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Centro de Referencia Perinatal Oriente
Facultad de Medicina, Universidad de Chile

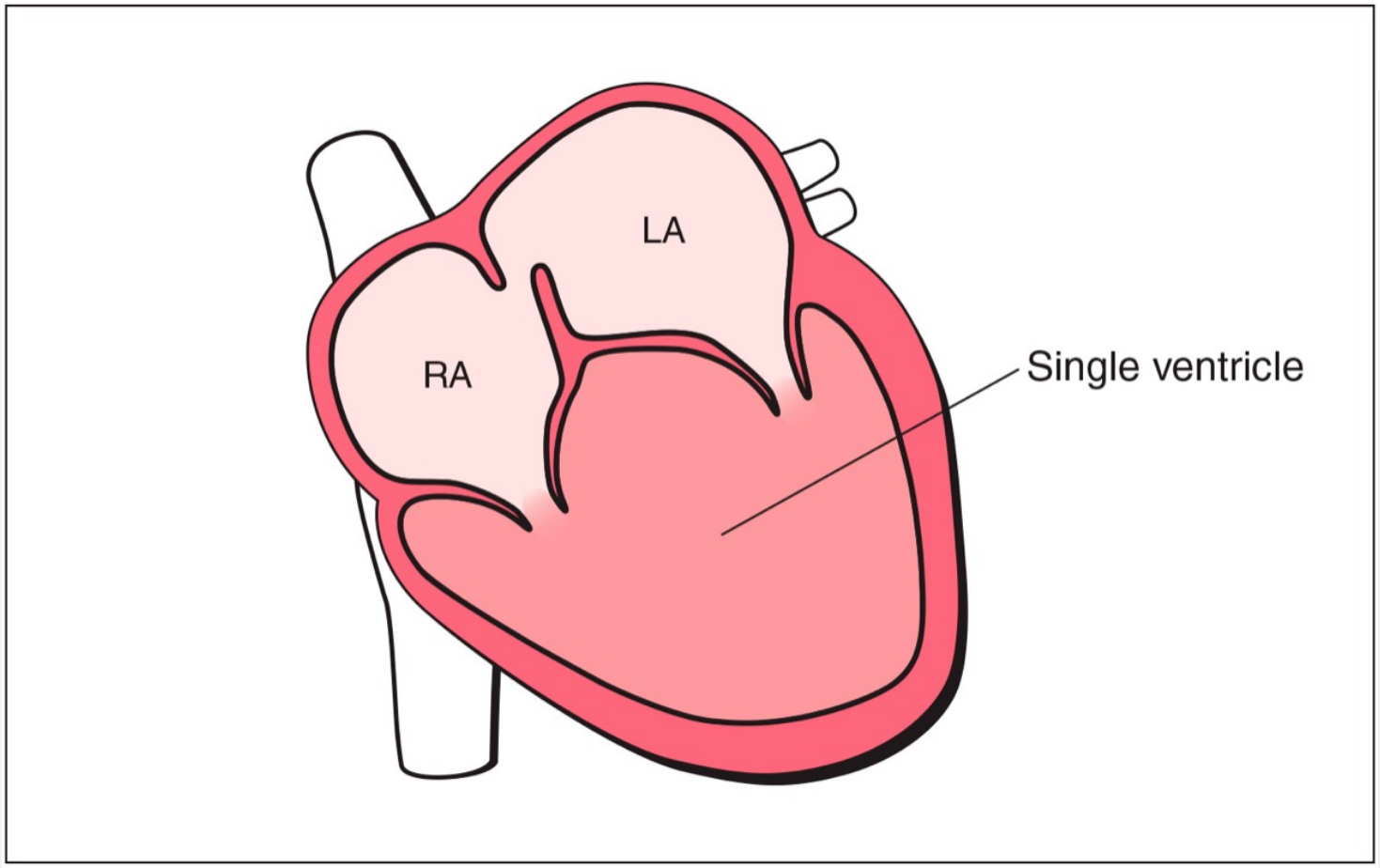
Ventrículo único de doble entrada

Dr. Pablo Silva Labarca
Programa de especialización Medicina Materno Fetal
Facultad de Medicina
Universidad de Chile

Introducción

- Se define como un grupo de anomalías en que dos aurículas desembocan en un único ventrículo dominante a través de una o dos válvulas AV.

- La morfología del ventrículo único puede variar y en general es izquierda, derecha o indeterminada.



Abuhamad, Alfred Z. A Practical Guide to Fetal Echocardiography, 3rd Edition. Wolters Kluwer Health.

Epidemiología

- Es una anomalía rara.
- Prevalencia de 0,1 a 0,08 por cada 1000 nacidos vivos.
- Corresponde a 1,25-1,8% de todas las cardiopatías congénitas.

Yagel, S., Silverman, N.H., & Gembruch, U. (Eds.). (2019). Fetal Cardiology: Embryology, Genetics, Physiology, Echocardiographic Evaluation, Diagnosis, and Perinatal Management of Cardiac Diseases, Third Edition (3rd ed.). CRC Press.

Embriología

- <u>2 teorías:</u>

- Fallo en la correcta alineación entre el septo infundibular de salida con el resto del septo interventricular

- El septo infundibular es una estructura con un origen embriológico diferente a la del resto del tabique y hay una falla en la formación del septo interventricular posterior.

Galindo A, Gratacos E, Martinez J. Cardiología Fetal. 1era edición: Editorial Marbán. Madrid- España. 2015

Anatomía y Fisiopatología

- Existencia de un único ventrículo aumentado de tamaño denominado ventrículo dominante que conecta total o mayoritariamente con las dos aurículas.
- Se observa una o dos válvulas AV desembocando en un único ventrículo.
- La morfología del ventrículo dominante debe establecerse a partir del patrón de trabeculación.
- Habitualmente existe una cámara de salida rudimentaria que se ubica en la pared anterosuperior del ventrículo y carece de conexión con la aurícula ipsilateral.

Galindo A, Gratacos E, Martinez J. Cardiología Fetal. 1era edición: Editorial Marbán. Madrid- España. 2015

Anatomía y Fisiopatología

- La cámara de salida puede estar conectada con el ventrículo a través del foramen bulboventricular y habitualmente esta conectada también con el tracto de salida.
- Los tractos de salida habitualmente están en malposición y pueden salir del ventrículo único o de la cámara de salida.
- Cuando están en concordancia ventrículo-arterial se conoce como corazón de Holmes.
- Tambien puede existir un grado variable de obstrucción de los tractos de salida.

Galindo A, Gratacos E, Martinez J. Cardiología Fetal. 1era edición: Editorial Marbán. Madrid- España. 2015

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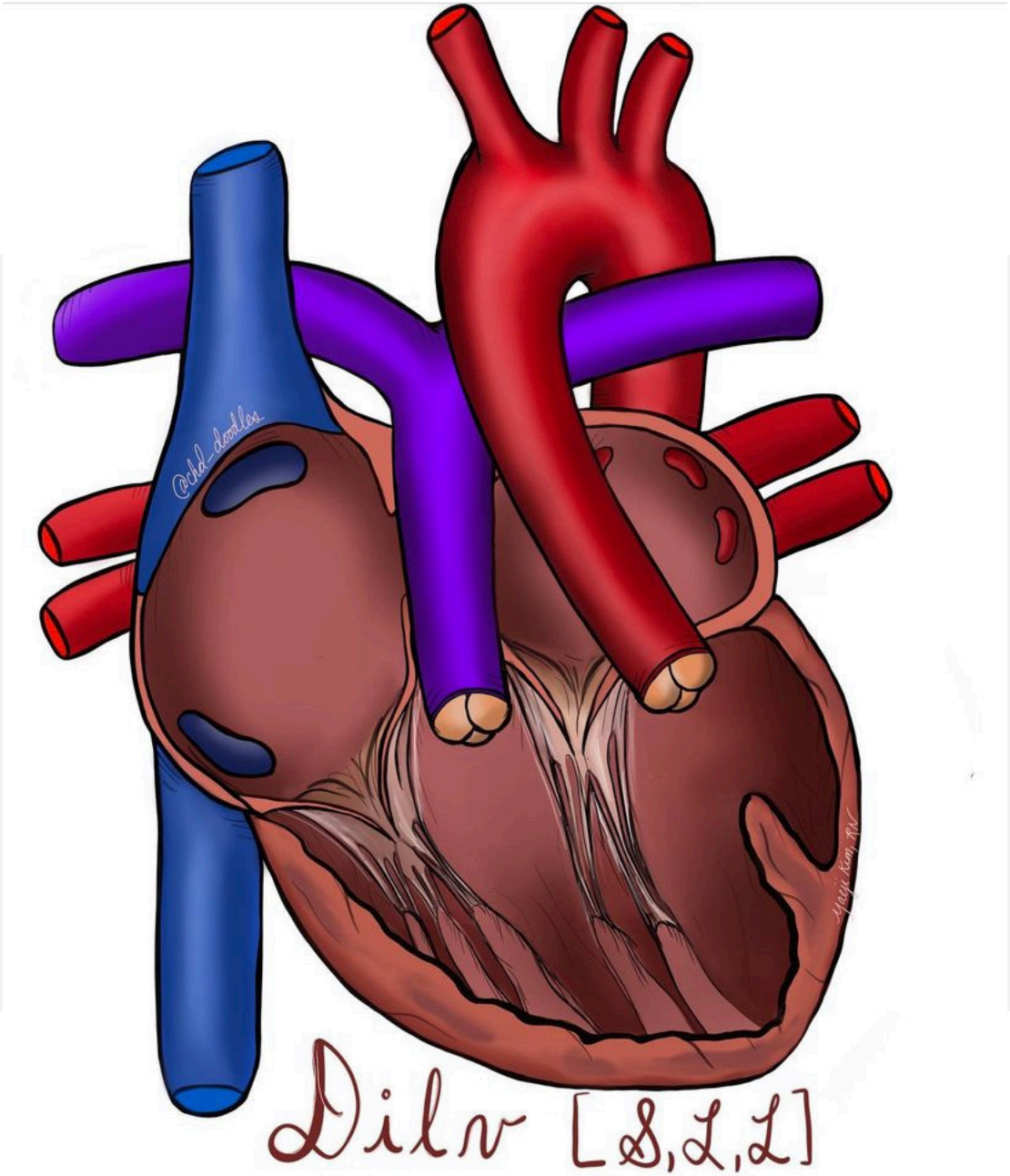

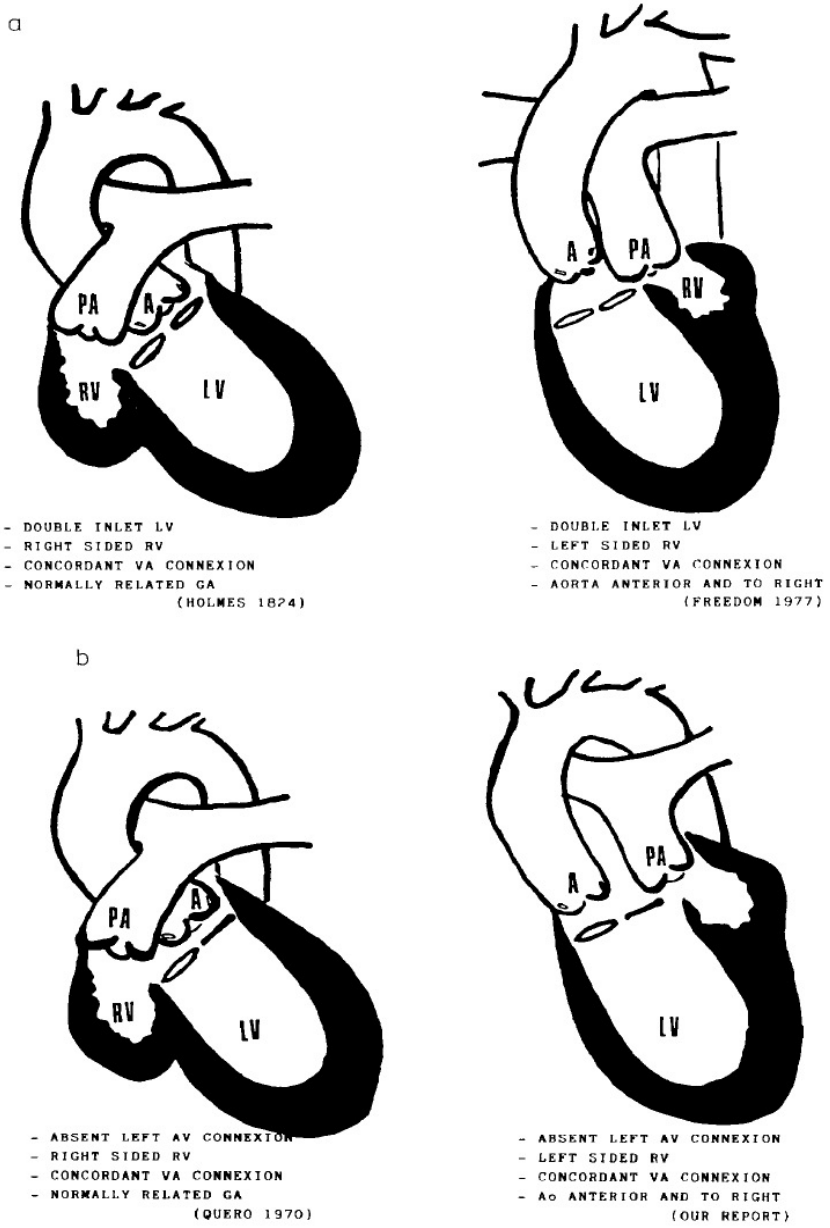

Fig. 4. Diagrams summarizing the variable morphology of univentricular atrioventricular connexion with concordant ventriculo-arterial connexion. (a) Double inlet to a dominant left ventricle (LV), right-sided rudimentary right ventricle (RV), concordant ventriculo-arterial connexion and normally related great arteries (GA); double inlet to a dominant left ventricle (LV), left-sided rudimentary right ventricle (RV), concordant ventriculo-arterial (VA) connexion, aorta anterior and to the right. (b) Absent left atrioventricular (AV) connexion, right-sided rudimentary right ventricle (RV), concordant ventriculo-arterial (VA) connection, normally related great arteries (GA); absent left atrioventricular (AV) connexion with left sided rudimentary right ventricle (RV), concordant ventriculo-arterial connexion, aorta anterior and to the right, our report.

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- Corazón de Holmes

Clasificación

- Tipo ventrículo izquierdo con/sin ventrículo derecho rudimentario.
  - 60-80% de los casos.
- Tipo ventrículo derecho con/sin ventrículo izquierdo rudimentario.
  - 5-25% de los casos.
- Tipo indeterminado o indiferenciado.
  - <5% de los casos.

Yagel, S., Silverman, N.H., & Gembruch, U. (Eds.). (2019). Fetal Cardiology: Embryology, Genetics, Physiology, Echocardiographic Evaluation, Diagnosis, and Perinatal Management of Cardiac Diseases, Third Edition (3rd ed.). CRC Press.

Formas Anatómicas DILV

- S-L-L: Cámara accesoria a la izquierda del ventrículo dominante con una conexión ventrículo-arterial discordante. (**Forma más frecuente**).

- S-D-N: Cámara accesoria a la derecha del ventrículo dominante con conexión ventrículo-arterial correcta.

- S-D-D: Cámara accesoria a la derecha del ventrículo dominante con conexión ventrículo-arterial discordante.

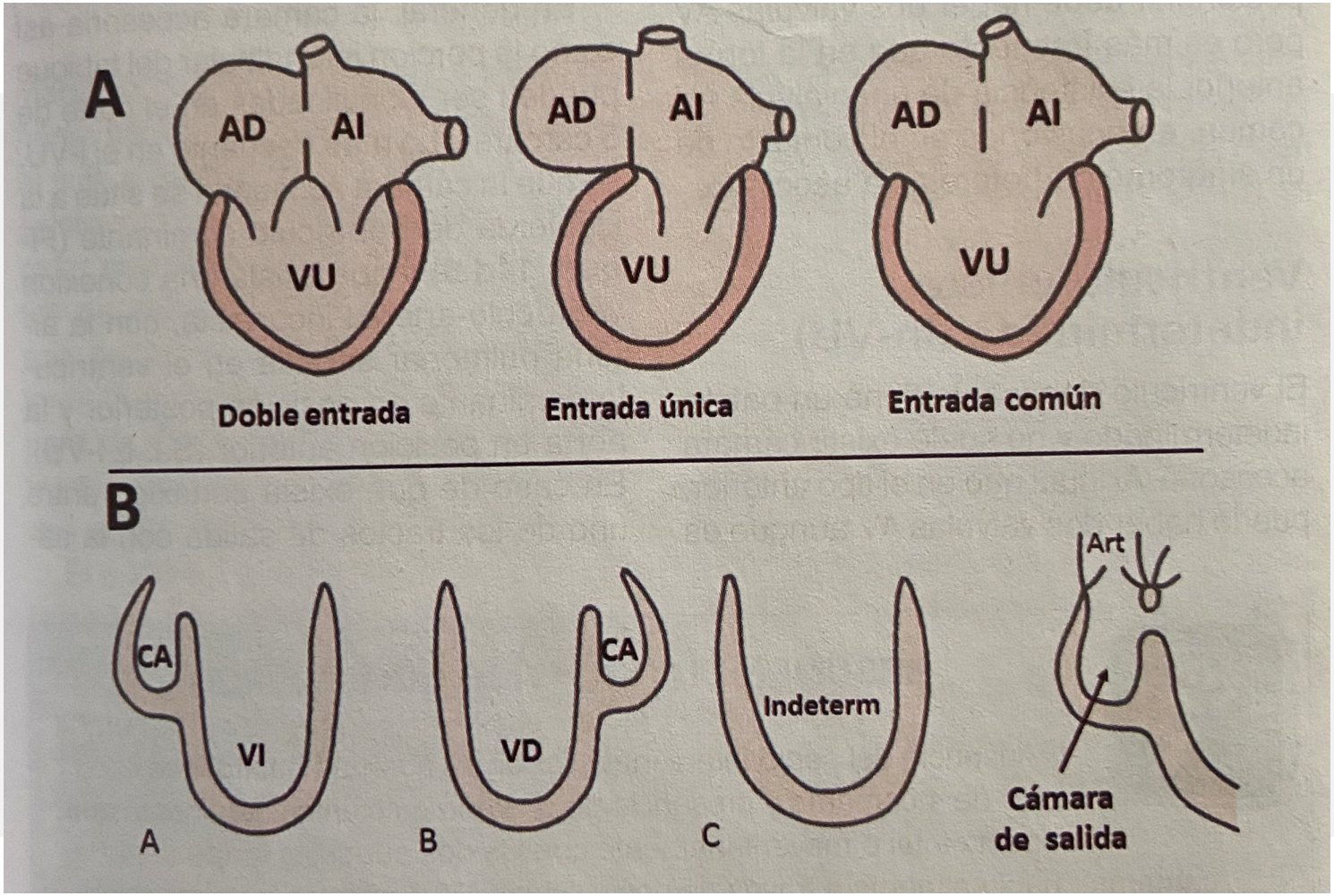
Galindo A, Gratacos E, Martinez J. Cardiología Fetal. 1era edición: Editorial Marbán. Madrid- España. 2015

Otras Formas Anatómicas

- **<u>Ventrículo único derecho:</u>**
  - Es menos frecuente y se caracteriza por ventrículo con trabeculaciones musculares del ventrículo derecho.
  - Puede haber 2 válvulas AV pero es mas común que haya una.
  - La cámara accesoria es más difícil de identificar porque en general se encuentra por posterior.

- **<u>Ventrículo único indeterminado:</u>**
  - El ventrículo dominante es indeterminado y no hay cámara accesoria.
  - Puede haber 2 válvulas AV pero lo más común es que haya una.

Galindo A, Gratacos E, Martinez J. Cardiología Fetal. 1era edición: Editorial Marbán. Madrid- España. 2015


A
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VU
Doble entrada
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Entrada única
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de salida

Diagnóstico prenatal

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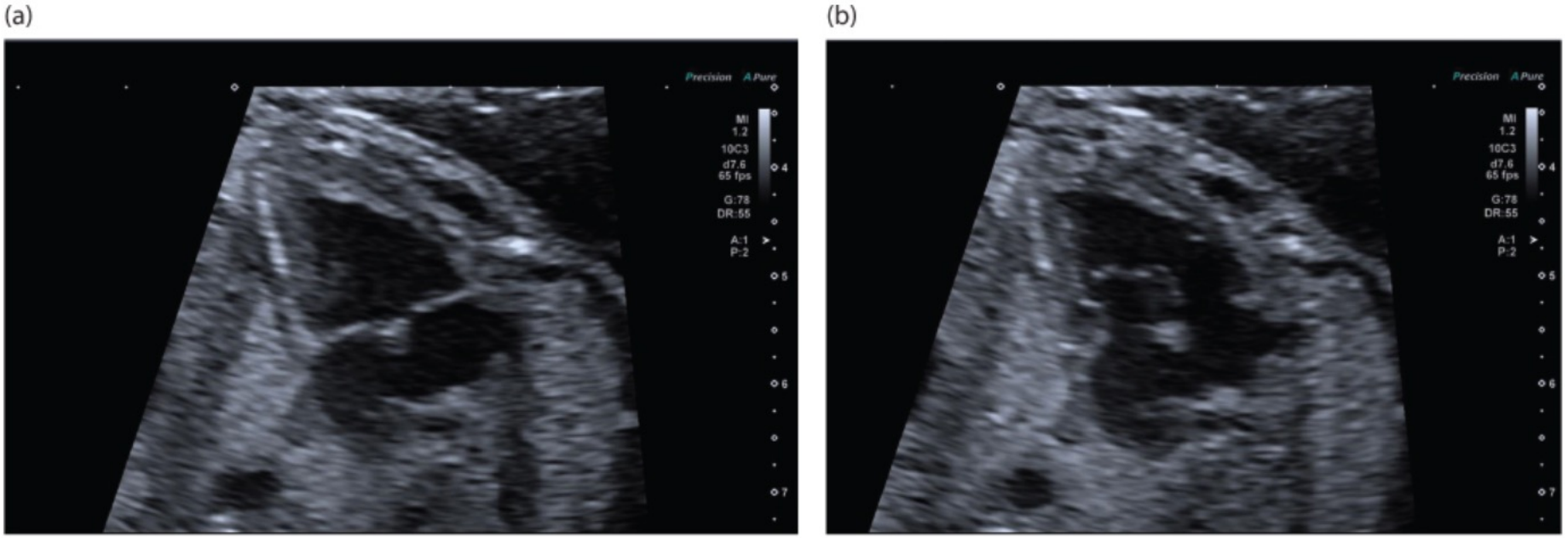

DIRV 22+5 semanas

Yagel, S., Silverman, N.H., & Gembruch, U. (Eds.). (2019). Fetal Cardiology: Embryology, Genetics, Physiology, Echocardiographic Evaluation, Diagnosis, and Perinatal Management of Cardiac Diseases, Third Edition (3rd ed.). CRC Press.

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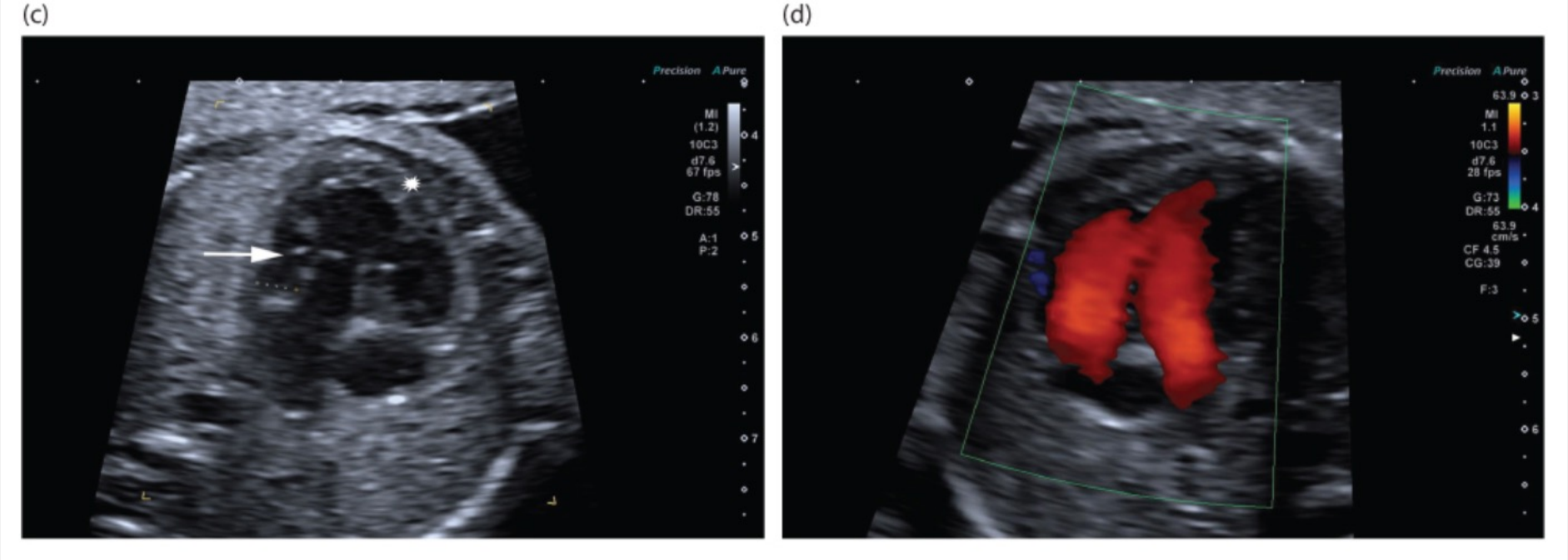

DIRV 22+5 semanas

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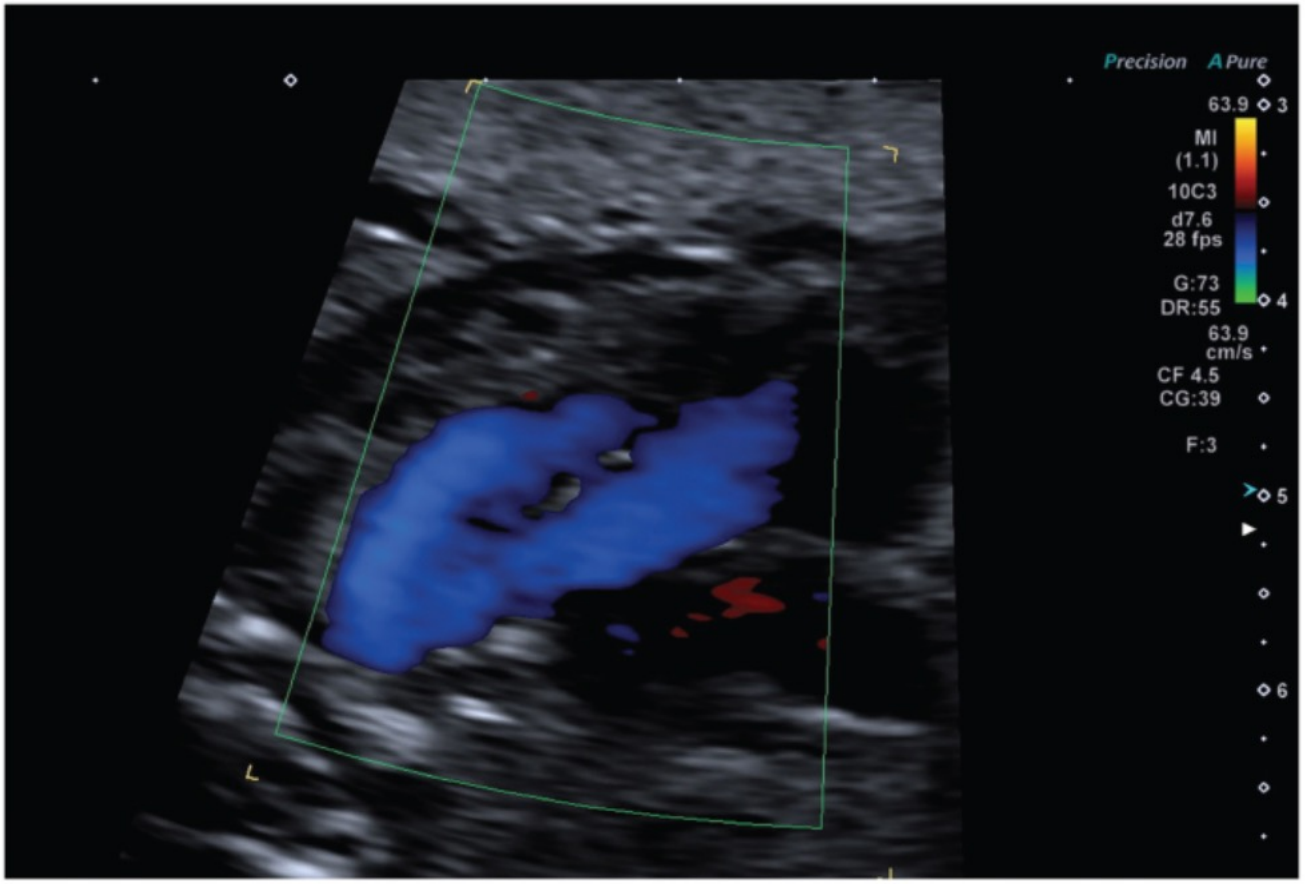
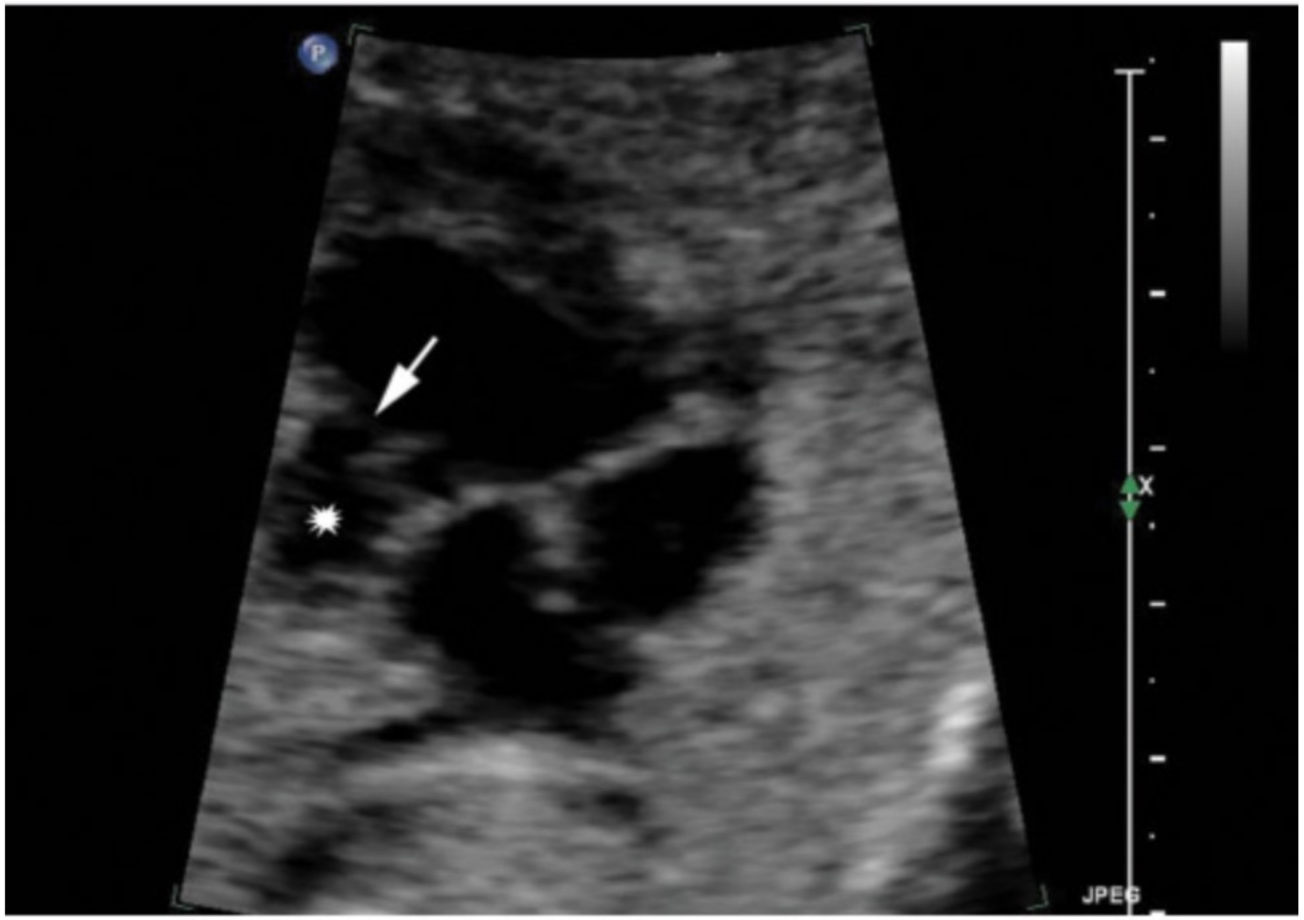
(e)



DIRV 22+5 semanas, D-TGA

Yagel, S., Silverman, N.H., & Gembruch, U. (Eds.). (2019). Fetal Cardiology: Embryology, Genetics, Physiology, Echocardiographic Evaluation, Diagnosis, and Perinatal Management of Cardiac Diseases, Third Edition (3rd ed.). CRC Press.

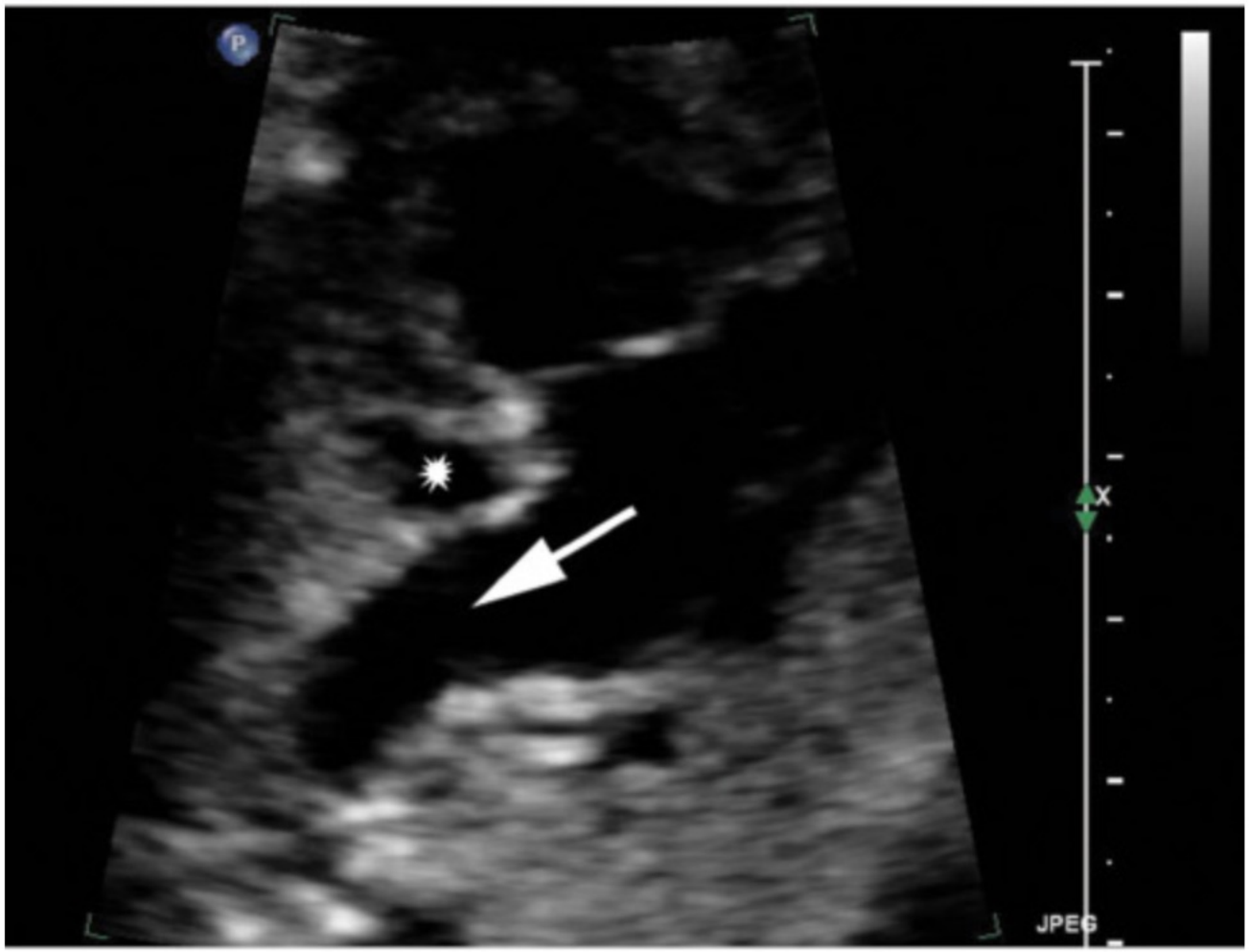
(a)



DILV 20+1 semanas

Yagel, S., Silverman, N.H., & Gembruch, U. (Eds.). (2019). Fetal Cardiology: Embryology, Genetics, Physiology, Echocardiographic Evaluation, Diagnosis, and Perinatal Management of Cardiac Diseases, Third Edition (3rd ed.). CRC Press.

(b)



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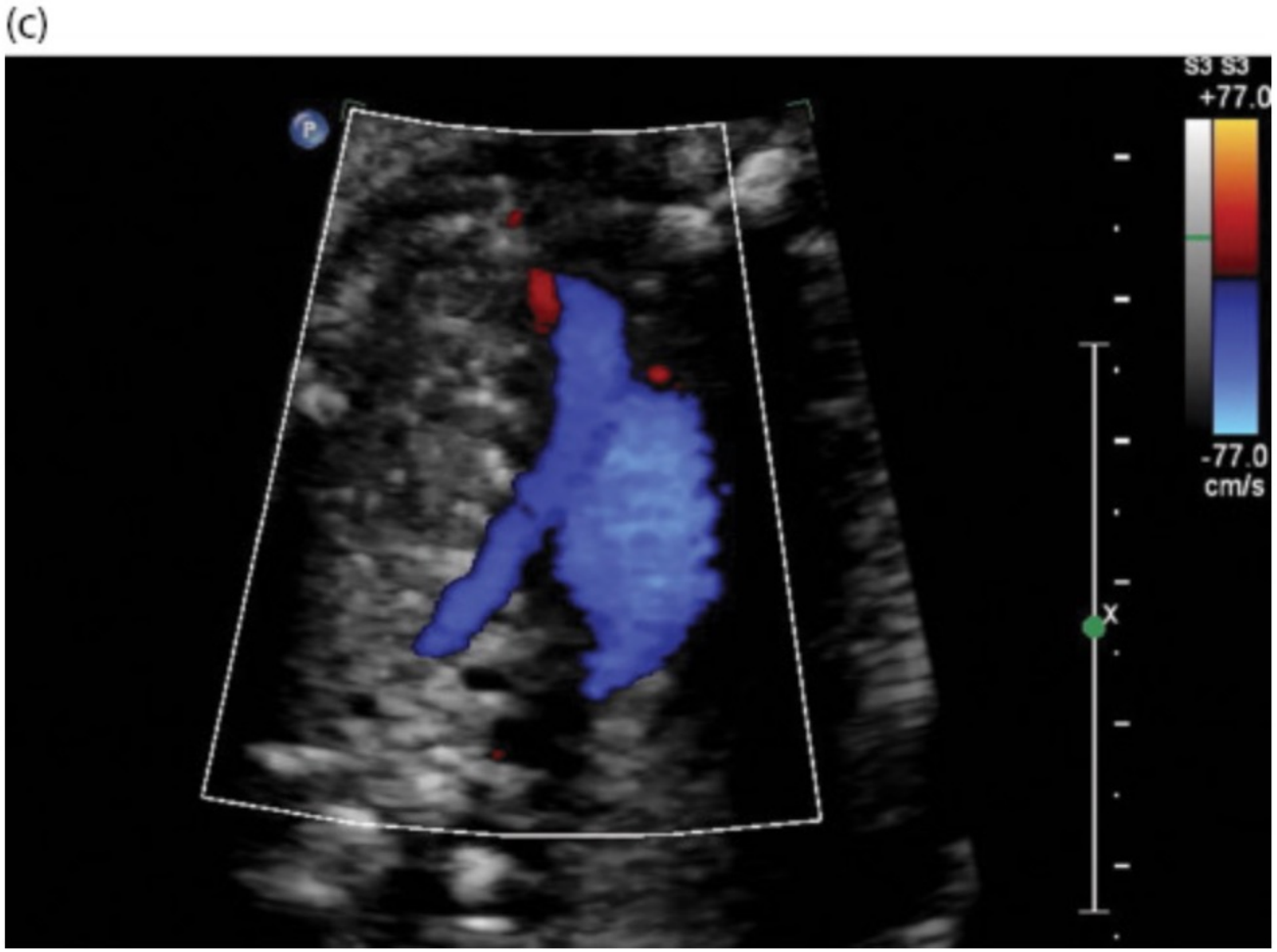
DILV 20+1 semanas, L-TGA, CoA

Yagel, S., Silverman, N.H., & Gembruch, U. (Eds.). (2019). Fetal Cardiology: Embryology, Genetics, Physiology, Echocardiographic Evaluation, Diagnosis, and Perinatal Management of Cardiac Diseases, Third Edition (3rd ed.). CRC Press.



DILV 20+1 semanas, L-TGA, CoA

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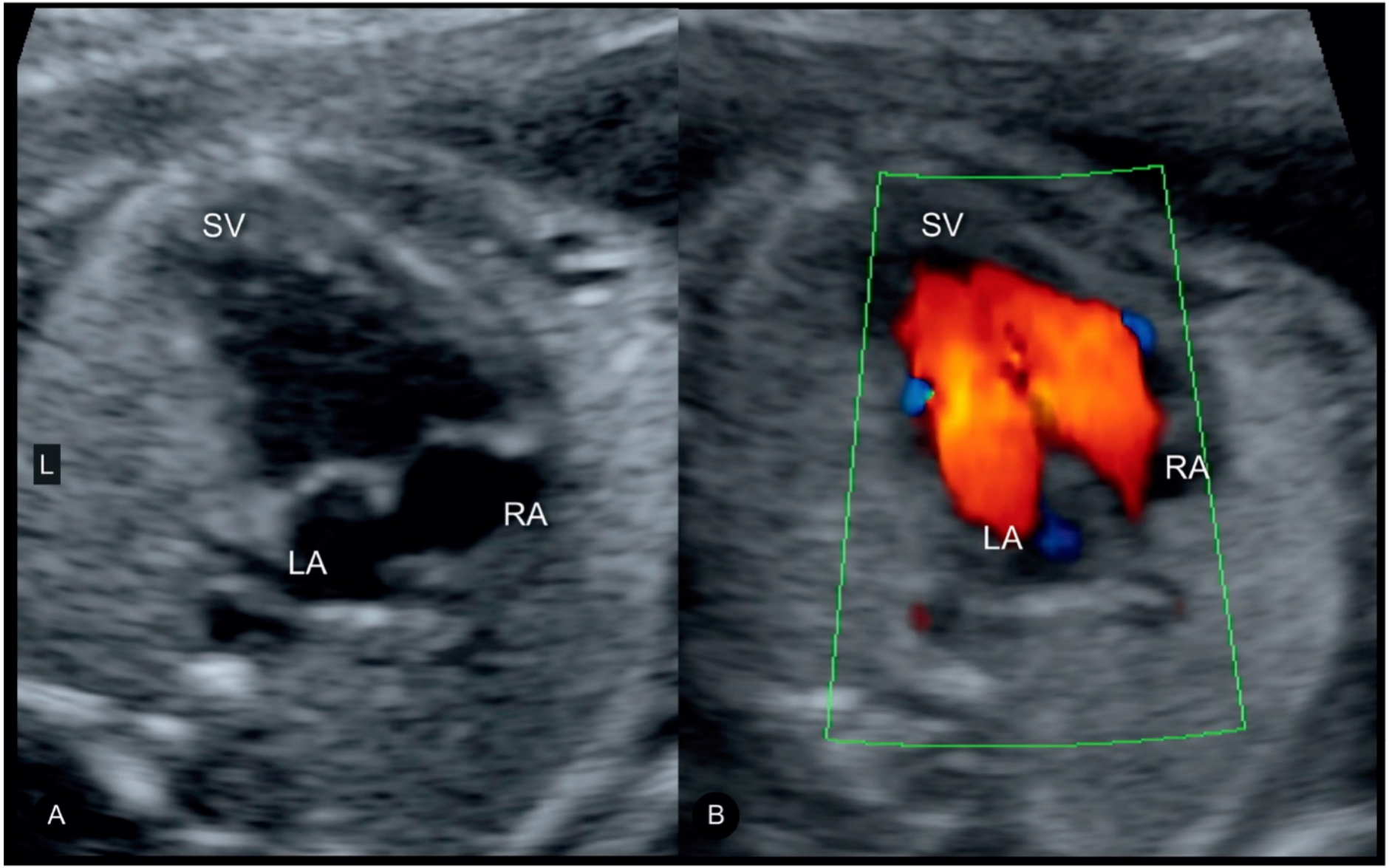
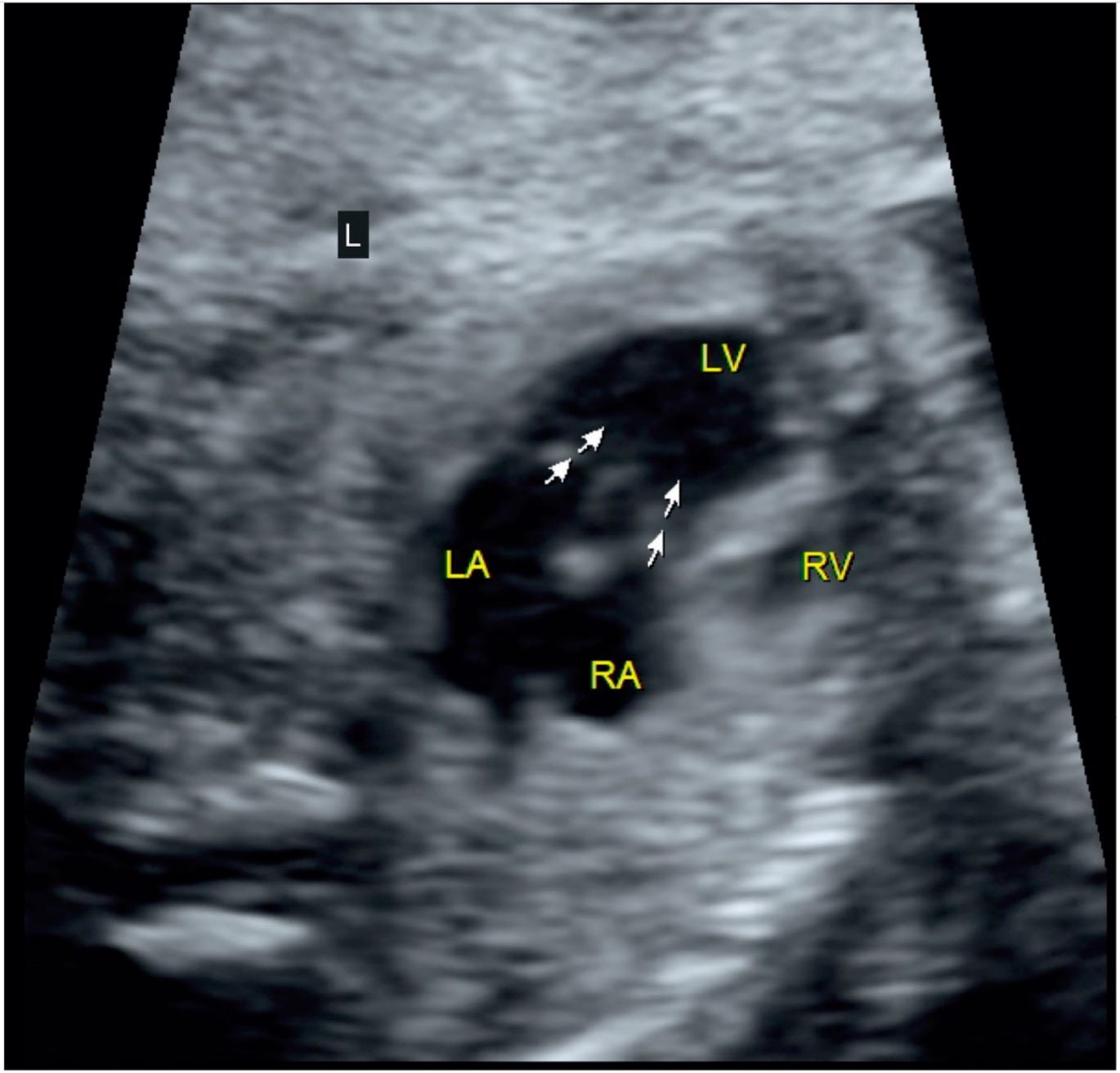


Four-chamber views in gray scale **(A)** and color Doppler **(B)** in a fetus with a double inlet ventricle. Note the presence of right (RA) and left (LA) atria and a single ventricle (SV) in **A**. **B** shows, in color Doppler, blood flow from the RA and LA through two respective atrioventricular valves into the SV. L, left.



Four-chamber view in gray scale in a fetus with a double inlet ventricle. Note that the right (RA) and left (LA) atria drain through two distinct atrioventricular valves into the left ventricle (LV). There is a rudimentary right ventricle (RV) as an outlet chamber drained from the LV. L, left.

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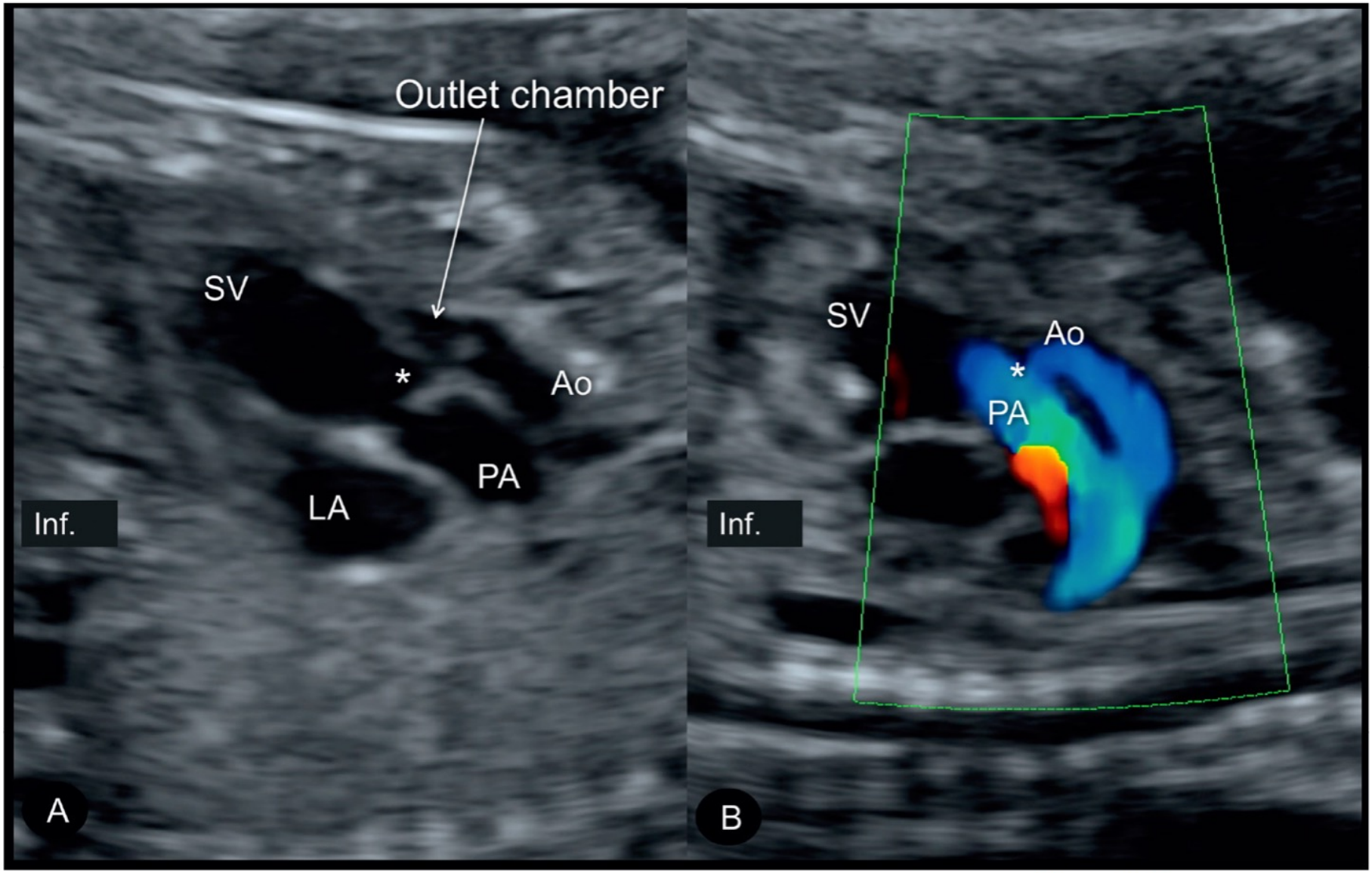
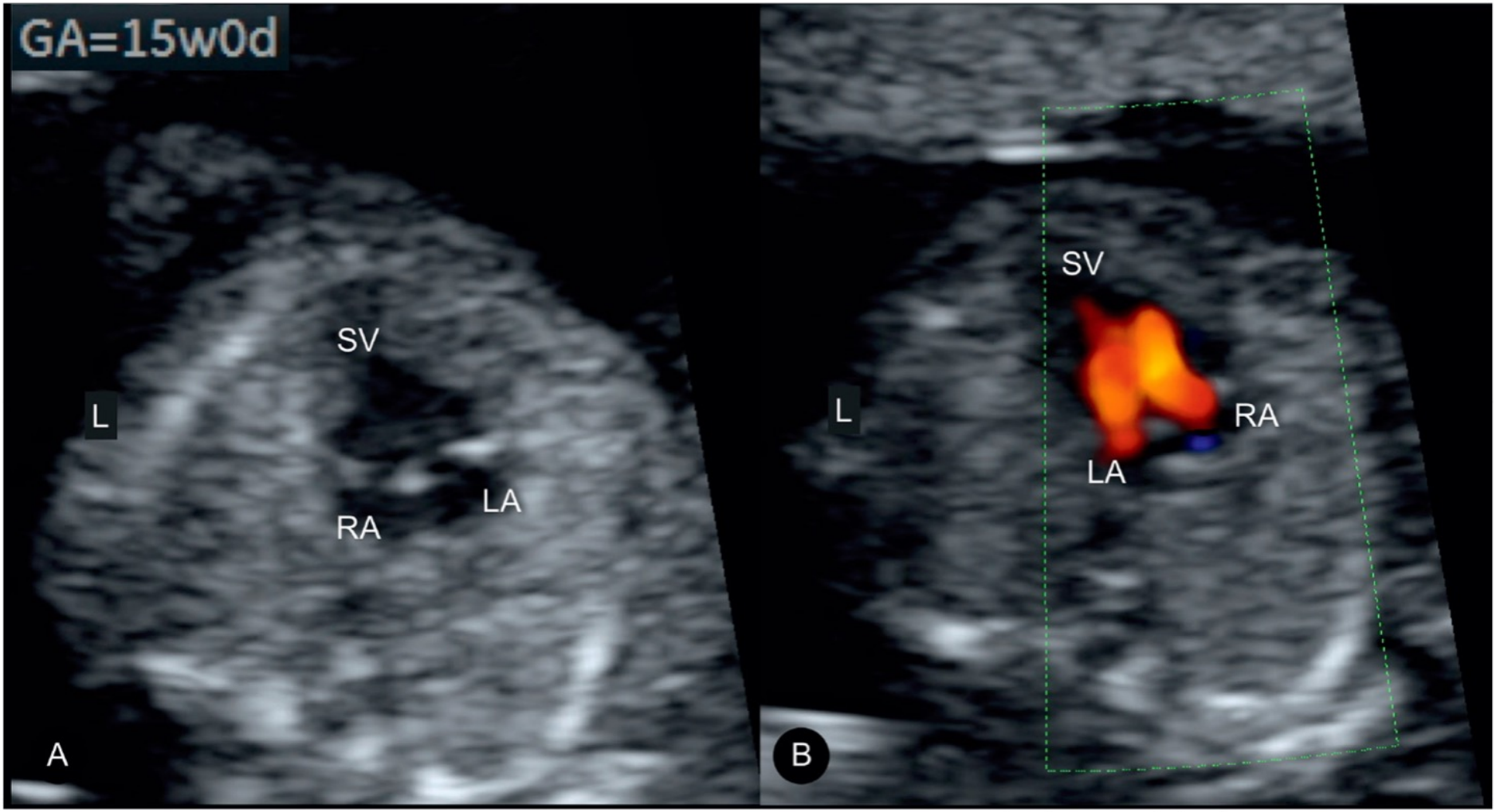
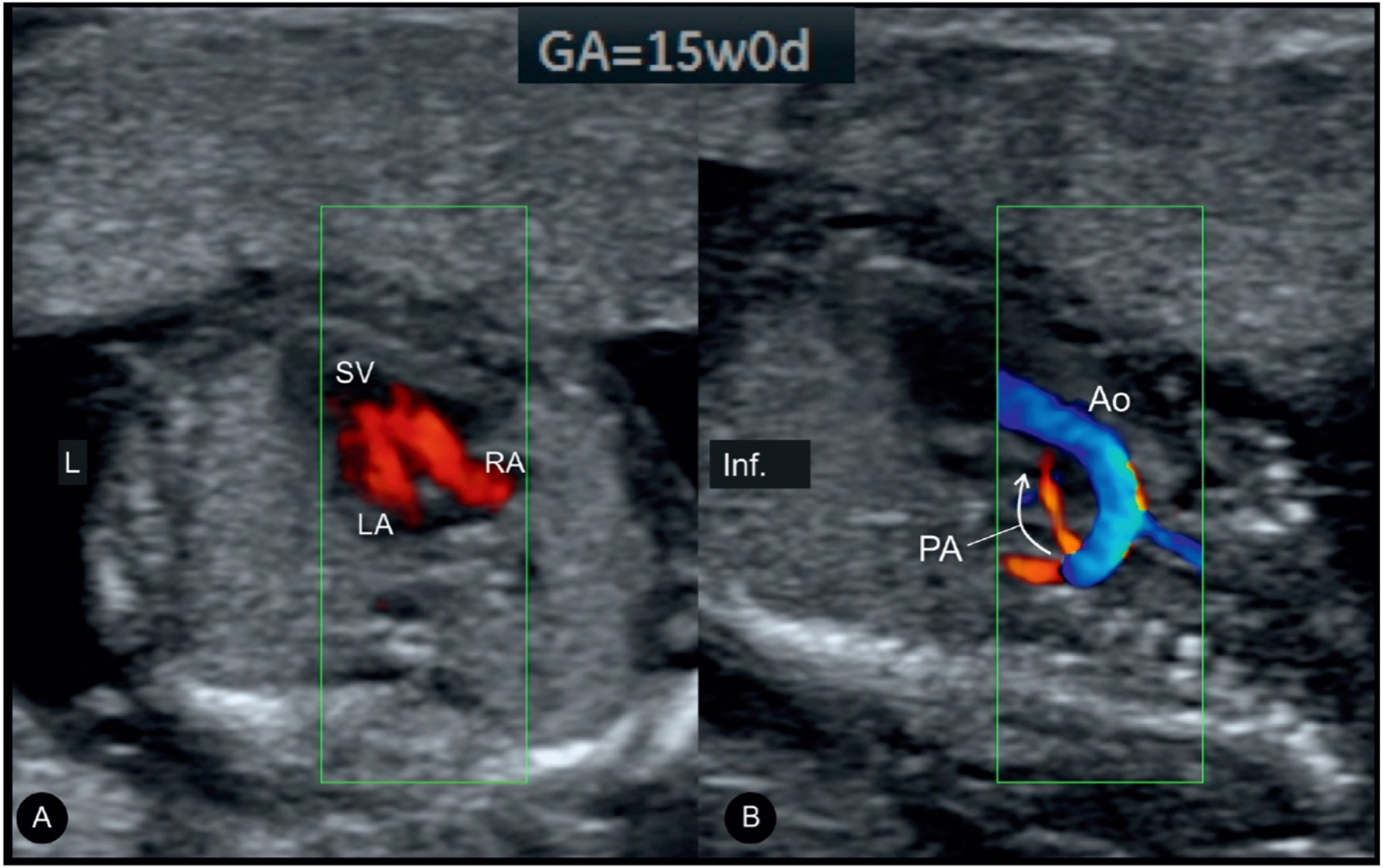


Long-axis views in gray scale **(A)** and color Doppler **(B)** in the same fetus shown in Figure 19.4 with a double inlet ventricle (SV) and a rudimentary outlet ventricle. The rudimentary outlet ventricle is connected with the SV through a ventricular septal defect (*asterisk*), called bulboventricular foramen. Aorta (Ao) and pulmonary artery (PA) arise in parallel orientation. Note that the Ao is smaller than the PA, due to the small size of the ventricular septal defect. Aortic coarctation was diagnosed after birth. Inf., inferior.



Fetus at 15 weeks' gestation with a double inlet ventricle, with both right (RA) and left (LA) atria draining through two respective atrioventricular valves into a single ventricle (SV). **A** is in gray scale and **B** is in color Doppler. L, left.



Four-chamber **(A)** and longitudinal **(B)** views in color Doppler in a fetus at 15 weeks' gestation with a double inlet ventricle (same fetus as in Fig. 19.6). Note in **A** that the right (RA) and left (LA) atria drain through two respective atrioventricular valves into a single ventricle (SV). The longitudinal plane in **B** reveals the presence of pulmonary atresia. The pulmonary artery (PA) is hypoplastic, demonstrates retrograde flow (*arrow*), and is located posterior to the aorta (Ao). Inf., inferior; L, left.

Corazón de Holmes

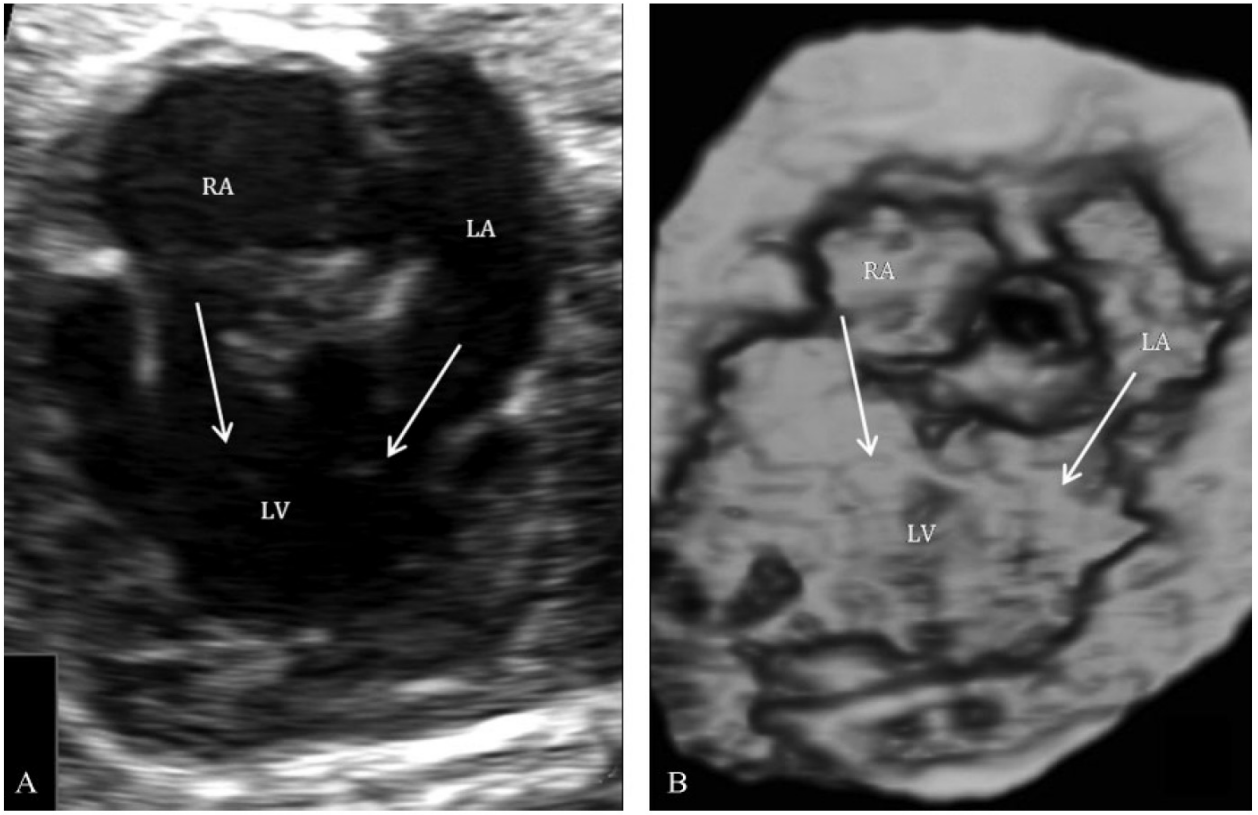


Figure 3. Holmes heart (case 3) showing both AV valves connected to a morphological left ventricle (A). Three-dimensional rendered image of the heart emphasizing the complete absence of the interventricular septum (B). LA, left atrium; LV, left ventricle; RA, right atrium.

Weichert J, Axt-Fliedner R, Gembruch U, Hartge DR. Holmes heart--a simple antenatal diagnosis of a complex cardiac anomaly? Fetal echocardiographic findings and review. Congenit Heart Dis. 2013 Nov-Dec;8(6):579-84.

Corazón de Holmes

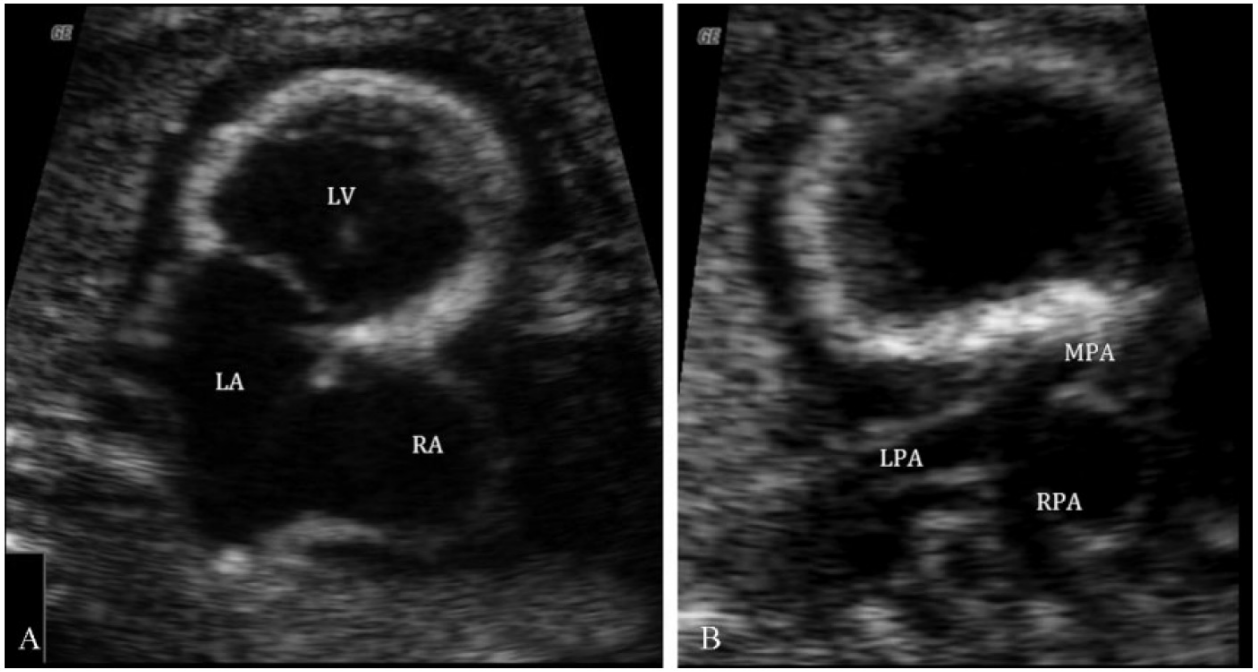


Figure 2. Transverse section of the fetal thorax (case 2) showing a markedly dilated heart with an apparent endocardial fibroelastosis of the left ventricle. The right AV valve is thickened and dysfunctional (A). (B) Depicts the obstruction of the right outflow tract with a small main pulmonary artery and its branches. LA, left atrium; LV, left ventricle; LPA, left pulmonary artery; MPA, main pulmonary artery; RPA, right pulmonary artery; RA, right atrium.

Weichert J, Axt-Fliedner R, Gembruch U, Hartge DR. Holmes heart--a simple antenatal diagnosis of a complex cardiac anomaly? Fetal echocardiographic findings and review. Congenit Heart Dis. 2013 Nov-Dec;8(6):579-84.

Corazón de Holmes

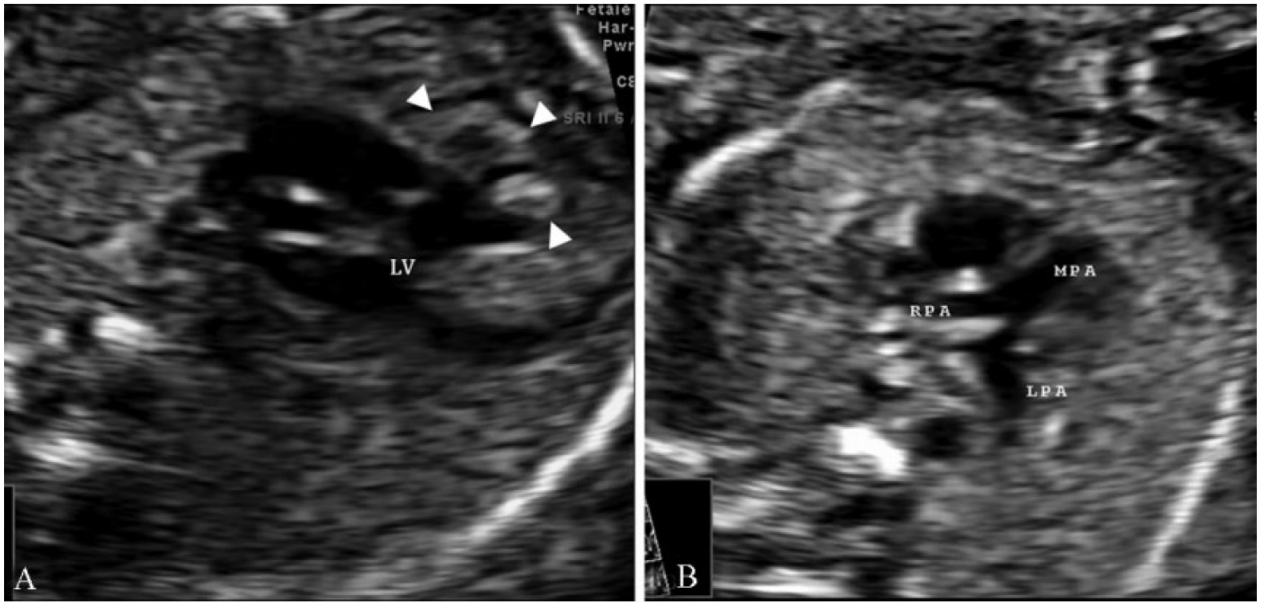


Figure 4. Transverse oblique view (case 3) demonstrating the right-sided rudimentary outlet chamber (arrowheads) adjacent to a morphological left ventricle (A). Small right outflow tract arising off the anteriorly located outlet chamber via the foramen bulboventriculare (B). LV, left ventricle; MPA, main pulmonary artery; RPA, right pulmonary artery; RA, right atrium.

Weichert J, Axt-Fliedner R, Gembruch U, Hartge DR. Holmes heart--a simple antenatal diagnosis of a complex cardiac anomaly? Fetal echocardiographic findings and review. Congenit Heart Dis. 2013 Nov-Dec;8(6):579-84.

Anomalías asociadas

- Las anomalías extracardíacas y cromosómicas pueden existir pero son raras.

- Entre las anomalías descritas en las distintas series se encuentran:
  - Higroma quístico
  - Hernia diafragmática congénita
  - Sindrome de Klinefelter
  - Sindrome de DiGeorge
  - Sindrome de Goldenhar
  - Trisomía 18

Yagel, S., Silverman, N.H., & Gembruch, U. (Eds.). (2019). Fetal Cardiology: Embryology, Genetics, Physiology, Echocardiographic Evaluation, Diagnosis, and Perinatal Management of Cardiac Diseases, Third Edition (3rd ed.). CRC Press.

Diagnóstico diferencial

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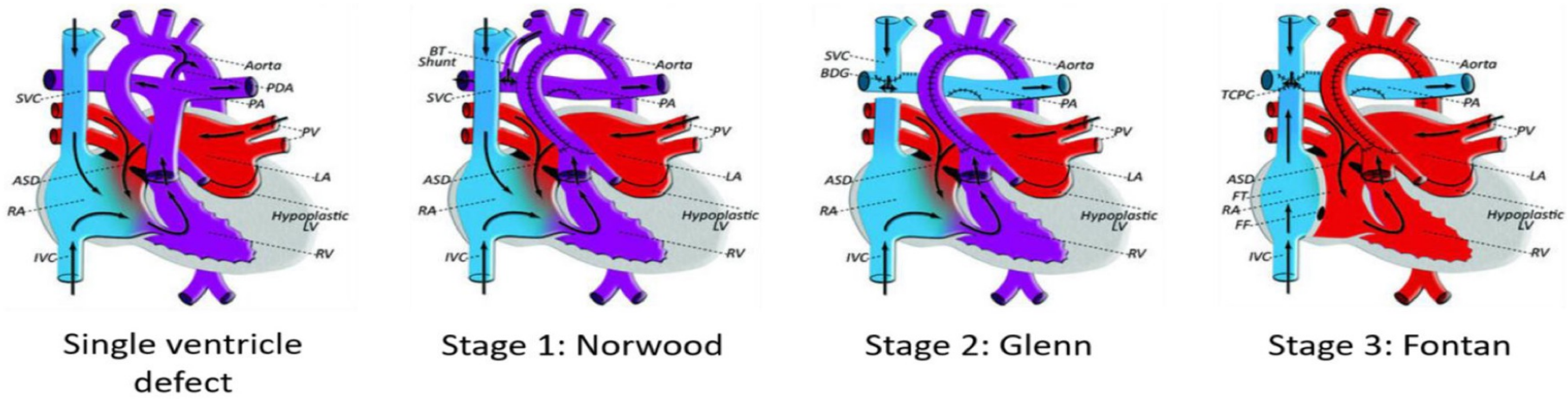
TABLE 19.1 Cardiac Anomalies That May Show a *Single Ventricle* on Fetal Echocardiography

- Hypoplastic left heart syndrome
- Pulmonary atresia with intact septum
- Atrioventricular septal defect (large or unbalanced)
- Single ventricle in right and left isomerism
- Corrected transposition with tricuspid atresia
- Mitral atresia with ventricular septal defect
- Double inlet ventricle
- Tricuspid atresia with ventricular septal defect

Abuhamad, Alfred Z. A Practical Guide to Fetal Echocardiography, 3rd Edition. Wolters Kluwer Health.

Tratamiento postnatal



Single ventricle defect | Stage 1: Norwood | Stage 2: Glenn | Stage 3: Fontan

Fig. 1.
Staged palliation for single ventricle congenital heart defects. Blood volume is colored by blue (deoxygenated) and red (oxygenated). ASD: atrial septal defect, BDG: bidirectional Glenn anastomosis, BT: Blalock-Taussig, FF: Fontan fenestration, FT: Fontan tunnel, IVC/SVC: inferior/superior vena cava, LA: left atrium, LV: left ventricle, PA: pulmonary artery, PDA: patent ductus arteriosus, PV: pulmonary vein, RA: right atrium, RV: right ventricle, TCPC: total cavopulmonary connection

Trusty PM, Slesnick TC, Wei ZA, Rossignac J, Kanter KR, Fogel MA, Yoganathan AP. Fontan Surgical Planning: Previous Accomplishments, Current Challenges, and Future Directions. J Cardiovasc Transl Res. 2018 Apr;11(2):133-144.

Resultados perinatales

¿Qué dice la evidencia?

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Ultrasound Obstet Gynecol 2015; **45**: 657–663
Published online 23 April 2015 in Wiley Online Library (wileyonlinelibrary.com). **DOI:** 10.1002/uog.14634

Perinatal outcome after prenatal diagnosis of single-ventricle cardiac defects

R. S. BEROUKHIM*†, K. GAUVREAU†, O. J. BENAVIDEZ*†, C. W. BAIRD‡, T. LAFRANCHI† and W. TWORETZKY†

**Department of Pediatric/Congenital Cardiology, Massachusetts General Hospital for Children, Boston, MA, USA; †Department of Cardiology, Boston Children's Hospital, Boston, MA, USA; ‡Department of Cardiac Surgery, Boston Children's Hospital, Boston, MA, USA*

Beroukhim RS, Gauvreau K, Benavidez OJ, Baird CW, LaFranchi T, Tworetzky W. Perinatal outcome after prenatal diagnosis of single-ventricle cardiac defects. Ultrasound Obstet Gynecol. 2015 Jun;45(6):657-63.

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Table 1 Characteristics of 312 fetuses diagnosed prenatally with single-ventricle cardiac defects

Characteristic	*Value*
Dominant left ventricle	104 (33)
Tricuspid atresia	46/104 (44)
Double-inlet left ventricle	30/104 (29)
Pulmonary atresia with intact ventricular septum	18/104 (17)
Single left ventricle	5/104 (5)
Left dominant atrioventricular canal defect	3/104 (3)
TGA with straddling tricuspid valve	2/104 (2)
Dominant right ventricle	208 (67)
Standard-risk HLHS	150/208 (72)
High-risk HLHS*	50/208 (24)
Right dominant atrioventricular canal	8/208 (4)
Twin gestation	29 (9)
Hydrops	6 (2)
Prenatal genetic diagnosis and/or other major malformation	26 (8)
GA at first fetal echocardiogram (weeks)	21 (15–41)
GA at delivery (weeks)	38 (31–41)
Birth weight (kg)	3.1 (1.4–4.5)
Age of survivors at follow-up (years)	7.2 (1.9–15.1)

Data are given as *n* (%) or median (range). *HLHS with highly restrictive or intact atrial septum, mitral stenosis with aortic atresia and/or coronary artery sinusoids. GA, gestational age; HLHS, hypoplastic left heart syndrome; TGA, transposition of the great arteries.

Ultrasound Obstet Gynecol 2015; **45**: 657–663
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Table 2 Prenatal outcome of patients diagnosed with single-ventricle cardiac defects before and after 24 weeks' gestation, according to presence of dominant left (LV) or right (RV) ventricle

Prenatal outcome	*All cases (n = 312)*	*Dominant LV (n = 104)*	*Dominant RV (n = 208)*
Termination of pregnancy	98 (31)	32 (31)	66 (32)
In-utero demise	12 (4)	4 (4)	8 (4)
Lost to follow-up after echo before 24 weeks' gestation	9 (3)	3 (3)	6 (3)
Lost to follow-up after echo after 24 weeks' gestation	3 (1)	—	3 (1)
Live birth	190 (61)	65 (63)	125 (60)

Data are given as *n* (%). $P = 0.91$ for comparison of prenatal outcome between dominant LV *vs* dominant RV patients. echo, echocardiography.

Beroukhim RS, Gauvreau K, Benavidez OJ, Baird CW, LaFranchi T, Tworetzky W. Perinatal outcome after prenatal diagnosis of single-ventricle cardiac defects. Ultrasound Obstet Gynecol. 2015 Jun;45(6):657-63.

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Table 3 Postnatal outcome of all liveborn patients with single-ventricle cardiac defects according to presence of dominant left (LV) or right (RV) ventricle

Postnatal outcome	*All cases (n = 190)*	*Dominant LV (n = 65)*	*Dominant RV (n = 125)*	P†
Comfort care	5 (3)	1 (2)	4 (3)	< 0.001
Heart transplant for failed palliation	8 (4)	1 (2)	7 (6)	< 0.001
Death or transplant after intent to treat	52 (27)	8 (12)	44 (35)	< 0.001
Transplantation-free survival after Fontan procedure	129 (68)	56 (86)	73 (58)	< 0.001
Lost to postnatal follow-up before Fontan procedure	4 (2)	—	4 (3)	< 0.001
Age of transplantation-free survivors at follow-up (years)*	7.3 (1.9–15.1)	7.0 (2.6–15.1)	7.4 (1.9–14.8)	0.68

Data are given as *n* (%) or median (range). **n* = 129: excluding those lost to follow-up. †Postnatal outcome compared between those with dominant LV and dominant RV.

Beroukhim RS, Gauvreau K, Benavidez OJ, Baird CW, LaFranchi T, Tworetzky W. Perinatal outcome after prenatal diagnosis of single-ventricle cardiac defects. Ultrasound Obstet Gynecol. 2015 Jun;45(6):657-63.

Ultrasound Obstet Gynecol 2015; **45**: 657–663
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Perinatal outcome after prenatal diagnosis of single-ventricle cardiac defects

R. S. BEROUKHIM*†, K. GAUVREAU†, O. J. BENAVIDEZ*†, C. W. BAIRD‡, T. LAFRANCHI† and W. TWORETZKY†

**Department of Pediatric/Congenital Cardiology, Massachusetts General Hospital for Children, Boston, MA, USA; †Department of Cardiology, Boston Children's Hospital, Boston, MA, USA; ‡Department of Cardiac Surgery, Boston Children's Hospital, Boston, MA, USA*

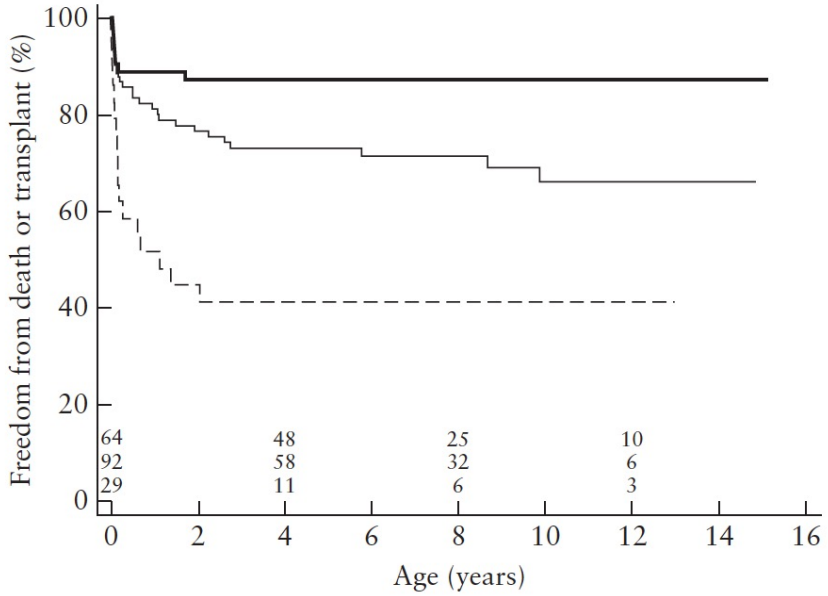

Figure 2 Kaplan–Meier survival plot showing difference in postnatal transplantation-free survival after Fontan procedure of all live births with prenatal diagnosis of dominant LV (——, $n = 65$) and dominant RV categorized into high-risk (– – –, $n = 30$) or standard-risk (——, $n = 95$) hypoplastic left heart syndrome. Log rank test $P < 0.001$. Values at bottom of plot indicate number of patients alive and not lost to follow-up at each time point, displayed in same order as curves.

Beroukhim RS, Gauvreau K, Benavidez OJ, Baird CW, LaFranchi T, Tworetzky W. Perinatal outcome after prenatal diagnosis of single-ventricle cardiac defects. Ultrasound Obstet Gynecol. 2015 Jun;45(6):657-63.

Outcome of Fetuses and Infants With Double Inlet Single Left Ventricle

Edythe B.C. Tham, MBBS[a,*], Rachel Wald, MD[a,b], Doff B. McElhinney, MD[a], Alim Hirji, MD[b], Donna Goff, MD[c], Pedro J. Del Nido, MD[a], Lisa K. Hornberger, MD[c], Lynne E. Nield, MD[b], and Wayne Tworetzky, MD[a]

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Table 1
Prenatal characteristics and anatomic subtypes

Variable	Live Births (n = 43)	Termination/ Neonatal Death (n = 21)	p Value
Gestation at diagnosis (weeks)	26 ± 7	19 ± 3	<0.001
Extracardiac anomalies or chromosomal anomalies	0	5 (24%)	0.01
Anatomic subtypes			
1 AV valve*	12 (28%)	4 (19%)	NS
2 AV valves	29 (67%)	18 (86%)	NS
Systemic obstruction†	21 (49%)	8 (38%)	NS
Pulmonary obstruction	12 (28%)	9 (43%)	NS
No obstruction	9 (21%)	6 (29%)	NS

Data are expressed as mean ± SD or as number (percentage).

* One AV valve was severely hypoplastic or atretic but remained in connection with the single left ventricle.

† Two patients with bilateral outflow tract obstructions were analyzed in the systemic obstruction group.

Tham EB, Wald R, McElhinney DB, Hirji A, Goff D, Del Nido PJ, Hornberger LK, Nield LE, Tworetzky W. Outcome of fetuses and infants with double inlet single left ventricle. Am J Cardiol. 2008 Jun 1;101(11):1652-6.

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Table 2
Postnatal anatomic characteristics across institutions

Variable	Institution			Total	p Value
	A	B	C		
1 AV valve	9 (16%)	20 (27%)	8 (44%)	37 (25%)	0.06
2 AV valves	47 (84%)	55 (73%)	10 (56%)	112 (75%)	NS
Systemic obstruction					
Arch hypoplasia	29 (52%)	20 (27%)	6 (33%)	55 (37%)	0.004
Isolated coarctation	3 (5%)	10 (13%)	2 (11%)	15 (10%)	NS
Pulmonary obstruction	8 (14%)	35 (47%)	5 (28%)	48 (32%)	0.001
No obstruction	16 (29%)	11 (15%)	4 (22%)	31 (21%)	NS
Associated cardiac anomalies	20 (36%)	49 (65%)	7 (39%)	76 (51%)	0.001
Noncardiac congenital or chromosomal anomalies	1 (2%)	11 (13%)	3 (17%)	15 (10%)	0.02

Tham EB, Wald R, McElhinney DB, Hirji A, Goff D, Del Nido PJ, Hornberger LK, Nield LE, Tworetzky W. Outcome of fetuses and infants with double inlet single left ventricle. Am J Cardiol. 2008 Jun 1;101(11):1652-6.

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Table 3
Type and number of surgical procedures (n = 149)

Procedure	No.
Norwood/Damus	63 (42%)
Other neonatal surgery	62 (42%)
Blalock-Taussig shunt	27 (18%)
Pulmonary artery band alone	17 (11%)
Blalock-Taussig shunt and pulmonary artery band	3 (2%)
Coarctation repair	
With arterial switch	2 (1%)
With Blalock-Taussig shunt	2 (1%)
Pulmonary artery band and coarctation repair	8 (8%)
Total anomalous pulmonary venous return repair	1 (0.7%)
Atrial septectomy alone	1 (0.7%)
Interventional: bilateral pulmonary artery band and ductus arteriosus stent	1 (0.7%)
BDCPA	102 (68%)
Fontan	89 (60%)
Total surgeries	316

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Outcome of Patients With Double-Inlet Left Ventricle or Tricuspid Atresia With Transposed Great Arteries

Yueh-Tze Lan, MD,* Ruey-Kang Chang, MD, MPH,* Hillel Laks, MD†
Los Angeles, California

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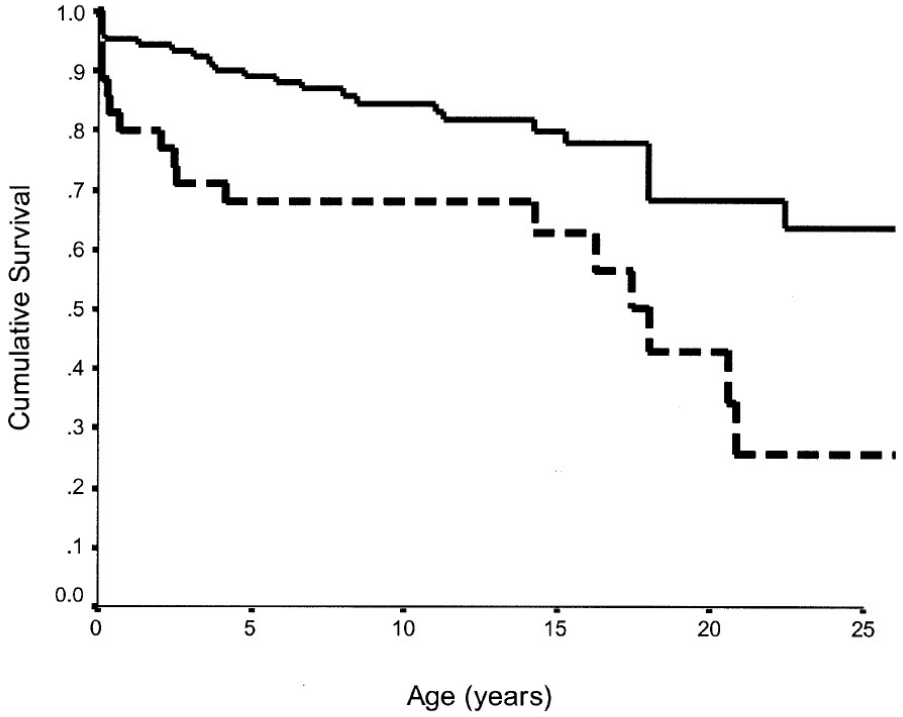

Figure 2. Kaplan-Meier survival curve for all-cause mortality of patients with double-inlet left ventricle up to 25 years of age (n = 105; **solid line**) and patients with tricuspid atresia with transposed great arteries (n = 35; **broken line**).

- 140 pacientes
  - 105 DILV
- Sobrevida
  - 5 años 89%
  - 15 años 80%
  - 25 años 63%

Lan YT, Chang RK, Laks H. Outcome of patients with double-inlet left ventricle or tricuspid atresia with transposed great arteries. J Am Coll Cardiol. 2004 Jan 7;43(1):113-9.

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Los Angeles, California

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Table 2. Characteristics of Patients With a DILV and Patients With TA-TGA

	DILV Group (n = 105)	TA-TGA Group (n = 35)	p Value
Gender (M/F)	77/28	26/9	0.91
Mortality	24 (23)	17 (49)	0.007
Tachyarrhythmia	21 (20)	13 (37)	0.069
Complete heart block	21 (20)	3 (9)	0.20
Pacemaker placement	31 (30)	7 (20)	0.38
Arch anomaly	30 (29)	11 (31)	0.91
Pulmonary atresia/stenosis	44 (42)	11 (31)	0.37
Pulmonary artery banding	72 (69)	17 (49)	0.054
DKS procedure	38 (36)	13 (37)	0.92
BVF/subaortic resection	36 (34)	8 (23)	0.29

Data are presented as the number (%) of patients.

DILV = double-inlet left ventricle; TA-TGA = tricuspid atresia with transposed great arteries; other abbreviations as in Table 1.

Lan YT, Chang RK, Laks H. Outcome of patients with double-inlet left ventricle or tricuspid atresia with transposed great arteries. J Am Coll Cardiol. 2004 Jan 7;43(1):113-9.

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Los Angeles, California

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Table 4. Multivariate Analysis for Mortality Risk Factors

Independent Variables	Odds Ratio*	95% CI	p Value
Demographic variables			
Male gender (female)	0.59	0.21–1.58	0.29
Born after 1990 (before 1990)	1.11	0.37–3.27	0.86
Anatomic variables			
TA-TGA (DILV)	1.54	0.39–6.09	0.53
Presence of PA/PS (no PA/PS)	0.11	0.02–0.73	0.02
Coarctation of aorta (no coarctation)	3.02	0.80–11.4	0.10
AV valve atresia (no AV valve atresia)	2.05	0.56–7.46	0.28
Surgical variables			
BVF/subaortic resection	1.61	0.47–5.49	0.45
Pulmonary artery banding	0.15	0.03–0.61	0.01
DKS procedure	0.61	0.15–2.51	0.49
Pacemaker placement	4.53	1.56–13.14	0.006
Tachyarrhythmia	4.52	1.40–14.64	0.01

*Odds ratios for mortality were calculated using the groups in parenthesis as the reference (odds ratio = 1.0).

AV = atrioventricular; CI = confidence interval; other abbreviations as in Tables 1 and 2.

Lan YT, Chang RK, Laks H. Outcome of patients with double-inlet left ventricle or tricuspid atresia with transposed great arteries. J Am Coll Cardiol. 2004 Jan 7;43(1):113-9.

DOI: 10.1002/pd.4821 PRENATAL DIAGNOSIS

ORIGINAL ARTICLE

Prenatal diagnosis of functionally univentricular heart, associations and perinatal outcomes

Aline Wolter[1]*, Sina Nosbüsch[1], Andreea Kawecki[1], Jan Degenhardt[1], Christian Enzensberger[2], Oliver Graupner[3], Carina Vorisek[1], Hakan Akintürk[4], Can Yerebakan[4], Markus Khalil[5], Dietmar Schranz[5], Jochen Ritgen[6], Rüdiger Stressig[6] and Roland Axt-Fliedner[1]

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Table 1 **Cardiac diagnosis in 155 fetuses with dominant RV and LV**

Dom LV	*n* = 52	Dom RV	*n* = 103
TA	17 (32.7%)	HLHS (high-risk HLHS standard risk HLHS)	63 (61.2%) 17/63 (26.9%) 46/63 (73.1%)
DILV	24 (46.1%)	HLHC	9 (8.7%)
Heterotaxy	2 (3.8%)	DIRV	2 (1.9%)
Unbalanced AVSD	3 (5.8%)	Heterotaxy	11 (10.7%)
PA:IVS	6 (11.5%)	Unbalanced AVSD	13 (12.6%)
		Other	5 (4.9%)

Dom LV, dominant left ventricle; Dom RV, dominant right ventricle; TA, tricuspid valve atresia; DILV, double inlet left ventricle; AVSD, atrioventricular septal defect; PA:IVS, pulmonary valve atresia and intact ventricular septum; HLHS, hypoplastic left heart syndrome; HLHC, hypoplastic left heart complex; DIRV, double inlet right ventricle.

Wolter A, Nosbüsch S, Kawecki A, Degenhardt J, Enzensberger C, Graupner O, Vorisek C, Akintürk H, Yerebakan C, Khalil M, Schranz D, Ritgen J, Stressig R, Axt-Fliedner R. Prenatal diagnosis of functionally univentricular heart, associations and perinatal outcomes. Prenat Diagn. 2016 Jun;36(6):545-54.

Table 2 **Perinatal characteristics of patients with functionally univentricular heart**

	Dom LV *n* = 52	Dom RV *n* = 103	All *n* = 155	*p* Value
Mean maternal age (years)	32.0 (20–43)	31.2 (17–46)	31.5 (17–46)	n.s.
Diagnosis CHD (wks)	23.7 (12.3–39.3)	24.0 (11.7–36.7)	23.8 (11.7–39.3)	n.s.
Abnormal karyotype (71 known karyotypes, 70/71 prenatally known)	3/70 (4.3%)	9/70 (12.9%) +1 postpartum	12/70 (17.1%) 13/71 (18.3%)	n.s. n.s.
Extracardiac malformation	5 (9.6%)	10 (9.7%)	15 (9.7%)	n.s.
EFE	1 (1.9%)	14 (13.6%)	15 (9.7%)	0.02
TOP	5 (9.6%)	13 (12.6%)	18 (11.6%)	n.s.
Birth weight (g)	2947 (552–4430)	3056 (565–4230)	3017 (552–4420)	n.s.
Gestational age at delivery (wks)	37.7 (23.0–42.9)	38.3 (28.7–41.3)	38.1 (23.0–42.9)	n.s.
SGA	5/45 neonates (11.1%)	13/83 neonates (15.7%)	18/128 neonates (14.0%)	n.s.
Delivery<37 wks	4/45 neonates (8.9%)	13/83 neonates (15.7%)	17/128 neonates (13.2%)	n.s.
Delivery<34 wks	3/45 neonates (6.7%)	2/83 neonates (2.4%)	5/128 neonates (3.9%)	n.s.
pH art.	7.32 (6.90–7.46)	7.30 (6.89–7.47)	7.30 (6.89–7.47)	n.s.
APGAR 1 min	8	8	8	n.s.
APGAR 5 min	8	9	9	n.s.
APGAR 10 min	9	9	9	n.s.
Caesarean section (%)	41.7%	51.5%	48.1%	n.s.

Data are given as n (%) or mean (range).

Dom LV, dominant left ventricle; Dom RV, dominant right ventricle; CHD, congenital heart disease; wks, weeks of gestation; n.s., not significant; EFE, endocardial fibroelastosis; TOP, termination of pregnancy; SGA, small for gestational age.

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ORIGINAL ARTICLE

Prenatal diagnosis of functionally univentricular heart, associations and perinatal outcomes

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Table 5 **Postnatal outcome of 115 newborns with intention to treat**

Postnatal outcome	Dom LV $n=41$	Dom RV $n=74$	All $n=115$	P value
Intervention <48 h postpartum	3 (7.3%)	19 (25.6%) (10/17 High-risk HLHS)	22 (19.1%)	0.02
Urgent intervention <48 h postpartum	3 (7.3%)	16 (21.6%) (10/17 High-risk HLHS)	19 (16.5%)	0.07
Reanimation	0	3 (4%) (2/17 High-risk HLHS)	3 (2.6%)	n.s.
HTX listed	1 (2.4%)	7 (9.5%) (3/17 High-risk HLHS)	8 (7.0%)	n.s.
Death < 30 d	1 (2.4%)	10 (13.5%) (4/17 [23.5%] High-risk HLHS)	11 (9.6%)	n.s. (0.02 for High-risk HLHS)
30-d survival	40 (97.6%)	64 (86.5%) (13/17 [76.5%] High-risk HLHS)	104 (90.5%)	n.s. (0.02 for High-risk HLHS)

Dom LV, dominant left ventricle; Dom RV, dominant right ventricle; n.s., not significant; HTX, heart transplantation.

Wolter A, Nosbüsch S, Kawecki A, Degenhardt J, Enzensberger C, Graupner O, Vorisek C, Akintürk H, Yerebakan C, Khalil M, Schranz D, Ritgen J, Stressig R, Axt-Fliedner R. Prenatal diagnosis of functionally univentricular heart, associations and perinatal outcomes. Prenat Diagn. 2016 Jun;36(6):545-54.

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European Journal of Cardio-Thoracic Surgery 0 (2017) 1–7
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ORIGINAL ARTICLE

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Improved long-term outcomes in double-inlet left ventricle and tricuspid atresia with transposed great arteries: systemic outflow tract obstruction present at birth defines long-term outcome

Lotte C. Franken[a,b,†], Manouk Admiraal[a,b,†], Charlotte E. Verrall[a,c], Diana Zannino[d], Julian G. Ayer[a,c,e], Ajay J. Iyengar[c,d,f,g], Andrew D. Cole[a], Gary F. Sholler[a,c,e], Yves D'Udekem[c,d,f,g] and David S. Winlaw[a,c,e,*]

Franken LC, Admiraal M, Verrall CE, Zannino D, Ayer JG, Iyengar AJ, Cole AD, Sholler GF, D'Udekem Y, Winlaw DS. Improved long-term outcomes in double-inlet left ventricle and tricuspid atresia with transposed great arteries: systemic outflow tract obstruction present at birth defines long-term outcome. Eur J Cardiothorac Surg. 2017 Jun 1;51(6):1051-1057.

Table 1: Patient characteristics

Variable	All (*n* = 211)	DILV (*n* = 152)	TA-TGA (*n* = 59)
Male	129 (61%)	92 (61%)	37 (63%)
CHW patient (others RCH)	101 (48%)	70 (47%)	31 (53%)
Additional morphologies			
Subpulmonary stenosis	53 (25%)	41 (27%)	12 (20%)
Pulmonary atresia	19 (9%)	14 (9%)	5 (8%)
Coarctation of aorta	54 (26%)	39 (26%)	15 (25%)
Interrupted arch	12 (6%)	7 (5%)	5 (8%)
Valve abnormalities	32 (15%)	27 (18%)	5 (8%)
Operative characteristics			
First palliation	*n* = 207	*n* = 150	*n* = 57
Single stage Fontan procedure	19 (9%)	15 (10%)	4 (7%)
PA banding	116 (56%)	82 (55%)	34 (60%)
With arch repair	41/116 (35%)	31/82 (38%)	10/34 (29%)
Without arch repair	75/116 (65%)	51/82 (62%)	24/34 (71%)
Norwood	20 (10%)	11 (7%)	9 (16%)
Shunt	29 (14%)	23 (15%)	6 (11%)
Age at first palliation in days[a]	*n* = 202	*n* = 145	*n* = 57
Median [range]	26.5 [1–11461]	26 [1–11461]	29 [1–1631]
PA banding	*n* = 114, 26.5 [1–11461]	*n* = 80, 25.5 [1–11461]	*n* = 34, 28 [2–1029]
Norwood type procedure	*n* = 20, 5.5 [2–170]	*n* = 11, 7 [2–170]	*n* = 9, 4 [2–87]
Shunt	*n* = 28, 5 [1–2191]	*n* = 22, 4 [1–2191]	*n* = 6, 173 [1–557]
Staging with BDG	*n* = 138 (65%)	*n* = 102 (67%)	*n* = 36 (61%)
Age at BDG in months, mean (SD)	26.67 (39.98)	27.69 (43.34)	23.76 (28.65)
DKS	85/190 (45%)	60/138 (43%)	25/52 (48%)
Aortic arch repair	64/205 (31%)	44/149 (30%)	20/56 (36%)
Revision of arch repair	10/64 (16%)	7/44 (16%)	3/20 (15%)
Systemic outflow tract obstruction			
SOTO present at birth	*n* = 208, 13 (6%)	*n* = 151, 7 (5%)	*n* = 57, 6 (11%)
SOTO developed over time	*n* = 169, 65 (38%)	*n* = 117, 51 (44%)	*n* = 52, 14 (27%)
Resection SOTO			
BVF enlargement	*n* = 184, 26 (14%)	*n* = 132, 19 (14%)	*n* = 52, 7 (13%)
Subaortic resection	*n* = 184, 3 (2%)	*n* = 132, 3 (2%)	*n* = 52, 0 (0%)
Fontan completion	170 (83%)	135 (91%)	35 (62%)
Missing	7	4	3

DILV: double inlet left ventricle; TA-TGA: tricuspid atresia-transposed great arteries; CHW: Children's Hospital at Westmead; RCH: Royal Children's Hospital; DKS: Damus-Kaye-Stansel; SOTO: systemic outflow tract obstruction; PA: pulmonary artery; BDG: Bi-directional Glenn; BVF: bulboventricular foramen.
[a]Distribution is highly skewed. Appropriate to report medians for this variable.

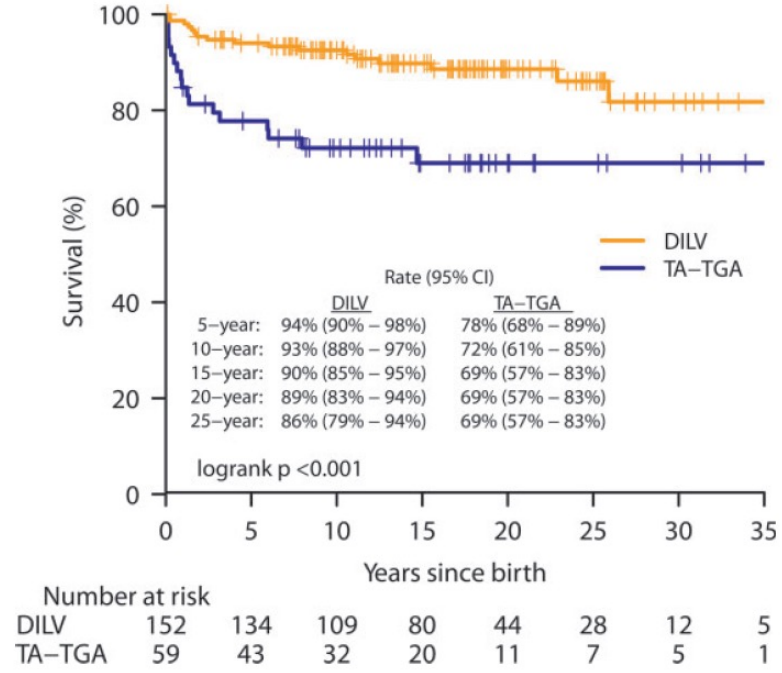

Figure 2: Kaplan–Meier curves of overall survival according to morphologic subgroup. TA-TGA patients have a worse early outcome with a 5-year survival of 78% vs. 94% for the DILV cohort ($P < 0.001$). DILV: double inlet left ventricle; TA-TGA: tricuspid atresia-transposed great arteries.

Table 4: Overall survival–multivariable Cox model ($n = 208$, 34 deaths)

Variable	Level	HR (95% CI)	*P*-value (HR = 1)
Primary diagnosis—DILV	No (ref)	1	
	Yes	0.32 (0.16–0.63)	0.001
SOTO initial	No (ref)	1	
	Yes	3.54 (1.36–9.20)	0.01

HR: hazard ratio; CI: confidence interval; DILV: double inlet left ventricle; SOTO: systemic outflow tract obstruction.

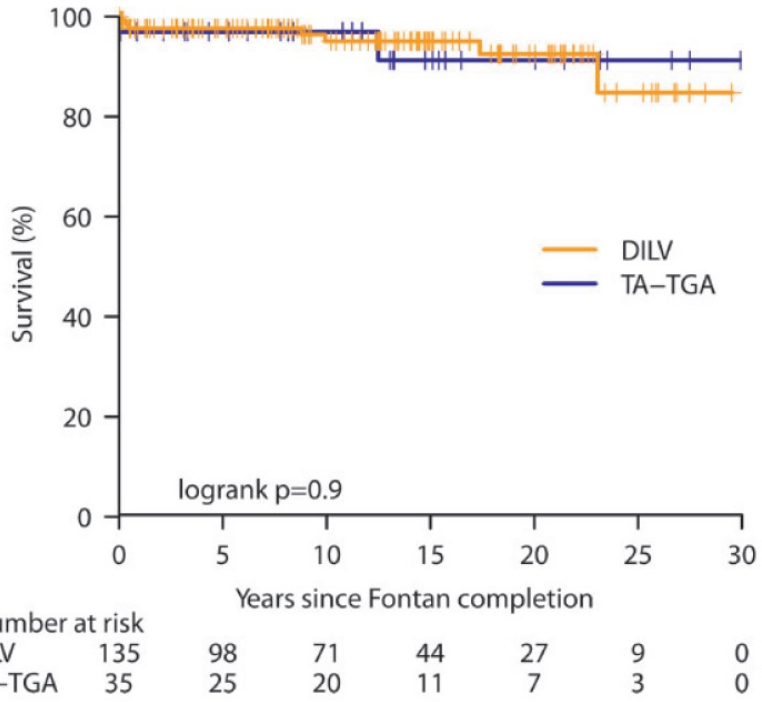

Figure 4: Survival of the TA-TGA subgroup is not different to DILV after Fontan completion. TA-TGA: tricuspid atresia-transposed great arteries; DILV: double inlet left ventricle.

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Centro de Referencia Perinatal Oriente
Facultad de Medicina, Universidad de Chile

Ventrículo único de doble entrada

Dr. Pablo Silva Labarca
Becado Medicina Materno Fetal
Universidad de Chile