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

Terapia Fetal en Cardiopatías Congénitas

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Introducción

- Las cardiopatías congénitas son las afecciones mas comunes en la vida prenatal.
- Incidencia 6-12/1000 nacidos vivos.
- La mayoría de las malformaciones cardiacas ocurren tempranamente en el desarrollo y permanecen estables en el embarazo.
- Un pequeño grupo de cardiopatías pueden ser evolutivas a lo largo de la gestación y se podrían beneficiar de la terapia fetal.

Terapia Fetal en Cardiopatías Congénitas

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- **<u>Estenosis Aórtica Severa</u>**
  - Síndrome de Hipoplasia de Ventrículo Izquierdo
- **<u>Atresia Pulmonar con septum interventricular intacto</u>**
  - Síndrome de Hipoplasia de Ventrículo Derecho
- **<u>Síndrome de Hipoplasia de Ventrículo Izquierdo con Septum atrial intacto o restrictivo</u>**

Freud LR, Tworetzky W. Fetal interventions for congenital heart disease. Curr Opin Pediatr. 2016 Apr;28(2):156-62.

Objetivos Terapia Fetal

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Estenosis Aórtica Severa / SHVI	Lograr circulación biventricular
Atresia Pulmonar con septum interventricular intacto / SHVD	Lograr circulación biventricular
SHVI / Septum atrial intacto/restrictivo	Mejorar sobrevida

Freud LR, Tworetzky W. Fetal interventions for congenital heart disease. Curr Opin Pediatr. 2016 Apr;28(2):156-62.

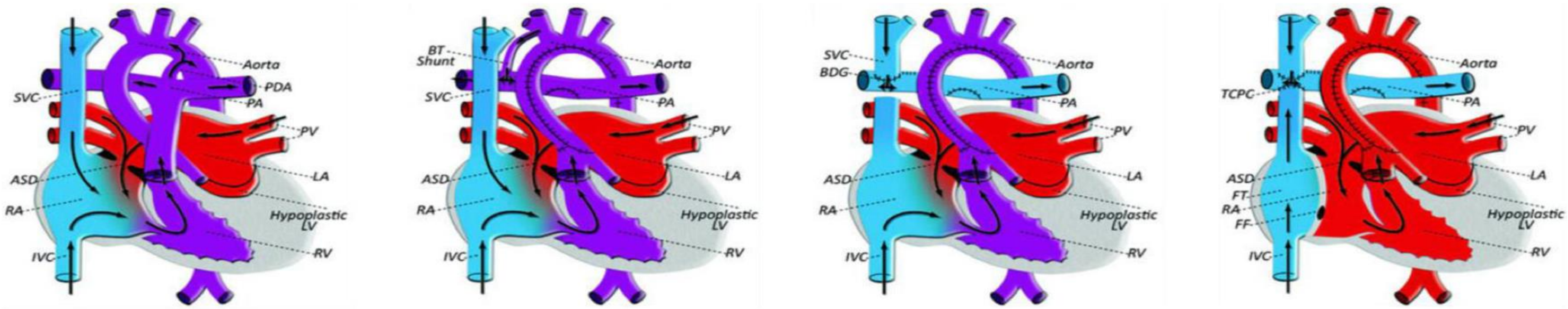
Estenosis Aórtica Severa - SHVI

- La estenosis aórtica severa diagnosticada en 2do trimestre con ventrículo izquierdo normal o dilatado puede evolucionar a SHVI.

- **Fisiopatología**
  - *VI bombea contra una postcarga elevada*
  - *Dilatación y disfunción ventricular*
  - *Regurgitación mitral*
  - *Disminución de la perfusión miocárdica*
  - *Muerte celular y desarrollo de tejido cicatricial*
  - *Detención del desarrollo de VI*

Schidlow DN, Tworetzky W, Wilkins-Haug LE. Percutaneous fetal cardiac interventions for structural heart disease. Am J Perinatol. 2014 Aug;31(7):629-36.

Cirugía en SHVI



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.

EAo/SHVI

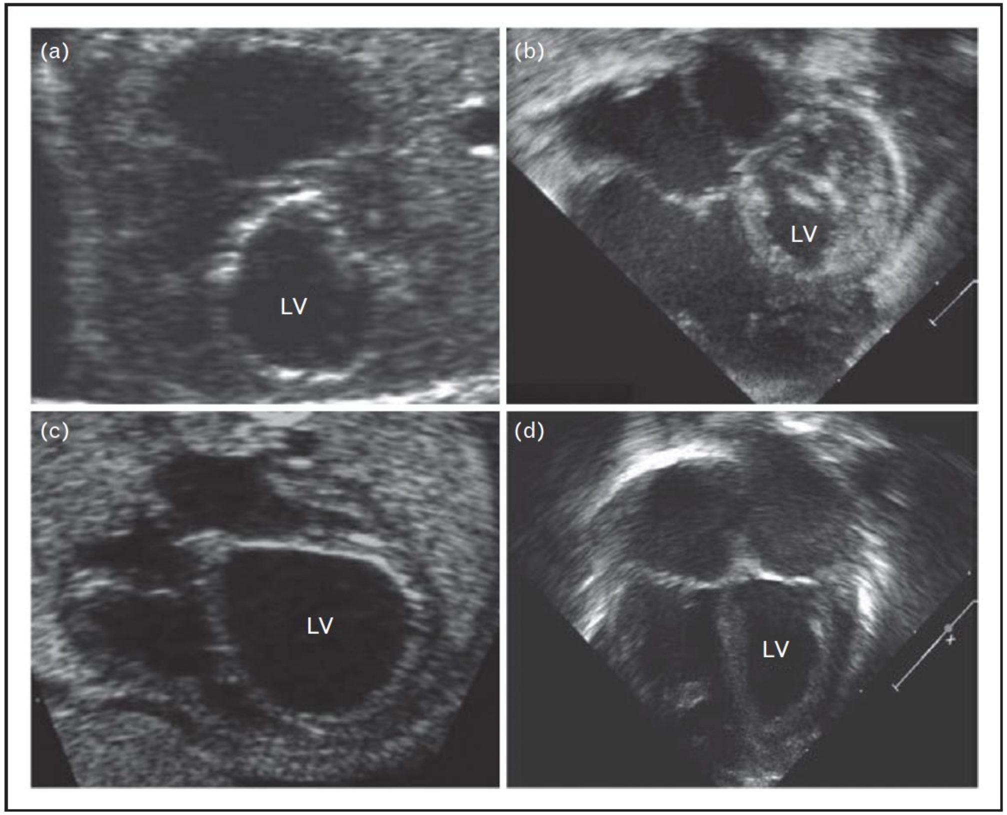


FIGURE 2. Four-chamber views of the heart are demonstrated for two patients, with the preintervention fetal echocardiograms on the left and the postnatal echocardiograms on the right. The patient depicted in the first row (images a and b) had a technically unsuccessful fetal aortic valvuloplasty and was managed as HLHS after birth, whereas the patient in the second row (images c and d) had a technically successful procedure and a biventricular outcome. (Reprinted with permission from [20■].) LV, left ventricle.

Freud LR, Tworetzky W. Fetal interventions for congenital heart disease. Curr Opin Pediatr. 2016 Apr;28(2):156-62.

Ecocardiografía fetal en EAo/SHVI

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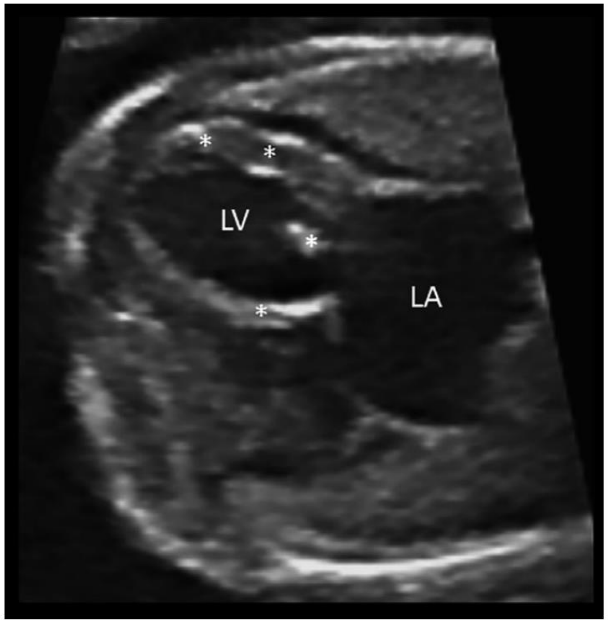

FIGURE 2 Typical appearance of the myocardium in aortic stenosis with evolving hypoplastic left heart syndrome. The left ventricle is dilated. There are multiple areas of increased echogenicity consistent with left ventricular endocardial fibroelastosis. LA = left atrium; LV = left ventricle; *endocardial fibroelastosis

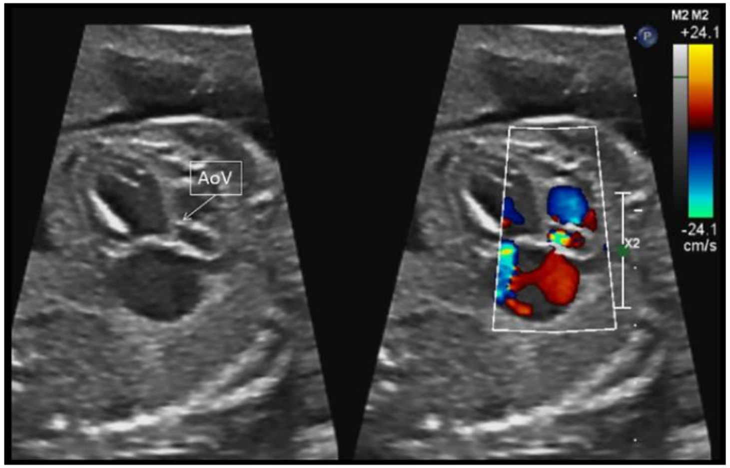

FIGURE 1 Color compare image of aortic stenosis with evolving hypoplastic left heart syndrome. The left panel demonstrates a two-dimensional image of a thickened and doming aortic valve. The right panel demonstrates a small jet of antegrade aortic valve flow consistent with severe aortic stenosis. AoV = aortic valve

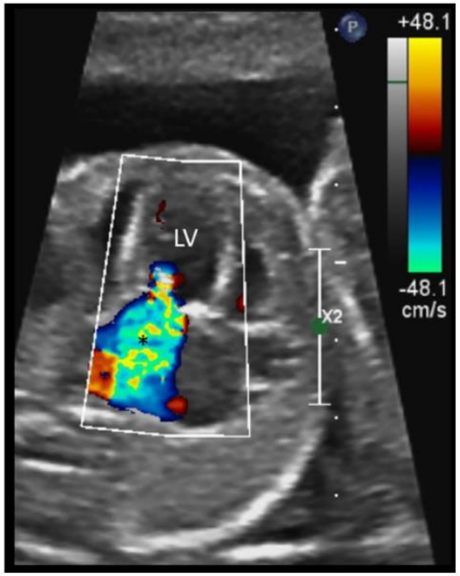

FIGURE 4 Significant mitral regurgitation in a fetus with aortic stenosis and evolving hypoplastic left heart syndrome. LV = left ventricle; *mitral regurgitation

Schidlow DN, Tworetzky W, Wilkins-Haug LE. Percutaneous fetal cardiac interventions for structural heart disease. Am J Perinatol. 2014 Aug;31(7):629-36.

Ecocardiografía fetal en EAo/SHVI

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TABLE 1 Key elements of echocardiographic assessment to identify evolving hypoplastic left heart syndrome (eHLHS)

LV long-axis dimension Z-score
LV short-axis dimension Z-score
Aortic valve annulus Z-score
Mitral valve annulus Z-score
Mitral valve regurgitation peak gradient
Aortic valve stenosis peak gradient
Assessment of ventricular systolic function
Direction of transverse aortic arch flow

Schidlow DN, Freud L, Friedman K, Tworetzky W. Fetal interventions for structural heart disease. Echocardiography. 2017 Dec;34(12):1834-1841.

EAo/SHVI

Selección de Pacientes

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- ¿Puede la ecocardiografía fetal identificar aquellos fetos que van a desarrollar SHVI?
- ¿Se puede realizar una valvuloplastía aórtica de manera segura y exitosa en esos fetos?
- ¿Una valvuloplastía aórtica fetal exitosa puede restaurar un flujo adecuado que permita el desarrollo del VI?

Tworetzky W, Wilkins-Haug L, Jennings RW, van der Velde ME, Marshall AC, Marx GR, Colan SD, Benson CB, Lock JE, Perry SB. Balloon dilation of severe aortic stenosis in the fetus: potential for prevention of hypoplastic left heart syndrome: candidate selection, technique, and results of successful intervention. Circulation. 2004 Oct 12;110(15):2125-31.

EAo/SHVI

Selección de Pacientes

- Elementos que sugieren progresión a SHVI
  - Flujo retrógrado en arco aórtico transverso
  - Flujo izquierda-derecha en foramen oval
  - Onda E/A mitral monofásica
  - Disfunción sistólica de VI

Schidlow DN, Freud L, Friedman K, Tworetzky W. Fetal interventions for structural heart disease. Echocardiography. 2017 Dec;34(12):1834-1841.

EAo/SHVI

Selección de Pacientes

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TABLE 2 Threshold scoring system to estimate potential for biventricular outcome in mid-gestation fetuses with AS and evolving hypoplastic left syndrome (HLHS)

LV long-axis Z-score > 0
LV short-axis Z-score > 0
Aortic annulus Z-score > −3.5
MV annulus Z-score > −2
MV or AS maximum gradient 20

One point assigned for each of the above parameters. A threshold score ≥ 4 had 100% sensitivity, 53% specificity, 38% positive predictive value, and 100% negative predictive value for identifying fetuses that had a biventricular outcome from the time of birth without intermediate univentricular palliation.[41]

Schidlow DN, Freud L, Friedman K, Tworetzky W. Fetal interventions for structural heart disease. Echocardiography. 2017 Dec;34(12):1834-1841.

Técnica Valvuloplastía aórtica fetal

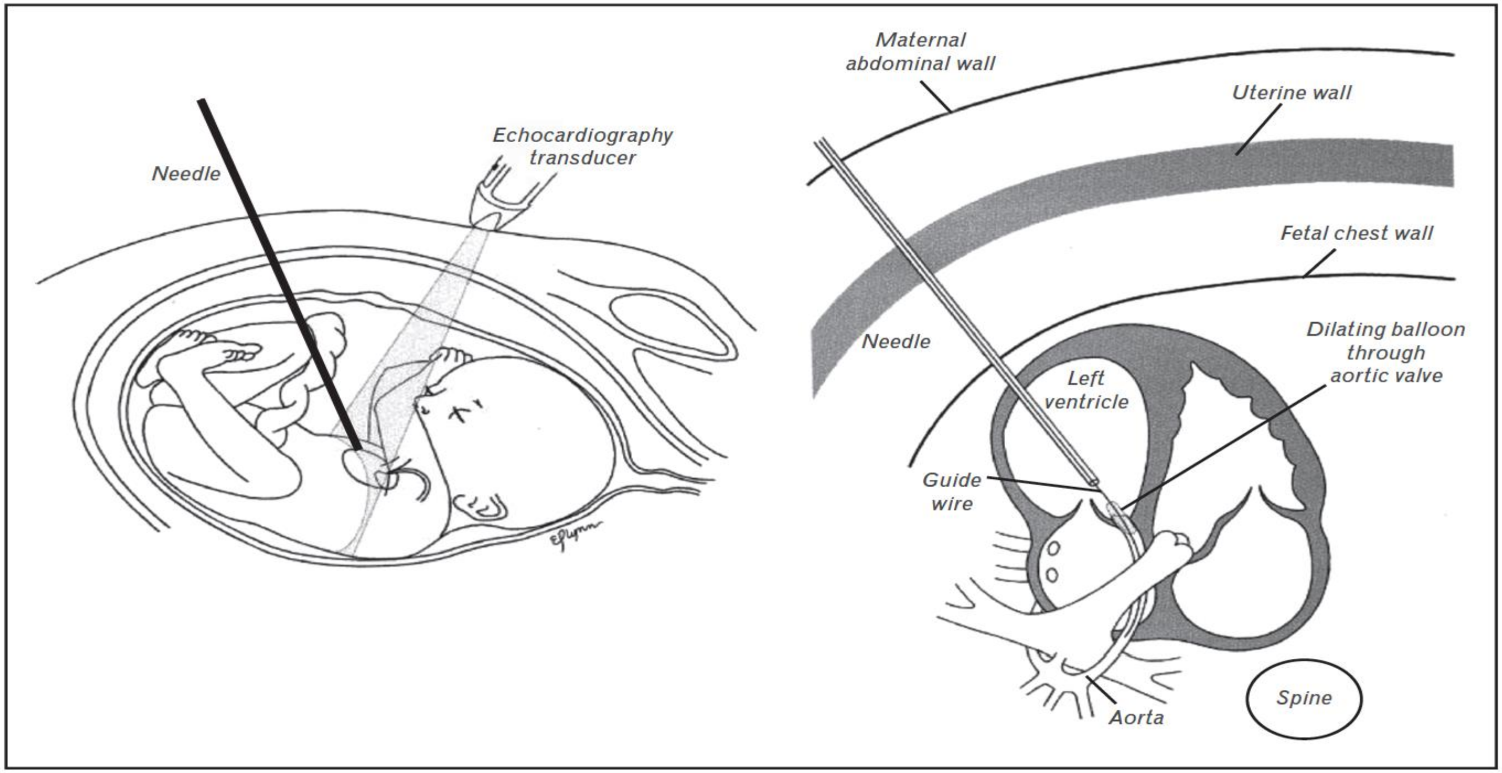

FIGURE 1. Ideal fetal position is demonstrated, with the fetal left chest anterior. The cannula has an unobstructed pathway from the maternal abdomen to the left ventricular (LV) apex. Once the LV apex is punctured, the guide wire and coronary angioplasty catheter are positioned for balloon dilation across the aortic valve. (Reprinted with permission from [11].).

Freud LR, Tworetzky W. Fetal interventions for congenital heart disease. Curr Opin Pediatr. 2016 Apr;28(2):156-62.

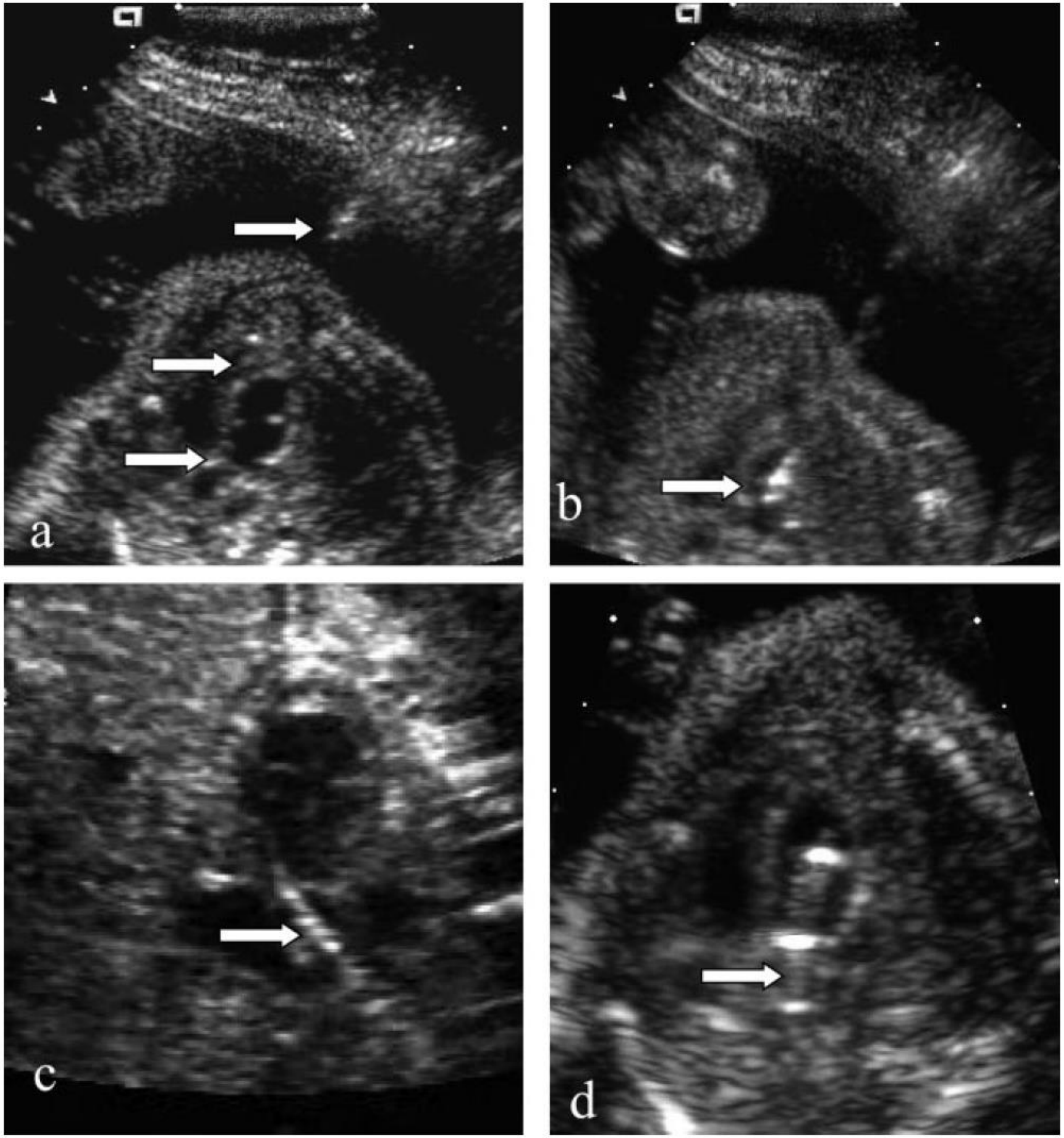

Figure 2. Ultrasound images of technically successful percutaneous in utero aortic valvuloplasty. a, LV before intervention. Four-chamber view demonstrating echogenic LV, apex-forming RV, and alignment of needle (arrow), LV apex (arrow), aortic valve, and ascending aorta (arrow). b, Cannula in LV. After LV puncture with cannula, LV appears smaller. Cannula can be seen as echo-bright line in LV, with its tip directed at aortic valve (arrow). c, Guidewire across aortic valve in ascending aorta (arrow). d, Balloon across valve. Correct position of balloon across aortic valve is confirmed by external measurements of catheter and by ultrasound imaging. Balloon inflation appears as movement of bright reflection along line of LV outflow track and across level of valve (arrow).

Tworetzky W, Wilkins-Haug L, Jennings RW, van der Velde ME, Marshall AC, Marx GR, Colan SD, Benson CB, Lock JE, Perry SB. Balloon dilation of severe aortic stenosis in the fetus: potential for prevention of hypoplastic left heart syndrome: candidate selection, technique, and results of successful intervention. Circulation. 2004 Oct 12;110(15):2125-31.

Congential Heart Disease

Balloon Dilation of Severe Aortic Stenosis in the Fetus

Potential for Prevention of Hypoplastic Left Heart Syndrome

Candidate Selection, Technique, and Results of Successful Intervention

Wayne Tworetzky, MD; Louise Wilkins-Haug, MD, PhD; Russell W. Jennings, MD;
Mary E. van der Velde, MD; Audrey C. Marshall, MD; Gerald R. Marx, MD; Steven D. Colan, MD;
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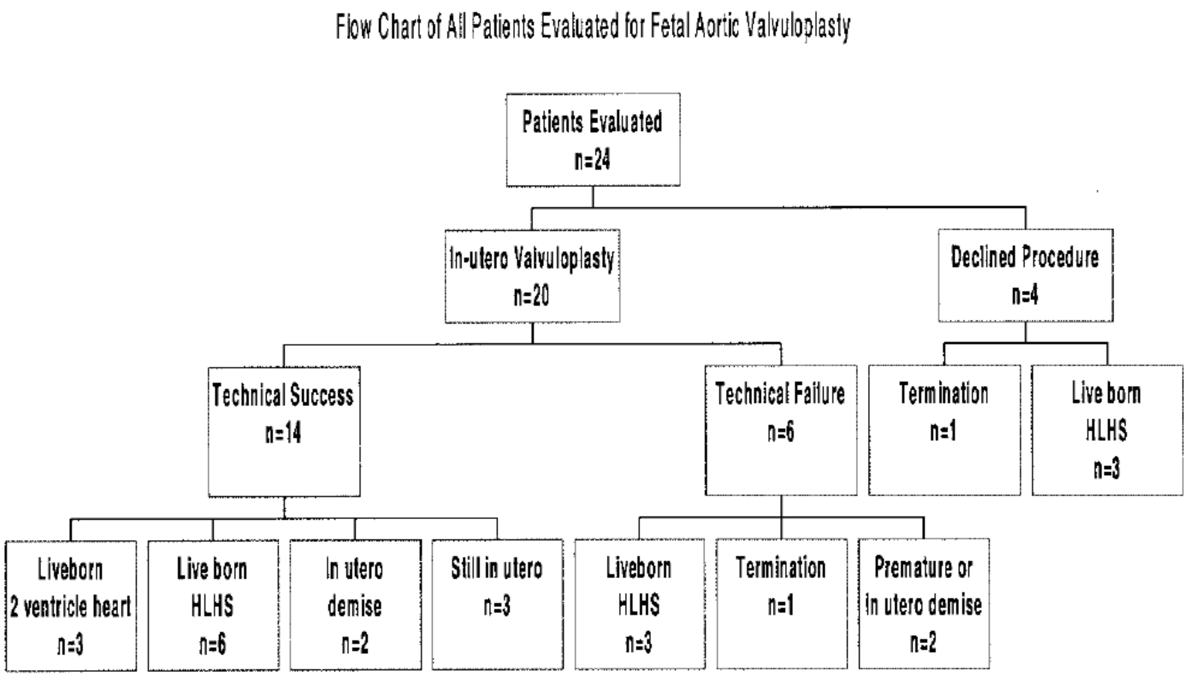

Figure 6. Flow chart showing perinatal outcomes of all 24 patients initially considered candidates for in utero aortic valvuloplasty.

Tworetzky W, Wilkins-Haug L, Jennings RW, van der Velde ME, Marshall AC, Marx GR, Colan SD, Benson CB, Lock JE, Perry SB. Balloon dilation of severe aortic stenosis in the fetus: potential for prevention of hypoplastic left heart syndrome: candidate selection, technique, and results of successful intervention. Circulation. 2004 Oct 12;110(15):2125-31.

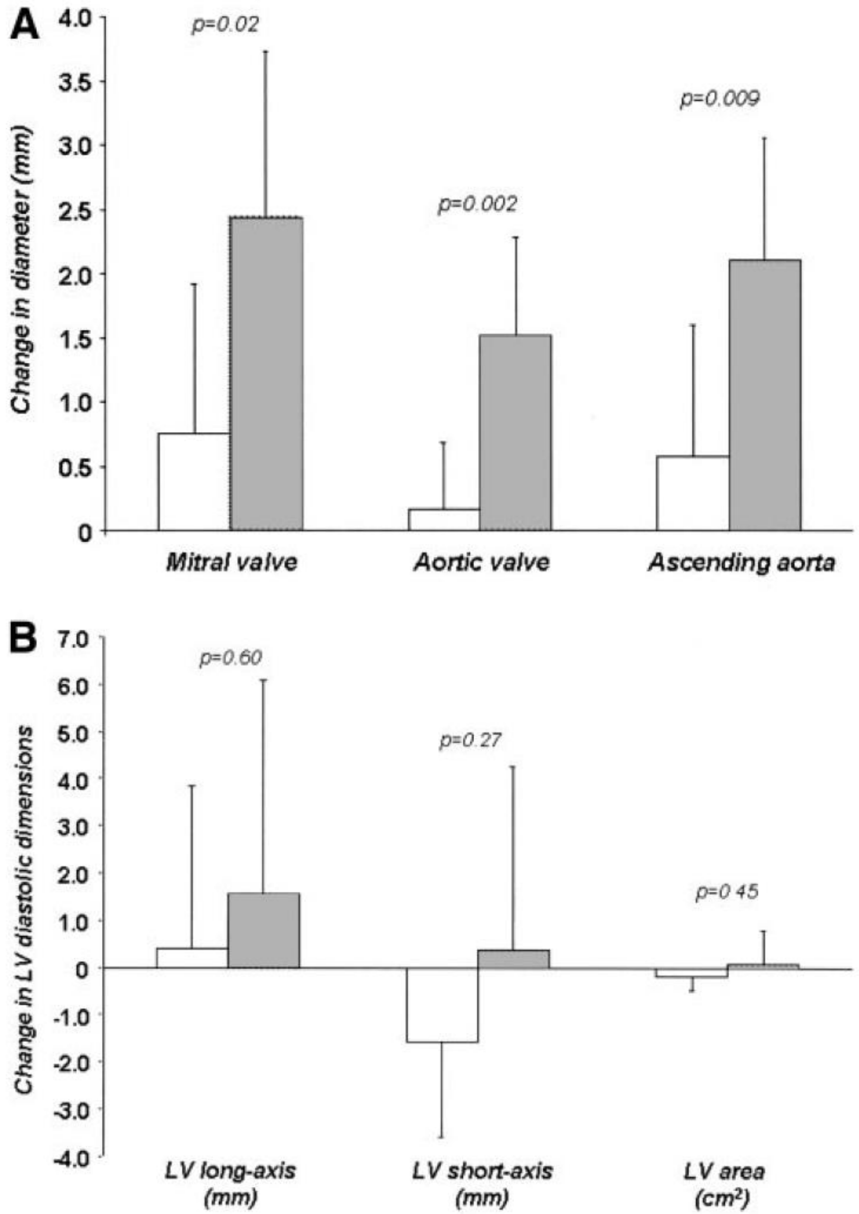

Figure 5. A, Change in dimension of left heart structures (mitral and aortic valves and ascending aorta) in fetuses that had technically successful in utero aortic valvuloplasty (gray bars) vs those with unsuccessful procedure and those that declined procedure (white bars). Only fetuses with pregnancies carried to near-term delivery (>33 weeks' gestation) were included. Data reflect first and last measurements made during gestation (n=15 for data in this figure; of original 24 patients evaluated, 3 are still in utero, 3 died in utero, 1 was born prematurely, and 2 pregnancies were terminated). B, Change in LV diastolic dimensions (long axis, short axis, and cross-sectional area) of fetuses that had technically successful in utero aortic valvuloplasty (gray bars) compared with those with unsuccessful procedure and those that declined procedure (white bars). Only fetuses with pregnancies carried to near-term delivery (>33 weeks' gestation) were included. Data reflect first and last measurements made during gestation (n=15 for data in this figure; of original 24 patients evaluated, 3 are still in utero, 3 died in utero, 1 was born prematurely, and 2 pregnancies were terminated).

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Tworetzky W, Wilkins-Haug L, Jennings RW, van der Velde ME, Marshall AC, Marx GR, Colan SD, Benson CB, Lock JE, Perry SB. Balloon dilation of severe aortic stenosis in the fetus: potential for prevention of hypoplastic left heart syndrome: candidate selection, technique, and results of successful intervention. Circulation. 2004 Oct 12;110(15):2125-31.

Ultrasound Obstet Gynecol 2007; **30**: 715–720
Published online 3 September 2007 in Wiley InterScience (www.interscience.wiley.com). **DOI:** 10.1002/uog.5132

Changes in left heart hemodynamics after technically successful *in-utero* aortic valvuloplasty

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Table 1 Doppler and left heart functional variables in fetuses undergoing technically successful *in-utero* aortic valvuloplasty ($n = 26$)

Echocardiographic variable	*Pre-intervention*	*Post-intervention*	P
Biphasic MV inflow	5/25 (20%)*	21/23 (91%)*	< 0.001
MV inflow duration Z-score	−3.1 ± 1.8	−0.3 ± 0.9	< 0.001
MV inflow duration indexed to cardiac cycle length	0.29 ± 0.12	0.46 ± 0.08	0.001
Heart rate (bpm)	144 ± 13	136 ± 11	0.004
Heart rate Z-score	−0.2 ± 1.1	−0.6 ± 1.4	NS
Moderate or severe MV regurgitation	14/26 (54%)	5/23 (22%)*	0.02
LV ejection fraction (%)	19 ± 10	39 ± 14	< 0.001
Antegrade aortic valve color Doppler jet width (mm)	1.0 ± 0.3	3.9 ± 1.3	< 0.001
Aortic valve jet width indexed to aortic annulus diameter	0.37 ± 0.10	0.88 ± 0.31	< 0.001
Antegrade flow in transverse arch	0/25 (0%)*	17/26 (65%)	< 0.001
Bidirectional flow across foramen ovale	0/26 (0%)	6/25 (24%)*	0.01

Data are presented as mean ± SD or frequency (%). *Color and/or pulsed Doppler interrogation of the mitral valve and the aortic arch was not performed in up to three cases (different measurements in different patients) for some of the measurements. Statistical analyses for each variable only included patients with both pre- and post-intervention data. LV, left ventricle; MV, mitral valve; NS, not significant.

Selamet Tierney ES, Wald RM, McElhinney DB, Marshall AC, Benson CB, Colan SD, Marcus EN, Marx GR, Levine JC, Wilkins-Haug L, Lock JE, Tworetzky W. Changes in left heart hemodynamics after technically successful in-utero aortic valvuloplasty. Ultrasound Obstet Gynecol. 2007 Oct;30(5):715-20.

Table 2 Doppler and left heart functional variables in controls and fetuses undergoing technically unsuccessful *in-utero* aortic valvuloplasty ($n = 18$)

Echocardiographic variable	*Baseline*	*Late gestation*	P
Biphasic MV inflow	1/16 (6%)*	2/11 (18%)*	NS
MV inflow duration *Z*-score	-3.1 ± 1.9	—*	
MV inflow duration indexed to cardiac cycle length	0.36 ± 0.18	—*	
Heart rate (bpm)	146 ± 15	140 ± 11	0.08
Heart rate *Z*-score	-0.4 ± 2.5	-0.4 ± 0.7	NS
Moderate or severe MV regurgitation	7/17 (41%)*	5/12 (42%)*	NS
Antegrade aortic valve color Doppler jet width (mm)	1.4 ± 0.8†	2.4 ± 1.6†	0.07
Aortic valve jet width indexed to aortic annulus diameter	0.54 ± 0.27†	0.73 ± 0.61†	NS
Antegrade flow in transverse arch	0/18 (0%)	0/18 (0%)	NS
Bidirectional flow across foramen ovale	0/18 (0%)	0/18 (0%)	NS

Data are presented as mean ± SD or frequency (%). *Color and/or pulsed Doppler interrogation of the mitral valve and the aortic arch was not performed for some of the measurements. Only variables with data available for at least 50% of control fetuses were included. Follow-up data were available in less than 50% of fetuses for MV inflow duration; these data are not presented. For four patients, the MV inflow pattern could be distinguished from Doppler evaluation of the LV outflow tract, but inflow duration could not be calculated due to the inability to distinguish the baseline clearly and consistently. †$n = 14$. LV, left ventricle; MV, mitral valve; NS, not significant.

Selamet Tierney ES, Wald RM, McElhinney DB, Marshall AC, Benson CB, Colan SD, Marcus EN, Marx GR, Levine JC, Wilkins-Haug L, Lock JE, Tworetzky W. Changes in left heart hemodynamics after technically successful in-utero aortic valvuloplasty. Ultrasound Obstet Gynecol. 2007 Oct;30(5):715-20.

Congenital Heart Disease

Predictors of Technical Success and Postnatal Biventricular Outcome After In Utero Aortic Valvuloplasty for Aortic Stenosis With Evolving Hypoplastic Left Heart Syndrome

Doff B. McElhinney, MD; Audrey C. Marshall, MD; Louise E. Wilkins-Haug, MD, PhD; David W. Brown, MD; Carol B. Benson, MD; Virginia Silva, MSN; Gerald R. Marx, MD; Arielle Mizrahi-Arnaud, MD; James E. Lock, MD; Wayne Tworetzky, MD

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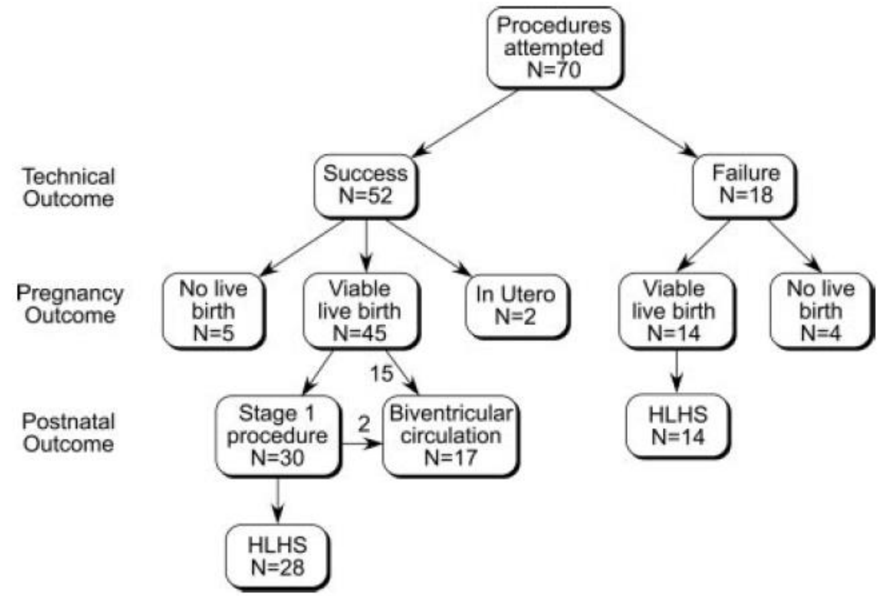

Figure 1. Flow diagram depicting procedural, pregnancy, and postnatal outcomes among 70 fetuses that underwent attempted prenatal aortic valvuloplasty for AS with evolving HLHS.

74% de éxito
30% circulación biventricular
40% complicaciones

McElhinney DB, Marshall AC, Wilkins-Haug LE, Brown DW, Benson CB, Silva V, Marx GR, Mizrahi-Arnaud A, Lock JE, Tworetzky W. Predictors of technical success and postnatal biventricular outcome after in utero aortic valvuloplasty for aortic stenosis with evolving hypoplastic left heart syndrome. Circulation. 2009 Oct 13;120(15):1482-90.

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Table 3. Comparison of Fetuses That Underwent Technically Successful and Unsuccessful In Utero Aortic Valvuloplasty

Variable	Technically Successful Intervention (n=52)	Technically Unsuccessful Intervention (n=18)	*P*
Gestational age, wk	23.8±2.1	24.6±3.6	0.26
Anatomic features			
Aortic annulus diameter, mm	2.91±0.45	3.01±0.82	0.56
Aortic annulus diameter *Z* score	−2.5±0.9	−2.6±1.0	0.60
Ascending aorta diameter, mm	4.18±1.08	3.76±1.61	0.20
Ascending aorta diameter *Z* score	−0.5±1.9	−1.7±2.0	0.03
LV long-axis dimension, cm	1.88±0.36	1.57±0.63	0.01
LV long-axis dimension *Z* score	0.9±1.8	−1.2±1.6	<0.001
LV short-axis dimension, cm	1.26±0.33	1.20±0.42	0.57
LV short-axis dimension *Z* score	3.0±2.6	1.8±1.7	0.09
LV sphericity index	0.66±0.11	0.79±0.13	<0.001
RV long-axis dimension, cm	1.84±0.25	1.76±0.49	0.38
RV long-axis dimension *Z* score	1.9±1.6	0.2±1.7	0.01
Physiological features			
LV ejection fraction, %	22.8±11.4	20.1±10.6	0.39
"High" LV pressure (n=29), n (%)	24 (49)	5 (36)	0.38
Moderate or severe MR (n=38), n (%)	29 (56)	9 (50)	0.67
Procedural factors, n (%)			
Limited laparotomy (n=19)	14 (27)	5 (28)	0.94
Transplacental access (n=22)	16 (32)	6 (35)	0.80

All fetuses that underwent intervention are included, regardless of pregnancy or postnatal outcome. Data are presented as mean±SD.

McElhinney DB, Marshall AC, Wilkins-Haug LE, Brown DW, Benson CB, Silva V, Marx GR, Mizrahi-Arnaud A, Lock JE, Tworetzky W. Predictors of technical success and postnatal biventricular outcome after in utero aortic valvuloplasty for aortic stenosis with evolving hypoplastic left heart syndrome. Circulation. 2009 Oct 13;120(15):1482-90.

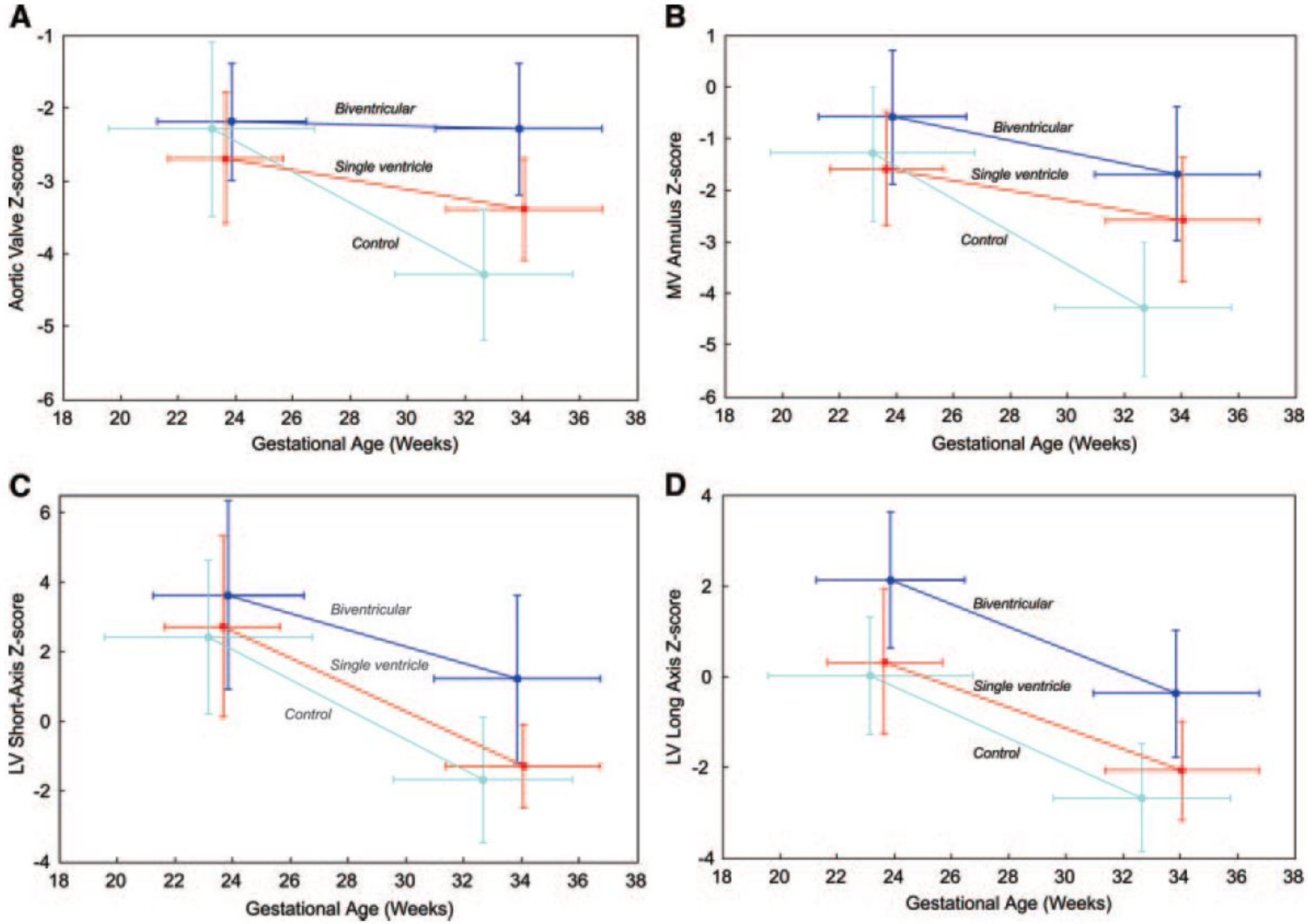

Figure 2. Mean±SD *Z* scores of the aortic annulus (A), MV annulus (B), LV short axis (C), and LV long axis (D) at the time of prenatal intervention and at the latest follow-up fetal echocardiogram with adequate data in fetuses with a biventricular outcome after technically successful intervention (from birth or after initial univentricular staging), fetuses with a univentricular outcome after technically successful intervention (single ventricle), and comparison fetuses (control).

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McElhinney DB, Marshall AC, Wilkins-Haug LE, Brown DW, Benson CB, Silva V, Marx GR, Mizrahi-Arnaud A, Lock JE, Tworetzky W. Predictors of technical success and postnatal biventricular outcome after in utero aortic valvuloplasty for aortic stenosis with evolving hypoplastic left heart syndrome. Circulation. 2009 Oct 13;120(15):1482-90.

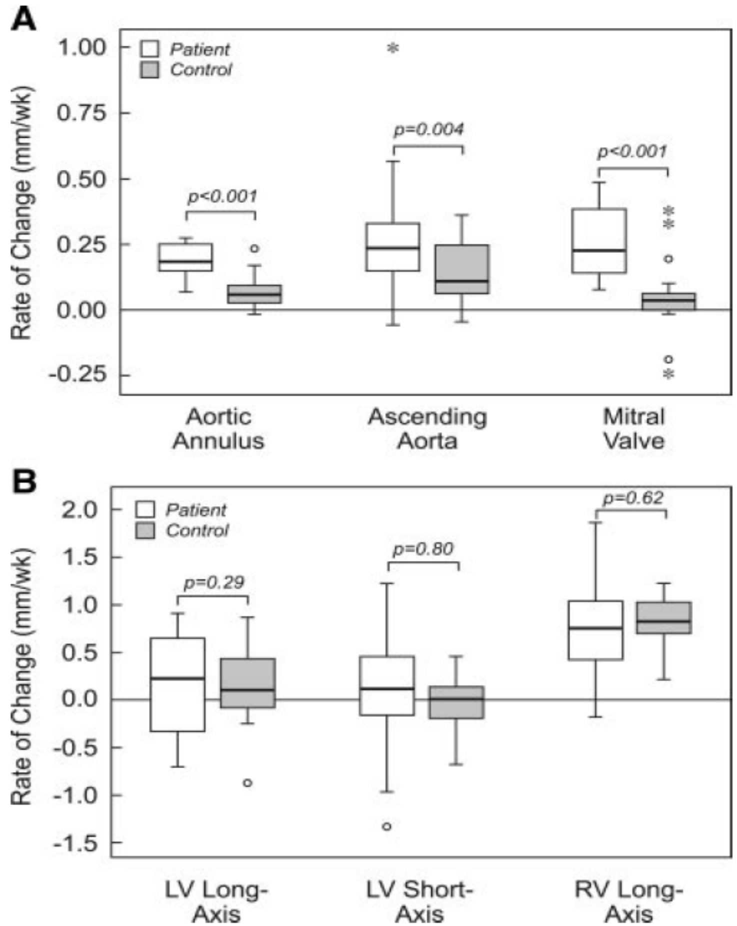

Figure 3. Box plots depicting the rate of change in aortic annulus, ascending aorta, and MV dimensions (A) and LV long- and short-axis and RV long-axis dimensions (B) in fetuses that underwent successful intervention and comparison group fetuses. All measures are expressed in millimeters per week, with positive numbers indicating an increase in size and negative numbers indicating a decrease. For each plot, the central line represents the median rate of change; box, the interquartile range; and whisker bars, 95% confidence intervals. Outliers are shown individually.

McElhinney DB, Marshall AC, Wilkins-Haug LE, Brown DW, Benson CB, Silva V, Marx GR, Mizrahi-Arnaud A, Lock JE, Tworetzky W. Predictors of technical success and postnatal biventricular outcome after in utero aortic valvuloplasty for aortic stenosis with evolving hypoplastic left heart syndrome. Circulation. 2009 Oct 13;120(15):1482-90.

Table 4. Preintervention Variables Associated With Postnatal Biventricular or Single-Ventricle Outcome After In Utero Aortic Valvuloplasty

	Biventricular Outcome (n=17)	All Attempted Interventions, Single-Ventricle Outcome (n=51)	Technically Successful Interventions, Single-Ventricle Outcome (n=33)	*P*
Gestational age, wk	23.9±2.6	24.0±2.6	23.7±2.0	0.95,* 0.68†
Aortic annulus diameter *Z* score	−2.2±0.8	−2.7±0.9	−2.7±0.9	0.03,* 0.02†
Ascending aorta diameter *Z* score	0.6±1.9	−1.4±1.8	−1.2±1.6	<0.001,* 0.002†
LV long-axis dimension *Z* score	2.1±1.5	-0.2±1.8	0.3±1.6	<0.001,* <0.001†
LV short-axis dimension *Z* score	3.6±2.7	2.4±2.3	2.7±2.6	0.09,* 0.28†
LV sphericity	0.62±0.09	0.72±0.13	0.69±0.12	0.004,* 0.04†
MV annulus diameter *Z* score	−0.6±1.3	−1.6±1.3	−1.6±1.1	0.009,* 0.01†
RV long-axis dimension *Z* score	1.7±1.8	1.1±1.2	1.4±1.1	0.13,* 0.46†
Female, n (%)	7 (41)	9 (18)	7 (21)	0.06,* 0.14†
"High" LV pressure, n (%)	13 (76)	15 (29)	10 (30)	0.006,* 0.004†
Moderate or severe MR, n (%)	10 (59)	27 (53)	18 (54)	0.67,* 0.77†
MV inflow time (msec)	124±38	115±40	118±45	0.43,* 0.62†
MV inflow time *Z* score	−2.9±1.6	−3.2±1.7	−3.1±1.9	0.47,* 0.67†
Restrictive PFO/intact atrial septum, n (%)	4 (24)	4 (8)	2 (6)	0.10,* 0.16†
Acute postdilation AR (moderate or greater), n (%)	5 (29)	NA	14 (42)	0.45†

PFO indicates patent foramen ovale. Includes patients with a biventricular outcome from birth or after intermediate univentricular palliation. Biventricular outcome vs single-ventricle outcome *including or †excluding technically unsuccessful interventions. Data are presented as mean±SD unless otherwise indicated.

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McElhinney DB, Marshall AC, Wilkins-Haug LE, Brown DW, Benson CB, Silva V, Marx GR, Mizrahi-Arnaud A, Lock JE, Tworetzky W. Predictors of technical success and postnatal biventricular outcome after in utero aortic valvuloplasty for aortic stenosis with evolving hypoplastic left heart syndrome. Circulation. 2009 Oct 13;120(15):1482-90.

Análisis multivariado

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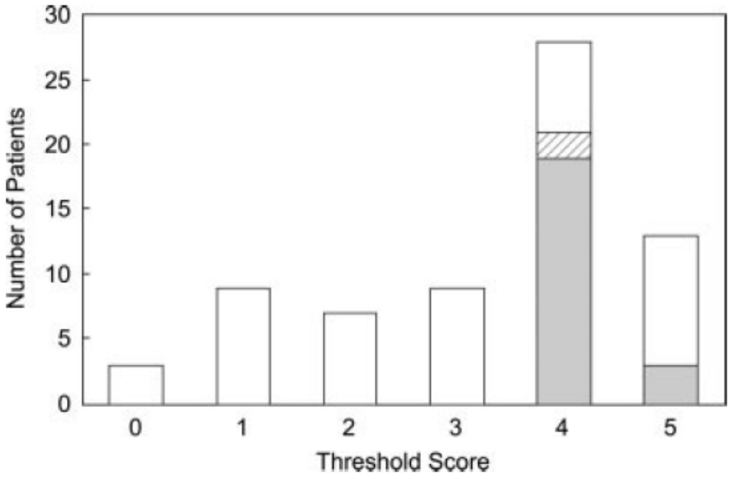

Figure 4. Distribution of threshold scores according to biventricular or univentricular outcome postnatally. White shading indicates fetuses with a univentricular outcome; gray shading, those with a biventricular outcome from birth; and hatched shading, the 2 that initially underwent univentricular staging and were later converted to a biventricular circulation.

Threshold score ≥4 (≥4 of the following)
- LV long-axis *Z* score >0
- LV short-axis *Z* score >0
- Aortic annulus *Z* score >−3.5
- MV annulus *Z* score >−2
- MR or AS maximum systolic gradient ≥20 mm Hg

Sensibilidad 100%
Especificidad 53%
VPP 38%
VPN 100%

McElhinney DB, Marshall AC, Wilkins-Haug LE, Brown DW, Benson CB, Silva V, Marx GR, Mizrahi-Arnaud A, Lock JE, Tworetzky W. Predictors of technical success and postnatal biventricular outcome after in utero aortic valvuloplasty for aortic stenosis with evolving hypoplastic left heart syndrome. Circulation. 2009 Oct 13;120(15):1482-90.

Table 1. Previous and Current Selection Guidelines for Fetal Aortic Valvuloplasty

1. Dominant cardiac anatomic anomaly is valvar AS with all of the following
   - Decreased mobility of valve leaflets
   - Antegrade Doppler color flow jet across aortic valve smaller than the valve annulus diameter
   - No or minimal subvalvar LV outflow obstruction
2. Evolving HLHS
   - LV function qualitatively depressed
     - AND EITHER
   - Retrograde or bidirectional flow in the transverse aortic arch (between the first 2 brachiocephalic vessels) at any time during the cardiac cycle
     - OR two of the following:
   - Monophasic MV inflow (Doppler profile of MV inflow without discrete E and A waves)
   - Left-to-right flow across atrial septum or intact atrial septum (bulging left to right)
   - Bidirectional flow in pulmonary veins
3. Potential for a technically successful procedure and biventricular outcome postnatally
   - Criteria used for most of the patients in the present study (all 3 of the following)
     - LV long-axis *Z* score ≥ -2
     - LV function qualitatively depressed but generating at least a 10 mm Hg pressure gradient across aortic valve or 15 mm Hg MR jet gradient
     - MV diameter *Z* score > -3
   - Modified criteria based on the findings of the present study
     - Unequivocal AS (vs aortic atresia)
     - LV long-axis *Z* score > -2
     - Threshold score ≥ 4 (≥ 4 of the following)
       - LV long-axis *Z* score > 0
       - LV short-axis *Z* score > 0
       - Aortic annulus *Z* score > -3.5
       - MV annulus *Z* score > -2
       - MR or AS maximum systolic gradient ≥ 20 mm Hg

CERPO

McElhinney DB, Marshall AC, Wilkins-Haug LE, Brown DW, Benson CB, Silva V, Marx GR, Mizrahi-Arnaud A, Lock JE, Tworetzky W. Predictors of technical success and postnatal biventricular outcome after in utero aortic valvuloplasty for aortic stenosis with evolving hypoplastic left heart syndrome. Circulation. 2009 Oct 13;120(15):1482-90.

Predictors of Technical Success and Postnatal Biventricular Outcome After In Utero Aortic Valvuloplasty for Aortic Stenosis With Evolving Hypoplastic Left Heart Syndrome

Doff B. McElhinney, MD; Audrey C. Marshall, MD; Louise E. Wilkins-Haug, MD, PhD; David W. Brown, MD; Carol B. Benson, MD; Virginia Silva, MSN; Gerald R. Marx, MD; Arielle Mizrahi-Arnaud, MD; James E. Lock, MD; Wayne Tworetzky, MD

CERPO

- El éxito de la valvuloplastía se logró en aprox. ¾ de los casos.
- Un menor largo del VI se asoció con mayor riesgo de falla de la técnica.
- Existe un riesgo importante de óbito.
- La regurgitación aórtica es común y generalmente mejora o desaparece antes del nacimiento.
- El VI no crece después de una valvuloplastía exitosa pero contribuye al gasto cardiaco.
- Una combinación de características anatómicas y fisiológicas del corazón izquierdo puede identificar aquellos fetos con potencial de lograr una circulación biventricular.

McElhinney DB, Marshall AC, Wilkins-Haug LE, Brown DW, Benson CB, Silva V, Marx GR, Mizrahi-Arnaud A, Lock JE, Tworetzky W. Predictors of technical success and postnatal biventricular outcome after in utero aortic valvuloplasty for aortic stenosis with evolving hypoplastic left heart syndrome. Circulation. 2009 Oct 13;120(15):1482-90.

Congenital Heart Disease

Fetal Aortic Valvuloplasty for Evolving Hypoplastic Left Heart Syndrome

Postnatal Outcomes of the First 100 Patients

Lindsay R. Freud, MD; Doff B. McElhinney, MD; Audrey C. Marshall, MD; Gerald R. Marx, MD; Kevin G. Friedman, MD; Pedro J. del Nido, MD; Sitaram M. Emani, MD; Terra Lafranchi, NP-C; Virginia Silva, RN; Louise E. Wilkins-Haug, MD, PhD; Carol B. Benson, MD; James E. Lock, MD; Wayne Tworetzky, MD

CERPO

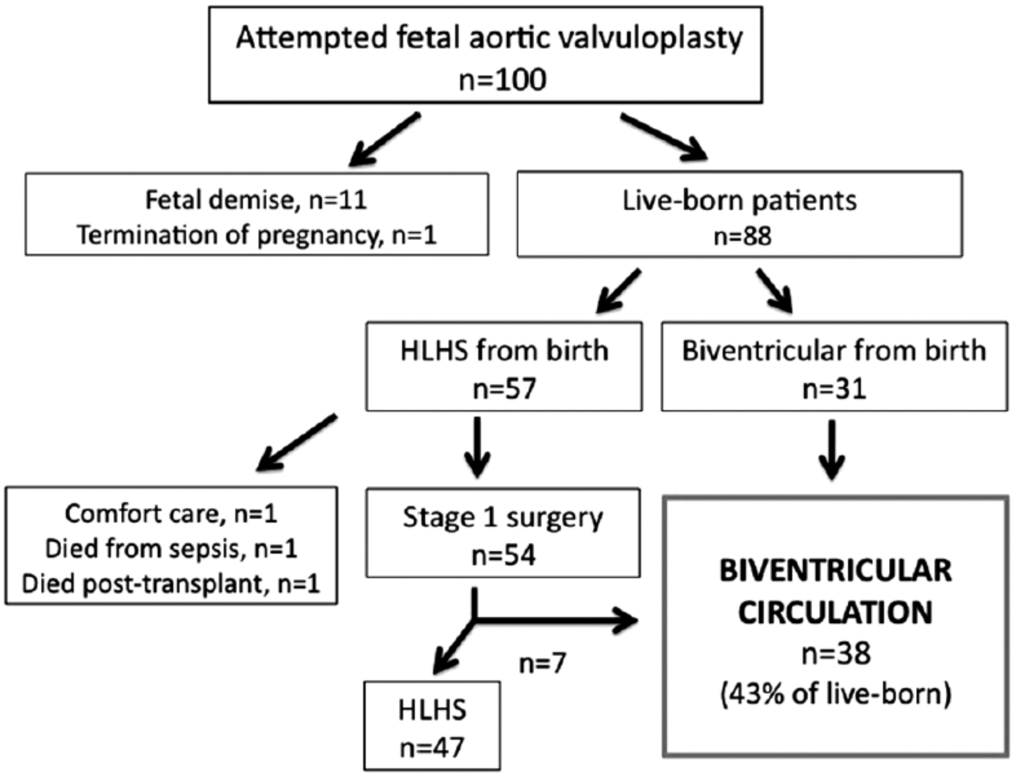

Figure 2. Flow diagram summarizing postnatal management and outcomes for the entire 100-patient cohort. HLHS indicates hypoplastic left heart syndrome.

Técnica exitosa en 77 pacientes

Freud LR, McElhinney DB, Marshall AC, Marx GR, Friedman KG, del Nido PJ, Emani SM, Lafranchi T, Silva V, Wilkins-Haug LE, Benson CB, Lock JE, Tworetzky W. Fetal aortic valvuloplasty for evolving hypoplastic left heart syndrome: postnatal outcomes of the first 100 patients. Circulation. 2014 Aug 19;130(8):638-45.

Sobrevida post valvuloplastía

CERPO

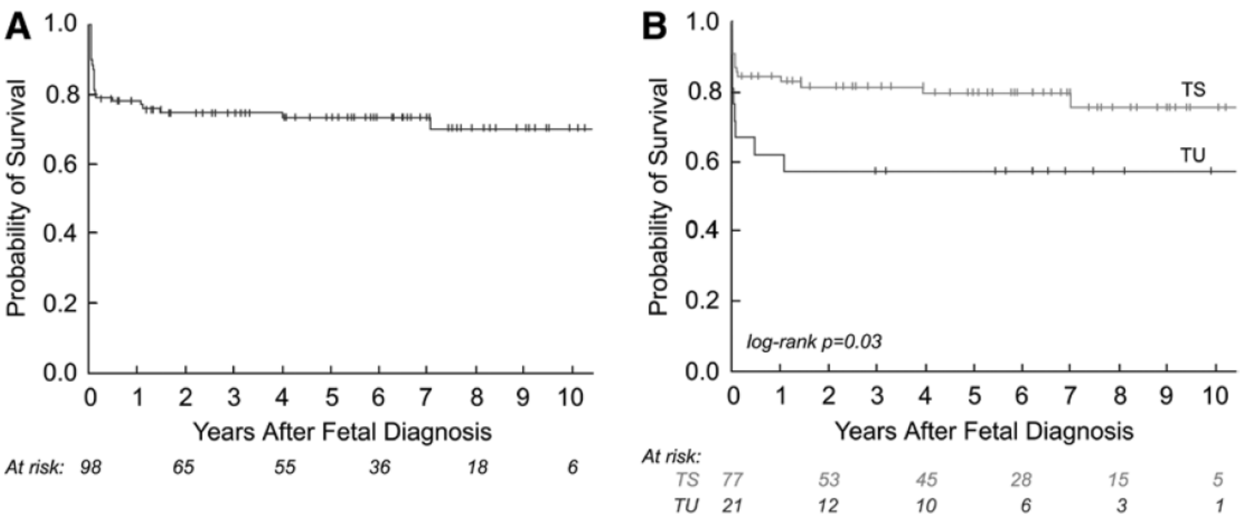

Figure 1. Kaplan–Meier curves depicting intention-to-treat analyses from the date of fetal intervention. **A**, Survival of all fetuses. **B**, Survival of all fetuses stratified by technical success – technically successful (TS) vs technically unsuccessful (TU).

Freud LR, McElhinney DB, Marshall AC, Marx GR, Friedman KG, del Nido PJ, Emani SM, Lafranchi T, Silva V, Wilkins-Haug LE, Benson CB, Lock JE, Tworetzky W. Fetal aortic valvuloplasty for evolving hypoplastic left heart syndrome: postnatal outcomes of the first 100 patients. Circulation. 2014 Aug 19;130(8):638-45.

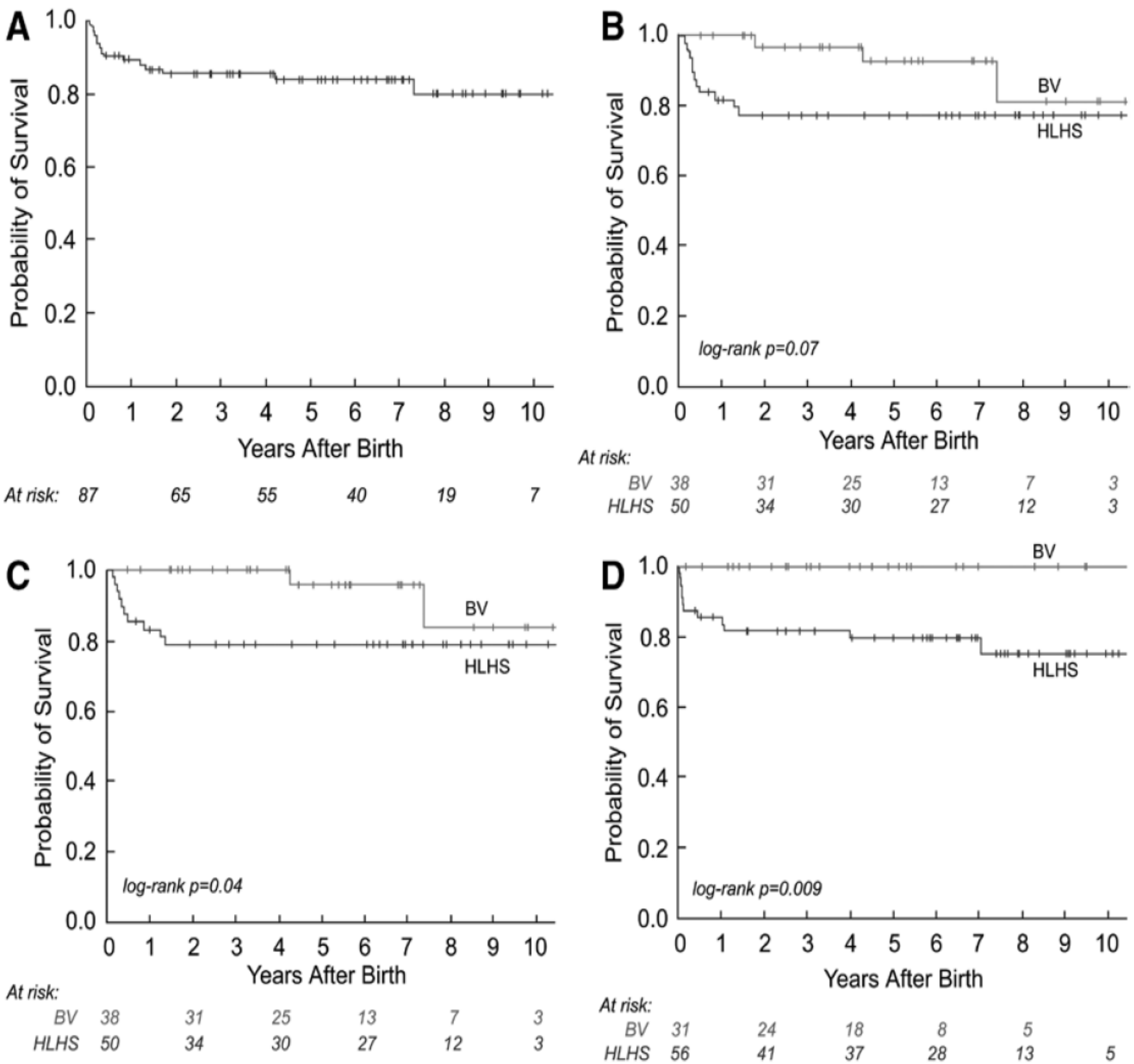

Figure 4. Kaplan–Meier curves depicting survival from the time of birth. **A**, All-cause mortality in the live-born cohort. **B**, All-cause mortality between hypoplastic left heart syndrome (HLHS) and biventricular (BV) outcome groups. **C**, Cardiac mortality between HLHS and BV outcome groups (excluding the 2 noncardiac deaths: 1 in the HLHS and 1 in the BV outcome group). **D**, Cardiac mortality between HLHS and BV outcome groups based on the initial postnatal management strategy.

Freud LR, McElhinney DB, Marshall AC, Marx GR, Friedman KG, del Nido PJ, Emani SM, Lafranchi T, Silva V, Wilkins-Haug LE, Benson CB, Lock JE, Tworetzky W. Fetal aortic valvuloplasty for evolving hypoplastic left heart syndrome: postnatal outcomes of the first 100 patients. Circulation. 2014 Aug 19;130(8):638-45.

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ORIGINAL ARTICLE

WILEY PRENATAL DIAGNOSIS

CERPO

Early hemodynamic changes after fetal aortic stenosis valvuloplasty predict biventricular circulation at birth

Aaron R. Prosnitz[1] | Monika Drogosz[1] | Audrey C. Marshall[1] | Louise E. Wilkins-Haug[2] | Carol B. Benson[3] | Lynn A. Sleeper[1] | Wayne Tworetzky[1] | Kevin G. Friedman[1]

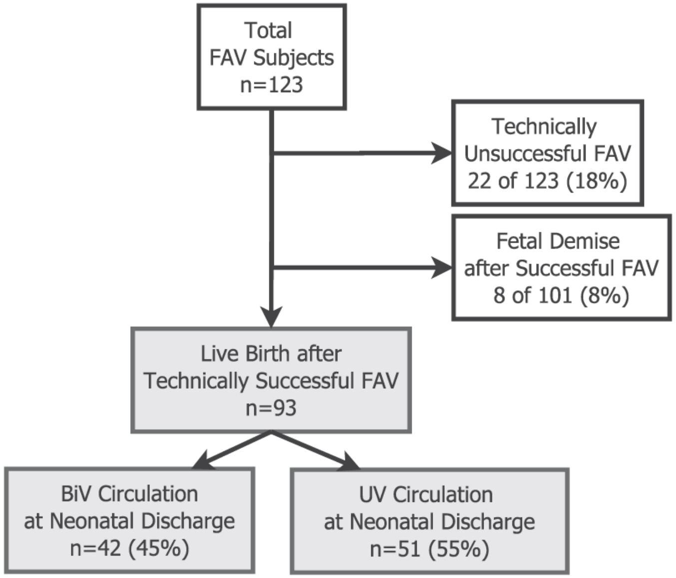


FIGURE 1 Derivation of analysis cohort. Among all subjects undergoing FAV for fetal aortic stenosis with high-risk features of evolving HLHS, those with technically unsuccessful procedures and/or fetal demise after FAV were excluded. The remaining 93 subjects were live-born after a technically successful FAV. Within this analysis cohort, 42 subjects (45%) achieved BiV circulation at neonatal discharge. Abbreviations: BiV (biventricular), FAV (fetal aortic valvuloplasty), HLHS (hypoplastic left heart syndrome), UV (univentricular)

Prosnitz AR, Drogosz M, Marshall AC, Wilkins-Haug LE, Benson CB, Sleeper LA, Tworetzky W, Friedman KG. Early hemodynamic changes after fetal aortic stenosis valvuloplasty predict biventricular circulation at birth. Prenat Diagn. 2018 Mar;38(4):286-292.

TABLE 2 Paired data sets of early fetal hemodynamic changes after FAV

Variable	Pre-FAV	Post-FAV	Change	*P*-Value
Any antegrade systolic transverse aortic arch flow[a] (*n* = 91)	0 (0%)	59 (65%)	65%	<0.001
Bidirectional FO flow (*n* = 81)	9 (11%)	22 (27%)	16%	0.002
Change in FO flow (*n* = 81):				
Unchanged left-to-right flow	----	----	59 (73%)	
New bidirectional flow	----	----	15 (19%)	
Unchanged bidirectional flow	----	----	7 (9%)	
Mitral inflow pattern (*n* = 82):				
Biphasic	15 (18%)	32 (39%)	21%	0.003
Partially fused	11 (13%)	15 (18%)		
Monophasic	56 (68%)	35 (43%)		
Change in mitral inflow pattern (*n* = 82):				
Worsened to monophasic or partially fused, or unchanged monophasic	----	----	37 (45%)	
Unchanged partially fused or biphasic	----	----	12 (15%)	
Improved to partially fused or biphasic	----	----	33 (40%)	
MV inflow time (msec) (*n* = 75)	105 [81, 150]	155 [133, 190]	32 [8, 78]	<0.001
MV inflow time z-score (*n* = 75)	−3.4 [−4.4, −1.8]	−1.5 [−2.5, −0.2]	1.2 [0.3, 3.0]	<0.001
MV inflow index (msec/bpm) (*n* = 75)	0.75 [0.58, 1.06]	1.06 [0.89, 1.36]	0.26 [0.03, 0.53]	<0.001
Mitral regurgitation (> mild) (*n* = 93)	49 (53%)	37 (40%)	−13%	0.017
LV end-diastolic volume (mL) (*n* = 93)	2.23 [1.53, 3.73]	2.22 [1.35, 3.21]	−0.06 [−0.19, 0.06]	0.013
LV end-diastolic volume z-score (*n* = 89)	1.78 [0.72, 3.04]	1.26 [0.25, 2.94]	−0.26 [−1.3, 0.97]	<0.001
LV sphericity (*n* = 88)	0.68 [0.59, 0.75]	0.66 [0.57, 0.75]	−0.02 [−0.09, 0.05]	0.16
LV ejection fraction (%) (*n* = 87)	23 [18, 32]	28 [22, 41]	6.9 [−0.4, 14.7]	<0.001
Change in LV ejection fraction (≥ 5%) (*n* = 87)			54 (62%)	
LV systolic pressure (mm Hg) (*n* = 84)	45.9 [35, 60.3]	40.1 [32.1, 55]	−3.6 [−10.9, 3.2]	0.001
Aortic jet width (mm) (*n* = 83)	1.3 [1, 1.5]	2.8 [2.4, 3.1]	1.5 [1.1, 1.9]	<0.001
Aortic jet width index (jet width/annulus) (*n* = 82)	0.44 [0.35, 0.52]	0.88 [0.75, 0.97]	0.41 [0.31, 0.54]	<0.001
Aortic regurgitation (< mild) (*n* = 92)	0 (0%)	31 (34%)	34%	<0.001

All values listed as "median, [25th percentile, 75th percentile]" or "number of subjects (% of the total)." All data displayed in pre-FAV and post-FAV columns are limited to data pairs that allow for complete-case analysis. The Change column displays either the pre-FAV percentage subtracted from the post-FAV percentage for categorical variables, or the median of the paired differences for continuous variables.

Abbreviations: FAV, fetal aortic valvuloplasty; LV, left ventricle; MV, mitral valve; FO, patent foramen ovale.

[a]Among the 59 subjects with antegrade systolic arch flow, 36 (61%) had partially antegrade flow and 23 (39%) had exclusively antegrade flow.

CERPO

Prosnitz AR, Drogosz M, Marshall AC, Wilkins-Haug LE, Benson CB, Sleeper LA, Tworetzky W, Friedman KG. Early hemodynamic changes after fetal aortic stenosis valvuloplasty predict biventricular circulation at birth. Prenat Diagn. 2018 Mar;38(4):286-292.

TABLE 3 Univariate models for BiV circulation as a function of early post-FAV hemodynamic changes

Variable	Observations	OR	95% CI	*P*-Value
Post-FAV systolic transverse aortic arch flow:				<0.001
Exclusively retrograde	32/91	Ref	----	----
Partially antegrade	36/91	9.80	2.84–34.85	<0.001
Exclusively antegrade	23/91	19.83	4.88–80.54	<0.001
New bidirectional FO flow	15/81	5.14	1.47–17.97	0.010
Change in mitral inflow pattern:				<0.001
Unchanged monophasic or worsened	37/82	Ref	----	----
Unchanged fused or biphasic	12/82	4.17	1.04–16.62	0.043
Improved to fused or biphasic	33/82	3.21	1.20–8.54	0.020
Change in MV inflow time (msec)	75	1.00	0.99–1.00	0.32
Change in MV inflow time z-score	75	0.89	0.73–1.09	0.27
Change in MV inflow index (msec/bpm)	74	0.48	0.17–1.37	0.17
Change in mitral regurgitation (> mild)	93	1.33	0.55–3.20	0.53
Change in LV end-diastolic volume (mL)	93	0.95	0.64–1.42	0.82
Change in LV end-diastolic volume z-score	93	1.01	0.77–1.32	0.95
Change in LV sphericity	88	3.49	0.20–60.66	0.39
Change in LV ejection fraction (%)	87	1.05	1.01–1.10	0.016
Improved LV ejection fraction (≥ 5%)	54/87	3.10	1.24–7.76	0.016
Change in LV systolic pressure (mm Hg)	84	0.97	0.93–1.01	0.11
Change in aortic jet width (per 1-mm increase)	83	1.17	0.60–2.28	0.64
Change in aortic jet width index (jet width/annulus)	82	0.87	0.11–6.94	0.90
Increasing AR from ≤ mild to ≥ moderate	31/91	0.77	0.32–1.85	0.56

"Ref" signifies the reference group for the estimated odds ratios. "Change" is defined as post-FAV minus pre-FAV.

Abbreviations: AR, aortic regurgitation; FAV, fetal aortic valvuloplasty; LV, left ventricle; OR, odds ratio; FO, patent foramen ovale.

Prosnitz AR, Drogosz M, Marshall AC, Wilkins-Haug LE, Benson CB, Sleeper LA, Tworetzky W, Friedman KG. Early hemodynamic changes after fetal aortic stenosis valvuloplasty predict biventricular circulation at birth. Prenat Diagn. 2018 Mar;38(4):286-292.

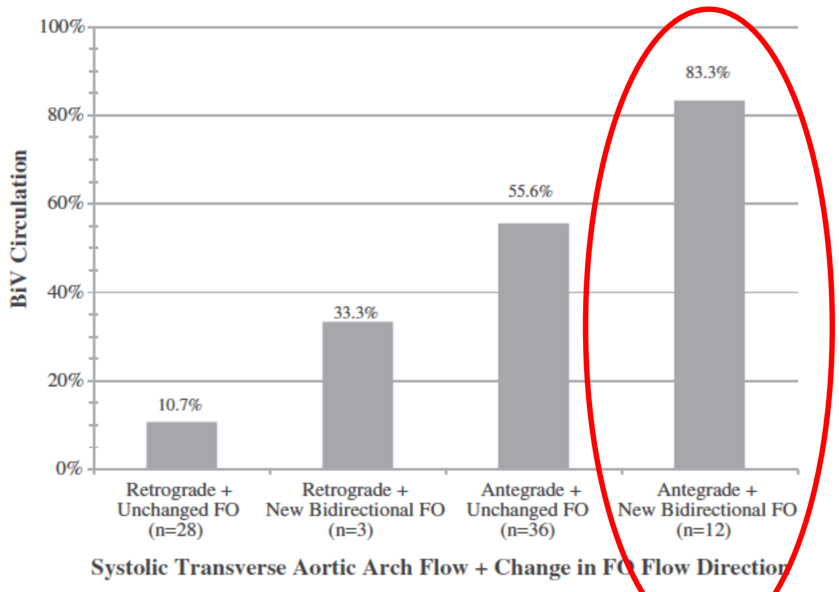


FIGURE 2 Post-FAV hemodynamic changes predict circulation status at neonatal discharge. After FAV for fetal aortic valve stenosis, there are several acute hemodynamic changes that signify increased forward flow through the left ventricle. Key changes are increasing degree of antegrade systolic flow across the transverse aortic arch and improving direction of flow across the FO, both of which play a role in predicting the chance of BiV circulation after birth. This bar chart depicts the percentage of subjects with BiV circulation at neonatal discharge (n = 34/79) by systolic transverse aortic arch flow direction after FAV (ie, exclusively retrograde versus any amount of antegrade systolic flow) and by the presence of new post-FAV bidirectional FO flow versus unchanged flow direction (either left-to-right or bidirectional). Abbreviations: BiV (biventricular), FAV (fetal aortic valvuloplasty), FO (patent foramen ovale)

CERPO

TABLE 4 Multivariate model for BiV circulation as a function of early post-FAV hemodynamic changes (n = 79; 34 events (BiV circulation); c-statistic = 0.791)

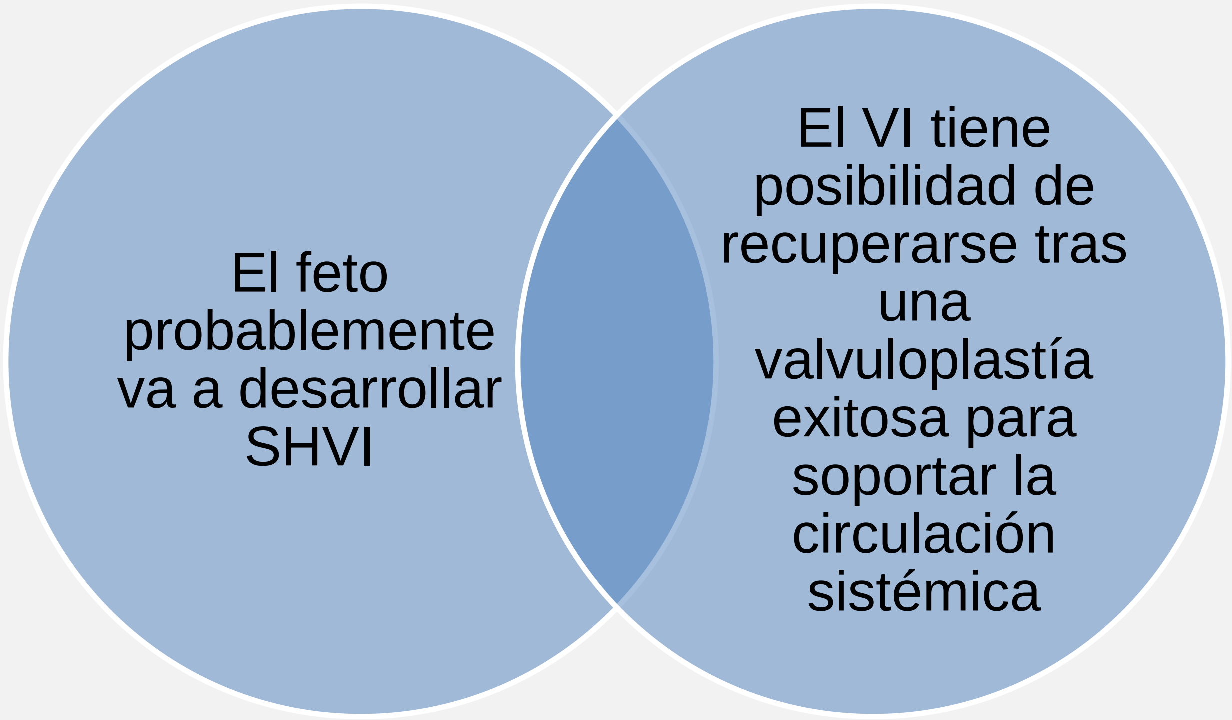
Variable	OR	95% CI	*P*-Value
Systolic transverse aortic arch flow:			<0.001
Exclusively retrograde	Ref	----	----
Partially antegrade	9.01	2.49–32.57	0.001
Exclusively antegrade	14.63	2.98–72.15	0.001
New bidirectional FO flow	3.75	0.89–15.80	0.07

"Ref" signifies the reference group for the estimated odds ratios.

Abbreviations: FAV, fetal aortic valvuloplasty; OR, odds ratio; FO, patent foramen ovale.

Prosnitz AR, Drogosz M, Marshall AC, Wilkins-Haug LE, Benson CB, Sleeper LA, Tworetzky W, Friedman KG. Early hemodynamic changes after fetal aortic stenosis valvuloplasty predict biventricular circulation at birth. Prenat Diagn. 2018 Mar;38(4):286-292.

Criterios de selección



Prosnitz AR, Drogosz M, Marshall AC, Wilkins-Haug LE, Benson CB, Sleeper LA, Tworetzky W, Friedman KG. Early hemodynamic changes after fetal aortic stenosis valvuloplasty predict biventricular circulation at birth. Prenat Diagn. 2018 Mar;38(4):286-292.

Ultrasound Obstet Gynecol 2018; **52**: 212–220
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Improved technical success, postnatal outcome and refined predictors of outcome for fetal aortic valvuloplasty

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KEYWORDS: fetal cardiac intervention; fetal cardiology; hypoplastic left heart syndrome

CERPO

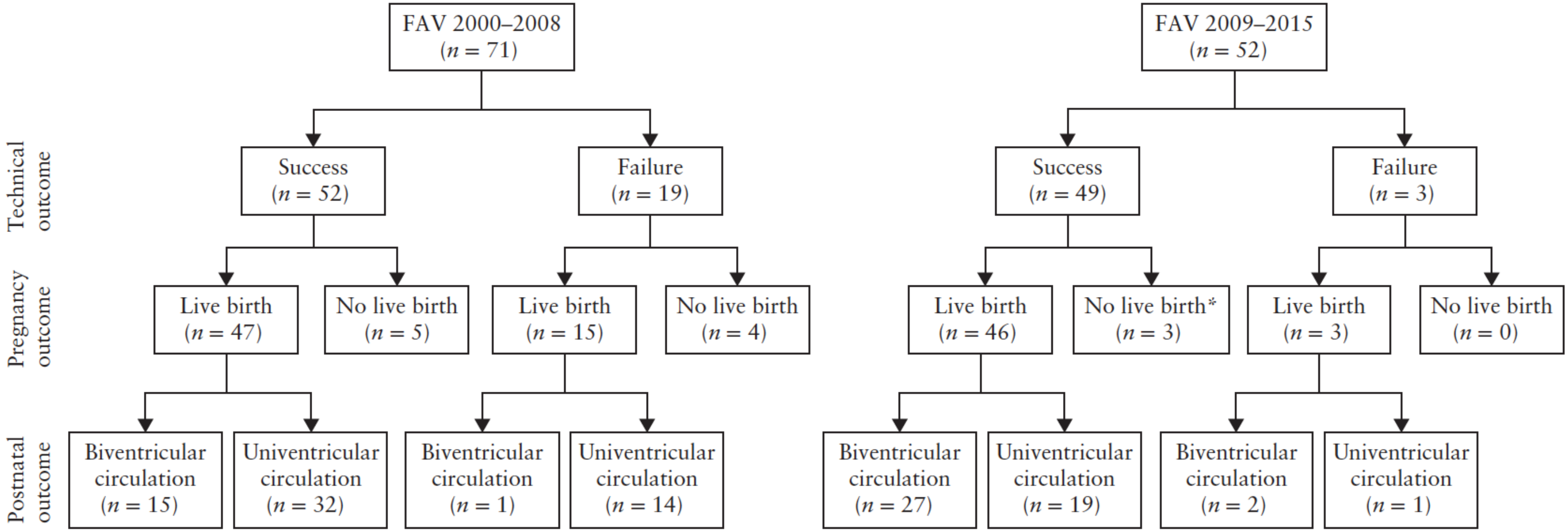


Figure 1 Procedural, pregnancy and postnatal outcomes of fetuses undergoing fetal aortic valvuloplasty (FAV), according to whether procedure was performed in early (2000–2008) or later (2009–2015) period. *Includes one patient who was born prematurely and had no postnatal cardiac intervention.

Friedman KG, Sleeper LA, Freud LR, Marshall AC, Godfrey ME, Drogosz M, Lafranchi T, Benson CB, Wilkins-Haug LE, Tworetzky W. Improved technical success, postnatal outcome and refined predictors of outcome for fetal aortic valvuloplasty. Ultrasound Obstet Gynecol. 2018 Aug;52(2):212-220.

Modelos predictivos

CERPO

Table 2 Factors associated with biventricular outcome ($n = 42$) in liveborn neonates following technically successful fetal aortic valvuloplasty ($n = 90$*), based on multivariate analysis

Factor	*Odds ratio (95% CI)*	P
Ascending aorta dimension *Z*-score	1.95 (1.27–2.99)	0.002
LV pressure (per 5-mmHg increase)	1.37 (1.12–1.67)	0.002
LV long-axis dimension *Z*-score	1.56 (1.02–2.40)	0.041
Mitral valve inflow time *Z*-score	1.79 (1.19–2.69)	0.005

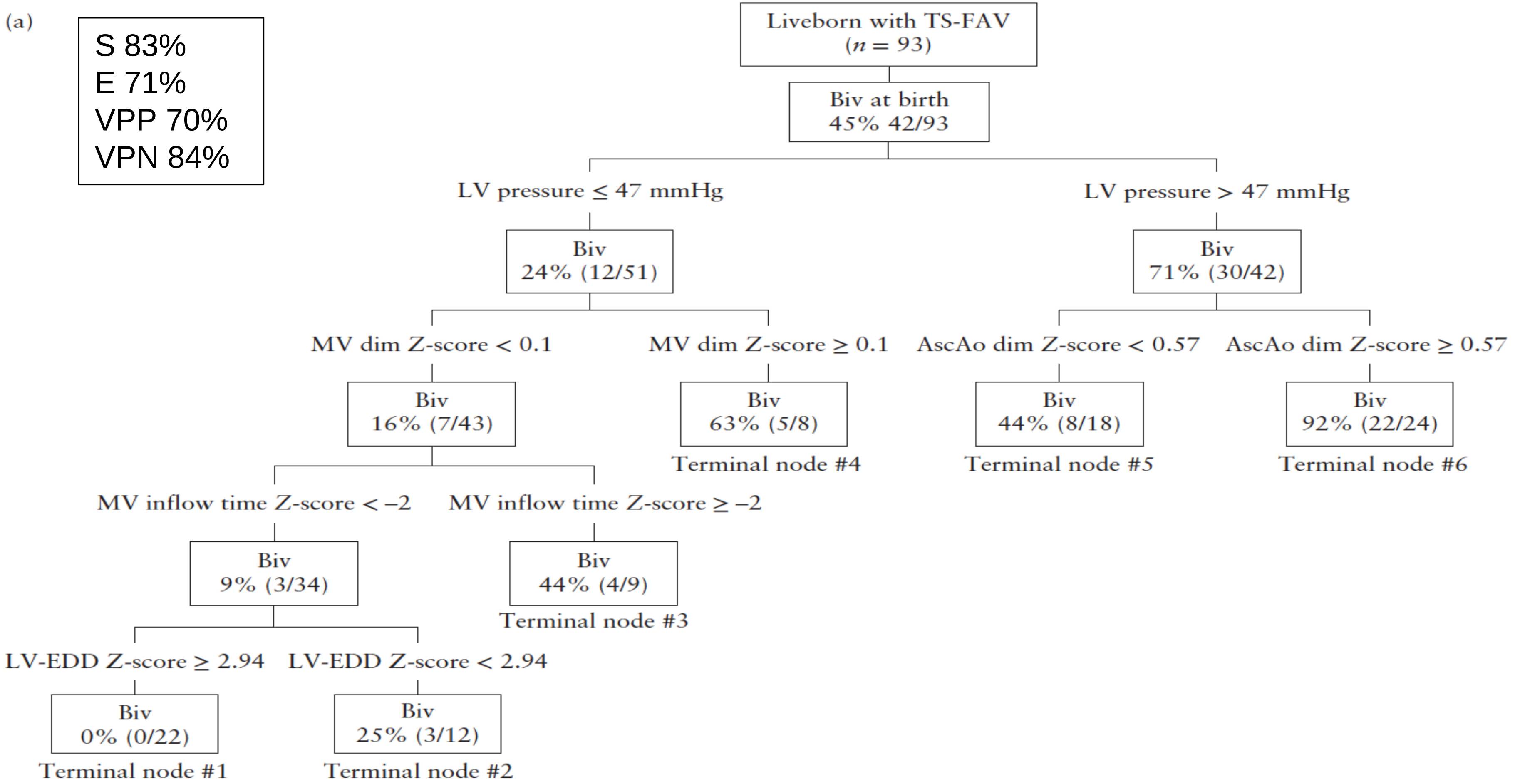
C-statistic = 0.91. *Data not available for three cases. LV, left ventricle.

Friedman KG, Sleeper LA, Freud LR, Marshall AC, Godfrey ME, Drogosz M, Lafranchi T, Benson CB, Wilkins-Haug LE, Tworetzky W. Improved technical success, postnatal outcome and refined predictors of outcome for fetal aortic valvuloplasty. Ultrasound Obstet Gynecol. 2018 Aug;52(2):212-220.

Análisis CART

(a)

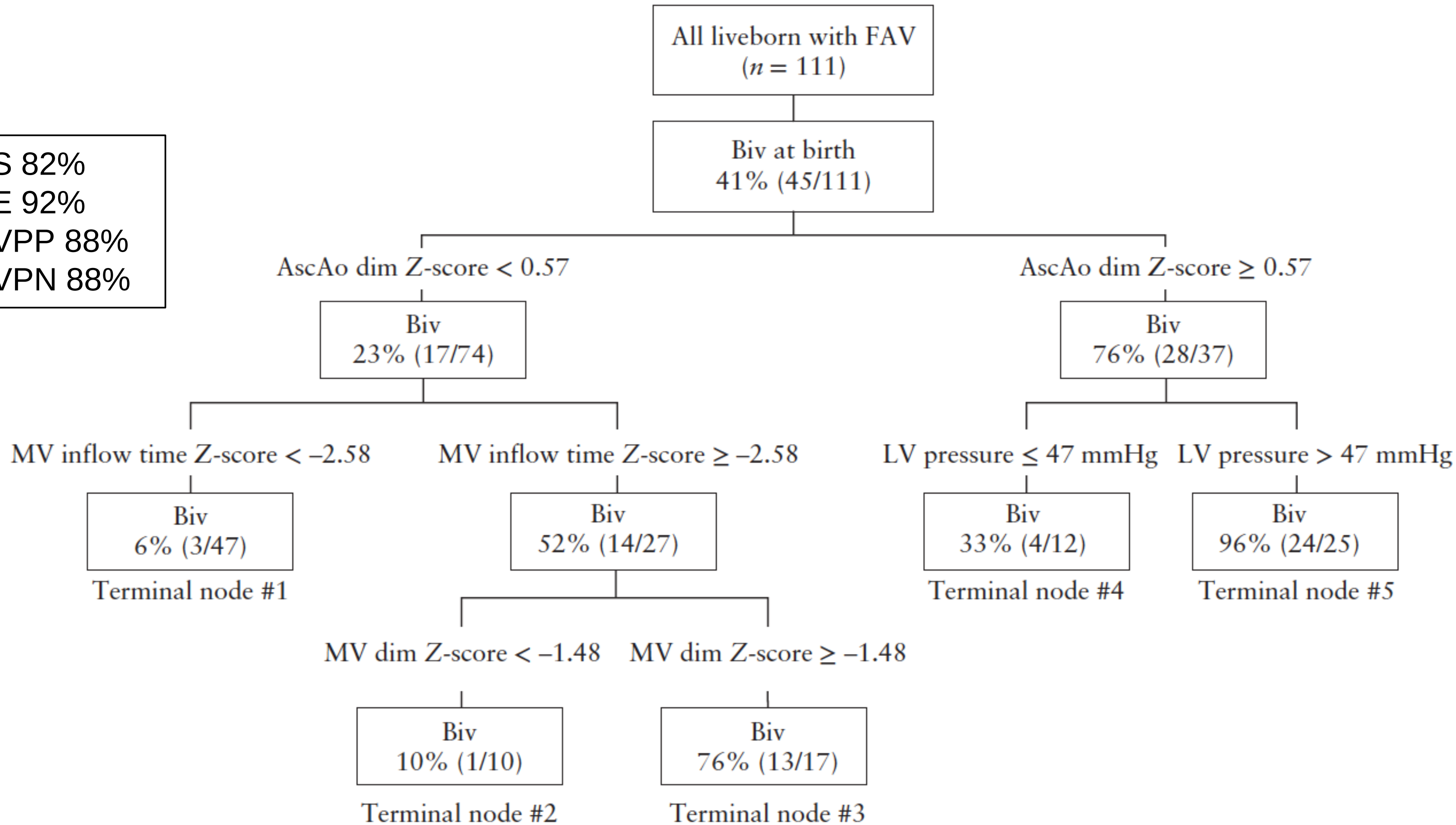
S 83%
E 71%
VPP 70%
VPN 84%



Friedman KG, Sleeper LA, Freud LR, Marshall AC, Godfrey ME, Drogosz M, Lafranchi T, Benson CB, Wilkins-Haug LE, Tworetzky W. Improved technical success, postnatal outcome and refined predictors of outcome for fetal aortic valvuloplasty. Ultrasound Obstet Gynecol. 2018 Aug;52(2):212-220.

S 82%
E 92%
VPP 88%
VPN 88%

(b)



Friedman KG, Sleeper LA, Freud LR, Marshall AC, Godfrey ME, Drogosz M, Lafranchi T, Benson CB, Wilkins-Haug LE, Tworetzky W. Improved technical success, postnatal outcome and refined predictors of outcome for fetal aortic valvuloplasty. Ultrasound Obstet Gynecol. 2018 Aug;52(2):212-220.

Atresia Pulmonar con Septum IV intacto / SHVD

- La atresia pulmonar con septum interventricular intacto puede evolucionar a SHVD.

- **<u>Fisiopatología</u>**
- *Atresia de infundíbulo del tracto de salida de VD*
- *Atresia válvula pulmonar*
- *Hipoplasia VD*
- *Fistulas ventrículo-coronarias*
- *Regurgitación tricuspídea*
- *Flujo retrogrado en ramas pulmonares por ductus arterioso*

Atresia Pulmonar con Septum IV intacto / SHVD

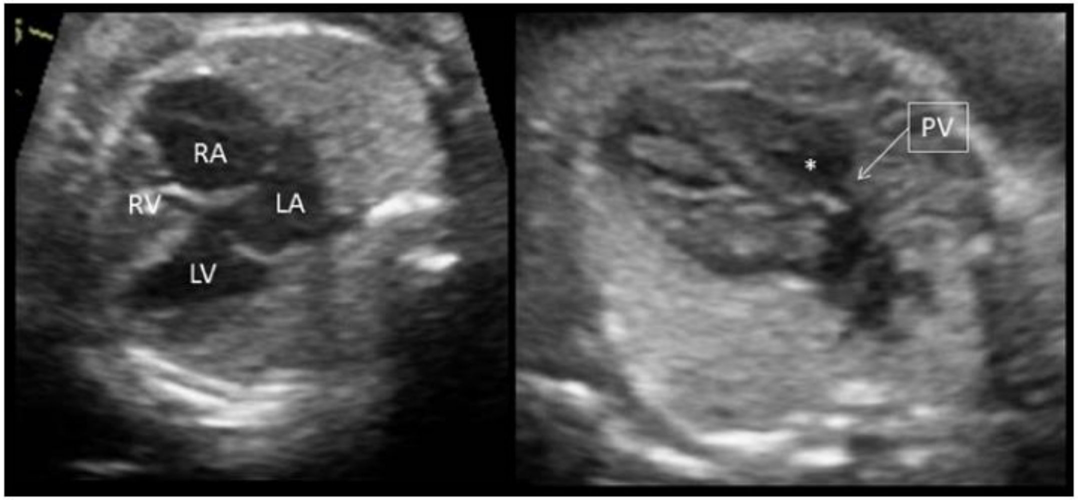


FIGURE 7 Appearance of a fetal heart with pulmonary atresia with intact ventricular septum at 25-wk gestation that may be a candidate for fetal cardiac intervention. There is mild tricuspid valve and right ventricular hypoplasia. Membranous pulmonary atresia and a patent right ventricular outflow tract are present. LA = left atrium; LV = left ventricle; PV = pulmonary valve; RA = right atrium; RV = right ventricle, *right ventricular outflow tract

Schidlow DN, Freud L, Friedman K, Tworetzky W. Fetal interventions for structural heart disease. Echocardiography. 2017 Dec;34(12):1834-1841.

Atresia Pulmonar con Septum IV intacto / SHVD

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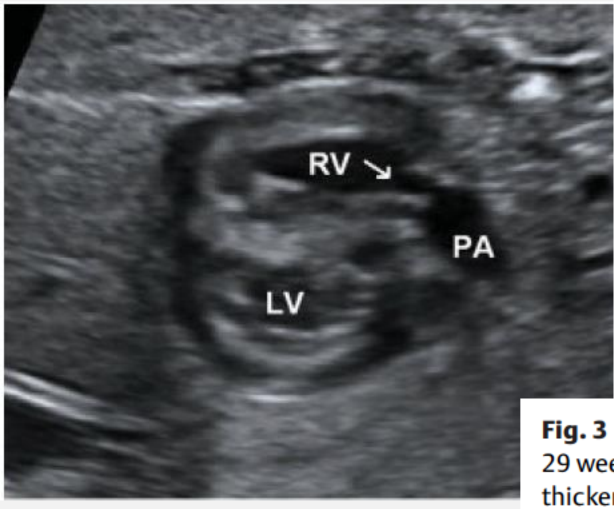

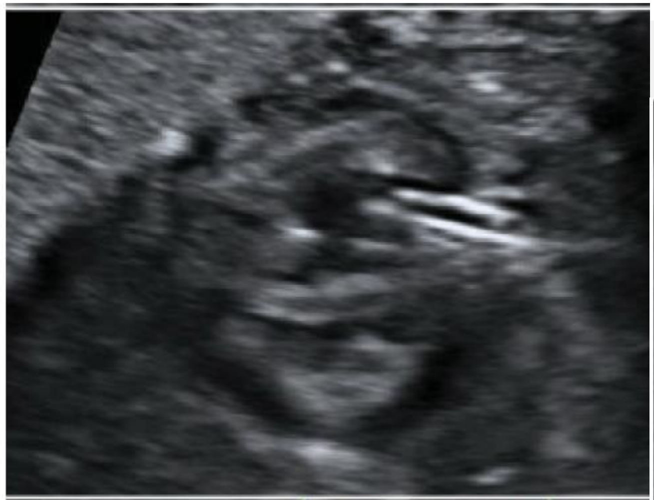

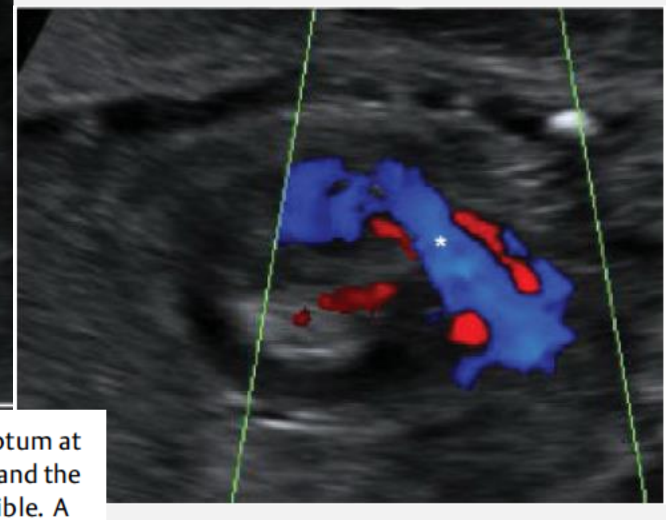

Fig. 3 Top panel: Pulmonary atresia with intact ventricular septum at 29 weeks gestational age. The right ventricle is hypertrophied, and the thickened, doming, and atretic pulmonary valve (arrow) is visible. A normally sized right pulmonary artery is visualized. Middle panel: After perforation of the pulmonary valve with a cannula, an angioplasty balloon is inflated in the right ventricular outflow tract and across the pulmonary valve. Bottom panel: Still-frame color Doppler image after pulmonary valve perforation and balloon angioplasty in the same patient. There is a broad jet of anterograde flow (asterisk) into the pulmonary artery. LV, left ventricle; RV, right ventricle; PA, main pulmonary artery.

Schidlow DN, Freud L, Friedman K, Tworetzky W. Fetal interventions for structural heart disease. Echocardiography. 2017 Dec;34(12):1834-1841.

Atresia Pulmonar con Septum IV intacto / SHVD

Selección de pacientes

CERPO

TABLE 3 Features prompting consideration of fetal intervention for pulmonary atresia with intact ventricular septum (PA/IVS)

Membranous pulmonary atresia
Intact or highly restrictive ventricular septum
Tricuspid valve annulus Z-score <2.5
Identifiable but qualitatively small right ventricle

Schidlow DN, Freud L, Friedman K, Tworetzky W. Fetal interventions for structural heart disease. Echocardiography. 2017 Dec;34(12):1834-1841.

Atresia Pulmonar con Septum IV intacto / SHVD

CERPO

Fetal pulmonary valvuloplasty for critical pulmonary stenosis or atresia with intact septum

Gerald Tulzer, Wolfgang Arzt, Rodney C G Franklin, Pamela V Loughna, Rudi Mair, Helena M Gardiner

Progressive stenosis of the semilunar valves in utero can be life threatening. We treated two fetuses with complete or almost complete pulmonary atresia and imminent hydrops (increased cardiothoracic ratio, pericardial effusion, holosystolic tricuspid regurgitation extending into diastole, and abnormal venous Dopplers). We dilated the pulmonary valve of two fetuses in utero at 28 and 30 weeks' gestation, through the mothers' abdomens. After the procedure, the fetuses had decreased signs of circulatory failure and gestation continued until near term. In the neonatal period, we did a repeat valvuloplasty with systemic-to-pulmonary arterial shunt. Both children (now aged 18 months and 12 months) now have biventricular circulation. Surgery on selected fetuses with semilunar valve stenosis or atresia, or both, can extend pregnancy and favourably change the postnatal surgical options.

Lancet 2002; **360:** 1567–68

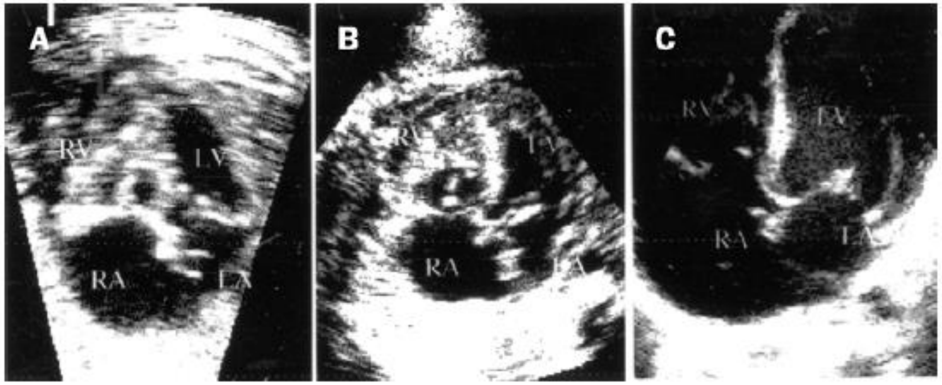

Figure 2: **Growth of the small right ventricle after successful balloon dilation of the pulmonary valve**
Images are serial four-chamber views of fetus A before procedure (A), 6 weeks after procedure (B), and after birth (C). RV=right ventricle. LV=left ventricle. RA=right atrium. LA=left atrium.

Tulzer G, Arzt W, Franklin RC, Loughna PV, Mair R, Gardiner HM. Fetal pulmonary valvuloplasty for critical pulmonary stenosis or atresia with intact septum. Lancet. 2002 Nov 16;360(9345):1567-8.

Research Article

Fetal Intervention in Right Outflow Tract Obstructive Disease: Selection of Candidates and Results

E. Gómez Montes,[1] I. Herraiz,[1] A. Mendoza,[2] and A. Galindo[1]

[1] *Fetal Medicine Unit-Red de Investigación en Salud Materno-Infantil y del Desarrollo (SAMID), Department of Obstetrics and Gynaecology, Hospital Universitario "12 de Octubre", Universidad Complutense, 28041 Madrid, Spain*
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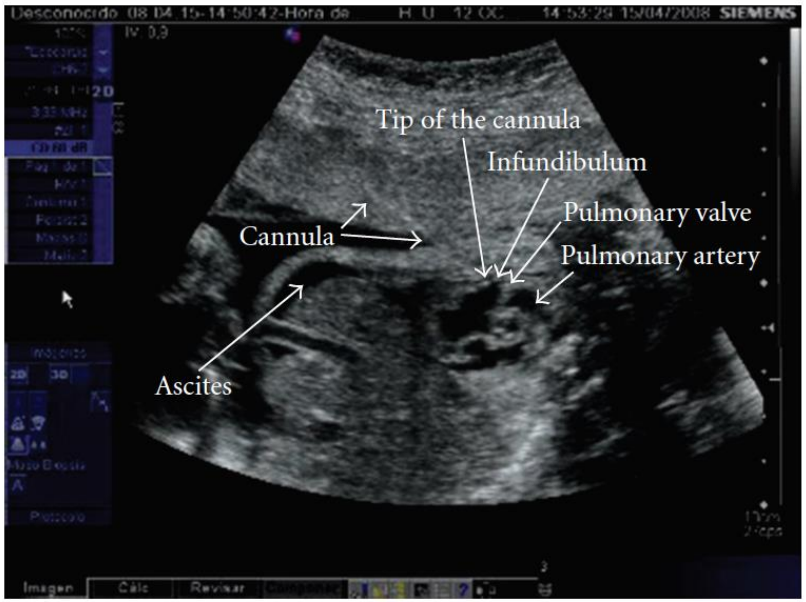

Figure 1: Cannula course during fetal cardiac intervention for pulmonary valvuloplasty, entering the right ventricle at the infundibulum, in the longitudinal view, with the cannula course parallel to the RV outflow tract.

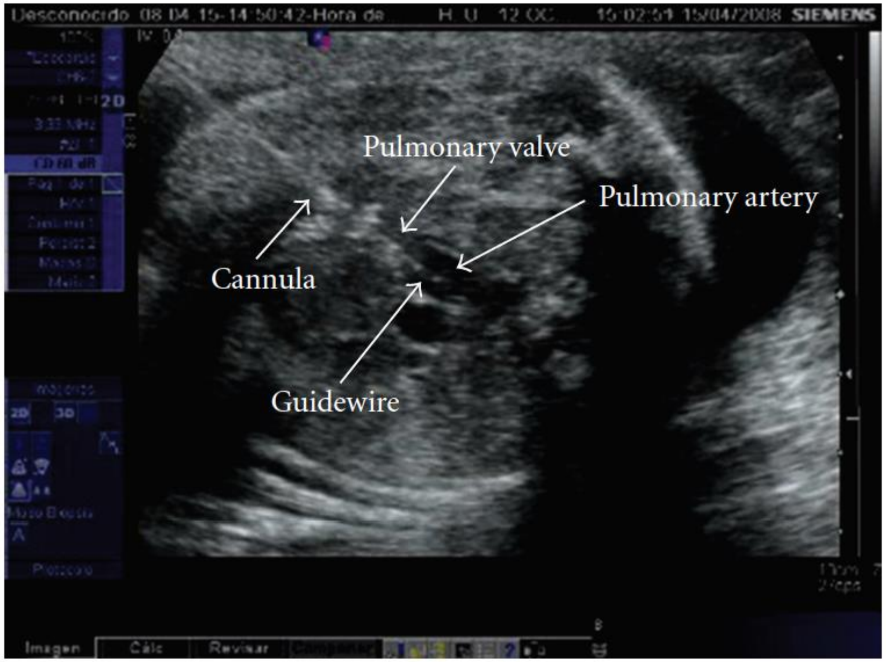

Figure 2: The guidewire has crossed the pulmonary valve, and it is inside the main pulmonary artery.

Gómez Montes E, Herraiz I, Mendoza A, Galindo A. Fetal intervention in right outflow tract obstructive disease: selection of candidates and results. Cardiol Res Pract. 2012;2012:592403.

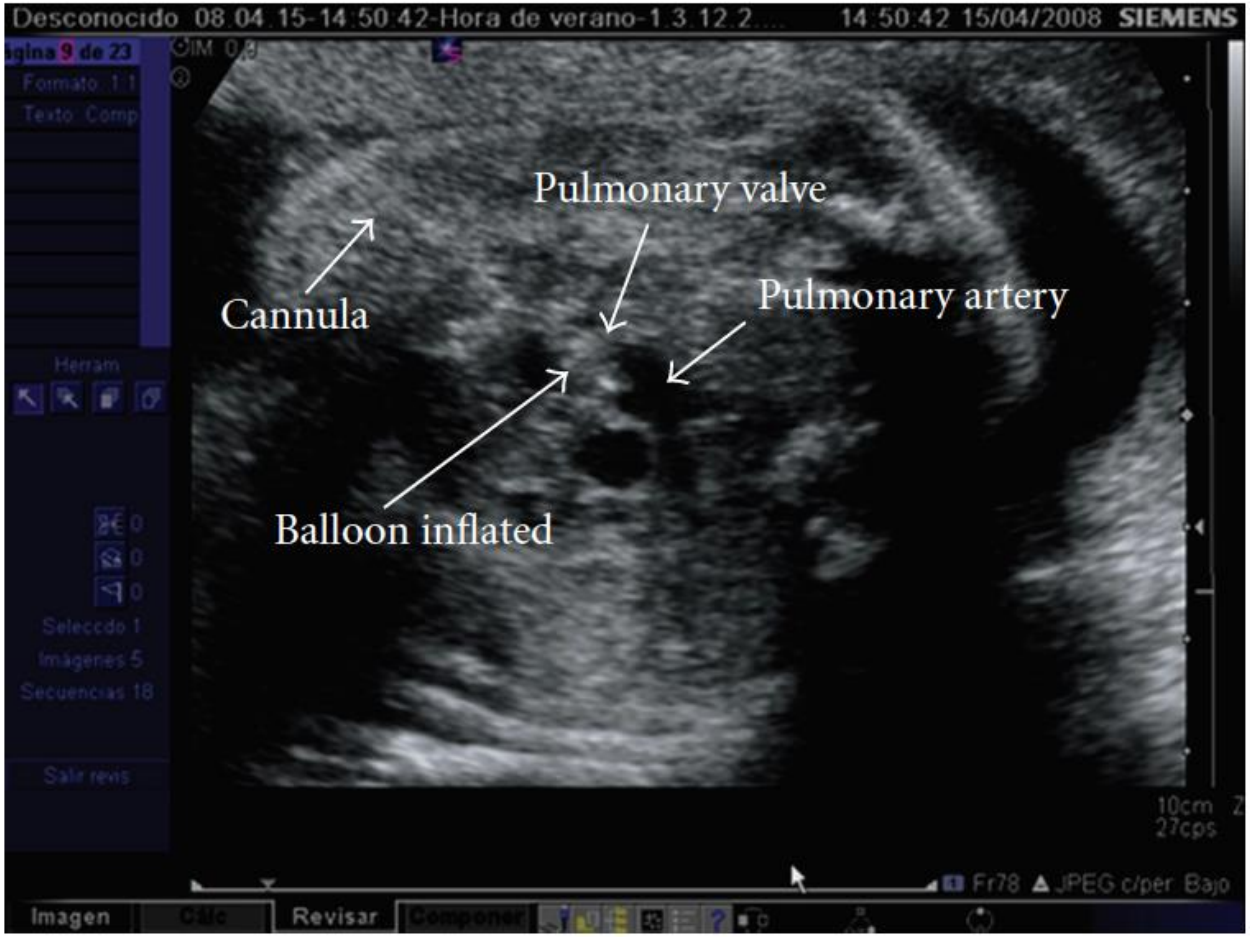

FIGURE 3: The coronary angioplasty balloon is inflated across the fetal pulmonary valve.

Gómez Montes E, Herraiz I, Mendoza A, Galindo A. Fetal intervention in right outflow tract obstructive disease: selection of candidates and results. Cardiol Res Pract. 2012;2012:592403.

Criterios de selección

Table 1: Criteria used for prenatal echocardiographic prediction of postnatal outcome for pulmonary atresia/critical pulmonary stenosis with intact ventricular septum (PA/CS-IVS).

Reference	Prenatal predictors of non-BV outcome	Test accuracy
Salvin et al. (2006) [22]	(i) TV *z*-score ≤ -3	Sn 92%, Sp 100%
Roman et al. (2007) [19]	(i) TV/MV ratio < 0.7 (ii) RV/LV length ratio < 0.6 (iii) Tricuspid inflow duration/cardiac cycle length ≤ 31.5% (iv) Presence of ventriculocoronary connections	3/4 criteria: Sn 100%, Sp 75%
Gardiner et al. (2008) [12]	(i) PV *z*-score < −1 or TV *z*-score < −3.4 before 23 weeks (ii) Median TV *z*-score < −3.95, before 26 weeks (iii) Median PV *z*-score < −2.8 and medium TV/MV ratio < 0.7 at 26–31 weeks (iv) Median TV *z*-score < −3.9 and medium TV/MV ratio < 0.59 after 31 weeks	(i) 2/2 criteria < 23 weeks: Sn 100%, Sp 80% (ii) 1 criteria < 26 weeks*: Sn 92%, Sp 100% (iii) 2/2 criteria at 26–31 weeks: Sn 100%, Sp 100% (iv) 1 criteria after 31 weeks: Sn 100%, Sp 100%
Gómez-Montes et al. (2011) [17]	(i) TV/MV ratio ≤ 0.83 (ii) RV/LV length ratio ≤ 0.64 (iii) PV/AV ratio ≤ 0.75 (iv) Tricuspid inflow duration/cardiac cycle length ≤ 36.5%	4/4 criteria: Sn 100%, Sp 100% 3/4 criteria: Sn 100%, Sp 92%

TV: tricuspid valve; MV: mitral valve; RV: right ventricle; LV: left ventricle; PV: pulmonary valve; AV: aortic valve; BV: biventricular; Sn: sensitivity; Sp: specificity.
*In combination with hemodynamical markers of right atrial pressure (severity of tricuspid regurgitation, waveform characteristics of the ductus venosus, and restriction of the interatrial septum) and the presence of coronary artery fistulae.

Gómez Montes E, Herraiz I, Mendoza A, Galindo A. Fetal intervention in right outflow tract obstructive disease: selection of candidates and results. Cardiol Res Pract. 2012;2012:592403.

Research Article

Fetal Intervention in Right Outflow Tract Obstructive Disease: Selection of Candidates and Results

E. Gómez Montes,[1] I. Herraiz,[1] A. Mendoza,[2] and A. Galindo[1]

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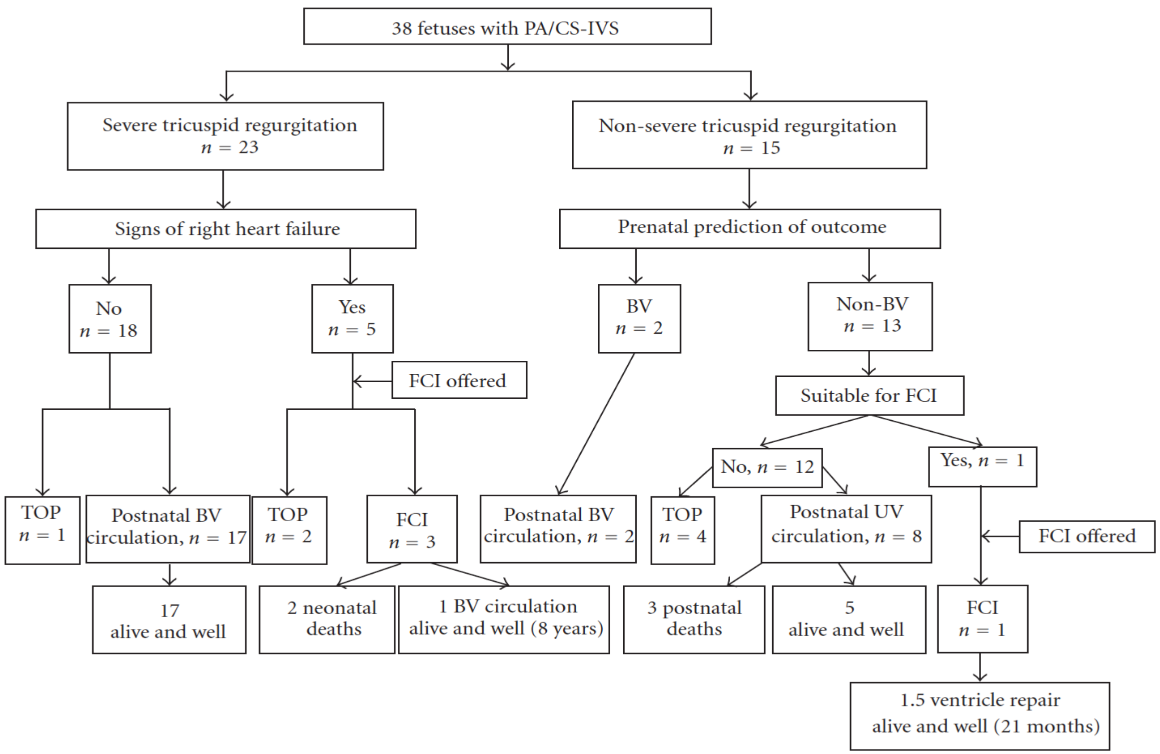


Figure 4: Flow chart showing the distribution of fetuses with pulmonary atresia/critical stenosis with intact ventricular septum (PA/CS-IVS) according to the process of selection for fetal cardiac intervention (FCI). BV, biventricular; UV, univentricular; TOP, termination of pregnancy.

Gómez Montes E, Herraiz I, Mendoza A, Galindo A. Fetal intervention in right outflow tract obstructive disease: selection of candidates and results. Cardiol Res Pract. 2012;2012:592403.

In Utero Valvuloplasty for Pulmonary Atresia With Hypoplastic Right Ventricle: Techniques and Outcomes

WHAT'S KNOWN ON THIS SUBJECT: Only 3 case reports have been published on fetal intervention for pulmonary atresia.

WHAT THIS STUDY ADDS: This is the first series on fetal PV perforation and dilation for treatment of PA/IVS and documents changes in right heart growth after successful fetal pulmonary valvuloplasty.

CONTRIBUTORS: Wayne Tworetzky, MD,[a,b] Doff B. McElhinney, MD,[a,b] Gerald R. Marx, MD,[a,b] Carol B. Benson, MD,[c] Roland Brusseau, MD,[d] Donna Morash, RN,[e] Louise E. Wilkins-Haug, MD, PhD,[f] James E. Lock, MD,[a,b] and Audrey C. Marshall, MD[a,b]

[a]Department of Cardiology, Children's Hospital Boston, Boston, Massachusetts; [b]Department of Pediatrics, Harvard Medical School, Boston, Massachusetts; Departments of [c]Radiology and [f]Obstetrics and Gynecology, Brigham and Women's Hospital and Harvard Medical School, Boston, Massachusetts; Departments of [d]Anesthesiology, Perioperative and Pain Medicine and [e]Surgery, Children's Hospital Boston and Harvard Medical School, Boston, Massachusetts

abstract

Tworetzky W, McElhinney DB, Marx GR, Benson CB, Brusseau R, Morash D, Wilkins-Haug LE, Lock JE, Marshall AC. In utero valvuloplasty for pulmonary atresia with hypoplastic right ventricle: techniques and outcomes. Pediatrics. 2009 Sep;124(3):e510-8.

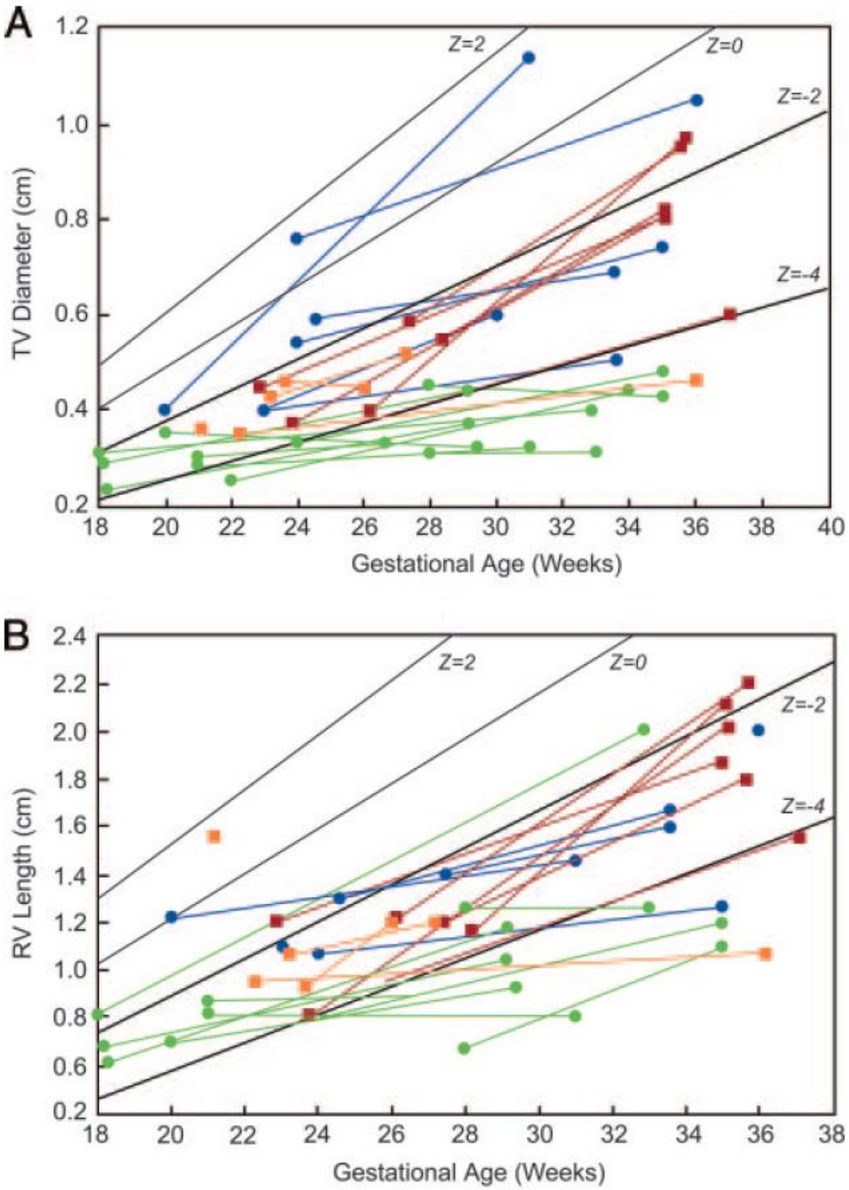
Resultados

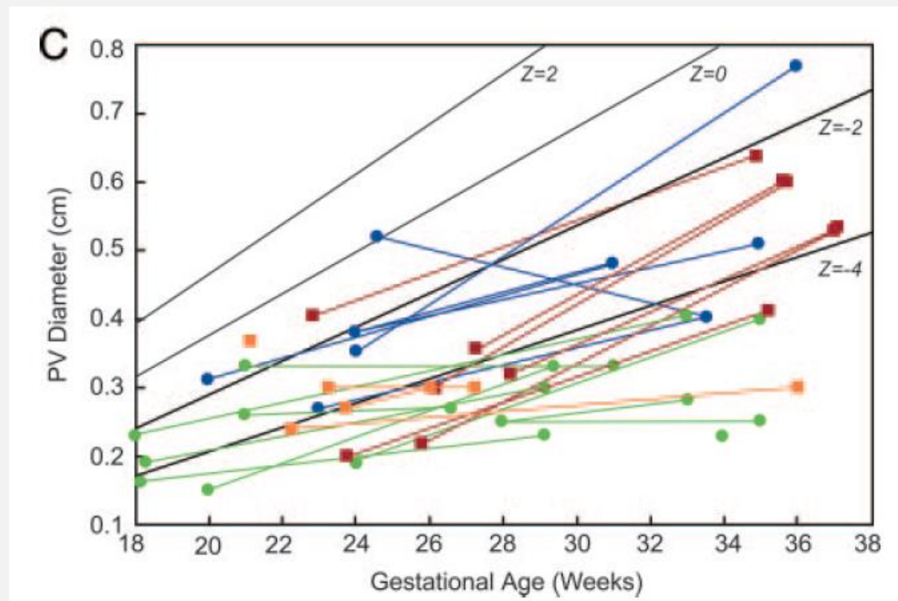
TABLE 1 Details of Prenatal Pulmonary Valvuloplasty

Patient No.	EGA, wk	Laparotomy Performed	Successful Introduction of Cannula Into RVOT	Successful Perforation of PV	Successful Balloon Dilation of PV	Antegrade Flow Present After Procedure	Treated for Hemodynamic Instability
1	24	Yes	Yes	No	No	No	Yes
2	22	Yes	Yes	No	No	No	Yes
3	21	No	No	No	No	No	Yes
4	24	Yes	Yes	Yes	No	Yes (+)	Yes
5	23	No	Yes	No	Yes	Yes (+++)	Yes
6	27	Yes	Yes	Yes	Yes	Yes (+++)	Yes
7	28	No	Yes	Yes	Yes	Yes (++)	No
8	24	Yes	Yes	Yes	Yes	Yes (+++)	No
9	26	Yes	Yes	Yes	Yes	Yes (++)	No
10	26	No	Yes	Yes	Yes	Yes (++)	No

EGA indicates estimated gestational age; RVOT, right ventricular outflow tract.

Tworetzky W, McElhinney DB, Marx GR, Benson CB, Brusseau R, Morash D, Wilkins-Haug LE, Lock JE, Marshall AC. In utero valvuloplasty for pulmonary atresia with hypoplastic right ventricle: techniques and outcomes. Pediatrics. 2009 Sep;124(3):e510-8.





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Azul = biventricular
Verde = Univentricular
Rojo = Intervención exitosa
Naranjo = Intervención no exitosa

Tworetzky W, McElhinney DB, Marx GR, Benson CB, Brusseau R, Morash D, Wilkins-Haug LE, Lock JE, Marshall AC. In utero valvuloplasty for pulmonary atresia with hypoplastic right ventricle: techniques and outcomes. Pediatrics. 2009 Sep;124(3):e510-8.

Fetal Diagnosis and Therapy

Original Paper

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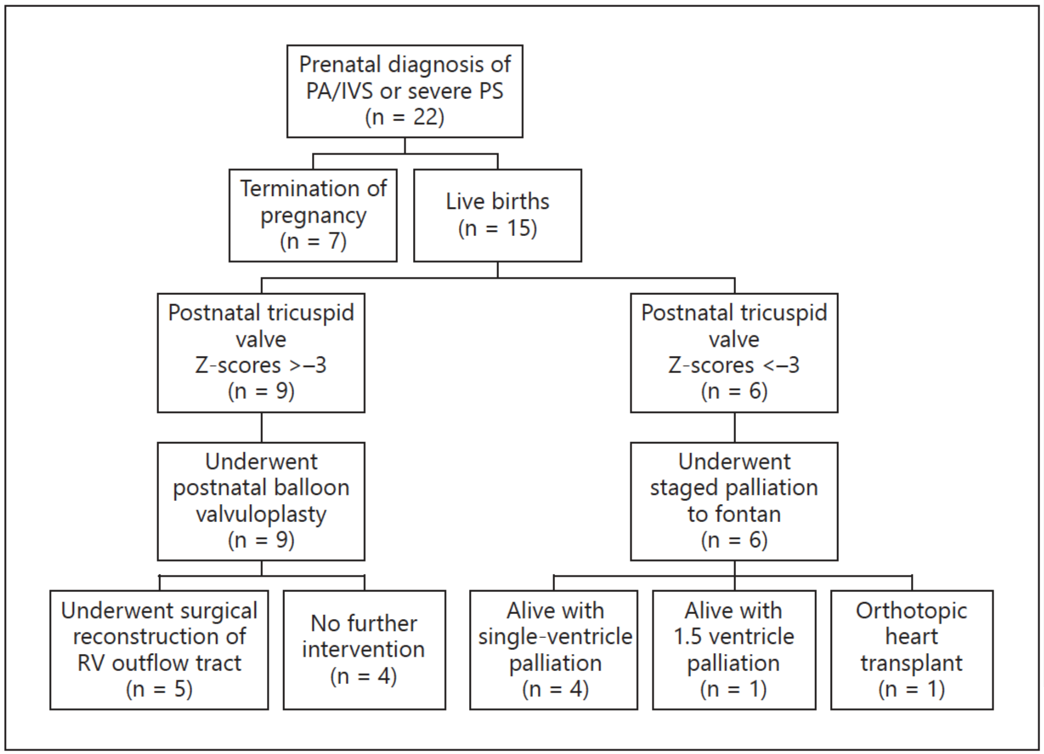
CERPO

Prenatal Tricuspid Valve Size as a Predictor of Postnatal Outcome in Patients with Severe Pulmonary Stenosis or Pulmonary Atresia with Intact Ventricular Septum

Alexander Lowenthal[a] Breniel Lemley[a] Alaina K. Kipps[a] Michael M. Brook[a] Anita J. Moon-Grady[a, b]

Departments of [a]Pediatrics and [b]Fetal Treatment Center, University of California, San Francisco, Calif., USA



Lowenthal A, Lemley B, Kipps AK, Brook MM, Moon-Grady AJ. Prenatal tricuspid valve size as a predictor of postnatal outcome in patients with severe pulmonary stenosis or pulmonary atresia with intact ventricular septum. Fetal Diagn Ther. 2014;35(2):101-7.

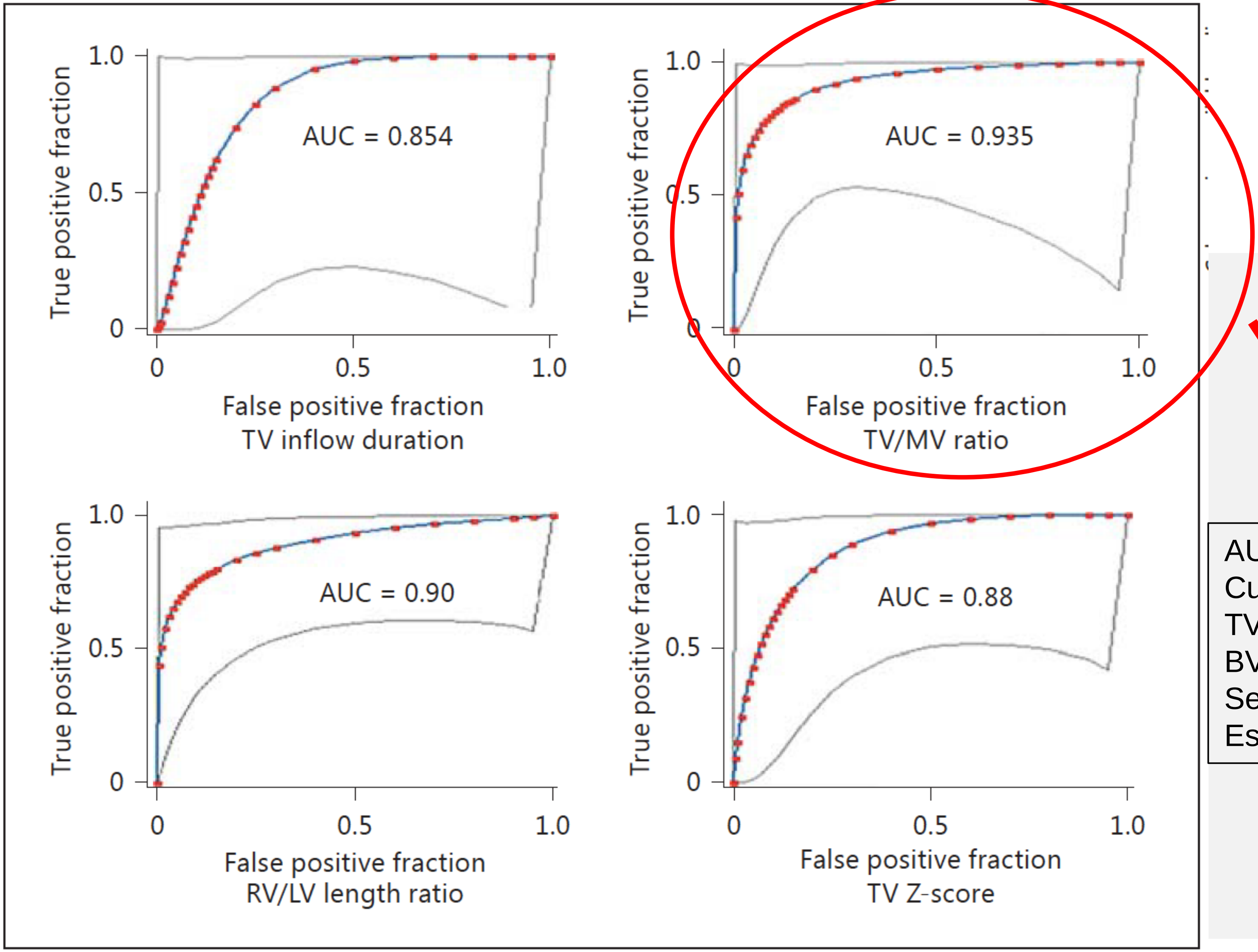
Table 1. Data derived from the initial echocardiograms

Fetal echocardiographic data	Neonatal TV Z >–3 (n = 9)	Neonatal TV Z <–3 (n = 6)	p value
GA at Dx, weeks (±SD)	27.1±4.8	29.5±3.8	NS
TV/MV ratio (±SD)	0.74±0.2	0.4±0.2	0.002
TV Z score (±SD)	–2.28±1.8	–6.45±2.8	0.003
TV inflow duration (±SD)	0.37±0.06	0.27±0.09	0.046
RV/LV length ratio (±SD)	0.72±0.2	0.38±0/1	0.004
None/mild TR	6	6	NS
Moderate/severe TR	4	0	see Discussion
PV forward flow	5	0	see Discussion
TV monophasic	1	4	NS
Abnormal systemic venous Dopplers	5	5	NS
Retrograde DA flow	7*	6	NS

DA = Ductus arteriosus; Dx = diagnosis; GA = gestational age; LV = left ventricle; MV = mitral valve annulus; NS = not significant; RV = right ventricle; TR = tricuspid regurgitation; TV = tricuspid valve annulus.

* Two fetuses had equivocal imaging on the initial study but on follow-up fetal examination and in our database had flow reversal in the ductus arteriosus.

Lowenthal A, Lemley B, Kipps AK, Brook MM, Moon-Grady AJ. Prenatal tricuspid valve size as a predictor of postnatal outcome in patients with severe pulmonary stenosis or pulmonary atresia with intact ventricular septum. Fetal Diagn Ther. 2014;35(2):101-7.



AUC 0.94
Cut-off > 0.63
TV Z-score >-3
BV outcome
Sensibilidad 78%
Especificidad 100%

Lowenthal A, Lemley B, Kipps AK, Brook MM, Moon-Grady AJ. Prenatal tricuspid valve size as a predictor of postnatal outcome in patients with severe pulmonary stenosis or pulmonary atresia with intact ventricular septum. Fetal Diagn Ther. 2014;35(2):101-7.

SHVI / Septum atrial intacto o restrictivo

- SHVI establecido sin potencial de circulación biventricular

- SHVI
  - Cierre anormal del foramen oval
  - Engrosamiento anormal del septum atrial
  - Septum primum anormalmente grande

- Septum restrictivo en 22% de los casos de SHVI
- 6% de los casos septum intacto

Schidlow DN, Tworetzky W, Wilkins-Haug LE. Percutaneous fetal cardiac interventions for structural heart disease. Am J Perinatol. 2014 Aug;31(7):629-36.
Schidlow DN, Freud L, Friedman K, Tworetzky W. Fetal interventions for structural heart disease. Echocardiography. 2017 Dec;34(12):1834-1841.

SHVI / Septum atrial intacto o restrictivo

CERPO

- **<u>Fisiopatología</u>**
- *Septum atrial intacto o restrictivo*
- *Aumento de presión en aurícula izquierda*
- *Daño en vasculatura y parénquima pulmonar*
- *“Arterialización de venas pulmonares”*

Schidlow DN, Tworetzky W, Wilkins-Haug LE. Percutaneous fetal cardiac interventions for structural heart disease. Am J Perinatol. 2014 Aug;31(7):629-36.

SHVI / Septum atrial intacto o restrictivo

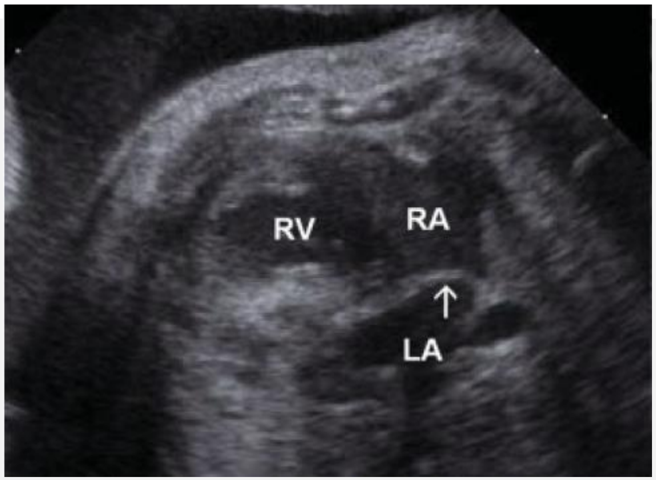
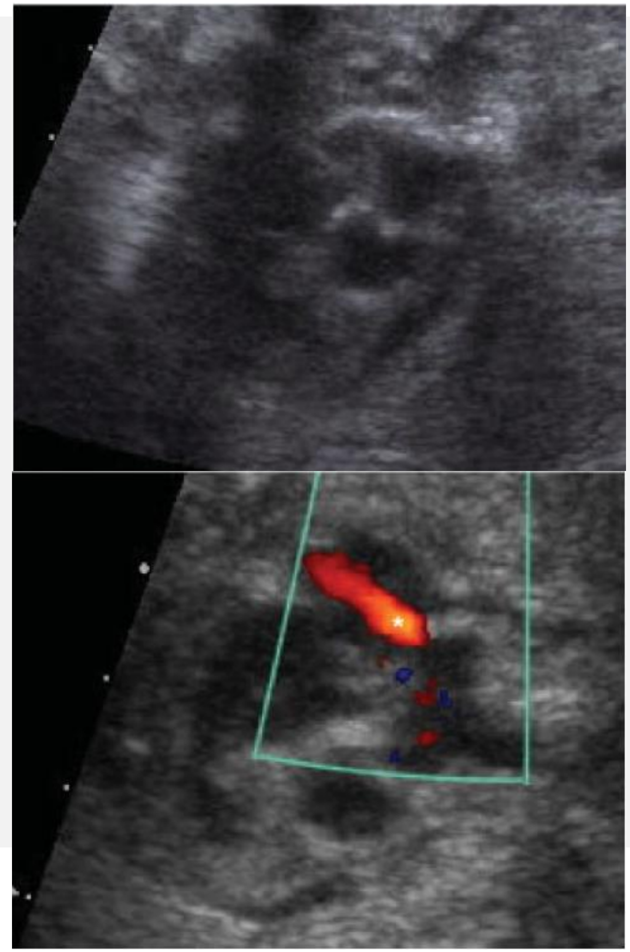


Fig. 2 Top panel: Hypoplastic left heart syndrome with intact atrial septum at 31 weeks gestational age. Severe left ventricular hypoplasia is noted. The left atrium is markedly dilated, and the thickened, hypertrophied septum primum (arrow) bulges into the right atrium. Middle panel: An inflated angioplasty balloon traverses the right atrium, atrial septum and into the left atrium. Bottom panel: Still-frame color Doppler image demonstrating a jet of left-to-right flow (asterisk) across the newly created atrial septal defect. LA, left atrium; RA, right atrium; RV, right ventricle.



Schidlow DN, Tworetzky W, Wilkins-Haug LE. Percutaneous fetal cardiac interventions for structural heart disease. Am J Perinatol. 2014 Aug;31(7):629-36.

SHVI / Septum atrial intacto o restrictivo

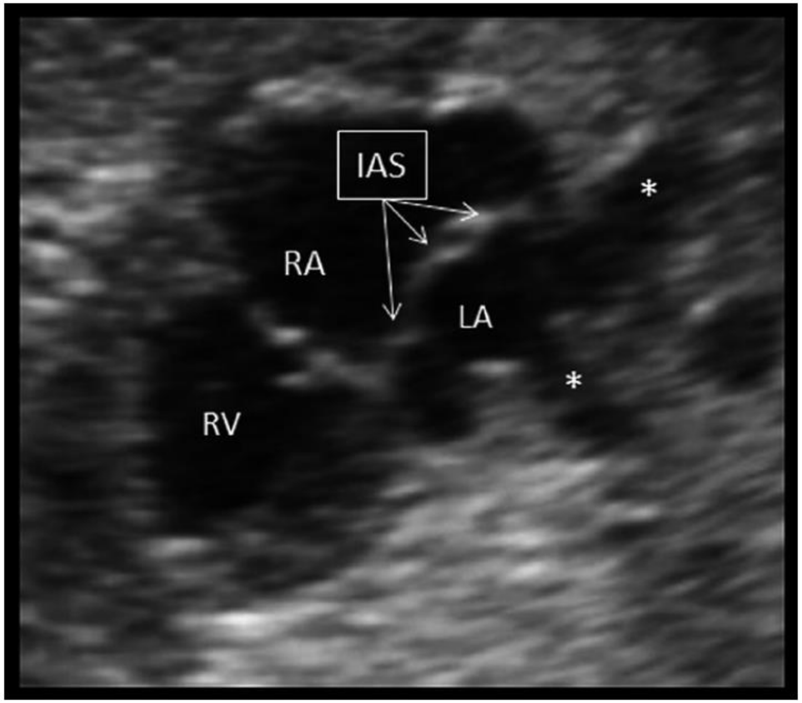


FIGURE 8 Hypoplastic left heart syndrome with intact atrial septum. The atrial septum is thickened and bows into the right atrium. Dilated pulmonary veins, consistent with left atrial hypertension, are present. IAS = intact atrial septum; LA = left atrium; RA = right atrium; RV = right ventricle, *pulmonary veins

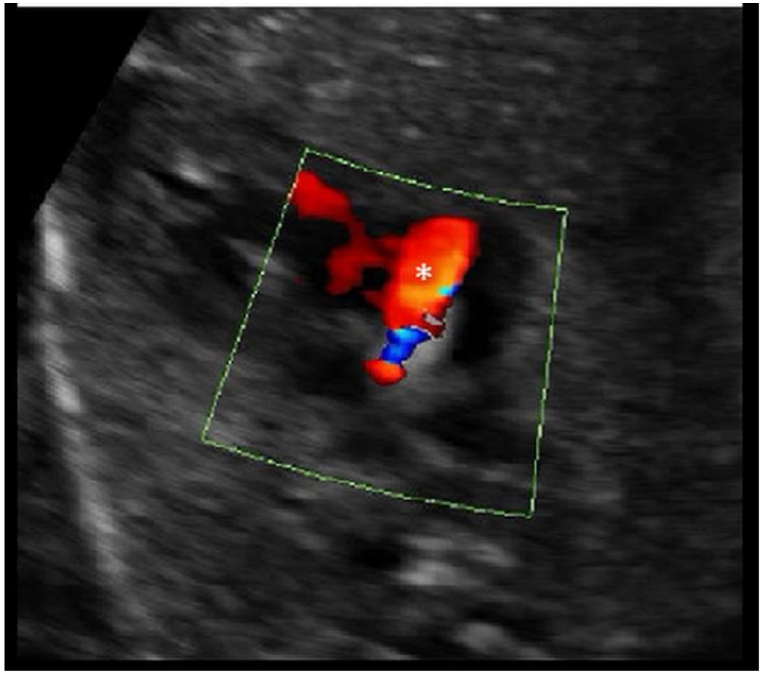


FIGURE 10 Appearance of hypoplastic left heart syndrome with intact atrial septum after stent angioplasty of the atrial septum. The top panel demonstrates a two-dimensional image of the atrial septum stent. The bottom panel demonstrates a relatively broad jet of accelerated left-to-right flow across the stented atrial septum. LA = left atrium; RA = right atrium; RV = right ventricle, *left atrium-to-right atrium color Doppler jet

SHVI / Septum atrial intacto o restrictivo

Técnica quirúrgica

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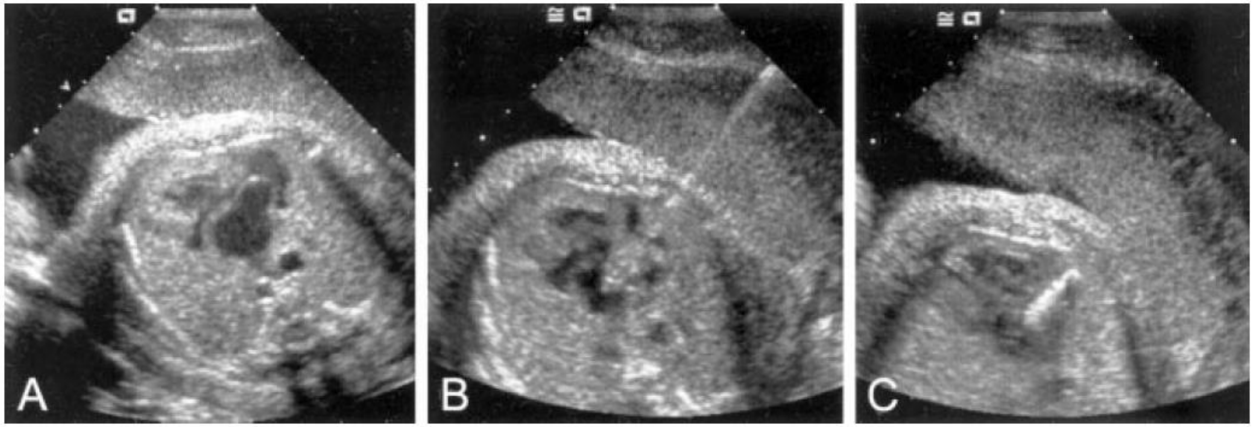

Figure 3. A–C, Transabdominal ultrasound image of heart in 1 fetus with dilated left atrium and thin, bulging atrial septum before atrial septal puncture (A), during septal puncture with Chiba needle (B), and during dilation with 3-mm coronary angioplasty balloon (C).

Marshall AC, van der Velde ME, Tworetzky W, Gomez CA, Wilkins-Haug L, Benson CB, Jennings RW, Lock JE. Creation of an atrial septal defect in utero for fetuses with hypoplastic left heart syndrome and intact or highly restrictive atrial septum. Circulation. 2004 Jul 20;110(3):253-8.

SHVI / Septum atrial intacto o restrictivo

Selección de pacientes

Table 2 Characteristics suggesting atrial restriction in HLHS

Intact atrial septum
$\leq$ 1-mm atrial septal defect
Prominent pulmonary vein flow reversal
Forward: reverse pulmonary vein velocity time integral ratio < 5

Abbreviation: HLHS, hypoplastic left heart syndrome.

Schidlow DN, Tworetzky W, Wilkins-Haug LE. Percutaneous fetal cardiac interventions for structural heart disease. Am J Perinatol. 2014 Aug;31(7):629-36.

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ISSN 0735-1097/04/$30.00
doi:10.1016/j.jacc.2004.01.033

CERPO

Intrauterine Pulmonary Venous Flow and Restrictive Foramen Ovale in Fetal Hypoplastic Left Heart Syndrome

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Lisa K. Hornberger, MD*†
Toronto, Ontario, Canada

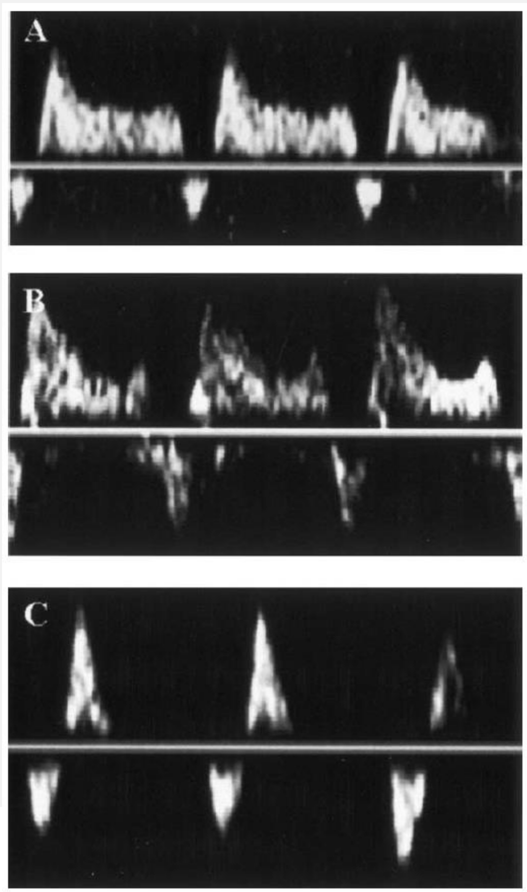
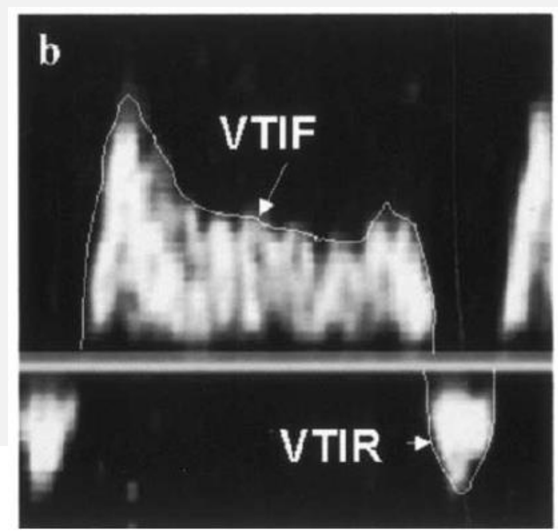


Figure 2. Three pulmonary vein flow patterns identified in fetuses with left heart obstruction. **(A)** Continuous forward flow with a small a-wave reversal (VTIR/VTIF ratio <0.18). **(B)** Continuous forward flow with an increased a-wave reversal (VTIR/VTIF ratio ≥0.18). **(C)** To-and-fro flow pattern with absent early diastolic forward flow. VTIF = velocity-time integral for forward flow; VTIR = velocity-time integral for reverse flow.



Taketazu M, Barrea C, Smallhorn JF, Wilson GJ, Hornberger LK. Intrauterine pulmonary venous flow and restrictive foramen ovale in fetal hypoplastic left heart syndrome. J Am Coll Cardiol. 2004 May 19;43(10):1902-7.

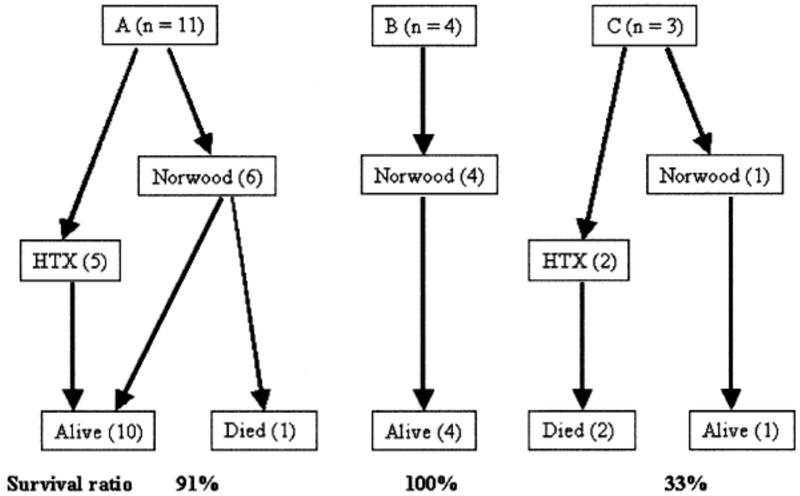


Figure 4. Outcome of patients with perinatal surgical intervention. Peri-operative survival was 91% in patients with type A, 100% with type B, and 33% with type C pulmonary vein flow. HTX = heart transplant.

Table 2. Comparison of Neonatal FO Size and Clinical Condition Between Fetuses With Different Pulmonary Venous Flow Patterns Identified in Utero

	Type A Flow (n = 12)	Type B Flow (n = 4)	Type C Flow (n = 3)	p Value*
Neonatal FO (mm)	4.5 ± 2.1	1.6 ± 1.6	0.0 ± 0.0	0.0015
PaO_2 (mm Hg)	52.0 ± 15.8	61.7 ± 25.9	30.7 ± 3.5	0.022
BE (mmol/l)	−2.6 ± 3.6	−4.3 ± 1.4	−12.2 ± 11.1	0.040
Ventilation	2	0	3	0.010
Inotropic support	2	0	2	0.112
Emergent septoplasty	0	0	3	0.001

*The Kruskal-Wallis test was used to compare continuous variables and the Fisher exact test for categorical variable. Data are presented as the mean value ± SD or number of patients.

FO = size of foramen ovale; other abbreviations as in Table 1.

Taketazu M, Barrea C, Smallhorn JF, Wilson GJ, Hornberger LK. Intrauterine pulmonary venous flow and restrictive foramen ovale in fetal hypoplastic left heart syndrome. J Am Coll Cardiol. 2004 May 19;43(10):1902-7.

Predictive Value of Fetal Pulmonary Venous Flow Patterns in Identifying the Need for Atrial Septoplasty in the Newborn With Hypoplastic Left Ventricle

Erik Michelfelder, MD; Carlen Gomez, MD; William Border, MBChB;
William Gottliebson, MD; Cheri Franklin, CNP

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TABLE 1. Median Fetal Pulmonary Venous Doppler Indices for Newborns Who Did or Did Not Require EAS

	(+) EAS (n=8)	(−) EAS (n=33)	*P*
S-wave velocity, cm/s	45 (21–75)	42 (16–71)	NS
D-wave velocity, cm/s	14 (4–26)	21 (7–40)	<0.04
S-wave/D-wave ratio	3.2 (2.0–5.0)	1.8 (1.0–5.8)	<0.001
A-wave velocity, cm/s	35 (13–54)	17 (7–41)	<0.001
A-wave duration, ms	92 (68–119)	63 (20–88)	<0.001
A-wave VTI, cm	2.5 (0.8–5.3)	0.6 (0.1–2.4)	0.0001
Forward flow VTI, cm	5.9 (3.1–10.8)	8.4 (3.1–14.5)	NS
Forward/reverse VTI ratio	3.4 (0.9–5.8)	13.6 (3.3–48.3)	<0.0001
A-wave/D-wave ratio	2.7 (1.3–6.8)	0.7 (0.2–4.3)	0.0001

Values in parentheses are ranges.

TABLE 2. ROC Analysis for PVD Indices With AUC ≥0.90

PVD Index	Area Under ROC Curve	95% Confidence Limits	"Optimal" Cutpoint Value
S/D-wave velocity ratio	0.90	0.79–1.00	>2.5
A-wave duration	0.93	0.84–1.00	>77 ms
A-wave VTI	0.94	0.85–1.00	>1.4 cm
A/D-wave velocity ratio	0.94	0.87–1.00	>1.35
Forward/reverse VTI ratio	0.98	0.94–1.00	<5.0

AUC indicates area under the curve.

Michelfelder E, Gomez C, Border W, Gottliebson W, Franklin C. Predictive value of fetal pulmonary venous flow patterns in identifying the need for atrial septoplasty in the newborn with hypoplastic left ventricle. Circulation. 2005 Nov 8;112(19):2974-9.

Predictive Value of Fetal Pulmonary Venous Flow Patterns in Identifying the Need for Atrial Septoplasty in the Newborn With Hypoplastic Left Ventricle

Erik Michelfelder, MD; Carlen Gomez, MD; William Border, MBChB;
William Gottliebson, MD; Cheri Franklin, CNP

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TABLE 3. Clinical Accuracy of PVD Doppler Indices in Predicting Need for EAS in the Newborn Period

PVD Index	Sensitivity	Specificity	PPV	NPV	Positive LH Ratio	Negative LH Ratio
A-wave duration >77 ms	0.88 (0.47–0.99)	0.82 (0.64–0.92)	0.54 (0.26–0.79)	0.96 (0.80–0.99)	4.8 (2.2–10.4)	0.15 (0.02–0.97)
S/D ratio >2.5	0.88 (0.47–0.99)	0.85 (0.67–0.94)	0.58 (0.29–0.84)	0.97 (0.80–0.99)	5.8 (2.5–13.5)	0.15 (0.02–0.93)
A/D ratio >1.35	0.88 (0.47–0.99)	0.91 (0.75–0.98)	0.70 (0.35–0.92)	0.97 (0.81–0.99)	9.6 (3.2–29.2)	0.14 (0.02–0.87)
A-wave VTI >1.4 cm	0.88 (0.47–0.99)	0.94 (0.78–0.99)	0.78 (0.40–0.96)	0.97 (0.82–0.99)	14.4 (3.7–56.7)	0.13 (0.02–0.84)
Forward/reverse VTI ratio <5	0.88 (0.47–0.99)	0.97 (0.82–0.99)	0.88 (0.47–0.99)	0.97 (0.82–0.99)	28.9 (4.1–203)	0.13 (0.02–0.80)

LH ratio indicates likelihood ratio; NPV, negative predictive value; PPV, positive predictive value. Values in parentheses are 95% CIs.

Michelfelder E, Gomez C, Border W, Gottliebson W, Franklin C. Predictive value of fetal pulmonary venous flow patterns in identifying the need for atrial septoplasty in the newborn with hypoplastic left ventricle. Circulation. 2005 Nov 8;112(19):2974-9.

Congenital Heart Disease

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Creation of an Atrial Septal Defect In Utero for Fetuses With Hypoplastic Left Heart Syndrome and Intact or Highly Restrictive Atrial Septum

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Summary of Procedures

Fetus	Gestational Age, wk	LA Size, mm	Septum	LA–RA Gradient, mm Hg	Balloon Diameter, mm	Result*	O_2 Saturation at Catheterization, %	LA Pressure, mm Hg	Outcome
1	34	20×22	Thin	≥9†	3.0	++	73	33	Alive
2	30	8×10	Thick	≥7†	2.75	+	58	28	Neonatal death
3‡	26	5×7	Thick	Intact	2.75	+	NA	NA	Fetal demise
4	26	8×10	Thin	8	3.0	++	NA	NA	Neonatal death
5‡	30	27×37	Thin	Intact	2.75	++	NA	NA	Neonatal death
6	29	6×15	Thick	Intact	NA	0	43		Alive
7	33	11×15	Thin	Intact	2.75	++	61	26	Neonatal death

RA indicates right atrium.
*Results are the following: 0, no new defect; +, new defect ≤1 mm; and ++, new defect >2 mm.
†Determined by interrogating decompressing vein.
‡Hydropic.

Marshall AC, van der Velde ME, Tworetzky W, Gomez CA, Wilkins-Haug L, Benson CB, Jennings RW, Lock JE. Creation of an atrial septal defect in utero for fetuses with hypoplastic left heart syndrome and intact or highly restrictive atrial septum. Circulation. 2004 Jul 20;110(3):253-8.

Potenciales beneficios

Mejorar la circulación venosa pulmonar postnatal

Atenuar los cambios anatómicos pulmonares

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Results of *in utero* atrial septoplasty in fetuses with hypoplastic left heart syndrome

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- 21 procedimientos
  - 19 técnicamente exitosos (90%)
  - Complicaciones 8/21 casos (38%)
  - ASD > 3mm en 6 fetos

- ASD > 3mm
  - Mayor saturación de oxigeno postnatal (p=0.001)
  - Menor necesidad de descompresión atrial previo a la cirugía estadio I. (p=0.001).

- Sobrevida total 58%
  - 5/12 (42%) en descompresión atrial de urgencia
  - 5/7 (86%) en estadio I de cirugía paliativa

Marshall AC, Levine J, Morash D, Silva V, Lock JE, Benson CB, Wilkins-Haug LE, McElhinney DB, Tworetzky W. Results of in utero atrial septoplasty in fetuses with hypoplastic left heart syndrome. Prenat Diagn. 2008 Nov;28(11):1023-8.

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