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CASE REPORT
Congenital heart disease

Clinical use of percutaneous mechanical circulatory assistance in a patient with end-stage right-sided heart failure and massive tricuspid insufficiency due to congenital heart disease: first-in-the-world case report

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Background Due to improvement in the management of patients with congenital heart disease (CHD), the likelihood of their survival to adulthood is increasing. A relevant population suffers end-stage right ventricular failure (RVF) in their 3rd–4th decade of life. Hence, heart transplantation is still gold standard of treatment of end-stage heart failure, mechanical circulatory assistance has become a valuable tool in the bridging to heart transplant or definitive therapy. Use of implantable short-term or long-term devices is reported by others. However, within this clinical context, presence of significant tricuspid regurgitation (TR) or CHD is used as exclusion criteria for insertion of a percutaneous right ventricular circulatory support.

Case summary We described a 36-year-old patient diagnosed with Ebstein's anomaly and severe TR who is admitted to hospital due to RVF refractory to standard medical treatment. After case presentation to the heart team, an Impella RP device insertion was scheduled, in spite of the presence of TR or CHD after evaluation of pulmonary valve competency and 3D reconstruction with virtual device implantation. During support, the patient improved clinically and haemodynamically. Due to device displacement to the right ventricle, it was bedside explanted after 30 days of support. After mechanical unloading during 30 days patients' right ventricle recovered partially, permitting patient to improve his functional class.

Discussion Although TR and CHD are exclusion criteria for the implantation of the Impella RP device, we report clinical experience in patient with Ebstein's anomaly and severe TR supported with percutaneous device as bridge to heart transplantation during 30 days.

Keywords Right ventricular failure • Percutaneous mechanical circulatory support • Percutaneous right ventricular assist device • Impella RP • Congenital heart disease • Ebstein's anomaly • Case report

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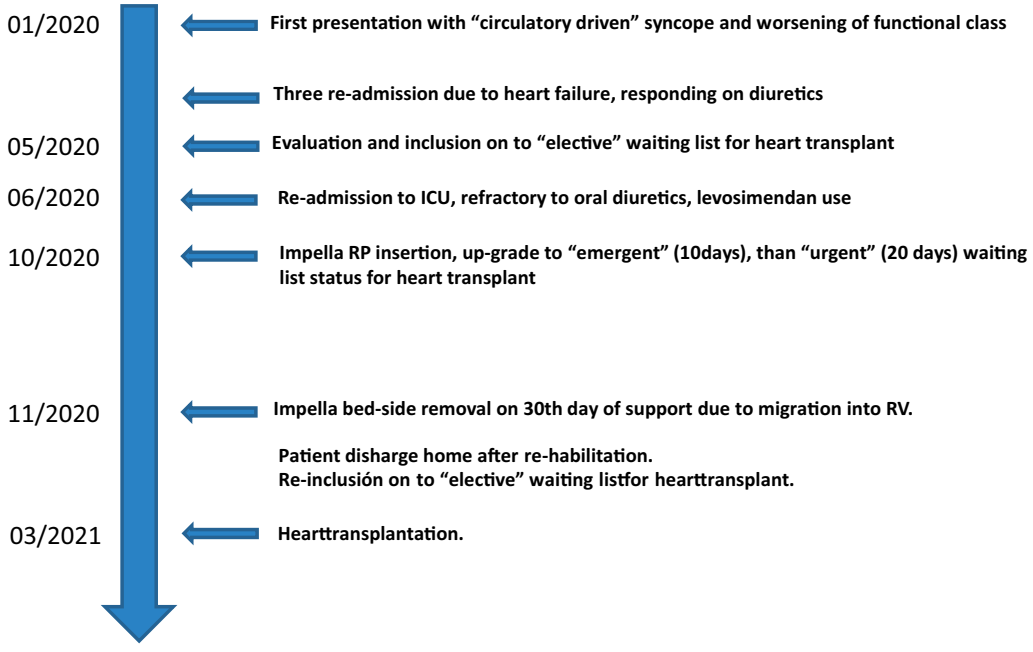
Learning points

- Impella RP implantation may be feasible in patients with congenital heart diseases as long as a meticulous planning is carried out (echocardiography evaluation of competency of pulmonary valve, computed tomography with 3D virtual reconstruction, and simulation of device position).
- Impella RP implantation is feasible in patients with Ebstein's anomaly.
- Impella RP implantation is feasible in patients with severe tricuspid regurgitation.

Introduction

Due to improvement in the management of patients with congenital heart disease (CHD), the likelihood of their survival to adulthood is increasing. A relevant proportion of them presents end-stage right ventricular failure (RVF) in their 3rd or 4th decade of life. Hence, heart transplantation is still gold standard of treatment of end-stage heart failure, mechanical circulatory support (MCS) became a valuable tool in the bridging to heart transplant or definitive therapy. Use of implantable short-term or long-term devices is reported by others. However, within this context, CHD itself and more, presence of significant tricuspid valve insufficiency are used as exclusion criteria for insertion of a percutaneous right ventricular circulatory support devices. We report clinical experience of percutaneous MCS with Impella RP (Abiomed Intl.) used as bridge to heart transplantation in patient with Ebstein's (type C) anomaly and severe tricuspid regurgitation due to end-stage RVF.

Timeline



Case presentation

A 36-year-old male with history of surgically corrected atrial septal defect in his childhood and Ebstein's anomaly (EA) was receiving periodic ambulatory levosimendan infusions due to congestive right-sided heart failure (RHF). Transthoracic echocardiogram (TTE) revealed features of EA Carpentier type D with severe right atrium (RA) dilation (36.5 cm^2), atrialization of right ventricle (atrialized RV 45.3 cm^2 and functional RV 12.4 cm^2) with severely reduced contractility, adherence to the underlying myocardium, and downward displacement of the septal (17 mm/m^2) and posterior leaflets with no outflow tract obstruction and massive tricuspid regurgitation (TR) (*Figure 1*). The left ventricle (LV) was normal, with leftward shift of the septum due to volume overload of RV.

Despite intermittent inotropic therapy, he presented in January 2020 in the emergency department with 'circulatory driven' syncope and worsening of functional class requiring three re-admissions for intravenous diuretic treatment of congestive RHF within 2 months. After evaluation, he was finally listed for heart transplant in elective status.

Later in June, his course was complicated by readmission to the intensive care unit again due to congestive RHF now refractory to medical treatment in INTERMACS profile 2. At admission, the patient was hypotensive with signs of systemic congestion, such as jugular insufficiency, peripheral oedema, and liver congestion. The NT-proBNP plasmatic level was 3500 pg/mL; other laboratory values are on *Table 1*. After presentation of the case to the 'Heart-Team', we evaluated options of mechanical circulatory support (MCS) between

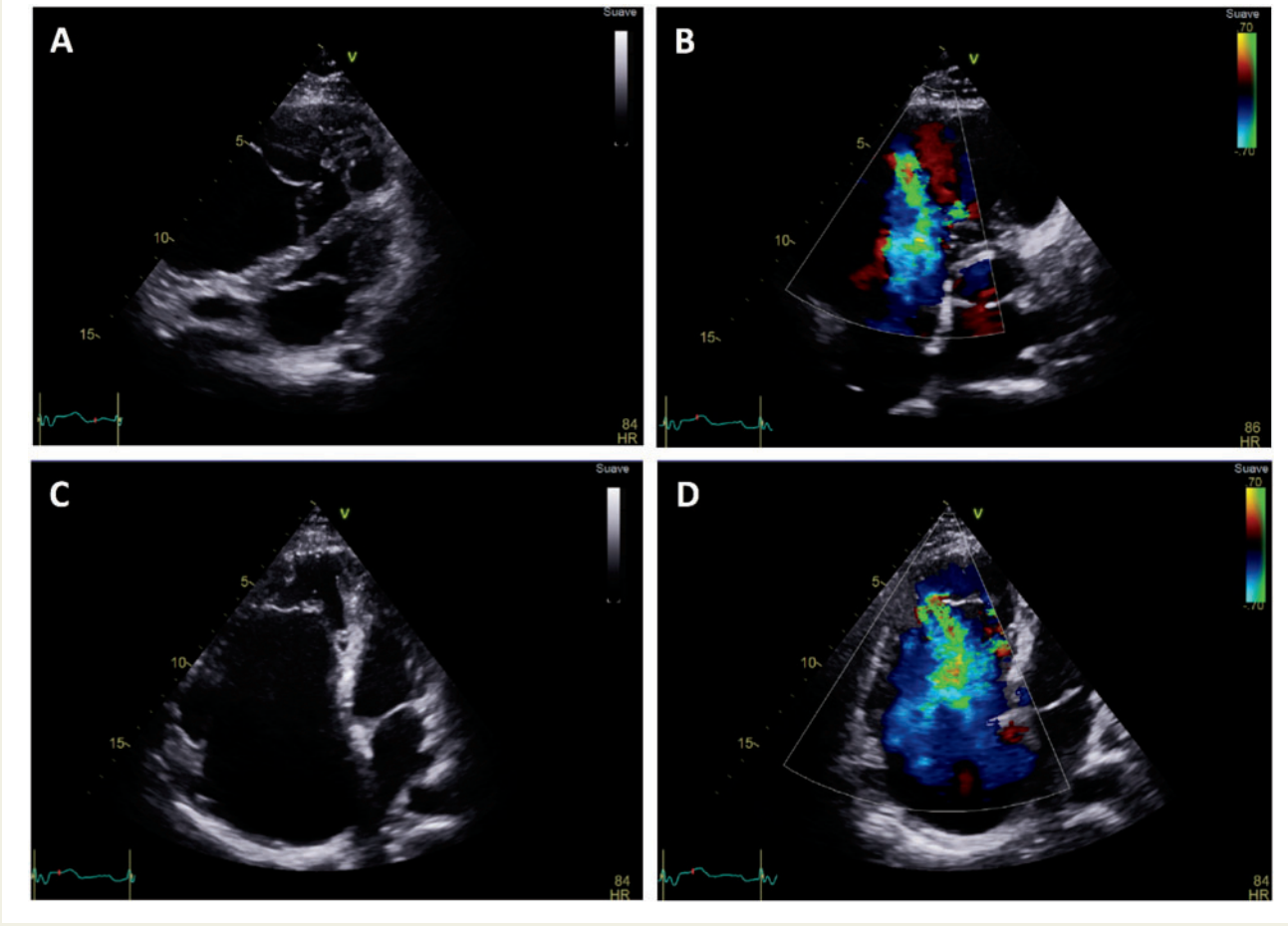


Figure 1 Echocardiographic views before Impella RP insertion. (*A*) Paraesternal short-axis transthoracic echocardiogram view shows a dilated right ventricle with leftward shift of the septum due to volume overload. (*B*) Paraesternal short-axis transthoracic echocardiogram view shows massive tricuspid regurgitation. (*C*) Apical four-chamber transthoracic echocardiogram view shows dilated atrialized RV and apical displacement of the septal and posterior tricuspid leaflets. (*D*) Apical four-chamber transthoracic echocardiogram view shows massive tricuspid regurgitation.

Table 1 Biochemical and haemodynamic measurements within 24 h before and after 15 days of initiation of Impella RP support device

Day	0	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15
Creatinine (mg/dL)	1.4	1.2	0.9	0.85	0.78	0.82	0.76	0.76	0.81	0.69	0.75	0.65	0.71	0.76	0.72	0.66
Bilirrubin (mg/dL)	2.5	2.6	2.84	2.33	1.97	2	1.82	1.53	1.77	1.63	1.43	1.45	1.39	1.52	1.29	1.1
LDH (U/L)	255	459	535	610	674	776	770	796	824	803	769	783	753	750	782	773
Platelets ($\times 10^3$/mL)	240	175	160	141	127	137	158	167	184	174	186	201	208	223	230	235
INR	1.6	1.53	1.42	1.3	1.27	1.18	1.13	1.13	1.14	1.1	1.09	1.1	1.15	1.1	1.13	1.1
UFH (IU/h)		1000	1000	250	350	350	400	400	400	400	400	350	300	300	300	350
APTT (s)	52.3	96.1	81.1	109	50	52	37.6	50.1	55.2	60.4	59.1	64.3	67.2	69.8	66.9	68.3
CVP (mmHg)	19	17	13	11	9	10	8	8	8	9	7	9	9	8	8	7
CI, Fick (L/min/m^2)		1.7	2.2	2.4	2.4	2.9	2.8	2.6	2.8	3	3.1	2.7	2.6	2.7	2.9	3.4
Flow (L/min)		3	3	3	3	3	3	3	3	3.1	3.1	3.1	3.1	3.1	3.1	3

APTT, activated partial thromboplastin time; CI, cardiac index; CO, cardiac output; CVP, central venous pressure; INR, international normalized ratio; LDH, lactate dehydrogenase; UFH, unfractionated heparin; IU, international units.

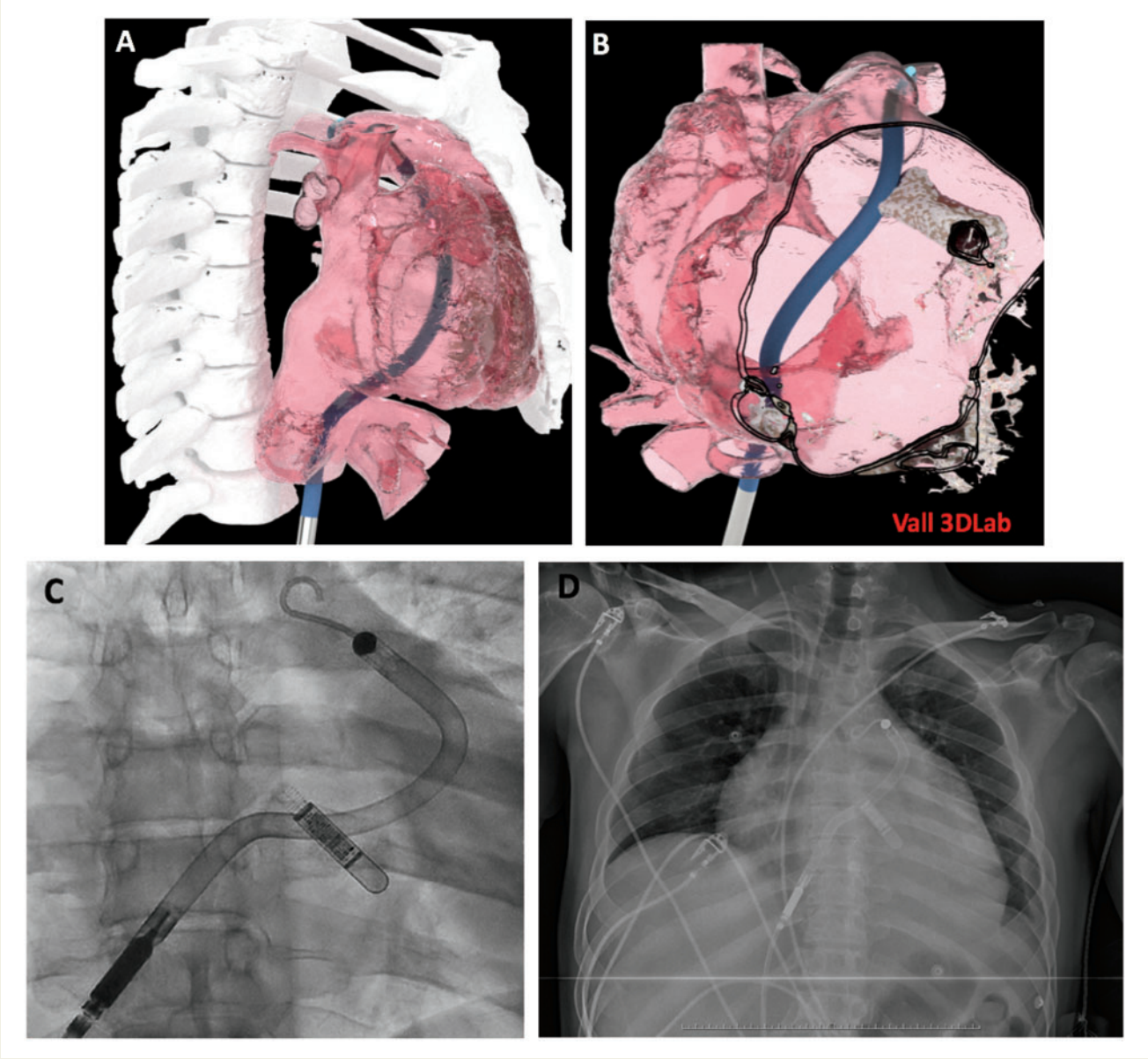


Figure 2 3D planning and reconstruction. (*A*) Lateral view showing the Impella RP inserted in the right ventricle, with the inlet within the right atrium and the outlet within the main PA. (*B*) The same view with the wall of right ventricle removed. Impella RP device *in situ* shown on fluoroscopy (*C*) and chest X-ray (*D*), the first day of insertion. The Impella RP inlet is located in the right atrium, and the outlet is situated in the main PA.

(i) surgical right ventricular assist device (RVAD) placement (re-sternotomy, direct connection of inflow and outflow cannulas to right atrium and Pulmonary Artery (PA), periprocedural bleeding and infection risk) and (ii) less invasive percutaneous MCS with Abiomed Impella RP (Abiomed Intl.) device.

A 3D biomodel of pulmonary artery, right ventricle, subvalvular apparatus, right atrium, and inferior vena cava was segmented from the end-diastolic phase of an electrocardiographically gated computed tomography scan using software Mimics innovation 23 (Materialise, Leuven, Belgium). Further, processing and modelling of Impella RP device was performed with Rhino 7 (Rhino, Robert McNeel and Associates for Windows, Washington DC, USA). A virtual Impella RP device was virtually implanted with outflow point above pulmonary valve. It was possible to place virtually device with location of outflow 27 mm above pulmonary valve in the pulmonary artery trunk. Based on the competence of pulmonary valve and suitability in the 3D reconstruction of the RV (*Figure 2A and B*), we decided to proceed with Impella RP implantation, having surgical RVAD options as back-up strategies in case of procedure failure. Regardless of the challenging anatomy, the device was correctly and uneventfully fluoroscopic-guided deployed, so as to position the inlet within the dilated RA and the outlet within the main PA (*Figure 2C*). Transplant status of the patient was improved based on country regulations to 'high urgent'. The patient experienced significant improvement in haemodynamics after Impella RP insertion (*Table 1*); cardiac index (CI) increased from 1.7 to 3 L/min/m^2 and central venous

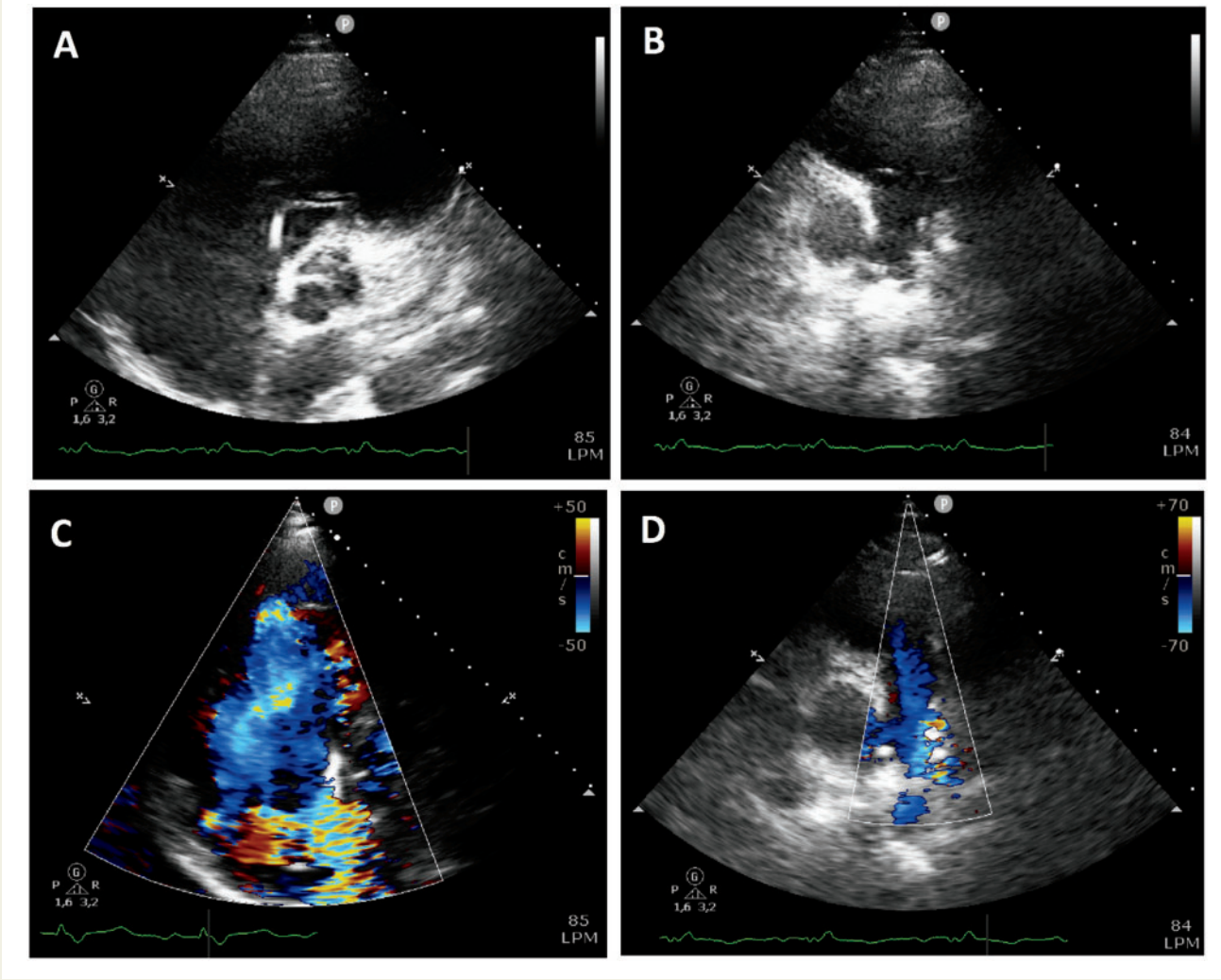


Figure 3 Echocardiographic views after the Impella RP insertion. (*A*) Paraesternal short-axis transthoracic echocardiogram view shows the Impella RP inlet within a severely dilated right atrium. (*B*) Paraesternal short-axis transthoracic echocardiogram view shows the Impella RP outlet in the main pulmonary artery. (*C*) Paraesternal short-axis transthoracic echocardiogram view shows persistent massive tricuspid regurgitation. (*D*) Paraesternal short-axis transthoracic echocardiogram view shows normal function of the pulmonic valve, with no pulmonary regurgitation.

pressure (CVP) decreased from 19 to 8 mmHg after 8 days with Impella RP support. Responsiveness to diuretics and laboratory values of end-organ function also improved (*Table 1*); diuretics were tapered down and serum creatinine and bilirubin steadily decreased (*Table 1*). Impella RP was set to motor level of P6 with approximately 3–3.2 L/min calculated flow [Impella motor purge required continued infusion of unfractionated heparin (UFH) 650 IU/h given through the pump motor]. Repeat TTE showed normal LV with leftward septal shift, dilated RA and RV with the Impella RP at the level of PA, massive TR and competent pulmonary valve with device placed (*Figure 3*). Systemic UFH was continued for the duration of Impella RP support with an activated partial thromboplastin time (aPTT) goal of 60–70 s. Similarly, platelets course and lactate plasma dehydrogenase daily controls were stable (*Table 1*). No thrombotic dysfunction nor device-related haemolysis occurred within 2 weeks of support. There were no patients or device-related complications, including absence of vascular thrombosis, local haemorrhage, vascular injury to the PA or displacement, as monitored by TTE, and chest X-ray within 2 weeks of support. At 15th day of support patient presented drop off in the calculated flow and increase of energy consumption with laboratory signs of haemolysis (platelets 80 $\times$ 10^3/mL and LDH 700 U/L). After 24 h volume loading and increase of anticoagulation to aPTT of 80 s with aim to decrease blood viscosity, we observed full normalization of all parameters (flow, energy consumption, platelets 200 $\times$ 10^3/mL, and LDH 230 U/L), excluding changed position of the pump, displaced more towards right ventricular outflow tract but still having outflow above pulmonary valve.

Based on clinical benefits for the patient, we decided to continue with ‘compassionate use’ of device apart of its certification. After 30 days of support device migrated to RV and after reposition failure, it was bed-side removed without any local complication. Patient kept stable and presented partial recovery of the RV function. Fortunately,

the patient's clinical situation remained stable and he could be still included on the elective transplant waiting list. On 10th December 2020, 3 weeks after Impella RP explant, the proBNP plasmatic level was 200 pg/mL. Finally, after 4 months in clinical stable situation without increased demand of diuretics and free of inotropic support, the patient underwent heart transplantation on 12th March 2021.

Discussion

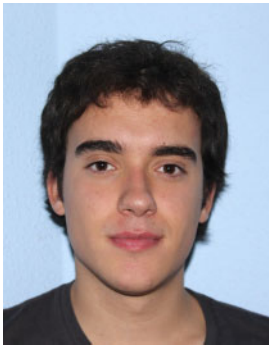
Because of improved surgical techniques and advances in the medical care of patients with congenital heart disease (CHD), the prevalence of CHD in adult patients has grown substantially over the past decade.[1] Congenital heart disease is a widely known aetiology of RHF, which is a dreadful and deleterious clinical problem consistently associated with poor outcomes. The management of RHF is often tricky and it can be even more intricate in the setting of CHD.[2,3] Over the last years, new options have emerged for the treatment of medically refractory RHF,[4] including cutting-edge percutaneous mechanical assist devices, such as the Impella RP (Abiomed, Danvers, MA, USA).

The Impella RP was approved by the Food and Drug Administration (FDA) on September 2017 for providing temporary support in patients with acute RHF. The RECOVER RIGHT study demonstrated the novel Impella RP was safe, easy to deploy, and reliably resulted in immediate haemodynamic improvement with reversal of shock and favourable survival.[5] The effectiveness of the promising Impella RP was later supported by Anderson *et al.*[6] and Cheung *et al.*[7] Ever since, the use of Impella RP has expanded greatly. Notwithstanding, subjects with existing CHD that would preclude the insertion of the device were excluded from these studies and lack of experience in this population is an issue. Likewise, structural tricuspid valve disease or TR represents a contraindication to Impella RP implantation according to the manufacturer. On the other hand, it has been noted recently that TR stands for a warning rather than an absolute contraindication.[8] For this reason, an accurate and individual assessment to evaluate the opportunity to support the failing RV is mandatory.

We have shown that Impella RP implantation is feasible in the setting of EA and that the haemodynamic effects of the device are not affected in the presence of TR as long as the pulmonary valve is competent. According to our experience, despite a backbreaking anatomy with prominent RV dilatation and structural and functional tricuspid valve disease, the Impella RP was correctly inserted under fluoroscopic guidance and resulted in increased CI and decreased CVP, with improvements in laboratory values of systemic end-organ perfusion. Moreover, there were no device-related complications during the certified time. We successfully managed one thrombotic dysfunction at Day 15th and been able to keep the patient stable during all times of MCS waiting for his donor heart. Hence, the presence of donor organ shortage did not allow us to transplant the patient within this time. However, the use of percutaneous MCS led to partial recovery of RV and gave us time to evaluate appropriate surgical strategy (short-term or implantable long-term ventricular assist device placement) in the case of heart failure recurrence.

To the best of our knowledge, this is the first-in-the-world case report of the percutaneous right ventricle mechanical circulatory support in a patient with CHD and massive TR.

Lead author biography



Dr M. García Gómez studied Medicine in Universidad Complutense de Madrid/Hospital General Universitario Gregorio Marañón. Dr García Gómez graduated in 2017 at the Universidad Complutense de Madrid (Spain). He is actually a third-year resident at Department of Cardiology at the Hospital Clínico Universitario (Valladolid). He is contributing to several research projects. His fields of interest are Critical Care Cardiology and Interventional Cardiology.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Slide sets: A fully edited slide set detailing these cases and suitable for local presentation is available online as Supplementary data.

Consent:The authors confirm that written consent for submission and publication of this case report, including images and associated text, has been obtained from the patient in line with COPE guidance.

Conflict of interest: None declared.

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