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Percutaneous transient occlusion of the transtricuspid flow: a new method to evaluate the right ventricle-dependent coronary circulation in pulmonary atresia with intact ventricular septum

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Abstract

Pulmonary atresia with an intact ventricular septum is characterised by heterogeneity in right ventricle morphology and coronary anatomy. In some cases, the presence of ventriculocoronary connections may promote coronary artery stenosis or interruption, and aortic diastolic pressure may not be sufficient to drive coronary blood flow. This requires a correct evaluation (currently done by angiography) which depends on whether the patient can be offered decompression of the right ventricle. To date, there is no objective method to do so, so we designed a percutaneous, transitory technique with the purpose of occluding the transtricuspid anterograde flow. The manoeuverer was performed in a 25-day-old female with pulmonary atresia with intact ventricular septum, right ventricle at suprasystemic level, and selective coronarography was not conclusive, the anterior descendant with stenosis in its middle third and from this point, thinner with to-fro flow. Occlusion was performed with a balloon catheter. We re-evaluated the coronary flow and the normalised anterior descendant flow. We hope that with this new method, we can give a more accurate diagnosis and determine the cases in which the coronary circulation is truly not right ventricle dependent to offer a greater number of patients biventricular or 1.5 ventricular repairs and thereby improve their quality of life and survival, the ones that turn out to be right ventricular dependant; offer them an early reference for cardiac transplant or in case it is not available to consider univentricular palliation knowing that this probably would not reduce the risk of ischaemia and/or death over time.

Background

Pulmonary atresia with intact ventricular septum is a rare form of CHD with an incidence classically reported of less than 1 in 10,000 live births, recently reported with an incidence of 4.1–4.6 in 100,000 in a 2021 study in Sweden.[1,2]

It was first described by Hunter in 1783 and then revisited by Peacock in 1869. The right ventricular outflow tract is imperforated, which can be either membranous or represented by a longer segment of muscular atresia, characterised by heterogeneity in the right ventricle morphology, its inlet portion, its functional size, and its coronary anatomy.[3] Many of these patients, approximately 31–68%[4] of the cases, have connections between the right ventricle and subepicardial coronary arteries; in some of these patients, this could be of vital importance and may be necessary to sustain adequate myocardial perfusion (right ventricular-dependent coronary circulation or RVDCC). This situation has been described in anywhere from 3 to 34% of patients presenting ventriculocoronary connections.[3]

In 1992, Giglia et al[4] conducted a retrospective study in which they mentioned the hypothesis that the survival of these patients could also depend on the RVDCC; since at first, the surgical management for this cardiopathy had focused primarily on the size of the right ventricle and on the number of right ventricular parts. In their study, 26 of 82 patients with PA-IVS had ventriculocoronary connections. Three groups were made: Group 1: ventriculocoronary connections without stenosis. Group 2: ventriculocoronary connections with stenosis of one coronary artery and Group 3: ventriculocoronary connections with stenosis of two coronary arteries, all groups with different outcomes. This demonstrated that it is very important to correctly evaluate the anatomy and perfusion of the coronary arteries.

The traditional and only way at the moment to evaluate this is by angiography (right ventriculography and coronarography); however, we consider that this method is very subjective because it depends on the interpretation of each evaluator, sometimes incorrectly

evaluated as described by Guleserian et al[5], who had two cases in which the angiography was misinterpreted preoperatively, leading to ischaemia and death.

In addition, there are different criteria taken by the different groups for the definition of RVDCC; one is the Boston group definition, and the other is the definition by the Houston group, who mention the definition as the presence of any coronary-cameral fistula with coronary obstruction, defined as severe stenosis, interruption, or atresia, proximal to the fistula and angiographic evidence of right ventricle perfusion of the myocardium through fistulous communication.[6] These are some of the most accepted definitions; however, we found that each centre has some variations in their criteria and definitions, and some even take to notice the size of the fistula, considering the ones that are too large as RVDCC, overall, as mentioned by Wright et al:[7] difficult to evaluate and to some degree subjective.

There have been efforts to address this situation, as reported by Loomba et al,[8] who described an anterograde coronary perfusion scoring system that is predictive of the need for transplantation and mortality.

The most objective report is the temporary decompression reported by Iwai et al. in Japan;[9] nevertheless, it implies a surgical procedure and cardiopulmonary bypass.

For these reasons, in the interventional cardiology department of the National Institute of Paediatrics of México, a percutaneous and transitory technique was designed with the purpose of evaluating coronary circulation in patients with pulmonary atresia with intact ventricular septum with ventriculocoronary connections in whom angiography could not determine if there was right ventricular-dependent coronary circulation.

Case presentation

A 25-day-old female was referred for cyanosis. At the physical examination, we found a continuous infraclavicular heart murmur and oxygen saturation of 70%. The chest X-ray showed a cardiothoracic ratio of 0.67 with decreased pulmonary vascularity (Fig. 1a). Electrocardiogram showed no sign of injury or ischaemia (Fig. 1b). Echocardiography: pulmonary atresia with intact ventricular septum with confluent pulmonary branches, a patent ductus arteriosus, and evidence of ventriculocoronary connections reported as probable right ventricular-dependent coronary circulation. Tricuspid valve annulus 9 mm (Z Score: –2.04). (Fig. 1c).

Cardiac catheterisation: RV pressure was 170/7-15 mmHg, left ventricle 118/0-12. Right ventriculography showed a small cavity, tripartite, with a small inlet tract but with a normal tricuspid valve annulus. The infundibulum was patent and narrow, the pulmonary valve had membranous atresia, and the pulmonary annulus was 4.5 mm (Z score: –3.08). The ventricular cavity was very hypertrophic with a moderate-sized ventriculocoronary connections with significant systolic retrograde flow in the anterior descendant, even so that we could see the sinus of Valsalva (Fig. 2). There was also a ductus and confluent normal-sized pulmonary branches.

The selective coronarography was not totally conclusive; the anterior descendant had light to moderate stenosis in its middle third and appeared to be thinner from this point (Fig. 3). In addition, there was an absence of contrast in systole, which indicated to-fro flow, and the circumflex artery (CX) showed no alterations (Fig. 4). The right coronary artery and posterior descendant showed anterograde flow.

To evaluate coronary dependency, we decided to perform transtricuspid flow occlusion with a 13.5 mm Z-5 atrial septostomy balloon catheter (Numed® 45 s Street West Cornwall, Ontario K6J1G3 Canada.) placing it in the inlet tract of the right ventricle, and we re-evaluated the coronary flow. The AD flow normalised, and there was no retrograde systolic flow (Figs. 5 and 6). The occlusion was maintained for 10 minutes, and there was no evidence of changes in the electrocardiogram or haemodynamic decompensation. This was considered confirmation of a non-RVDCC, so we decided, in spite of the size of the cavity, to try to decompress the right ventricle and open the pulmonary valve by radiofrequency, but the efforts were unsuccessful; therefore, we performed an atrioseptostomy, and a non-medicated coronary stent was placed in the ductus. Twenty days later, she underwent surgery for right ventricular decompression and outflow tract enlargement with a transannular patch, Blalock–Taussig–Thomas fistula placement with resection of the stent with duct ligation, and closure of the atrial defect with a fenestrated patch.

After surgery, she stayed in the cardiovascular ICU for 19 days with ventilatory assistance for 16 days. Echocardiogram post-surgery showed adequate flow through the right ventricle outflow tract, no evidence of residual stenosis, free pulmonary insufficiency, and preserved systolic function, without tricuspid regurgitation. After 2 months, the right ventricular cavity showed some enlargement. (Fig. 7)

Discussion

To date, the unification of criteria and definition of right ventricular-dependent coronary circulation has not been possible and is based on angiographic evaluation, which we consider is only a subjective method since it depends on the interpretation of each evaluator.

Since the determination of the right ventricular-dependent coronary circulation is crucial to the initial treatment and prognosis of the patients, we think that an objective method is needed more so in those cases where the angiography is not totally clear.

Some of the latest papers report that when the final repair is reached, survival reaches 95% regardless of the type of final procedure. These data, among those reported previously, suggest that coronary anomalies are important but difficult to assess in patients with pulmonary atresia with intact ventricular septum.[7]

It has been reported that performing fewer decompressions of the right ventricle has improved survival, giving the appearance that the increase in univentricular procedures is related to this,[7] but we do not know with certainty if these procedures are being performed in patients who truly have right ventricular-dependent coronary circulation. It will be important to follow these patients further into adulthood to determine if there is in fact an advantage for the patients with 1.5 ventricular over those with the Fontan procedure.[7] In addition, it is extremely rare for a patient with atresia of some of the coronary artery ostia to survive long time with univentricular physiology.[7]

The manoeuvre proposed could help to objectively determine the coronary anatomy and perfusion. It would have to include selective coronarography of both coronary arteries and right ventriculography prior to and during occlusion, complementing electrocardiogram evaluation looking for signs of lesion or ischaemia and the hemodynamic evaluation.

The 13.5-mm septostomy balloon is placed into the right ventricle cavity using the following technique: first, a

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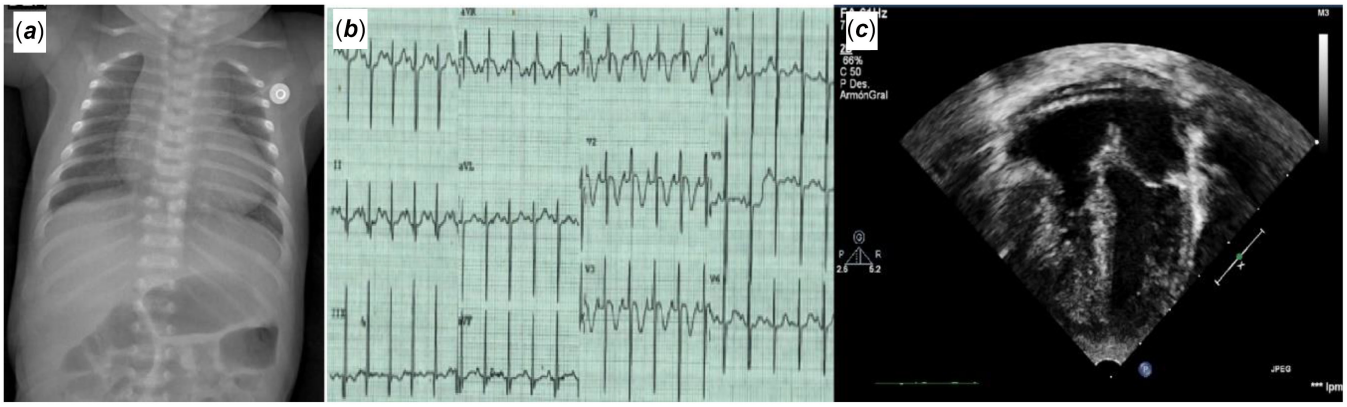

Figure 1. A: Chest X-ray: CTR of 0.67, decreased pulmonary vascularity. B: Electrocardiogram: no sign of injury or ischaemia. C: Echocardiogram: Apical-4-Chamber: Important RV hypertrophy with small cavity.

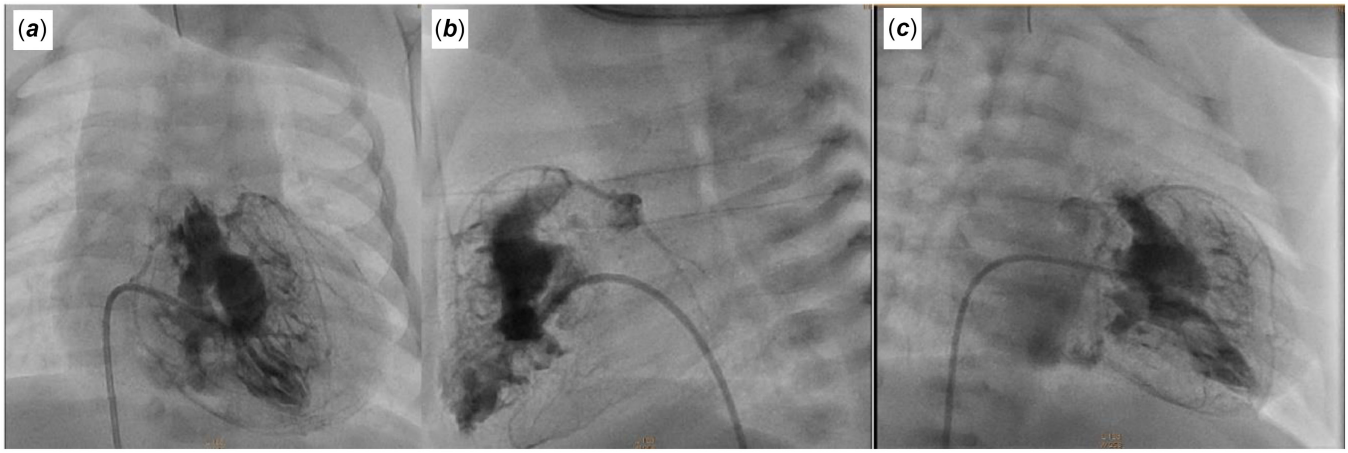

Figure 2. Right ventriculography A: Postero-anterior projection. B: Left lateral (LL) projection. C: Right anterior oblique (RAO) projection. We can see; RV tripartite, small trabecular portion, tricuspid valve annulus, VCCs and pulmonary atresia.

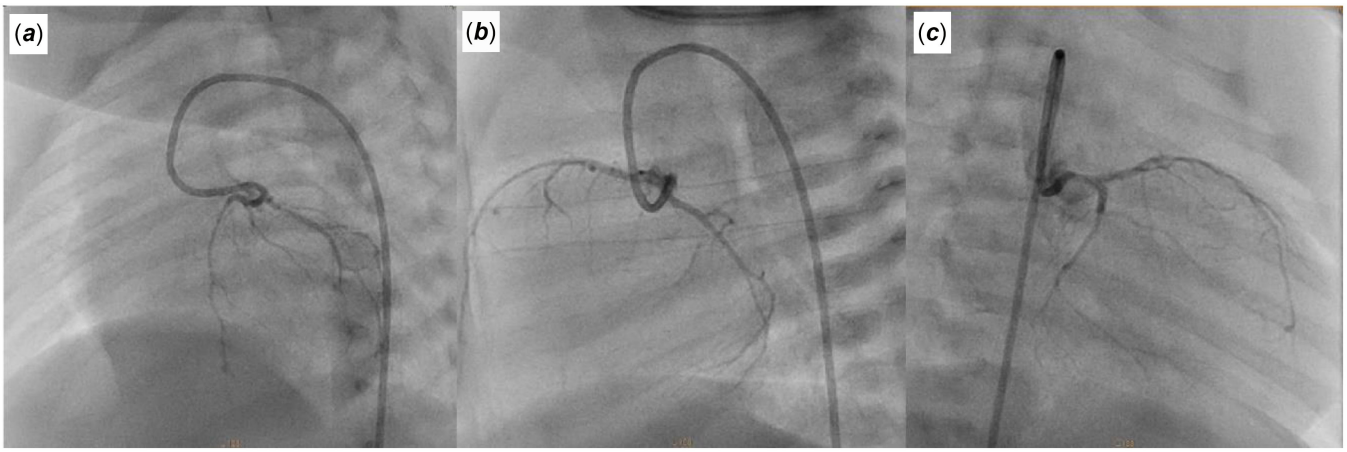

Figure 3. Left coronarography in telediastole. The AD with stenosis in its middle third, from this point we see a caliber decrease which is more evident in A. CX without alterations. A): 4-chamber (4-C) projection, B): LL projection, C): RAO projection.

multipurpose catheter is placed into the right ventricle cavity, then a 0.014" angioplasty wire is placed inside the multipurpose catheter into the right ventricular cavity. Then, the multipurpose catheter is withdrawn, and the balloon is advanced over the wire (be sure not to remove the wire when inserting it into the RV). At this point, it may be difficult to cross the tricuspid annulus, so it is suggested to move the balloon gently and rotate it on its axis and finally, place the balloon occluding the inflow

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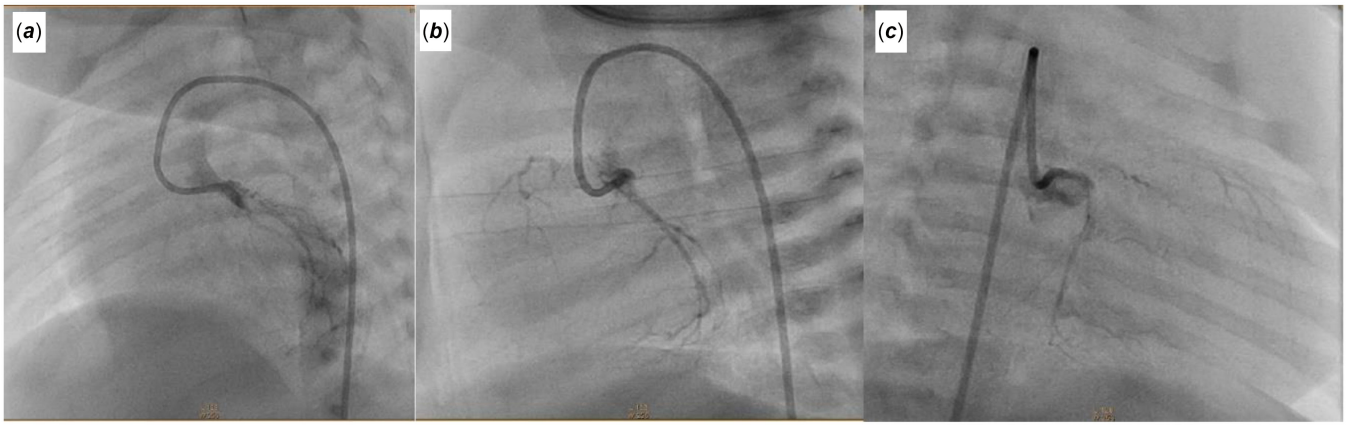

Figure 4. Same left coronarography in telesystole: We can see; absence of contrast in the AD which indicates a to-fro flow. CX without alterations. A): 4-C projection. B): LL projection, C): RAO projection.

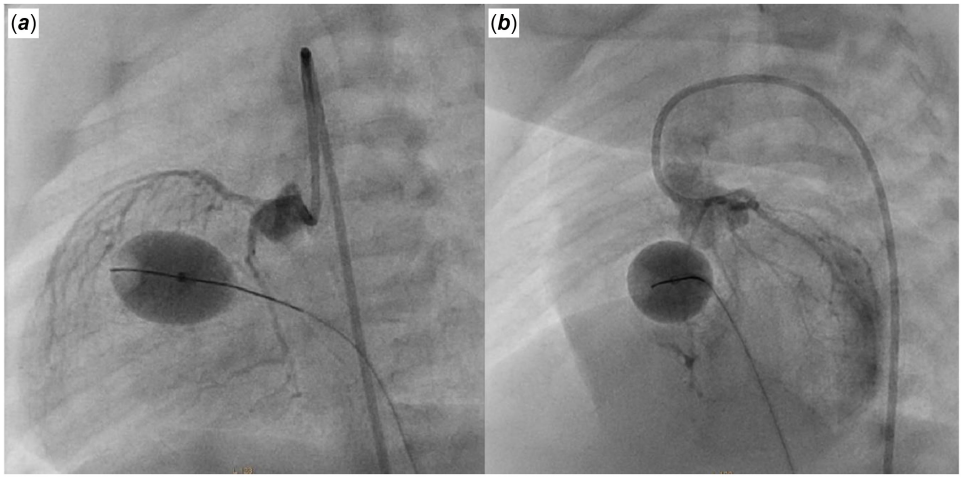

Figure 5. Left coronarography in telediastole with the transient occlusion of the transtricuspid flow. We can see the AD more clearly, and we can even see its diagonal branches better than before. A): LL projection, B): 4-C projection.

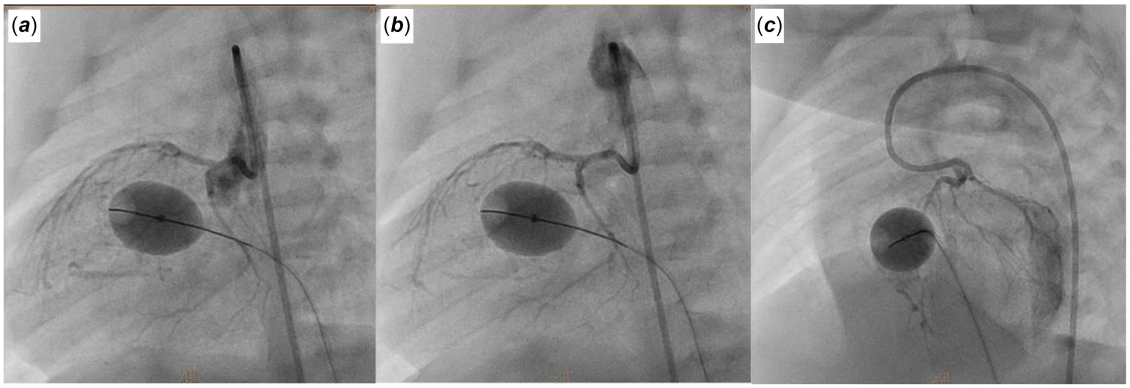

Figure 6. Same left coronarography. A) Protosystole LL Projection. B) Telesystole LL Projection. C) Telesystole 4-C Projection: we can see that the flow is totally anterograde during the occlusion with balloon and there is no evidence of to-fro flow.

portion of the RV. Balloon inflation is up to the millilitres recommended by the manufacturer (2 ml) or before if by echocardiography we observe that the balloon has totally occluded the trans-tricuspid flow, we recommend inflating the balloon as slowly as possible, trying not to get it trapped in the tricuspid valve apparatus.

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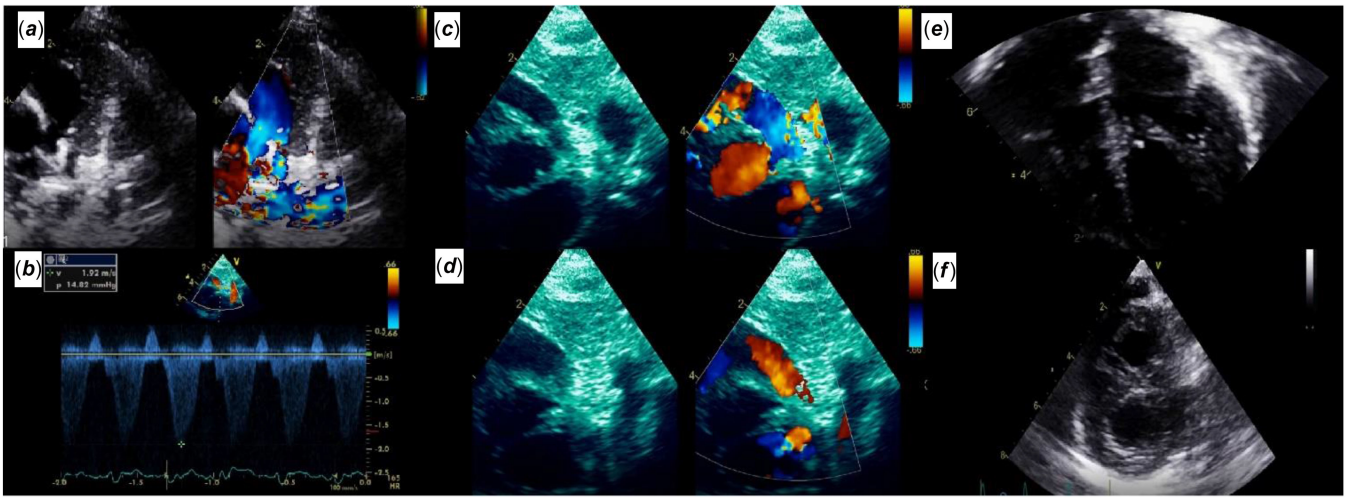

Figure 7. A. Parasternal short axis view (PSAX): Outflow tract with adequate flow. B: Pulsed Doppler; no stenotic gradient. C: PSAX: systole with anterograde flow. D: PSAX: diastole with free pulmonary insufficiency. E: Apical 4-Chamber: RV cavity larger than the beginning. F: PSAX at ventricular level; larger right ventricular cavity.

A possible complication that may concern physicians who perform this manoeuvre is injuring the tricuspid valve; however, this can be avoided by being aware of the deformation of the balloon catheter during inflation by fluoroscopy, and we can also use transthoracic echocardiography to ensure that the balloon is not trapped in the chordae tendineae of the tricuspid valve. In addition, we should not move the balloon during the manoeuvre. In our case, no insufficiency or lesion of the tricuspid valve apparatus was identified during follow-up. Another important point is not to obstruct flow in the interatrial septum with the balloon.

The manoeuvre gives the advantage of creating a temporary "decompression", removing the coronary flow that could be driven from the right ventricle and thus objectively evaluating if the anterograde flow from the aorta is sufficient to provide blood to the myocardium.

We are aware that in cases with right ventricular-dependent coronary circulation, the manoeuvre could lead to lesions, ischaemia, haemodynamic decompensation, or even death. In this situation, the advantage of the manoeuvre is that it can be immediately reversed, without the need for surgery or cardiopulmonary bypass; nevertheless, parents should always be informed of the complexity of the condition itself and the risks and benefits the manoeuvre would offer.

The occlusion could be of greater value in cases with doubt in the angiographic evaluation and the same way in cases such as the second group in Giglia's study[4], which may not meet some of the accepted criteria in the definitions for right ventricular-dependent coronary circulation, but that with the current evaluation could still have a bad outcome. The manoeuvre would give objectivity about the dependency, and in case, the manoeuvre is negative for ischaemia, support for the treating team to perform a decompression in those centres where the results are acceptable. Additionally, the centres that chose not to decompress and offer univentricular palliation would provide objectivity about the possibility that they could have acceptable long-term survival. However, in our institution, we prefer decompression in patients with non-right ventricular-dependent coronary circulation, but this could be controversial.

Conclusions

Finally, we think that with the conventional methods to evaluate the right ventricular-dependent coronary circulation, many patients are considered not candidates for decompression of the right ventricle, without the certainty of being truly dependent, leaving them with a univentricular physiology and depriving them of the opportunity for a biventricular physiology or at least 1.5 ventricular physiology.

We hope that with this new method, which would be complementary in cases where angiography is not totally conclusive, we can give a more accurate diagnosis and determine if right ventricular-dependent coronary circulation exists. In this situation, we can try to refer patients to a cardiac transplant centre or, if it is not available, consider univentricular palliation knowing that it is very probable that it will not reduce the risk of ischaemia and/or death over time.

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Competing interest. None.

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