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Percutaneous obliteration of the right ventricle to avoid coronary damage by sinusoids in patients with pulmonary atresia intact ventricular septum during staged single ventricle palliation

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Abstract

Background and Aims: Suprasystemic pressure waves can damage the coronary arteries resulting in myocardial ischemia and excess early mortality. We aimed to reduce the coronary pressure wave through the sinusoids by abolishing RV volume with percutaneous devices.

Methods and Results: Four patients with PA-IVS and coronary sinusoids from the hypertensive rudimentary RV were evaluated at a median age 26.6 months (range: 2.7–51.7). Right ventricle coronary dependent flow to the left ventricular myocardium was excluded. All four patients had dual perfusion with competitive flow from the RV through the sinusoids to the coronary arteries. Devices used were: Amplatzer vascular plug II of 10–16 mm; 27 coils (diameter 5–15 mm) in the oldest patient. Right ventricular angiography after cavity obliteration showed no more significant coronary perfusion through the sinusoids. There were no complications or deaths. Only minor and transient changes in the levels of troponin were observed. Coronary angiography at pre-Fontan evaluation showed no progress of coronary abnormalities in two patients.

Conclusion: In selected patients with functionally single left ventricle, obliteration of the hypertensive RV cavity by percutaneous devices is safe and abolishes the systolic pressure wave in coronary sinusoids. When performed early, this may halt coronary damage and avoid excess mortality.

KEYWORDS
coil, occlusion, pulmonary atresia intact septum, right ventricle, sinusoids, vascular plug

1 | INTRODUCTION

Pulmonary atresia with intact ventricular septum (PA-IVS) carries a high mortality and morbidity ranging from 1 to 5 year survival of 68–88% and 50–86.5%, respectively. Coronary artery abnormalities such as intramyocardial sinusoids, coronary artery obstruction,

stenosis, or even absence of connection of the coronary arteries from the aorta have been described in up to 60% of these patients; these subgroups have a higher early morbidity and mortality.[1–5] Right ventricle dependent coronary circulation (RVDCC) carries a very high risk peri- and post-operatively (inadequate myocardial perfusion during hypotension or heart massage, inadequate delivery/washout of cardioplegia) and adversely affects outcome long after single ventricular repair, most likely as a result of coronary related issues.[2]

Myocardial sinusoids with ventriculo-coronary connections are frequently observed and may be present in a significant proportion of children with PA-IVS, especially those with suprasystemic right ventricular pressures.[6] The suprasystemic systolic pressures wave induces changes such as dilatation of the sinusoids and intimal fibrodysplasia of the coronary arteries, frequently already during fetal life.[5–8] These may lead to retrograde perfusion of the coronary arteries with desaturated blood. Progressive intimal hyperplasia and medial hypertrophy follow and will progressively obstruct the lumen and reduce the vessel compliance, resulting in incapacity of the normal diastolic epicardial perfusion pressure to open the vessel and thereby requiring the suprasystemic pressure wave to provide flow though this coronary segment.[5–9] With time even the suprasystemic pressure wave will no longer be able to open that coronary segment, resulting in hypoperfusion and myocardial ischemia.

The aim of this intervention was to abolish the right ventricular pressure waves through the sinusoids by percutaneous obliteration of the right ventricular cavity.

2 | PATIENTS AND METHODS

This is a retrospective review of four cases performed from 2004 to 2018 after approval by the UZ Leuven Medical Ethics Committee. All patients had PA-IVS and coronary sinusoids from the hypertensive rudimentary RV. All patients were candidates for Fontan palliation;

the RV was considered too small without growth potential for even a one-and-a-half ventricular repair (median tricuspid valve z-score −3.7 [range −7.3 to 3.5], minute mono- to bipartite cavity with absence of outflow and/or apical parts). Demographic, clinical and imaging data was obtained from patient records.

Procedures were carried out under general anesthesia. Selective left and right coronary as well as right ventricle angiography was conducted in all patients; in this series selective coronary angiography while keeping the tricuspid valve open to confirm adequacy of the normal epicardial coronary perfusion was used in one patient. This was indirectly confirmed, since by splinting the tricuspid valve open during the procedure adequacy of normal epicardial coronary perfusion to the LV myocardium was verified. Right ventricle dependent coronary circulation (RVDCC) to the left ventricle was an exclusion criterion; competitive flow via sinusoids did not result in exclusion, neither the presence of sinusoids perfusing the right ventricular wall only. All four patients had dual perfusion with competitive flow from the RV through the sinusoids into both the right and left coronary arteries. One patient presented with coronary sinusoids connected to the circumflex right coronary artery causing large progressive aneurysmal dilatation up to 18 mm (previously reported).[10] Routine anticoagulation and prophylactic antibiotics were administered per local protocol. We aimed for abolishment of the pressure wave leaving a minimal residual right ventricle stroke volume, due to the theoretical risk of stasis-related thrombosis.

Following the procedure, all patients were monitored for arrhythmia or ischemic electrocardiographic (ECG) changes for 24 hr in the intensive care unit. Follow up cardiac enzymes and echocardiography was performed the day after the procedure. All patients received acetylsalicylic acid 1–2 mg/kg/d (lifelong) and clopidogrel 0.2 mg/kg/d for several months; patient 4 received clopidogrel for 2 years until the aneurysmal dilation of the coronary arteries had regressed to dimensions of less than 6 mm.

TABLE 1 Demographic and procedural data

Patient	1	2	3	4
Operation	Central shunt	BDG	BDG	Fontan
RV occlusion				
Age (mo)	2.7	46.0	7.2	51.7
Weight (kg)	5.5	12.0	6.6	16
RV dimensions				
Echocardiography	8 × 8	20 × 15	26 × 20	ND
Angiography	13.1 × 12.8	19.8 × 14.4	20.1 × 16.9	ND
Device type	AVP	AVP (n = 2)	AVP	Coil (n = 27)
Size (mm)	14	10, 16	12	5–15
Troponin T (ng/L)				
First	0.26	0.06	6.4	
After 24 hr	0.27	0.09	1.3	

Note: All procedures performed via femoral vein, except patient no. 4 who was performed via aneurysmal circumflex coronary artery. Device—AVP, Amplatzer vascular plug II (St Jude, Minnesota); BDG, bidirectional Glenn; ND, not done (coil occlusion).

3 | RESULTS

Procedures were performed at a median age and weight of 26.6 (2.7–51.7) months and 9.3 (5.2–16.0) kg, respectively. Additional data can be viewed in Table 1. The right ventricle was obliterated in one patient through the coronary artery with coils (no access though tricuspid valve after Fontan), in three through the tricuspid valve with vascular plugs (Figures 1–4). In the latter patients the plug was opened in the right ventricle; before release we monitored the ECG for several minutes to detect ischemic changes. In one patients (no. 1), the initial device was retrieved and changed for a larger size; in one patient 2 plugs were used. There was no correlation between angiographic or echocardiographic measurement and final device size (r = .88, p .31). Right ventricular angiography post obliteration demonstrated absence of significant coronary flow via sinusoids.

There were no deaths, nor evidence of ischemia nor arrhythmias. Only minor and transient changes in serum troponin-T levels were observed (Table 1). Predischarge echocardiography showed absence of right ventricle stroke volume and coronary flow via the sinusoids. During follow-up coronary angiography at pre-Fontan evaluation

FIGURE 1 Obliteration RV by Amplatzer vascular plug patient no. 1. Device post release in right ventricle

showed no evidence of disease progression in the coronary arteries of two patients (short follow-up in one). The oldest patient (no. 4) had a coronary angiogram performed 14 years after the procedure that showed no progress of the dystrophic lesions on the left anterior descending coronary artery—asymptomatic stenosis and myocardial bridge, with regression of the dilation of the circumflex artery were observed (Figure 4). Imaging, device placement, release and coronary angiography can be viewed in supplementary materials. Two children are still in follow-up and coronaries have not been evaluated yet. Patients are screened annually for coronary ischemia during treadmill exercise; we anticipate full evaluation once fully grown or if coronary ischemia is suspected.

4 | DISCUSSION

Ventriculo-coronary connections are common in PA-IVS and may play a role in the poor outcome of these patients.[7,8] The abnormal pulsatile perfusion of the coronary arteries with hypoxic desaturated blood at higher than systemic pressure may give rise to proliferative vasculitis.[6–8,11] The coronaropathy is progressive and unpredictable leading to coronary artery obstruction, ischemia and lethal arrhythmias. Apart from this, left ventricular function may also be impaired by the high pressure right ventricle.[12] This contributes to the early high and unpredictable mortality in this specific subgroup of patients with PA-IVS when compared to other patients with single ventricle.

This subgroup with coronary sinusoids has typically small hypoplastic ventricles with a single ventricle strategy being the only option.[13] In patients with coronary artery stenosis or RVDCC to the left ventricular myocardium, decompression of the right ventricle may involve myocardial infarction with fatal outcome—special care should be taken during surgery to maintain coronary perfusion.[8,14] Limited treatment options with varying degrees of success have been reported for this subgroup and consist of right ventricle plication, tricuspid valve narrowing or closure, or ligation of ventriculo-coronary connections; when the LV is significantly compromised, cardiac transplant may be the only option.[15–17]

Our results show that in highly selected patients obliteration of the right ventricle cavity by percutaneous means is feasible. Normal epicardial antegrade coronary arterial flow could be restored and/or

FIGURE 2 Obliteration RV patient no. 2. RV angiogram in (A) and device post release in (B). Note retrograde filling of left coronary artery via sinusoids during RV injection; no coronary artery obstruction seen. Left coronary patency demonstrated in (b) after RV occlusion by a 14 mm AVP II
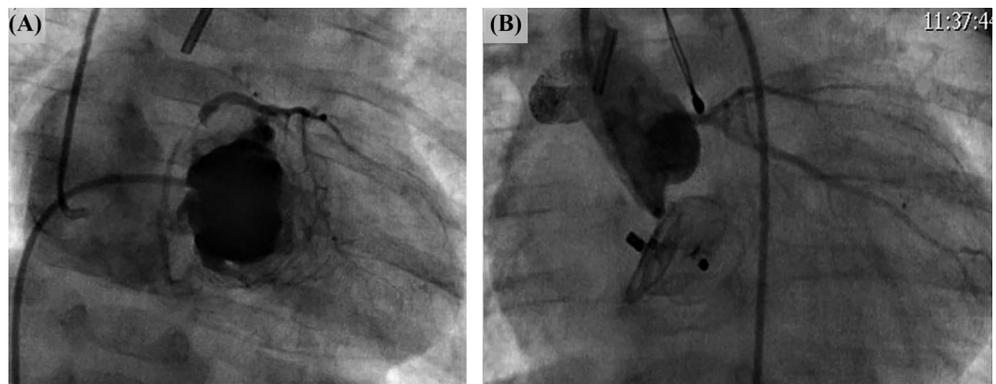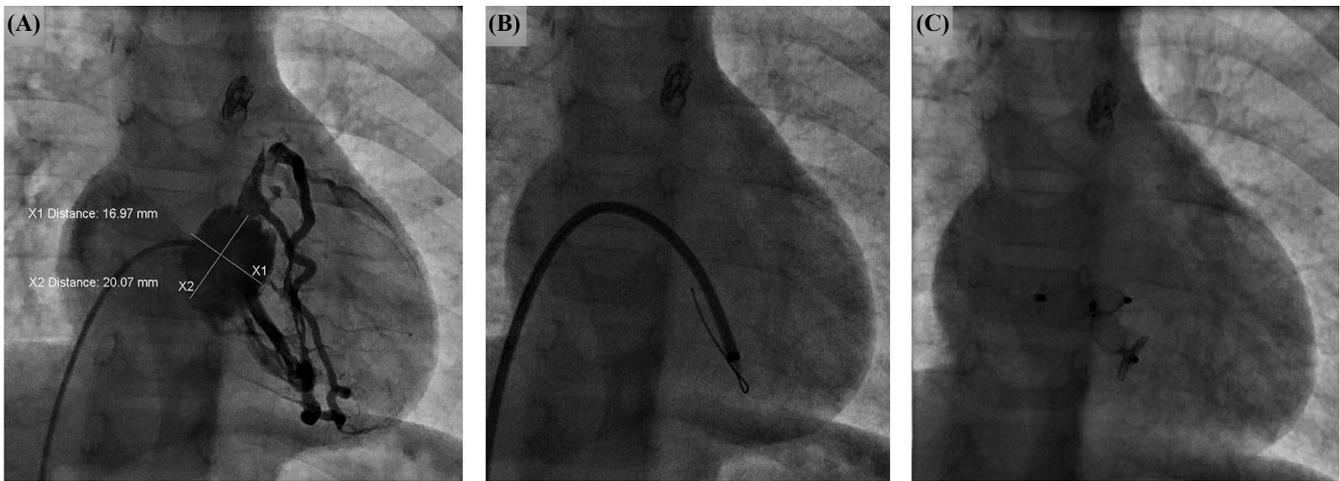
FIGURE 3 Obliteration RV patient no. 3. (A) Angiographic delineation and measurements of RV can be observed. Sinusoids with retrograde filling of left coronary artery can be seen. (B) Delivery sheath position stabilized with 0.014″ coronary guidewire curled in apex of RV. (C) Two AVP II devices (10 and 16 mm) released in the RV. RV, right ventricle; AVP, Amplatzer vascular plug
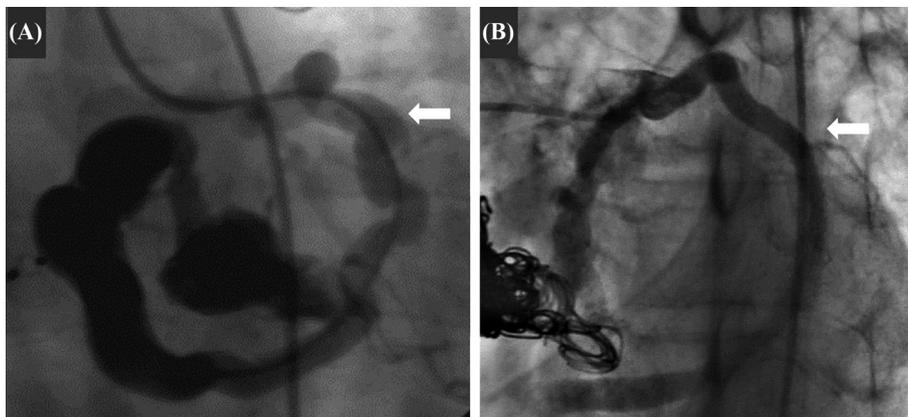

FIGURE 4 Coronary arteries patient no. 4. (A) Coronary arteriogram at before using 27 coils to obliterate the RV. Arrow indicates aneurysmal left anterior descending coronary artery. (B) Left coronary angiogram at age 17 years. The left anterior descending coronary artery (arrow) is dystrophic with asymptomatic stenosis and myocardial bridge

augmented by abolishment of right ventricle stroke volume. Our findings are in agreement with the findings of a surgical ligation series of right ventricle to coronary artery connections: it showed that myocardial oxygenation improved as antegrade coronary artery perfusion was restored.[17] Furthermore, similar to our experience during follow-up, the effect seemed to persist over time. They also experienced no major complications comparable to our limited experience in our series. Based on their results, the authors suggested that ventriculo-coronary connections should be taken down whatever repair strategy is envisaged in the future. It should be emphasized that we only performed the procedure if adequate epicardial coronary perfusion to the left ventricle could be demonstrated. The procedure is obviously not indicated when one and a half ventricle repair is contemplated or significant growth of the right ventricle anticipated. Furthermore, at this stage no clear clinical or functional benefits other than hypothetical benefits of eliminating competitive flow could be demonstrated in our small sample.

The oldest patient was referred late, but it appears that progression of coronary artery disease was halted or at least delayed. Long-term follow-up will establish whether this effect will be observed in the younger patients as well and these patients will be prospectively monitored. The authors speculate that in this select group of patients early obliteration of the right ventricular cavity may halt coronary disease progression and subsequently improve morbidity and mortality. This effect has previously been demonstrated by Yang et al where exclusion of the right ventricle improved left heart function and prevented myocardial ischemia.[18]

From a technical perspective, we found it helpful to use a delivery sheath 1French size larger than recommended for a particular device. This allowed us to place a safety coronary guidewire in the apex of the right ventricle which also provided stability during device placement (Figure 3). Controlled release devices were preferred since it allowed us to test for stability and adequacy of obliteration before release. It is difficult to select the ideal size of device and there was poor correlation with RV dimensions. Generally, we preferred to use the tricuspid valve-apical length to select size of device. However, it is clear that one must be prepared to change device if RV obliteration is inadequate. We did not mind a minimal remaining RV stroke volume, provided the systolic pressure wave was abolished—this also reduces risk of thrombosis.

5 | CONCLUSION

In very selected patients, obliteration of the RV cavity by percutaneous devices is safe and abolishes the pressure wave in coronary sinusoids. When performed early, this may halt coronary damage and avoid excess mortality.

CONFLICT OF INTEREST

The authors have no conflict of interest to declare.

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SUPPORTING INFORMATION

Additional supporting information may be found online in the Supporting Information section at the end of this article.

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