

**Figure 2.** A: T2-weighted sequence showing an increase in intensity in the lateral wall of the left ventricle, corresponding to tissue edema. B: T1-weighted sequence showing delayed enhancement affecting the subepicardium and sparing the subendocardium.

creatine kinase-MB fraction was 79  $\mu$ g/L. The remaining blood test results were normal. An echocardiogram showed normal anatomy of the coronary arteries, a moderately dilated left ventricle (end-diastolic diameter: 61 mm [Z-score=2.48]), an ejection fraction of 45% and moderate mitral failure. An MRI scan showed an ejection fraction of 50% with a normal end-diastolic volume. A delayed enhancement study showed a pattern of patchy subepicardial enhancement in the lateral wall. An area of increased signal intensity was visible in the T2-weighted MRI image, which was suggestive of edema (Fig. 2).

The patient's course was favorable; his systolic function returned to normal with a decrease in markers of damage. The suspected diagnosis was acute myocarditis. At admission, the results of polymerase chain reaction of blood and nasopharyngeal aspirate were negative for viruses; therefore the causal agent was not identified.

The third patient was a 10-year-old girl who presented to the emergency department after experiencing 4 episodes of oppressive chest pain, radiating to her arm. Each episode lasted approximately 1 h. An echocardiogram showed moderate ST-elevation of 1 mm in LII, LIII and aVF, with troponin T at 9.12  $\mu$ g/L and creatine kinase-MB fraction at 272  $\mu$ g/L. An echocardiogram showed normal coronary artery anatomy, a hypertrophic left ventricle (septum 10 mm [Z-score=2.77]; posterior wall 9 mm [Z-score=2.13]), which was not dilated, with an ejection fraction of 65%. T2-weighted MRI scan showed subepicardial areas of increased signal intensity in the free wall of the left ventricle. The delayed enhancement sequences showed generalized, subepicardial enhancement of the

left ventricle, compatible with acute myocarditis. Polymerase chain reaction testing of blood and nasopharyngeal aspirate was negative for viruses. The patient's course was favorable.

Precordial pain is a presentation of acute myocarditis and, although uncommon in children, should be included in the differential diagnosis.

The usefulness of MRI in these patients has been previously reported.<sup>6</sup> The clinical course is usually favorable and the most commonly described causative agent is Parvovirus B19. In the cases reported here, the diagnostic utility of MRI should be emphasised as it allowed catheterization to be avoided in our patients. It should be performed as an emergency procedure and, if inconclusive, a coronary angiography should be conducted to rule out coronary disease.

Ferran Gran,<sup>a,\*</sup> Amparo Castellote,<sup>b</sup> Laia Vega,<sup>a</sup> Dimpna Albert,<sup>a</sup> Queralt Ferrer,<sup>a</sup> and Joan Sanchez-De-Toledo<sup>c</sup>

<sup>a</sup>Unidad de Cardiología Pediátrica, Hospital Universitario de la Vall d'Hebron, Universidad Autónoma de Barcelona, Barcelona, Spain <sup>b</sup>Servicio de Radiología Pediátrica, Hospital Universitario de la Vall d'Hebron, Universidad Autónoma de Barcelona, Barcelona, Spain <sup>c</sup>Unidad de Cuidados Intensivos Pediátricos, Hospital Universitario de la Vall d'Hebron, Universidad Autónoma de Barcelona, Barcelona, Spain

\* Corresponding author: E-mail address: fgran@vhebron.net (F. Gran).

Available online 7 August 2013

#### REFERENCES

- 1. Kühl U, Schultheiss HP. Myocarditis in children. Heart Failure Clin. 2010;6: 483–96.
- 2. Sagar S, Liu PP, Cooper LT. Myocarditis. Lancet. 2012;379:738-47.
- Tschöpe C, Bock CT, Kasner M, Noutsias M, Westermann D, Schwimmbeck PL, et al. High prevalence of cardiac Parvovirus B19 infection in patients with isolated left ventricular diastolic dysfunction. Circulation. 2005;111:879–86.
- Simpson KE, Canter CE. Acute myocarditis in children. Expert Rev Cardiovasc Ther. 2011;9:771–83.
- Friedrich MG, Sechtem U, Schulz-Menger J, Holmvang G, Alakija P, Cooper LT, et al. Cardiovascular magnetic resonance in myocarditis: A JACC white paper. J Am Coll Cardiol. 2009;53:1475–87.
- 6. Laraudogoitia Zaldumbide E, Pérez-David E, Larena JA, Velasco del Castillo S, Rumoroso Cuevas JR, Onaindía JJ. Utilidad de la resonancia magnética cardiaca en el diagnóstico de los pacientes con síndrome coronario agudo y coronarias normales. Rev Esp Cardiol. 2009;62:976–83.

http://dx.doi.org/10.1016/j.rec.2013.05.012

# Fenestration Closure After Fontan Surgery. Contributions of Percutaneous Interventionism

# Cierre de fenestración tras la cirugía de Fontan. Aportaciones del intervencionismo percutáneo

## To the Editor,

The Fontan procedure is the final step in surgery for patients with a single ventricle. The hemodynamic changes that occur after the procedure can have a negative impact on the immediate outcome due to a sudden increase in pulmonary artery pressure. Fenestration of the Fontan circuit during surgery is therefore a common procedure in high-risk patients,<sup>1</sup> although systemic saturation may decrease as a result. The development of percutaneous implantation devices has enabled fenestration closure without the need for further surgery when hemodynamic conditions allow.<sup>2,3</sup>

Here, we analyze our experience of percutaneous closure of fenestrations in the extracardiac circuit after the Fontan procedure, taking into account the properties of the new occlusion devices available. In addition, we study the changes in pulmonary artery

1 PAIS 8 17 123 87 99 11 12 96/45 (62) 97/40 (56) 5 V510 mm 1 1 1   2 PA+TA 15 50 160 93 96 14 15 97/60 (73) 95/50 (73) 3 PF0 18 mm 1 1 1   3 TA+D-TGA 8 23 125 88 91 14 102/58 (75) 3 PF0 18 mm 1	Case	Underlying disease	Age, years	Weight, kg	Height, cm	Baseline saturation, %	Saturation after closure, %	Baseline PAP, mmHg	PAP after closure, mmHg	Baseline AoP, mmHg <sup>*</sup>	AoP after closure, mmHg*	Diameter of defect, mm	Device	Baseline FC	FC after closure
2 Ph-TA 15 50 160 93 96 14 15 96/16 3 PF0 18 mm 1   3 TA+D-TCA 8 23 125 88 95 18 55/67) 3 PF0 18 mm 1   4 TA 12 38 145 91 97 14 102/58 (73) 86/57 (67) 3 PF0 18 mm 1   5 HLV 5 16 102 85 67 95 44 (59) 8 767 (87) 95/767) 4 PF0 18 mm 1   6 PAIS 11 33 137 93 22 22 101/52 (68) 95/57 (77) 95/97 (77) 45 PF0 18 mm 1   7 AT+PS 11 33 137 93 97 12 85/57 (67) 95/97 (77) 45 PF0 18 mm 1 1   7 AT+PS 13 140 89 23 160 18 mm 1	1	PAIS	8	17	123	87	66	11	12	96/45 (62)	97/47 (66)	5	VS 10 mm	П	Ι
3 TAHD-TGA 8 23 125 88 95 67 3 P60 8 1   4 TA 12 38 145 91 97 14 102/58 (73) 88/57 (67) 4 P60 8 1   5 HU 5 16 102 85 167 15 95/4 (59) 8 75 16 17 1   6 PAIS 10 32 136 92 98 27 16 1 4 1	2	PA+TA	15	50	160	93	96	14	15	97/60 (73)	95/59 (72)	ŝ	PFO 18 mm	I	Ι
4 TA 12 38 145 91 97 14 102/58 (73) 88/57 (67) 4 FP0 18 mm 1   5 HLV 5 16 102 85 98 16 15 88/57 (67) 95/44 (59) 8 751 mm 1   6 PAIS 10 32 136 92 98 22 22 101/52 (88) 99/52 (70) 5 FP0 18 mm 1   7 AT+PS 11 33 137 93 97 23 24 90/60 (70) 87/60 (70) 4 FP0 18 mm 1   8 AT 11 33 137 93 97 23 106 70 87/60 (70) 87/60 (70) 87/60 (70) 87/60 (70) 87/60 (70) 87/60 (70) 1   9 AT+PS 11 83 90 90/50 (70) 87/60 (70) 87/60 (70) 87/60 (70) 87/60 (70) 87/60 (70) 87/60 (70) 87/60 (70) 87/60 (70) <t< td=""><td>£</td><td>TA+D-TGA</td><td>8</td><td>23</td><td>125</td><td>88</td><td>95</td><td>18</td><td>18</td><td>92/62 (76)</td><td>86/53 (67)</td><td>ŝ</td><td>PFO 18 mm</td><td>Π</td><td>Ι</td></t<>	£	TA+D-TGA	8	23	125	88	95	18	18	92/62 (76)	86/53 (67)	ŝ	PFO 18 mm	Π	Ι
5 HLV 5 16 102 85 16 15 88/57 (67) 95/44 (59) 8 V5 12 mm 1   6 PAIS 10 32 136 92 98 22 22 101/52 (68) 99/52 (70) 5 PF0 18 mm 1   7 AT+PS 15 50 157 90 97 15 88/57 (67) 96/52 (70) 5 PF0 18 mm 1   8 TA 11 33 137 93 97 23 24 90/60 (70) 87/60 (70) 4 PF0 18 mm 1   9 AT+PS 11 43 140 89 21 23 108/73 (89) 102/70 (85) 3.7 Cribiform AS 11   10 VS 11 11 87 94 10 18 1 16   10 VS 12 89 21 23 108/73 (89) 102/70 (85) 3.7 Cribiform AS 11	4	TA	12	38	145	91	97	14	14	102/58 (73)	88/57 (67)	4	PFO 18 mm	Ι	Ι
6 PAIS 10 32 136 92 98 22 101/52 (68) 99/52 (70) 5 PF0 18 mm 1   7 AT+PS 15 50 157 90 975 (67) 90/59 (71) 45 PF0 18 mm 1   8 TA 11 33 137 93 97 15 96/50 (70) 87/60 (70) 47 PF0 18 mm 1   9 AT+PS 11 43 140 89 95 157 96/52 (70) 96/51 (69) 3.7 PF0 18 mm 1   10 VS 17 53 157 90 98 21 23 700 (85) 3.7 Cribiform AS 11 1   11 DRVO+unelated VS 9 24 157 99/54 (69) 100/54 (69) 3.7 Cribiform AS 11 1   11 DRVO+unelated VS 9 24 10 100/54 (69) 3.7 Cribiform AS 11   12 TA+	ъ	HLV	ъ	16	102	85	98	16	15	88/57 (67)	95/44 (59)	8	VS 12 mm	I	П
7 MT+P5 15 50 157 90 71 4.5 86/51 (67) 4.5 76.0 8 mm 1   8 TA 11 33 137 93 97 23 24 90/60 (70) 87/60 (70) 4 PF0 18 mm 1   9 AT+P5 11 43 140 89 95 15 96/51 (69) 3.7 PF0 18 mm 1   10 V5 17 53 157 90 96/51 (69) 3.7 PF0 18 mm 1   11 V5 92 15 16 15 99/54 (69) 100/54 (69) 3 Ductus occluder 1   12 TA+PS 9 24 125 80 92 15 99/54 (69) 3 Ductus occluder 1   12 TA+PS 9 24 15 99/54 (69) 100/54 (69) 3 Ductus occluder 1   12 TA+PS 1 23 24 99/54 (69) <td>9</td> <td>PAIS</td> <td>10</td> <td>32</td> <td>136</td> <td>92</td> <td>98</td> <td>22</td> <td>22</td> <td>101/52 (68)</td> <td>99/52 (70)</td> <td>5</td> <td>PFO 18 mm</td> <td>Ι</td> <td>Ι</td>	9	PAIS	10	32	136	92	98	22	22	101/52 (68)	99/52 (70)	5	PFO 18 mm	Ι	Ι
8 TA 11 33 137 93 97 23 24 90/60 (70) 87/60 (70) 47 PF0 18 mm 1   9 AT+PS 11 43 140 89 55 15 96/52 (70) 96/51 (69) 3.7 PF0 18 mm 1   10 VS 17 53 157 90 98 21 102/70 (85) 3.3 Cribiform AS 11   11 DRVO+unrelated VS 9 21 15 99/54 (69) 100/54 (69) 3 Ductus occluder 1   12 TA+PS 9 24 122 80 92 21 99/56 (67) 90/56 (72) 4 PF0 18 mm 1   13 TA+VS 9 24 15 110/80 (95) 86/56 (67) 3 Ductus occluder 1   14 TA+VS 1 22 24 10 90/56 (67) 3 Ductus occluder 1   15 TA+VS 10 1	7	AT+PS	15	50	157	06	97	15	15	88/57 (67)	90/59 (71)	4.5	PFO 18 mm	П	II
9 MT+PS 11 43 140 89 55 15 96 51 66/51 66/51 66/51 66/51 66/51 66/51 66/51 66/51 66/51 66/51 66/51 66/51 66/51 70 7	8	TA	11	33	137	93	97	23	24	90/60 (70)	87/60 (70)	4	PFO 18 mm	Ι	Ι
10 VS 17 53 157 90 98 21 23 108/73 (89) 102/70 (85) 3.3 Cribiform AS III   11 DRVO+unrelated VS 9 21 115 87 94 16 15 99/54 (69) 100/54 (69) 3 Ductus occluder 1   12 TA+PS 9 24 122 80 92 21 29/56 (57) 4 PF0 18 mm 1   13 TA+VS+D-TGA 7 22 117 90 96 66 67 2 PF0 25 mm 1   AoP, aortic pressure: AS, atrial shunt; D-TGA, D-transposition of the great arteries; DRVO, double right ventricular outflow; FC, functional class; HLV, hypoplastic left ventricle; PA, pulmonary atresia; PAIS, pulmonary atresia;	6	AT+PS	11	43	140	89	95	15	15	96/52 (70)	96/51 (69)	3.7	PFO 18 mm	Ι	Ι
11 DRVO+unrelated VS 9 21 115 87 94 16 15 99/54 (69) 300/54 (69) 3 Ductus occluder 1   12 TA+PS 9 24 122 80 92 21 99/61 (67) 90/56 (67) 4 PF0 18 mm 11   13 TA+VS+D-TGA 7 22 117 90 96 161 65 66 2 PF0 25 mm 1   AoP, aortic pressure: AS, atrial shunt; D-TGA, D-transposition of the great arteries; DRVO, double right ventricular outflow; FC, functional class; HIV, hypoplastic left ventricle; PA, pulmonary atresia; PAIS, pulmonary atresia; PAIS	10	NS	17	53	157	06	98	21	23	108/73 (89)	102/70 (85)	3.3	Cribiform AS	Ш	III
12 TA+PS 9 24 122 80 92 21 29/61 (67) 90/56 (72) 4 PF0 18 mm II   13 TA+VS+D-TGA 7 22 117 90 15 110/80 (95) 86/56 (67) 2 PF0 25 mm I   AoP, aortic pressure; AS, atrial shunt; D-TGA, D-transposition of the great arteries; DRVO, double right ventricular outflow; FC, functional class; HJV, hypoplastic left ventricle; PA, pulmonary atresia; PAIS, pulmonary atresia;	11	DRVO + unrelated VS	6	21	115	87	94	16	15	99/54 (69)	100/54 (69)	3	Ductus occluder	Ι	Ι
13 TA+VS+D-TGA 7 22 117 90 96 15 15 110/80 95/56 67.7 2 PF0 25 mm 1   AoP, aortic pressure; AS, atrial shunt; D-TGA, D-transposition of the great arteries; DRVO, double right ventricular outflow; FC, functional class; HLV, hypoplastic left ventricle; PA, pulmonary artesia; PAIS, pulmonary artesia	12	TA+PS	6	24	122	80	92	21	21	99/61 (67)	90/56 (72)	4	PFO 18 mm	П	Ι
AoP, aortic pressure; AS, atrial shunt; D-TGA, D-transposition of the great arteries; DRVO, double right ventricular outflow; FC, functional class; HLV, hypoplastic left ventricle; PA, pulmonary atresia; PAIS, pulmonary a	13	TA+VS+D-TGA	7	22	117	06	96	15	15	110/80 (95)	86/56 (67)	2	PFO 25 mm	Ι	Ι
intact septum: PAP. mean pulmonary artery pressure: PFO. patent foramen ovale: PS. pulmonary stenosis: SV. single ventricle: TA. tricuspid atresia: VS. ventricular shunt.	AoP, aort intact sei	tic pressure; AS, atrial shu ptum: PAP. mean pulmor	nt; D-TG/ 1arv arter	A, D-transpo: v pressure:	sition of the PFO. patent	great arteries; DR foramen ovale: PS	VO, double rigl 3. pulmonary s	nt ventricular outfl tenosis: SV. single	ow; FC, funct ventricle: T/	ional class; HLV, A. tricuspid atres	hypoplastic left ia: VS. ventricul	ventricle; PA, ar shunt.	pulmonary atresia; F	AIS, pulmonary	atresia with

| | | | | | | | | | | **| | |** 

Systolic/diastolic (mean)

pressure, as well as oxygen saturation. Fourteen patients were included. Fenestration was indicated during the procedure because high-risk criteria were identified during catheterization performed prior to the Fontan procedure.<sup>4.5</sup> One patient was excluded from the analysis because no contrast flow was observed in the baseline angiography during the procedure.

Procedures were performed under general anesthetic. An artery was cannulated (4 Fr) for monitoring. Venous cannulation was performed (6, 7, or 8 Fr) for the therapeutic intervention, which was performed with monitoring by transesophageal echocardiography in 9 cases and intracardiac echocardiography in 5. After the procedure, the therapeutic regimen comprised low-molecular-weight heparin, acetylsalicylic acid for at least 6 months, and endocarditis prophylaxis with cefuroxime axetil for 1 week.

The quantitative data were expressed as means (standard deviation). We performed a Student t test for paired data to compare the mean pulmonary pressures and systemic saturation in the same patient.

The mean (standard deviation) duration of follow-up was 45 (41) months. No major complications were recorded during the procedures. Prior therapeutic catheterization had been performed in 8 of the 13 patients to close systemic-pulmonary collaterals. Of the 13 patients analyzed, 11 had systemic left ventricle and 2 systemic right ventricle (Table). None of the patients died during follow-up or were readmitted to hospital for cardiac causes.

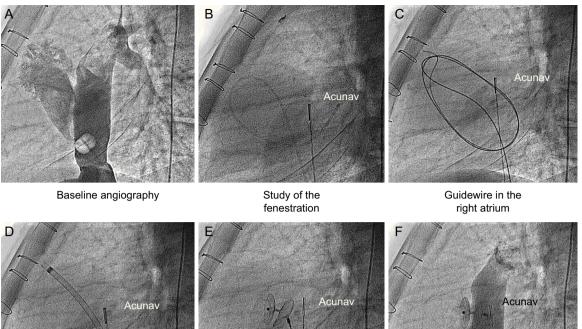
In all cases, Amplatzer occluders were used. The device type was chosen in accordance with the postsurgical anatomy, which was assessed by computed tomography prior to the procedure. Multislice computed tomography was used to locate the site of fenestration and select the angiographic view that would best visualize the defect, sliced perpendicular to the largest diameter of the defect. Once catheterization had been performed, contrast was injected and the maximum diameter of fenestration was estimated, in accordance with the calibration marked with the catheter (Figure). Computed tomography and transesophageal or intracardiac echocardiography complemented these measures and contributed to better selection of the device size. In addition to the maximum diameter of the defect, the distance between the atrial chamber and the internal edge of the conduit was taken into account. The type of Amplatzer occluder was selected in accordance with the anatomical configuration. Devices for patent foramen ovale were used for narrower fenestration while those for ventricular shunts and patent ductus arteriosus were chosen for the wider ones (Table).

In 10 cases, the device for patent foramen ovale was used (18 mm in 9 patients and 25 mm in 1 patient). In 2 patients, a muscular ventricular shunt device was used, while in the remaining patient, a ductus closure device was chosen. Oxygen saturation increased significantly after closure of the fenestration (89% [3.6] vs 96% [2.0]; *P*<.01) without any evidence of a significant increase in pulmonary artery pressure (17 [3.6] mmHg vs 17.2 [3.9] mmHg).

Patients with congenital heart disease in the form of single ventricle circulation will need to undergo surgery several times during their lives. The development of devices placed by percutaneous procedures can help avoid surgical procedures in some of these patients. Closure of the fenestration is necessary due to the long-term harmful effects of chronic hypoxemia.

The experience in our hospital suggests that a multidisciplinary approach in these patients is essential when designing therapeutic strategies and establishing the timing of the interventions. The development of percutaneous implantation devices has allowed greater flexibility in surgery, which can be adapted to the

Characteristics of the 13 Patients



Cannula in the right atrium

Amplatzer PFO 18 mm

Final result

Figure. A, Baseline angiography. B, Study of the fenestration guided by intracardiac echocardiography. C, Guidewire in the right atrium. D, Cannula in the right atrium. E, Release of patent foramen ovale closure device. F, Final angiography. PFO, patent foramen ovale.

hemodynamic conditions of each patient. Closure of the Fontan fenestration by catheterization is a safe and effective technique in these patients.

### Acknowledgments

We wish to acknowledge Dr. Ignacio Tejero, who passed away during the preparation of this letter. He had been involved in the treatment of these patients for past 20 years.

Marta Santisteban,<sup>a,\*</sup> Manuel Pan,<sup>a</sup> Miguel Romero,<sup>a</sup> Jaime Casares,<sup>b</sup> Elena Gómez,<sup>c</sup> and José Suárez de Lezo<sup>a</sup>

<sup>a</sup>Servicio de Cardiología, Hospital Universitario Reina Sofía, Córdoba, Spain

<sup>b</sup>Servicio de Cirugía Cardiovascular, Hospital Universitario Reina Sofía, Córdoba, Spain

<sup>c</sup>Servicio de Pediatría, Hospital Universitario Reina Sofía, Córdoba, Spain

\* Corresponding author.

E-mail address: marta\_santisteban@hotmail.com (M. Santisteban).

Available online 7 August 2013

### REFERENCES

- 1. Lemler MS, Scout WA, Leonard SR, Stromberg D, Ramaciotti C. Fenestration improves clinical outcome of de fontan procedure: a prospective, randomized study. Circulation. 2002;105:207-12
- 2. Bridges ND, Lock JE, Castañeda AR. Baffle fenestration with subsequent transcatheter closure. Modification of the Fontan operation for patients at increased risk. Circulation. 1990;82:1681-9.
- 3. Masura J, Borodacova L, Tittel P, Berden P, Podnar T. Percutaneous management of cyanosis in fontan patients using amplatzer occluders. Catheter Cardiovasc Interv. 2008;71:843-9
- 4. Mendoza A, Albert L, Ruiz E, Boni L, Ramos V, Velasco JM, et al. Operación de Fontan. Estudio de los factores hemodinámicos asociados a la evolución postoperatoria. Rev Esp Cardiol. 2012;65:356-62.
- Banka P, McElhinney DB, Bacha EA, Mayer JE, Gauvreau K, Geva T, et al. What is 5. the clinical utility of routine cardiac catheterization before a Fontan operation? Pediatr Cardiol. 2010;31:977-85.

http://dx.doi.org/10.1016/j.rec.2013.05.017