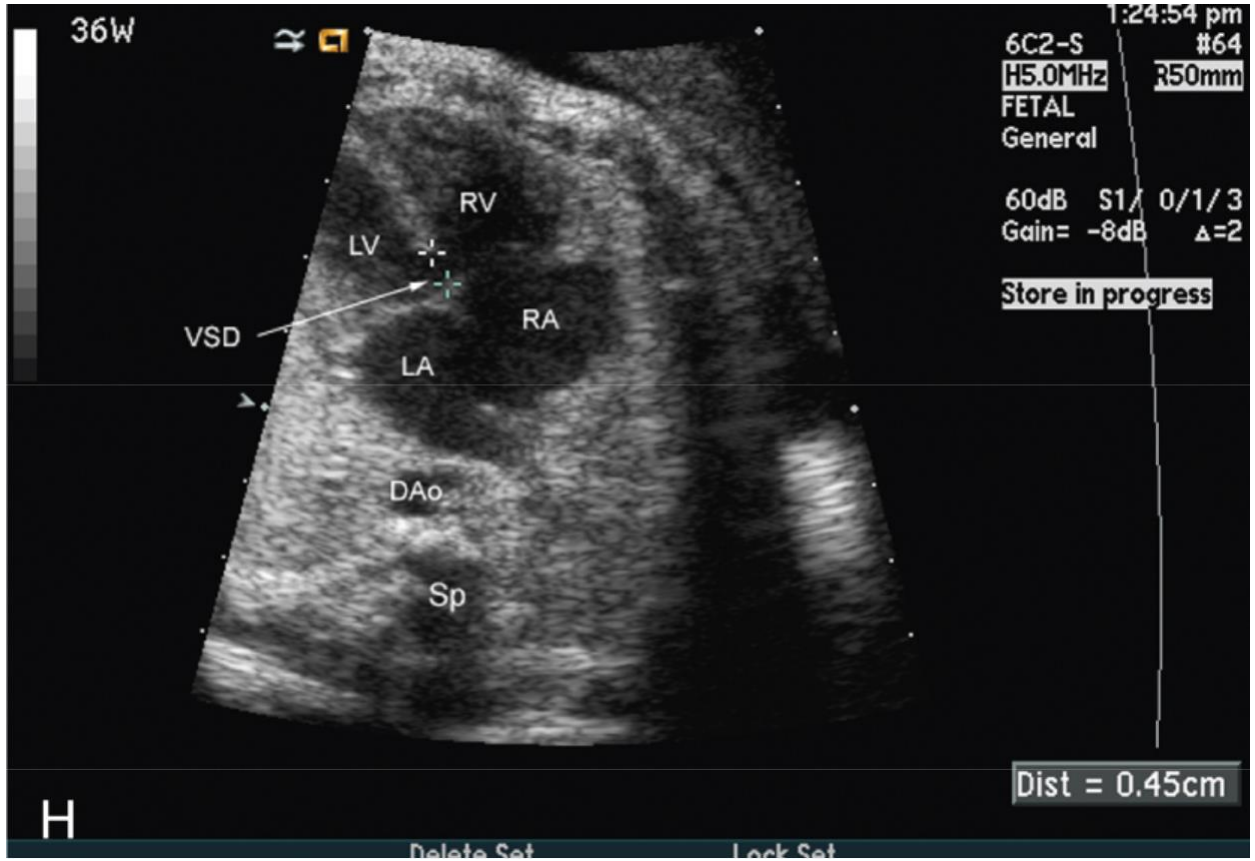
- Sagittal view of the parallel arches



Rychik, Jack. Zhiyun, Tian. *Fetal Cardiovascular Imaging: A Disease-Based Approach.* Elsevier Saunders, 2012.

D-TGA Fetal Echo Features

- Associated lesion: VSD



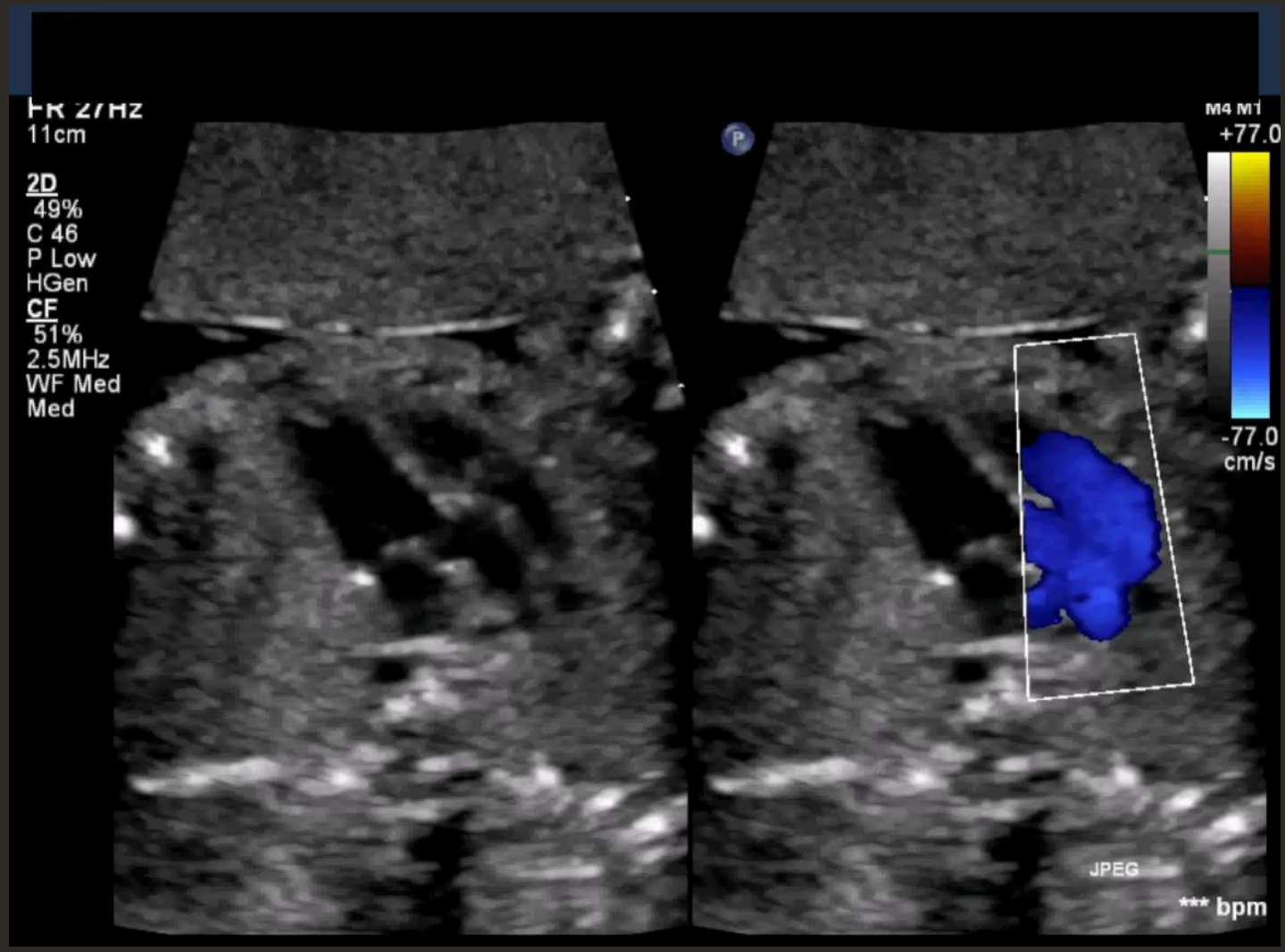
Rychik, Jack. Zhiyun, Tian. *Fetal Cardiovascular Imaging: A Disease-Based Approach.* Elsevier Saunders, 2012.

D-TGA Cases

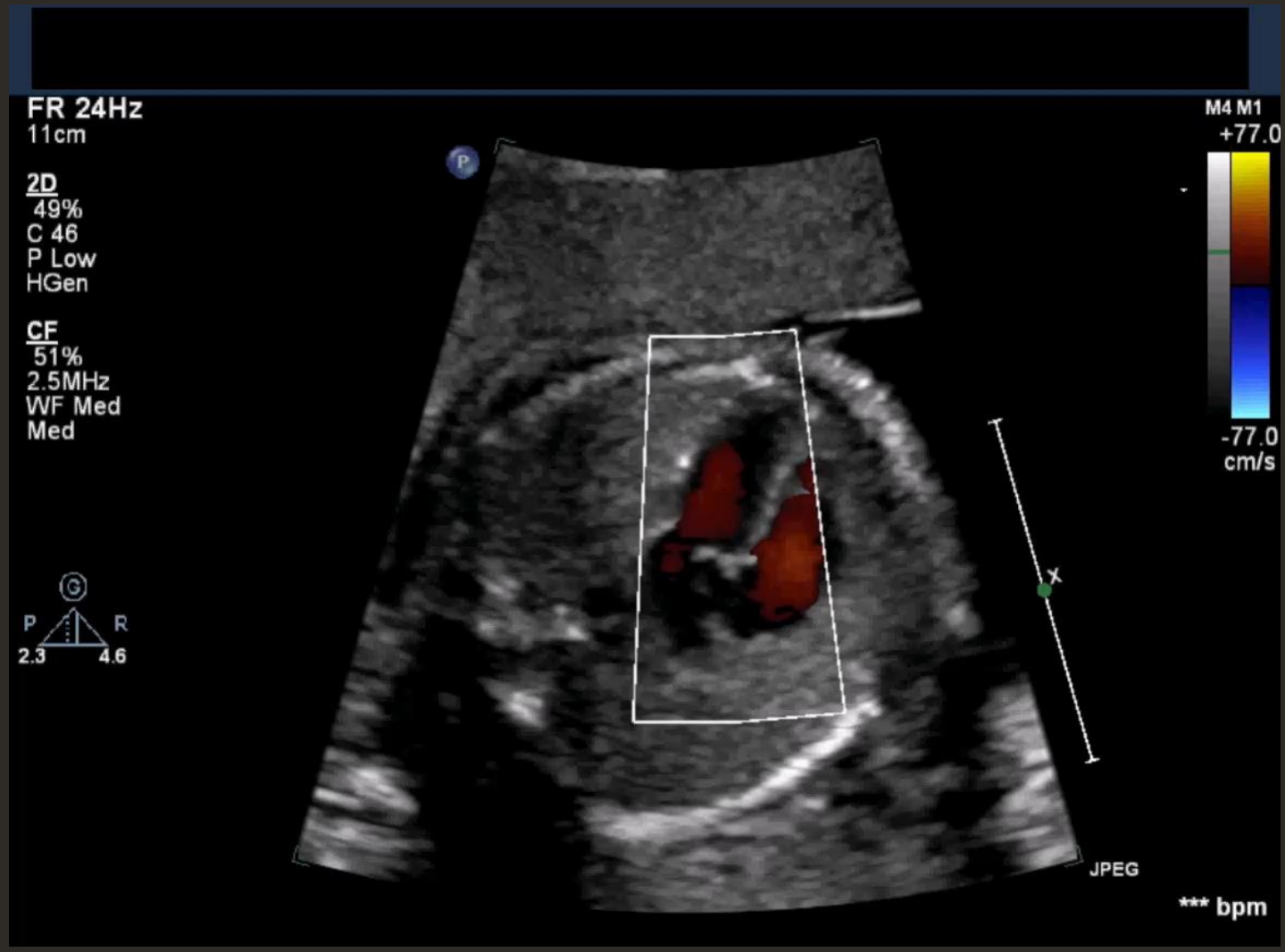
Rychik, Jack. Zhiyun, Tian. *Fetal Cardiovascular Imaging: A Disease-Based Approach.* Elsevier Saunders, 2012.

D-TGA/IVS parallel great arteries

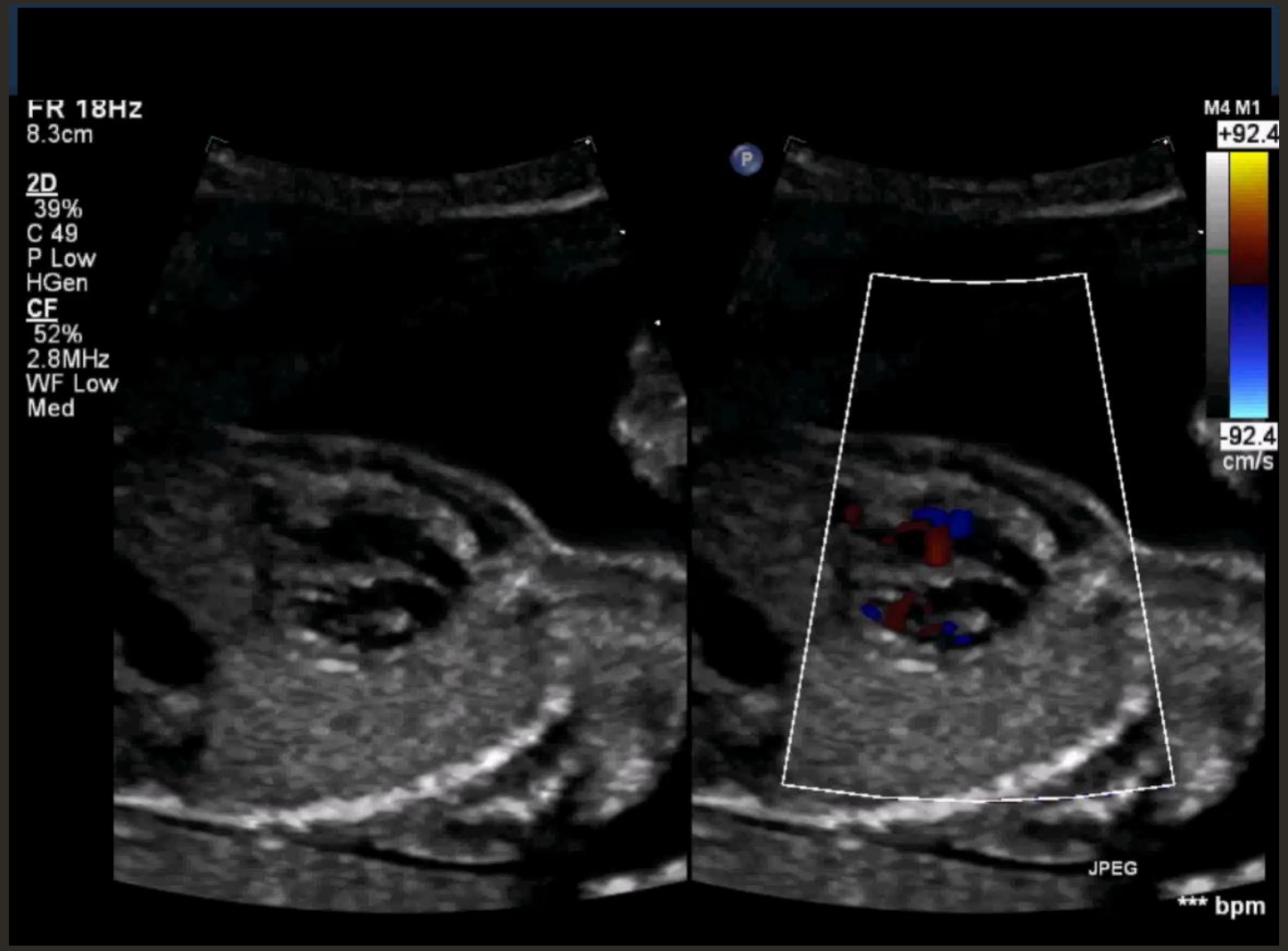
D-TGA/IVS parallel great arteries



D-TGA/IVS 4-chamber color sweep



D-TGA/IVS parallel great arteries to arches



Outcomes based on prenatal diagnosis of D-TGA

Detection of Transposition of the Great Arteries in Fetuses Reduces Neonatal Morbidity and Mortality

Damien Bonnet, Anna Coltri, Gianfranco Butera, Laurent Fermont, Jérôme Le Bidois, Jean Kachaner, and Daniel Sidi

Originally published 23 Feb 1999 | https://doi.org/10.1161/01.CIR.99.7.916 | Circulation. 1999;99:916–918

FULL TEXT ARTICLE

Prenatal Diagnosis of Transposition of the Great Arteries Reduces Postnatal Mortality: A Population-Based Study

Hazumu Nagata MD, Lauren Glick MD, Jane Lougheed MD, Michael Grattan MD, Tapas Mondal MD, Varsha Thakur MD, Steven M. Schwartz MD and Edgar Jaeggi MD

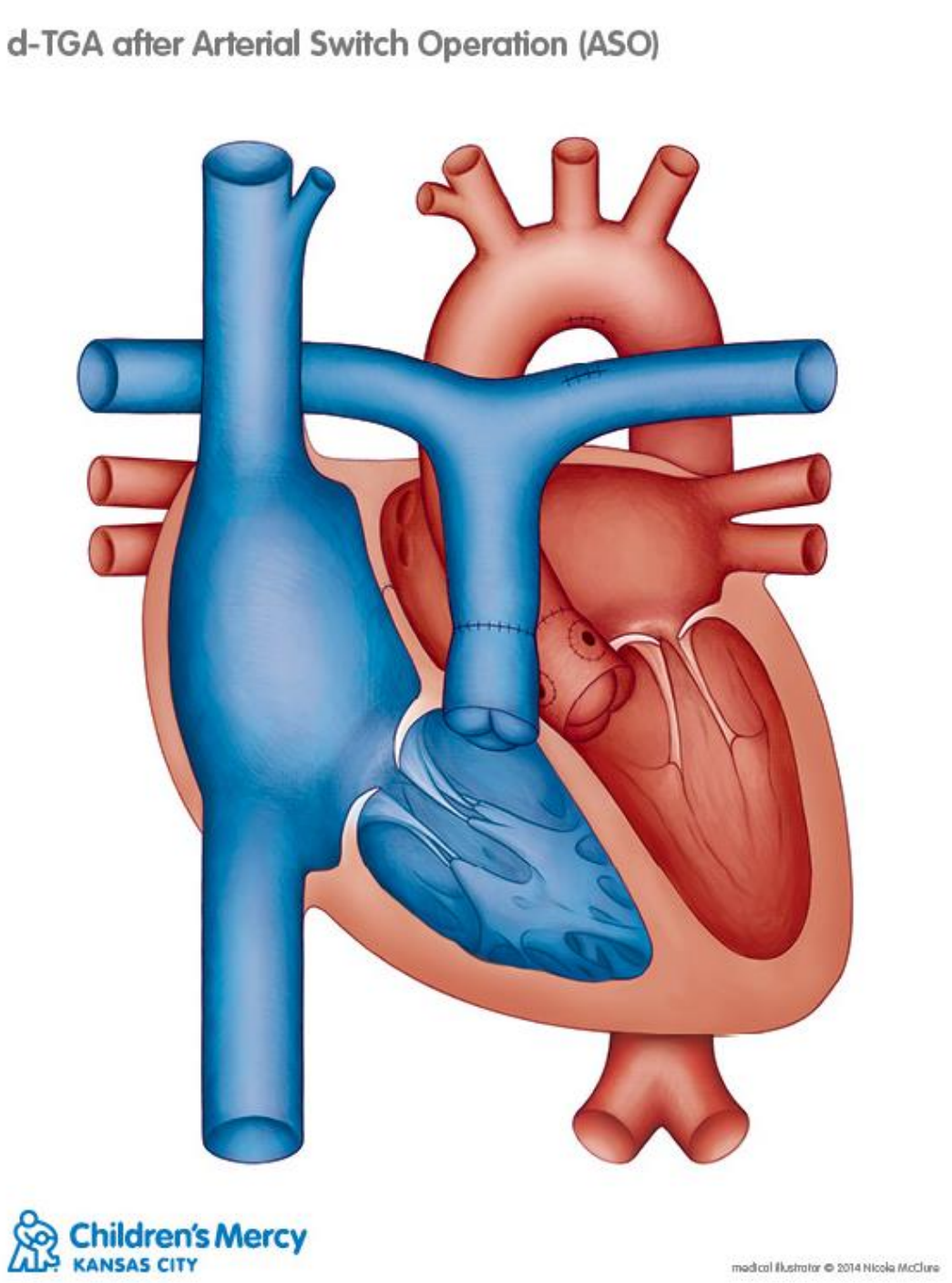
Canadian Journal of Cardiology, 2020-10-01, Volume 36, Issue 10, Pages 1592-1597, Copyright © 2020 Canadian Cardiovascular Society

Delivery Planning/Postnatal Management

- Potential for hemodynamic compromise and need for urgent intervention after delivery → delivery at referral center with experienced neonatologists and cardiologists
- Pulmonary and systemic circulations run in parallel rather than in series
- Pulmonary artery saturation > aortic saturation → severe hypoxemia
- Umbilical line placement and initiation of prostaglandin infusion

D-TGA Surgical Repair and Outcomes

- Arterial Switch Operation

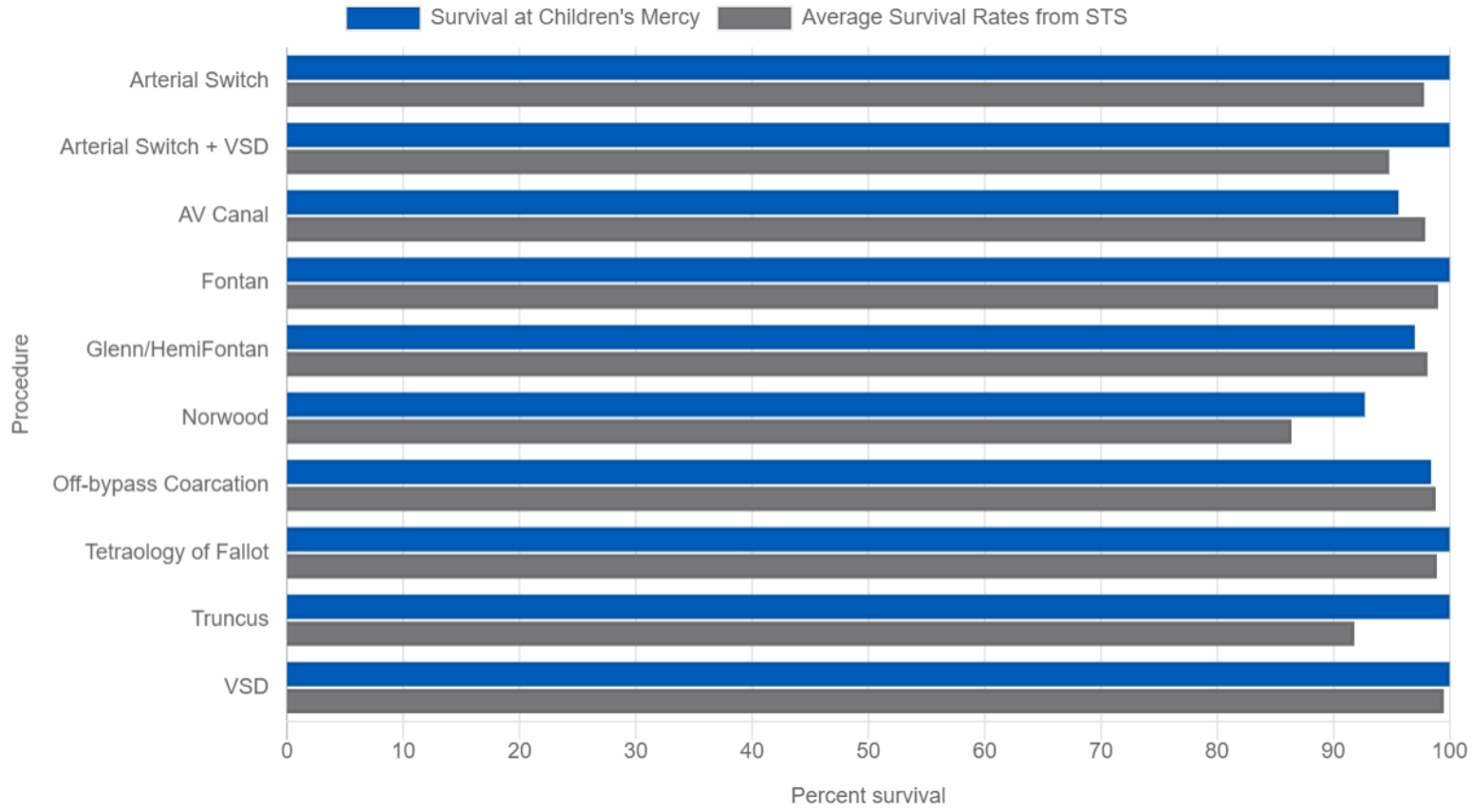


D-TGA Surgical Repair and Outcomes

- Arterial Switch Operation
- Long-term issues:
  - Supravalvar stenosis
  - Branch PA stenosis
  - Coronary artery issues
  - Neoaortic insufficiency

Outcomes by cardiovascular procedure at four years

Four-year Outcomes by STS Benchmark Operation



Four-year Median Post-Operative Length of Stay Days by STS Benchmark Operation

Thank you!

Texts

1. Allen, Hugh D., et al. *Moss and Adams' Heart Disease in Infants, Children, and Adolescents: Including the Fetus and Young Adult*. Wolters Kluwer Health, Lippincott Williams & Wilkins, 2013.
2. Park, Myung K., *Park's Pediatric Cardiology for Practitioners.* Mosby, 2014
3. Rychik, Jack. Zhiyun, Tian. *Fetal Cardiovascular Imaging: A Disease-Based Approach.* Elsevier Saunders, 2012.

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