Hyperacute flash pulmonary oedema after transcatheter pulmonary valve implantation: The melody of an overwhelmed left ventricle

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Received 24 January 2014; received in revised form 21 March 2014; accepted 27 March 2014
Available online 2 May 2014

Summary Percutaneous transcatheter Melody Valve implantation has achieved standard of care for the management of certain patients with right ventricular outflow tract dysfunction. With its widespread use, some rare and potentially fatal complications, such as right ventricular outflow tract rupture and coronary artery compression, have been reported. We report hyperacute flash pulmonary oedema after Melody Valve implantation for the first time in two patients and describe some possible predictors. © 2014 Elsevier Masson SAS. All rights reserved.

KEYWORDS
Transcatheter pulmonary valve; Melody; Congenital heart disease; Pulmonary oedema

Abbreviations: MRI, magnetic resonance imaging; TPV, transcatheter pulmonary valve.
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http://dx.doi.org/10.1016/j.acvd.2014.03.007
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MOTS CLÉS
Implantation percutanée d’une valve pulmonaire ; Melody ; Cardiopathies congénitales ; Cédème pulmonaire

Résumé L’implantation d’une valve Melody par voie percutanée est désormais considérée comme une technique standard dans la prise en charge des patients ayant une dystonie de la valve pulmonaire. Avec son utilisation croissante, des complications rares et potentiellement fatales telles que les ruptures de conduits ou les compressions coronaire commencent à être rapportées. Nous rapportons ici l’apparition d’un œdème du poumon hyperaigu après valvulation pulmonaire chez deux patients et en décrivons les mécanismes possibles.

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Background

Transcatheter pulmonary valve (TPV) replacement was reported in an animal model and then in humans for the first time in the year 2000 [1–3]. Percutaneous TPV implantation (Melody Valve; Medtronic, Minneapolis, MN, USA) is now an established therapy for the management of dysfunctional, incompetent and obstructed right ventricular outflow tract in eligible patients [4–6]. Excellent early, mid-term and even long-term success rates have been reported [7,8]. Coronary artery compression, conduit ruptures and pulmonary artery injury resulting from wire perforation and stent fracture are some procedural complications reported with TPV implantation [9–11]. We report unusual cases of hyperacute flash pulmonary oedema following uncomplicated TPV implantation in patients with standard indications and normal left ventricular systolic function.

Case 1

A 15-year-old male (weight 61 kg; height 165 cm) born with congenital stenotic bicuspid aortic valve underwent surgical aortic valve commissurotomy during the neonatal period. Subsequently, he underwent several palliative procedures, including percutaneous aortic dilation at 9 months and surgical mitral valve annuloplasty with cleft closure at 4 years. At 5 years of age he underwent a third balloon aortic valve dilation.

Owing to relentless progression despite balloon valvuloplasties, with high left ventricular pressures (190/0/18 mmHg), a transaortic valvular gradient of 80 mmHg and moderate insufficiency (aortic annulus 19 mm), he underwent the Ross procedure, with placement of a pulmonary homograft (16 mm), at 9 years of age; he also received optimised medical management with aspirin and beta-blockers.

Six years after the Ross procedure the patient presented with exertion dyspnoea and suprasystemic right ventricular systolic pressures of 125 mmHg, in contrast to a systemic pressure of 110 mmHg. Further evaluation showed severe calcific stenosis of the pulmonary homograft, with a maximum velocity of 6.8 m/s, a well-preserved left ventricular ejection fraction (70%), trivial mitral valve insufficiency, mild aortic valve insufficiency and poor exercise VO2 max. Cardiac magnetic resonance imaging confirmed these findings and cardiac catheterization showed a right ventricular end-diastolic pressure of 13 mmHg and a right ventricular systolic pressure of 110 mmHg, with 75 mmHg in the aorta during systole. Moreover, pulmonary pressure remained normal at 24/12 mmHg, with a mean of 16 mmHg. After ruling out possible coronary artery compression, the patient underwent dilation and stenting of the right ventricular outflow tract with a CPB234 stent mounted on a 20 mm BIB balloon (Numed Inc., Hopkinton, NY, USA). The immediate results were very gratifying, with a reduction in right ventricular to pulmonary artery pressure gradient. After 3 months, the patient underwent staged Melody Valve (22 mm) insertion. The Melody Valve was well deployed, with no complications or coronary artery compression and postprocedure haemodynamics showed a right ventricular pressure of 30/0/10 mmHg and a pulmonary artery pressure of 30/14 mmHg (mean 20 mmHg). The mean pulmonary capillary wedge pressures before and after Melody Valve insertion were 8 and 12 mmHg, respectively. During the 2-hour procedure, the patient received 1000 mL of serum and 80 mL of contrast dye. No volume expansion was needed during the catheterization.

Forty-five minutes after leaving the catheterization lab, while still in the recovery room, the patient developed sudden shortness of breath, coughing out massive amounts of pink frothy sputum, which made intubation very difficult, despite continuous suctioning. At this time, his systemic blood pressure was in the normal range (110/60/75 mmHg). Saturation dropped to 70% and a few litres of pink frothy fluid were suctioned in a brief period of time. The patient was finally intubated and ventilated using jet ventilation. During this time he coded with bradycardic cardiac arrest, but was successfully resuscitated with pressor support. A chest X-ray confirmed signs of severe cardiogenic pulmonary oedema (Fig. 1). After 5 days, the patient improved gradually with liberal diuresis and ventilation with positive end-expiratory pressure; he was extubated, watched for a day in the step down, and was subsequently mobilized and discharged from telemetry on aspirin and diuretics. Transthoracic echocardiography before discharge showed a well-seated transcatheter pulmonary valve with a reduction in right ventricular systolic pressure to 35 mmHg, a right ventricular to pulmonary artery maximum velocity of 2.3 m/s, good left ventricular systolic function and no signs of myocardial ischaemia. Cardiac magnetic resonance imaging (MRI) on subsequent follow-up at 3 years showed normal volumes and function of the right ventricle.
Case 2

A 13-year-old female (weight 26 kg; height 135 cm) who was born with syndromic tetralogy of Fallot associated with Scimitar syndrome underwent complete surgical repair at 8 months of age. A follow-up postoperative transthoracic echocardiogram showed no significant gradient over the right ventricle to pulmonary artery patch. However, there was turbulence over the reconnected right pulmonary vein, with a gradient of 6 mmHg. A pulmonary function test using a lung perfusion scan showed differential perfusion, with right lung perfusion of only 27% compared with 73% in the left lung. As the patient showed only minimal symptoms of shortness of breath, with normal activities and growth, the family opted for close follow-up on medication; she gradually improved with diuretics and oxygen therapy, which was successfully weaned off over time.

At 12 years of age, the patient developed exertion dyspnoea with nocturnal apnoea requiring reintroduction of home oxygen. Cardiac MRI showed severe dilatation of the right ventricle, with an indexed right ventricular end-diastolic volume of 164 mL/m² along with hypoplasia of the right pulmonary artery (Fig. 2). Invasive haemodynamic evaluation showed a mean right atrial pressure of 20 mmHg, a right ventricular end-diastolic pressure of 15 mmHg and a systolic pulmonary artery pressure of 65 mmHg (ventricularized pulmonary artery pressure 53/0/8 mmHg), with 78/48 mmHg (mean 59 mmHg) in the aorta. Angiography showed severe left pulmonary artery dilatation near its origin (25 mm), with massive pulmonary regurgitation. There was no forward flow in the right pulmonary vein; instead there was forward flow of blood from the main pulmonary artery to the right pulmonary artery during systole followed by backward flow of blood from the right to left pulmonary artery (and the right ventricle) and thus into the left atrium via the left pulmonary veins.

A team decision was made to replace the pulmonary valve by the transcatheter technique using the jailing technique [12]. At first, stenting of the right pulmonary artery was done with an intrastent 36 mm proximally and an intrastent 26 mm distally (EV3, Max LD). The procedure was staged to allow for seeding of the stents and thus reduce the risk of dislodgement. After 1 month, the patient underwent successful percutaneous Melody Valve insertion after receiving preprocedure diuretic therapy for 48 hours.

Cardiac catheterization data at the end of the procedure was as follows: mean right atrial pressure 12 mmHg; left pulmonary artery pressure 50/30 mmHg (mean 38 mmHg). The mean pulmonary capillary wedge pressures before and after Melody Valve insertion were 14 and 14 mmHg, respectively. During the 3.5-hour stay in the catheterization laboratory, the patient received 220 mL of fluid and 250 mL of contrast dye in total. No volume expansion was needed during the catheterization. In the recovery room, the patient received an additional 150 mL of fluid in 90 minutes.

Soon after the procedure, while still in the recovery unit, the patient developed acute respiratory distress with pulmonary oedema requiring high-pressure continuous positive airway pressure ventilation for 5 days. Her blood pressure rose from 119/69/85 mmHg to 135/84/95 mmHg, 1 hour after the procedure, but decreased to the normal range without any medication thereafter. A transthoracic echocardiogram showed good results, with a well-seated valve at the appropriate location, infrasystemic pulmonary artery pressures and good systolic function of the left ventricle, with the shortening fraction at 30%. Five days later, the patient was discharged home on aspirin and spironolactone. On subsequent follow-up at 2 years, the patient showed significant clinical improvement, with restoration of right ventricular volumes and size to near normal by cardiac MRI (Fig. 2).

Discussion

The two cases described in this report illustrate, for the first time, a rare, transient and reversible phenomenon of TPV implantation with hyperacute presentation. Taggart et al. reported congestive heart failure after Melody Valve implantation in a patient with an operated left ventricle with left ventricular to aortic graft and subacute onset of congestive symptoms 3 weeks after TPV implantation [13]. The two patients we have described had a catastrophic and near fatal presentation immediately after TPV implantation.

We hypothesize that TPV implantation in an unprepared, chronically underfilled left ventricle with a reverse Bernheim effect and in the absence of a shunt lesion may cause a sudden increase in cardiac output, with features of hyperacute— or rather catastrophic—diastolic heart failure. In the presence of severe pulmonary insufficiency, pulmonary
venous forward flow (and thus left ventricular filling) is considerably reduced (Fig. 3). With TPV, there is immediate augmentation of pulmonary venous flow, draining to the innocent, unprepared left ventricle, which may fail to accommodate all the returns. With no additional room, volume increase occurs at the expense of increase in pressure, which is reflected to the left atrium and thereby to the lungs, causing acute pulmonary oedema.

The reverse Bernheim effect and pericardial constraint causing anatomical and functional ventricular interdependence also deserve attention. Owing to sharing of transverse and oblique muscle fibres from the right ventricle to the left ventricle, a change in right ventricular geometry causes a change in the strain and rotation of the left ventricle [14]. With long-standing pulmonary insufficiency and right ventricular enlargement, the interventricular septum bulges to the left ventricular inflow and, furthermore, the stretched pericardium allows right ventricular enlargement by compromising left ventricular volumes (pericardial constraint). This may be particularly true in patients with frequent surgeries and pericardial thickening, although none of the patients described had any signs of constriction.

Fortunately, acute diastolic heart failure or, more aptly, overwhelmed left ventricle with flash pulmonary oedema, is a transient phenomenon that gets rectified once the left

Figure 2. Magnetic resonance imaging of Case 2: (A) before Melody Valve implantation; and (B) after Melody Valve implantation; see the extensive reduction of right ventricular outflow tract dimension.

Figure 3. Hypothesis for hyperacute flash pulmonary oedema immediately after Melody Valve implantation. iv: intravenous; LV: left ventricle; PEEP: positive end-expiratory pressure; RV: right ventricle; RVOT: right ventricular outflow tract.
ventricle gets self-trained and is ready for the challenge. A similar physiological increase in left ventricle preload is reported after secundum atrial septal defect closure in elderly patients with a stiff ventricle [15]. Invasive and non-invasive haemodynamic measurements during and after TPV implantation suggest favourable right ventricular loading conditions and improvement in left ventricular filling properties after TPV implantation over time [16–18]. In view of the two-stage procedure, we believe that the relief of the pulmonary regurgitation alone, with significant increase in antegrade pulmonary blood flow, may have been an important contributor to overperfusion pulmonary oedema.

The systemic pressure quite likely rose transiently after anaesthesia. The temporal profile of pulmonary oedema and high systemic pressure did not match; pulmonary oedema persisted despite systemic pressures not higher than before the intervention, suggesting no relation.

One may question why not all patients who qualify for the Melody Valve develop pulmonary oedema after TPV implantation; we still do not have a clear answer. In Case 1, pulmonary insufficiency developed after the surgical Ross procedure, but there are other concomitant factors that might suggest long-term left ventricular diastolic dysfunction, such as left ventricular outflow tract obstruction associated with left-sided anomalies and multiple operations. The clinical course of pulmonary oedema in Case 1 was quite aggressive, as one may expect from the hypothesis. In Case 2, with tetralogy of Fallot and Scimitar syndrome, the clinical course of pulmonary oedema was less aggressive, which may be due to the preprocedure priming with diuretics after lessons learned from Case 1. The overlapping features in both cases included left-sided anomalies, left-sided operation, age <15 years and severely elevated systolic right ventricular pressure (from right ventricular outflow tract obstruction as in Case 1 or from elevated pulmonary artery pressure as in Case 2), along with pulmonary insufficiency. The degrees of right ventricular pressure and dysfunction are also important to mention as they were present in both patients.

As a standard protocol, all patients receiving a Melody Valve at our institution go through the same selection process, which includes a transthoracic echocardiogram and MRI in the absence of contraindications. Serial transthoracic echocardiograms showed normal left ventricular diastology and normal left ventricular systolic functions. However, for obvious reasons, these could not be studied during resuscitation. We thought of contrast overload as a possible factor, but the total contrast use was 80 mL in Case 1 and 250 mL in Case 2. The total amounts of fluid infusion in Cases 1 and 2 were 1000 and 220 mL, respectively. At no point in time was volume expansion required in any patient during the procedure.

We observed this rare complication in only two patients out of more than 200 TPV implantation procedures. Having learned from this experience, we now evaluate multiple diastolic variables to identify major predictors (if any). We recommend detailed non-invasive study of left ventricular diastology before TPV implantation, along with planned preload reduction therapy in high-risk selected patients. We also recommend judicious use of contrast to reduce dye load and thus the need for high-volume intravenous fluids for hydration during intervention and in the immediate postoperative period. Once discharged, we encourage exercise rehabilitation to allow for further left ventricular training at higher cardiac outputs for such patients.

Conclusion

Hyperacute flash pulmonary oedema following TPV implantation is a rare and transient condition. We hypothesize that it is due to a sudden increase in cardiac output that overwhelms the left ventricle, which self-trains over time to handle the load. If unanticipated, this could be a stormy and potentially fatal condition. Nevertheless, it gives tremendous potential for one to study the dynamic diastology of the left ventricle after TPV implantation. Left-sided lesions with left-sided surgery, severely elevated systolic right ventricular pressure with pulmonary insufficiency and an unplanned procedure with no priming diuretics seem to be positive predictors based on our report. However, a multicentre study on the impact of TPV implantation on left ventricular diastology may improve our understanding.

Disclosure of interest

Younes Boudjemline acts as a proctor for Medtronic Inc. Other authors: none.

Acknowledgments

The authors thank Dr. R. Raimondi for reviewing the MRI data.

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