Clinical research
Arrhythmia/electrophysiology

Pulmonary vein isolation using an occluding cryoballoon for circumferential ablation: feasibility, complications, and short-term outcome

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Aims To assess safety, feasibility and short term outcome of pulmonary vein (PV) isolation in paroxysmal atrial fibrillation (AF) with a cryoballoon.

Methods We consecutively treated 57 patients with a double lumen 23 or 28 mm cryoballoon. The acute results, complications and follow-up over the first three months were analysed, using a comprehensive and intensive follow-up period.

Results During 57 procedures, 185 of 220 targeted PV’s were successfully isolated using the cryoballoon (84%) (balloon group, 33 patients). In 33 veins (15%) an additional segmental isolation (hybrid group, 24 patients) was necessary with a standard cryocatheter to achieve isolation. The average procedure times were respectively 211 ± 108 and 261 ± 83 minutes (NS), the average fluoroscopy times 52 ± 36 and 66 ± 33 minutes (NS). The number of balloon applications did not differ between both groups: respectively a median 9 (4–18) and 10 (5–17) (NS). We observed four phrenic nerve paralysis after ablation of the right superior PV: two resolved immediately after cessation of the cryoenergy, one recovered after 3 months, one persisted up to 6 months. A daily transtelephonic rhythm recording showed a significant drop in mean AF burden from 24% to 10%, 8% and 5% during the three consecutive months of follow-up (p < 0.01 versus baseline). No differences were observed between the treatment groups.

Conclusions Balloon cryoablation of the pulmonary veins with additional segmental isolation if necessary, is a good approach for patients presenting with paroxysmal AF, showing a significant reduction in AF burden after a single procedure. The major complication seems to be phrenic nerve paralysis after ablation of the right superior PV, but this is potentially reversible over several months.

KEYWORDS Clinical electrophysiology; Atrial fibrillation; Pulmonary vein isolation; Catheter ablation; Cryothermal; Balloon

Introduction
Pulmonary vein isolation (PVI), either segmental or circumferential, has become an important treatment of patients with atrial fibrillation (AF). Reports have been published that show up to 85% freedom of paroxysmal AF during long-term follow-up. A large number of different approaches and techniques exist. The procedure remains technically challenging with a significant number of complications such as thrombo-embolism, PV stenosis, atrio-esophageal fistulae, and left atrial (LA) flutter. Innovative new technologies are being developed to make isolation safer and easier.

Cryoablation has been promising because of low thrombogenicity and absence of PV stenosis, but the longer procedure and fluoroscopy times have limited this approach to segmental isolation. Recently, the development of balloon technology has opened the way for several novel approaches to isolation with new energy types (ultrasound, focused ultrasound, laser and cryotherapy). Cryoballoon experiments have been shown to be feasible and safe in animals. Our aim was to publish our initial experience in humans, not only to describe the procedural success rate but also to assess short-term efficacy in treatment of paroxysmal AF. We have adopted an intensive follow-up method, using daily event monitoring.

Methods
Patients
Patients with documented paroxysmal AF at two or more occasions were accepted as candidates. Exclusion criteria were LA dimension >50 mm measured in the parasternal long axis, as assessed with transthoracic echocardiography, valvular heart disease, and advanced age.
Procedure

All patients were treated with a double lumen cryoballoon (Arctic front, Cryocath, Montreal, Quebec, Canada; Figure 1). Both femoral veins and in some cases the left subclavian vein were used for venous access. A 10 F, intracardiac echocardiography (ICE) catheter (Flexview, EPMed) was introduced through the left femoral vein and positioned in the right atrium. A decapolar catheter was placed in the coronary sinus. After the first 10 cases, a double transseptal puncture was replaced by a single transseptal approach using a Brockenbrough needle, guided by both ICE and fluoroscopy. ICE was also used to ensure a posterior transseptal approach. A circular mapping catheter was advanced and positioned in the antrum of each PV to record the presence of PV potentials. After registration, the sheath was exchanged for a 14 F steerable sheath. The mapping catheter was exchanged for a 23 or 28 mm, 12 F balloon catheter, positioned over an exchange wire to occlude the ostium of each PV (Figure 2). Cryoenergy was given for 5 min per application. The applications per vein were directed towards the major side branches. Before targeting the right superior PV (RSPV), a quadrupolar catheter was positioned in the superior caval vein for continuous phrenic nerve stimulation during cryoapplication. At loss of capture, the ablation was instantaneously terminated. After targeting all PVs, the cryocatheter was exchanged for the circular mapping catheter to check for remaining electrical activity. If this registration showed persistence of the PV potentials, the cryoballoon was introduced again, trying to maximize wall contact at the location of the remaining potentials (as guided by the circular catheter, ICE, and fluoroscopy). If after this second ablation attempt the activity remained present, a conventional cryocatheter (Freezor Max, Cryocath) was used to perform a segmental isolation through the same transseptal puncture. If isolation could be achieved with the balloon, the patient was categorized as ‘balloon’, and if additional segmental isolation had to be performed, he was categorized as ‘hybrid’. These categories were included in further analysis. The day after the procedure, a transthoracic echocardiogram was made to exclude pericardial effusion and a chest X-ray to exclude pneumothorax and other thoracic complications.

All patients were treated with oral anticoagulation for at least 1 month before the procedure, aiming at an INR of 2.5–3.5. Two days before the procedure, patients were admitted and the oral anticoagulants were replaced by unfractionated heparin, aiming at a three times normal aPTT ratio. Two hours before the ablation, heparin was stopped. After venous puncture, and before transseptal puncture a 5000 IU heparin bolus was given. After transseptal puncture another 5000 IU heparin was given and a continuous titrated infusion of heparin was started. During the procedure, the activated clotting time was monitored every 30 min and kept above 350 s. After the procedure, the patients were treated with heparin and oral anticoagulants were restarted.

Anti-arrhythmic drug treatment after ablation

During the 3-month follow-up period after ablation, all patients were continued on the anti-arrhythmic medication they were taking before the ablation.

Follow-up method

Before ablation, patients were instructed to use an event recorder and to transmit daily at least one transtelephonic ECG strip at a fixed hour, and when symptoms were present. This was started 1 month before ablation and continued for 3 months afterwards. The heart rhythm on the ECG strips was coded as sinus rhythm, atrial flutter, atrial tachycardia, or AF. Atrial and ventricular premature beats and sinus tachycardia were coded, but are not reported, as they were infrequent. The heart rate during episodes of sinus rhythm was measured. Transmissions were coded as symptomatic or asymptomatic. The AF burden was defined as the percentage of days on which an AF episode was transmitted. Multislice CT scans were made before and at 3 months after ablation to evaluate the possible occurrence of PV stenosis as described before. PV stenosis was defined as a reduction of the diameter of more than 25%. Patients were seen at the outpatient clinic after 3 months.

Figure 1 Distal end of a 23 mm, 10.5 F, double lumen cryoballoon catheter (Cryocath) after inflation.

Figure 2 X-ray while contrast fluid is injected, showing occlusion of the right superior pulmonary vein with the cryoballoon catheter. Quadrupolar pacing catheter in the superior vena caval-right atrial junction. Multipolar mapping catheter in the coronary sinus. In the mid-right-atrium, an intracardiac echo catheter is visible.

Statistical analysis

Continuous variables are expressed as the mean value ± SD and were compared with the t-test. A $\chi^2$ test was used for categorical variables. Non-parametric tests were used when appropriate. The learning curve was analysed in blocks of 10 patients. Data pertaining to number of applications, procedure, and fluoroscopy times were documented for each subgroup. Statistical analysis of the hypothesis that procedure time and fluoroscopy times varied significantly between groups 1–5 was performed using the one-way analysis of variance.

Results

Patient data

A total number of 57 patients (44 male, 13 female), mean age 55 ± 9 years, underwent PVI with a cryoballoon. The mean LA dimension was 43 ± 7 mm. The mean LA volume
and in 2/57 patients (4%), three veins were targeted. In 5/57 patients (9%), two veins were targeted, with a median of 2 (1–7) applications to achieve complete electrical isolation. Remaining 33 veins (15%) were successfully isolated. When using both balloons (seven cases, 28 veins), successful balloon isolation was achieved in four procedures (57%) and 16 veins (57%). The remaining required additional use of a conventional cryocatheter.

The average procedure time was 232 ± 100 min and the average fluoroscopy time 58 ± 35 min for the entire population (Table 2). Adding an additional segmental isolation did not significantly prolong fluoroscopy or procedure times.

Procedures

During 57 procedures, registrations were made in 228 PVs, of which 220 showed PV potentials. All veins with potentials were targeted (left superior pulmonary vein (LSPV): n = 57, left inferior pulmonary vein (LIPV): n = 53, RSPV: n = 54, right inferior pulmonary vein (RIPV): n = 54]. and 218 were successfully isolated. The median number of balloon applications per vein was 2 (range 1–10), LSPV 3 (1–7), LIPV 3 (1–10), RSPV 2 (1–8), and RIPV 2 (1–6). A median of 9 (range 4–18) applications were given during the entire procedure. Of the 220 veins, 185 veins could be isolated using only the balloon (84%) in 32 patients (54%). There were no differences between the different veins: LSPV, 48 (84%), LIPV 43 (81%), RSPV 46 (82%), RIPV 48 (89%) (NS). In the remaining 33 veins (15%), a standard cryocatheter was used to perform additional segmental ablation with a median of 2 (1–7) applications to achieve complete electrical isolation (hybrid approach). The number of balloon applications did not significantly differ from the balloon group: 10 (5–17) (NS). In 17/57 patients (30%), only one vein had to be targeted with a median number of 2 (1–5) applications; in 5/57 patients (9%), two veins were targeted, and in 2/57 patients (4%), three veins were targeted.

When using a 23 mm cryoballoon (18 cases, 70 veins), 14 procedures (77%) and 52 veins (74%) were successful with just the balloon. With the 28 mm balloon (32 cases, 122 veins), 15 cases (47%) and 57 veins (47%) were successfully isolated. When using both balloons (seven cases, 28 veins), successful balloon isolation was achieved in four procedures (57%) and 16 veins (57%). The remaining required additional use of a conventional cryocatheter.

Complications

In this series, two severe complications required prolonged hospitalization. One patient experienced a left-sided hae-morrhax after haemorrhage due to puncture of the left subclavian vein. Another required surgical drainage of a pericardial effusion due to perforation of the left auriculum. In two cases, there were four cases of right phrenic nerve paralysis after application in the RSPV. At loss of phrenic nerve capture, ablation was immediately stopped. Two cases recovered after cessation of cryotherapy within the procedure. One recovered after 3 months (as documented with fluoroscopic evaluation of the diaphragm movement). One persisted for more than 6 months. The persistent phrenic nerve paralysis occurred during ablation with a 28 mm balloon, the others when ablating with a 23 mm balloon deep inside the RSPV.

One patient developed sustained atypical atrial flutter at 3 months after ablation. It responded to flecainide therapy and did not recur. Two patients complained of haemoptysis within the first month after the procedure.

### Table 2 Procedure and fluoroscopy times

<table>
<thead>
<tr>
<th>Balloon size</th>
<th>Balloon isolation</th>
<th>Hybrid isolation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Procedure (P = 0.06)</td>
<td>Rx (NS)</td>
<td>Veins</td>
</tr>
<tr>
<td>23 mm</td>
<td>162 ± 69</td>
<td>35 ± 30</td>
</tr>
<tr>
<td>28 mm</td>
<td>224 ± 130</td>
<td>64 ± 38</td>
</tr>
<tr>
<td>23 ± 28 mm</td>
<td>258 ± 67</td>
<td>68 ± 26</td>
</tr>
<tr>
<td>Total</td>
<td>211 ± 108</td>
<td>52 ± 36</td>
</tr>
</tbody>
</table>

F: frontal plane diameter; LIPV, left inferior pulmonary vein; LSPV, left superior pulmonary vein; NS, not significant; P-value, baseline vs. 3 months; RIPV, right inferior pulmonary vein; RSPV, right superior pulmonary vein; SD, standard deviation; T, transverse plane diameter.
Analysis of the learning curve

When we compared the 50 last procedures in groups of 10, it was evident that procedure and fluoroscopy times fell significantly (Figure 3). Procedure time fell from $375 \pm 87$ to $137 \pm 40$ min, fluoroscopy time from $105 \pm 30$ to $21 \pm 7$ min (both $P < 0.01$). Furthermore, the number of balloon applications decreased significantly (Table 3), whereas the proportion of patients requiring an additional segmental approach remained similar.

Event monitoring

All 57 patients who completed the follow-up, submitted daily rhythm strips 1 month before and 3 months after the intervention. They sent in additional strips at the time of complaints (Table 4). Before ablation, 981 ECG rhythm strips were available for analysis. The average heart rate in sinus rhythm was $65 \pm 9$ b.p.m. in the month before ablation. In the rhythm strips, AF was recorded 246 times (25%), yielding a median AF burden of 14%. After ablation, 3361 rhythm strips were transmitted and analysed. The average heart rate in sinus rhythm during the first, second, and third months was $68 \pm 8, 68 \pm 9$ and $66 \pm 8$ b.p.m., respectively. The mean heart rate differed significantly from baseline during follow-up ($P < 0.01$ for the first 2 months). Results of rhythm recordings (number of strips showing AF and calculated AF burden) are presented in Table 4. Overall, there was a significant reduction in AF burden from the first month on, persisting during the follow-up period. When comparing patients who experienced recurrence with the ones showing no recurrence, there was no significant difference in baseline burden: $0.29 \pm 0.31$ and $0.21 \pm 0.32$ (NS), respectively. Comparing the patients who underwent hybrid ablation with the balloon isolation patients showed no significant differences in AF burden during follow-up. The hybrid group, however, had a significant reduction in AF burden from the first month onwards, whereas the balloon group shows a clear trend in AF burden reduction during the first 2 months and becomes significant during the third month of follow-up (Table 5). Thirty-four (60%) patients never experienced a recurrence AF after the ablation.

Pulmonary vein diameter

All patients had multislice CT scans before and 3 months after ablation. No stenosis, as defined before, was seen at the evaluation at 3 months. Diameters are presented in Table 1.

Discussion

We present the data demonstrating the feasibility and efficacy of a cryoballoon in circumferential PVI. Circumferential

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Table 3 Results in five consecutive groups of 10 patients

<table>
<thead>
<tr>
<th>Patient number</th>
<th>1–10</th>
<th>11–20</th>
<th>21–30</th>
<th>31–40</th>
<th>41–50</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total number of veins</td>
<td>38</td>
<td>39</td>
<td>38</td>
<td>38</td>
<td>38</td>
<td>NS</td>
</tr>
<tr>
<td>Total number of veins with failed balloon isolation</td>
<td>11</td>
<td>7</td>
<td>4</td>
<td>4</td>
<td>4</td>
<td>NS</td>
</tr>
<tr>
<td>Total number of balloon applications</td>
<td>145</td>
<td>107</td>
<td>106</td>
<td>89</td>
<td>60</td>
<td>&lt;0.01</td>
</tr>
<tr>
<td>Total number of conventional cryocatheter applications</td>
<td>18</td>
<td>28</td>
<td>10</td>
<td>6</td>
<td>15</td>
<td>&lt;0.01</td>
</tr>
<tr>
<td>Patient number with additional cryoablation</td>
<td>5</td>
<td>4</td>
<td>4</td>
<td>4</td>
<td>4</td>
<td>NS</td>
</tr>
</tbody>
</table>

NS, not significant. P-value vs. baseline.

Table 4 Rhythm recording at baseline and during 3 months follow-up

<table>
<thead>
<tr>
<th></th>
<th>Baseline</th>
<th>1 month</th>
<th>2 months</th>
<th>3 months</th>
<th>Total FU</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of recordings</td>
<td>981</td>
<td>1174</td>
<td>1182</td>
<td>1005</td>
<td>3361</td>
</tr>
<tr>
<td>Mean heart rate ± SD</td>
<td>$65 \pm 9$</td>
<td>$68 \pm 8^*$</td>
<td>$68 \pm 9^*$</td>
<td>$66 \pm 8^*$</td>
<td></td>
</tr>
<tr>
<td>AF recordings (n)</td>
<td>246</td>
<td>108</td>
<td>77</td>
<td>45</td>
<td>230</td>
</tr>
<tr>
<td>Mean AF burden ± SD</td>
<td>$0.24 \pm 0.31$</td>
<td>$0.10 \pm 0.22^*$</td>
<td>$0.08 \pm 0.21^{***}$</td>
<td>$0.05 \pm 0.15^{***}$</td>
<td>$0.08 \pm 0.20$</td>
</tr>
<tr>
<td>Median AF burden (range)</td>
<td>$0.14 \text{ (0–1)}$</td>
<td>$0 \text{ (0–0.88)}$</td>
<td>$0 \text{ (0–1)}$</td>
<td>$0 \text{ (0–0.80)}$</td>
<td>$0 \text{ (0–1)}$</td>
</tr>
<tr>
<td>Patients with AF (n)</td>
<td>57</td>
<td>18</td>
<td>16</td>
<td>13</td>
<td>23</td>
</tr>
</tbody>
</table>

AF, atrial fibrillation; SD, standard deviation.

*P < 0.01 vs. baseline.

**P < 0.05 vs. baseline.

***P < 0.001 vs. baseline.
RF ablation has long been shown to yield a high success rate in the treatment of patients with paroxysmal AF, yet proves to be a cumbersome endeavour with high procedure and fluoroscopy times.\textsuperscript{3,5} RF applications in the left atrium are associated with several complications, including substantial mortality.\textsuperscript{4} Previous studies have adopted cryothermia in an attempt to minimize complications since it produces homogeneous lesions and keeps the endothelium intact, with a low thrombotic potential.\textsuperscript{25–30} Tissue adherence during the applications limits this approach to segmental PVI.\textsuperscript{19,31,32}

Several authors have tried applying balloon technology with both ultrasound and high energy focused ultrasound, proving its potential for circumferential ablation, but at a high complication cost.\textsuperscript{33,34} Combining the relatively safe cryothermal energy with a balloon is the next step towards making circumferential isolation of the PVs a simple and safe technique. After its feasibility had been proved in animal experiments,\textsuperscript{20,21} we are now publishing the first human data in this field.

**Acute success**

Our data show a high feasibility in obtaining complete PVI with the cryoballoon, but also show that in a number of cases this seems impossible and additional conventional cryocatheter ablation is required. We believe that anatomical features are the main reason for this. Some patients had oval or slit-like-shaped PV ostia and/or veins inserting onto the left atrium with a sharp angulation. In our experience, it posed more difficulties to occlude these veins with a spherical-shaped balloon. Although without reporting this, complete occlusion seems crucial in obtaining electrical isolation. We think that lack of blood flow allows the balloon to obtain lower temperatures. Incomplete occlusion, and blood flow warming the surface of the balloon, could produce reversible lesions.\textsuperscript{35–37} The learning curve also shows that over time the number of balloon applications falls, indicating that operator-dependent factors were present, along with simultaneous technical improvements of the device. The fact remains, however, that the lengthy cryoapplications add to the duration of the procedure and the use of an additional conventional catheter for a hybrid approach increases the cost of the overall procedure.

### Complications using cryoenergy

The most frequently seen complication in our series was phrenic nerve paralysis. This was also the major limitation in balloon catheters using different energy sources.\textsuperscript{33,34} Recently, it has been reported that this condition is temporary in the majority of the cases.\textsuperscript{38} Stimulating the phrenic nerve with superior caval vein pacing has proven to be a valuable precaution during isolation of the right superior vein in our series. The reversibility of lesions with short cryoenergy applications remains to be proven at this site. All but one of the phrenic nerve paralysis were seen while ablating with a 23 mm balloon deep inside the RSPV, and therefore we advise caution when using this balloon size in that region.

### Outcome data

An intensive follow-up, aimed at detecting asymptomatic recurrence, shows 60% freedom of AF. Several authors consider the first 3 months a blanking period in which recurrence is common, while the effect of the procedure is delayed.\textsuperscript{39–41} To our knowledge, this has never been proven for transvenous catheter ablation of AF, and certainly not when using cryothermal energy. Moreover, there are reports that early recurrence after RF ablation is indicative of long-term failure.\textsuperscript{42}

### Limitations

In our series, patients received anti-arrhythmic drugs before and after the procedure, which could be regarded as a limitation of the study. However, all of the patients had documented episodes of AF while taking their anti-arrhythmic drugs before ablation. Continuing the drug therapy can be considered as a way to reduce a potential bias due to changes in pharmacological treatment. The fact that the heart rate at month 3 was comparable to the baseline value underscores that the baseline autonomic situation was present again, without a change in AF occurrence vs. month 1 and 2. We are currently continuing our long-term follow-up with cessation of anti-arrhythmic drugs in patients who are free of AF after 3 months to further examine the recurrence rate in this group.

### Table 5  Atrial fibrillation burden

<table>
<thead>
<tr>
<th></th>
<th>n</th>
<th>PRE</th>
<th>1 M</th>
<th>2 M</th>
<th>3 M</th>
<th>POST</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Balloon</strong></td>
<td>33</td>
<td>Mean ± SD 0.19 ± 0.30</td>
<td>0.09 ± 0.23</td>
<td>0.08 ± 0.20</td>
<td>0.03 ± 0.08</td>
<td>0.06 ± 0.18</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Median (range) 0.06 (0–1)</td>
<td>0 (0–0.88)</td>
<td>0 (0–1)</td>
<td>0 (0–0.43)</td>
<td>0 (0–1)</td>
</tr>
<tr>
<td><strong>Hybrid</strong></td>
<td>24</td>
<td>Mean ± SD 0.31 ± 0.33</td>
<td>0.11 ± 0.22</td>
<td>0.11 ± 0.23</td>
<td>0.09 ± 0.21</td>
<td>0.10 ± 0.22</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Median (range) 0.20 (0–1)</td>
<td>0 (0–0.75)</td>
<td>0 (0–0.81)</td>
<td>0 (0–0.80)</td>
<td>0 (0–0.81)</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td>57</td>
<td>Mean ± SD 0.24 ± 0.31</td>
<td>0.10 ± 0.22</td>
<td>0.08 ± 0.21</td>
<td>0.05 ± 0.15</td>
<td>0.08 ± 0.20</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Median (range) 0.14 (0–1)</td>
<td>0 (0–0.88)</td>
<td>0 (0–1)</td>
<td>0 (0–0.80)</td>
<td>0 (0–1)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>P-value</td>
<td>&lt;0.01</td>
<td>&lt;0.01</td>
<td>&lt;0.01</td>
<td>&lt;0.01</td>
</tr>
</tbody>
</table>

1 M, first month after procedure; 2 M, second month after procedure; 3 M, third month after procedure; n, number; PRE, baseline; POST, 3-month total; SD, standard deviation. P-value vs. baseline.
Conclusion
We consider cryoablation with a balloon as a feasible initial approach for patients presenting with paroxysmal AF. The technique has an acceptable learning curve. The most frequent complication is phrenic nerve paralysis when ablating the RSPV, but this proved to be reversible in some of the cases.

Acknowledgements
Osama Soliman, MD, who was helpful in analysing left atrial volume data.

Conflict of interest: none declared.

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23. Lang RM, Bierig M, Devereux RB, Flachskampf FA, Foster E, Pelikka PA, Picard MH, Roman MJ, Seward J, Shewnes WS, Solomon SD, Spencer KT, Sutton MS, Stewar WJ. Recommendations for chamber quantification: a report from the American Society of Echocardiography’s Guidelines and Standards Committee and the Chamber Quantification Writing Group, developed in conjunction with the European Association of Echocardiography, a branch of the European Society of Cardiology. J Am Soc Echocardiogr 2005; 18:1440–1463.
Clinical vignette

Double-chambered right ventricle

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An asymptomatic 11-year old boy with a history of spontaneously regressed right ventricle (RV) rhabdomyoma was referred to our institution for cardiovascular magnetic resonance (CMR), because a recent transthoracic echocardiography (TTE) study raised the suspicion of double-chambered RV (DCRV). At the auscultation, a loud grade (4/6) systolic murmur was heard at precordium.

CMR revealed an aberrant fibro-muscular band (arrowheads) originating from the interventricular septum and dividing the RV cavity into a high-pressure chamber in the inlet part (IL-RV) and a low-pressure chamber in the outlet part (OL-RV) (Panels A and B). No other congenital defects have been visualized. The magnitude (Panel C) and phase-encoded flow (Panel D) images showed an accelerated blood flow at level of abnormal band and peak pressure gradient of 77 mmHg was measured. These findings confirmed the diagnosis of DCRV. This is principally a congenital abnormality, rare in the isolated form since it is often associated with ventricular septal defect or other congenital heart disorders. In our case, the muscular band was likely present at birth, although was probably concealed by tumour. It was minimally represented. Indeed, the obstruction may develop over time when progressive band hypertrophy occurs. The clinical presentation of DCRV differs from the absence of symptoms to syncope or angina according to the degree of obstruction. Typically, DCRV is diagnosed in childhood or adolescence. TTE has shown good accuracy for diagnosis in infants but not in adolescence and adults, rendering the use of CMR desirable. Surgery is the treatment of choice and it is advisable before RV dysfunction supervenes.

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