Transcatheter occlusion of sinus venosus atrial septal defect – a new therapeutic option?

Oclusão transcator de comunicação interatrial do tipo seio venoso – uma nova opção terapêutica?

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ABSTRACT – Background: Percutaneous treatment for atrial septal defects is utilized worldwide as first-line therapy. Its use is restricted to atrial septal defects localized within the confines of the oval fossa. All other atrial septal communications are, traditionally, submitted to surgery. Sinus venous atrial septal defect is the second most frequent type of atrial septal defects. The diameters are usually large and thus prone to right heart overload. Besides that, it is difficult to be diagnosed by transthoracic echocardiography, and often discovered at an advanced age. Sinus venous atrial septal defect patients are usually referred to surgical repair, because of unfavorable anatomic characteristics that prevent the use of atrial septal dedicated nitinol mesh devices. The objective of this article is to present a new transcatheter technique for occlusion of sinus venous atrial septal defect of the superior vena cava. Methods: All patients were selected by transthoracic echocardiography images and, after informed consent, were sent to attempt transcatheter occlusion. Procedures were performed under general anesthesia and orotracheal intubation. Selective contrast angiograms were obtained by right and left chamber catheterization. Selective contrast angiograms were performed in superior vena cava and right upper pulmonary vein. Simultaneous right upper pulmonary vein selective angiography was obtained after superior vena cava occlusion with large sizing balloons. The patients presenting persistent pulmonary vein flow to left atrium, despite superior vena cava occlusion, were submitted to sinus venous atrial septal defect closure by covered stents, implanted in the superior vena cava to repair the defect. Results: Four patients (two males) were selected for occlusion procedures. Age ranged from 30 to 53 years. Implant was possible in 50% of patients. In the remainder, superior vena cava occlusion showed right upper pulmonary vein flow stoppage at that level. The procedure was interrupted and patients electively sent to surgery. There were no residual shunts in patients treated, nor procedural complications. Conclusion: Transcatheter occlusion of sinus venous atrial septal defect is an appealing alternative to surgery. Longer follow-up and larger number of cases are warranted for establishing the efficacy and safety of this procedure.

Keywords: Interventional cardiology; Heart septal defects, atrial; Cardiac catheterization; Equipment and supplies

RESUMO – Introdução: O tratamento percutâneo para comunicação interatrial é usado mundialmente como primeira opção terapêutica. Seu uso é restrito a comunicações interatriais localizadas dentro dos limites da fossa oval. Todas as outras comunicações do septo atrial são tradicionalmente submetidas à cirurgia. A comunicação interatrial do tipo seio venoso é o segundo defeito mais frequente do septo atrial. Os diâmetros são geralmente grandes e propensos à sobrecarga cardíaca direita. Além disso, o diagnóstico por ecocardiografia transtorácica é difícil de ser realizado, sendo frequentemente descoberto em idade um pouco avançada. Os pacientes com comunicação interatrial do tipo seio venoso são geralmente encaminhados para correção cirúrgica, devido a características anatômicas desfavoráveis, que impedem o uso de dispositivos de malha de nitinol para fechamento de comunicação interatrial. O objetivo desse artigo é apresentar uma nova técnica transcator para oclusão de comunicação interatrial do tipo seio venoso na veia cava superior. Métodos: Todos os pacientes foram selecionados por imagens de ecocardiograma transesofágico e, após consentimento livre e informado, foram encaminhados para uma tentativa de oclusão transcator. Os procedimentos foram realizados sob anestesia geral e intubação orotracheal. Os traçados de pressão foram obtidos por cateterismo das câmaras direita e esquerda. Foram realizadas angiografias de contraste seletivo na veia cava superior e na veia pulmonar superior direita. A
angiografia seletiva simultânea da veia pulmonar superior direita foi obtida após a oclusão da veia cava superior com balões de maior tamanho. Os pacientes que apresentaram persistência do fluxo da veia pulmonar para o átrio esquerdo, apesar da oclusão da veia cava superior, foram submetidos ao fechamento da comunicação interatrial do tipo seio venoso por meio de stents cobertos, implantados na veia cava superior para corrigir o defeito. **Resultados:** Quatro pacientes (dois homens) foram selecionados para procedimentos de oclusão. A idade variou de 30 a 53 anos. O implante foi possível em 50% dos pacientes. No restante, a oclusão da veia cava superior mostrou interrupção do fluxo da veia pulmonar superior direita nesse nível. O procedimento foi interrompido, e os pacientes foram encaminhados eletronicamente para cirurgia. Não houve shunts residuais nos pacientes tratados, nem complicações relacionadas ao procedimento. **Conclusão:** A oclusão transcatheter da comunicação interatrial do tipo seio venoso é uma alternativa atraente à cirurgia. São necessários o acompanhamento mais longo e um número maior de casos para determinar a eficácia e a segurança desse procedimento.

**Descritores:** Cardiologia intervencionista; Comunicação interatrial; Cateterismo cardíaco; Equipamentos e provisões

**INTRODUCTION**

Interatrial communications correspond to 7% to 11% of all congenital cardiac defects. Its prevalence is 0.53 per 1,000 live births.1-3 Atrial septal defect (ASD) classification is based upon anatomic location in atrial septum. Sinus venosus ASD (SVASD) stands for less than 10% of all ASDs, and is localized outside the confines of the oval fossa (OF), immediately beneath the opening of superior vena cava (SVC) in right atrium (RA). As a consequence, the SVC overrides the communication and has bialtrial drainage (Figure 1). It is generally associated to anomalous drainage of right upper and/or middle lobe pulmonary veins, which can be connected to RA-SVC junction, or directly in the SVC wall.4-6 Surgical repair is the standard technique for correction of SVASD and, until recently, its only treatment option.7

A new transcatheter procedure using large covered stents implanted in SVC covering the interatrial communication was first related by Garg et al., in 2014, and subsequently modified by others.8-10 Deficient posterior wall of SVC is replaced by covered stent closing the atrial communication and redirecting the flow of right pulmonary veins to the left atrium (LA).

In this manuscript, the authors describe an initial single-center experience with transcatheter closure of SVASD and its immediate follow-up.

**METHODS**

**Patient selection**

All consecutive ASD patients referred for transcatheter occlusion were screened and had SVASD diagnosed by transesophageal echocardiography (TEE). The medical charts of SVASD patients were retrospectively analyzed.

This study was evaluated and approved by the Research Ethics Committee (CAAE 34608820.0.0000.5227).

**Inclusion criteria**

All consecutive patients diagnosed with SVASD were included in the study. Usually SVASD are large enough to produce right chamber overload; therefore, all patients were considered for treatment. All patients were selected by TEE imaging (Figure 2).

![Figure 1](image_url) On left panel transesophageal echocardiography in bicaval view, color Doppler shows bialtrial superior vena cava flow. Right side panel shows the same feature visualized on lateral superior vena cava angiography.
Exclusion criteria
Pulmonary hypertension (PH) with high pulmonary vascular resistance (PVR) and associated defects, as well as absence of right upper pulmonary vein (RUPV)-to-LA flow after occlusion of SVC, were exclusion criteria.

Transcatheter occlusion of SVASD was anatomically feasible in 50% of cases.

Procedure
All procedures were performed under general anesthesia and orotracheal intubation after, at least, 8-hour fasting. Unfractionated heparin (100IU/kg, up to a maximum dose of 10,000IU) was administered after obtaining venous access and TEE probe introduction. Intravenous 2g cefazolin was administered to all, followed by 1g, as single dose after 6 hours, in the intensive care unit (ICU).

Vascular access was obtained through right and left femoral vein puncture. In the first case, left femoral artery access was obtained as well. This was deemed unnecessary in the following procedures, and left arterial pressure was obtained by non-invasive methods only. Right and left chambers catheterization and pressure tracings were registered. Left chambers were reached through SVASD. Superior vena cava angiography was obtained, through right femoral vein approach, to ascertain its biatrial connection and diameter measurement. It was also important to rule out absence of left SVC (LSCV) and/or innominate vein, since it could impact on SVC diameter. Right upper pulmonary vein was reached via left femoral vein, and angiography was performed depicting flow to LA. A Super Stiff™ 0.035”/260mm guidewire was inserted into the right femoral and positioned high in the SVC. Balloon size was chosen to be 2 to 4mm larger than SVC diameter measured by angiography. Occluding balloon was, then, inflated and positioned to occlude SVC and SVASD, under TEE guidance. Right upper pulmonary vein angiography during balloon occlusion was performed to confirm persistence or absence of RUPV flow to LA.

In the event of free flow to LA, a covered large stent was implanted, expanded in SVC, and with its inferior edge placed as inferiorly as to completely close the ASD, guided by TEE. Flaring of inferior edge of the stent was performed to obtain better apposition to SVASD opening. Stents were crimped on double balloons (Balloon-In-Balloon, NuMed, Hopkinton, New York, USA) 2 to 4mm larger than the SVC angiographically measured diameter, introduced inside large-bore long Mullins sheaths, with appropriate internal diameter.

Control RUPV and SVC angiography, in addition to TEE images, were performed to ascertain SVASD proper occlusion.

All patients were followed overnight in intensive care unit and were discharged after control transthoracic echocardiography (TTE) the next morning. Patients in whom stent implant was not deemed possible were, electively, referred to surgical repair.

Follow-up
All patients were prescribed aspirin (100mg) for 6 months and clopidogrel (75mg) for 3 months. They were instructed to comply with prophylaxis for endocarditis, if necessary, for 6 months following the procedure. Control TTE was performed after 1 month and TEE in the third month.
RESULTS

From February 2017 to September 2018, a total of 59 consecutive patients were selected for transcatheter closure of ASDs and SVASD was diagnosed in four of them. Table 1 shows patient demographic data, and table 2, the procedure characteristics.

Table 1. Demographic data of four patients

<table>
<thead>
<tr>
<th>ID</th>
<th>Sex</th>
<th>Age (years)</th>
<th>Weight (kg)</th>
<th>RUPV connection</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>F</td>
<td>53</td>
<td>70</td>
<td>SVC-RA junction</td>
</tr>
<tr>
<td>2</td>
<td>M</td>
<td>45</td>
<td>80</td>
<td>SVC-RA junction</td>
</tr>
<tr>
<td>3</td>
<td>F</td>
<td>30</td>
<td>65</td>
<td>SVC wall</td>
</tr>
<tr>
<td>4</td>
<td>M</td>
<td>30</td>
<td>90</td>
<td>SVC wall</td>
</tr>
</tbody>
</table>

RUPV: right upper pulmonary vein; F: feminine; SVC: superior vena cava; RA: right atrium; M: male.

Sinus venosus atrial septal defect static diameter varied from 14 to 23 mm. Superior vena cava and SVASD were simultaneously occluded with large balloons. Right upper pulmonary vein was catheterized and selective angiography was obtained to analyze flow to LA during defect occlusion. Stent implantation was possible in the cases in which RUPV free flow to LA was maintained. In the remainder, procedure was interrupted without implant and patients were oriented to go to surgery.

To occlude and measure SVC diameter, large balloons were utilized: Z-Med™ II 22/40-mm (NuMed Hopkinton, New York) in the first case, 24-mm AGA Sizing Balloon (St Jude-Abbott, USA) in one and 24-mm, and 34-mm Lifetech Sizing Balloons (Lifetech Shenzhen, China) in the others.

In two instances (patients 3 and 4), pulmonary venous angiography did not show RUPV to LA flow after SVC balloon occlusion, and procedures were interrupted without stent implantation. In the other two patients, RUPV free flow to LA persisted (Figure 3). In one case (patient 1) a covered 8ZIG45 CP stent manually crimped over a balloon-in-balloon (BIB) 22/50 mm (NuMed, Hopkinton, New York) was implanted in the SVC, and in the other (patient 2), a covered 8ZIG45 CP stent pre-mounted on 24/50 mm BIB balloon was implanted (Figure 4). Both stents were superexpanded to achieve better adhesion to the borders of the SVASD. In both, stent was implanted in SVC, long enough to completely cover the inferior border of the defect.

Table 2. Procedural characteristics of four patients

<table>
<thead>
<tr>
<th>ID</th>
<th>Sizing balloon (mm)</th>
<th>SVASD (mm)</th>
<th>Stent/balloon (mm)</th>
<th>Result</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Z-Med™ 22/40</td>
<td>23</td>
<td>8ZIG45 CP stent + BIB 22/50</td>
<td>Effective</td>
</tr>
<tr>
<td>2</td>
<td>Lifetech 24</td>
<td>20</td>
<td>PM 8ZIG45 CP stent + BIB 24/50</td>
<td>Effective</td>
</tr>
<tr>
<td>3</td>
<td>AGA 24</td>
<td>14</td>
<td>No Unsuitable</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>Lifetech 34</td>
<td>18</td>
<td>No Unsuitable</td>
<td></td>
</tr>
</tbody>
</table>

SVASD: sinus venosus atrial septal defect; CP stent: Cheatham-Platinum stent; BIB: balloon-in-balloon.

Figure 3. Right upper pulmonary vein selective angiogram with simultaneous superior vena cava and atrial septal defect balloon occlusion. In the top panel right upper pulmonary vein, free flow to left atrium persists, despite of superior vena cava/atrial septal defect occlusion. Bottom panel, on the other hand, shows total interruption of right upper pulmonary vein-left atrium flow by complete obliteration of right upper pulmonary vein. In the first case stent implant was possible, and second patient was sent to surgery.
In one patient (patient 2), the stent did not properly adhere to the walls of SVC, slipping inferiorly into RA, during implant. Balloon was inflated capturing the stent, which was successfully repositioned.

Immediately after stent implant, no residual flow through SVASD was detected by TEE in both cases. Control TTE after 1 month and TEE after 3 months showed correctly positioned stent, and no residual flow through SVASD (Figure 5).

There were no significant complications and no deaths in this small case series.

**DISCUSSION**

Sinus venosus is the most posterior part of RA, were both caval veins are connected. Embryologically, it originates from the most distal portion of the cardiac tube, and is connected to the so-called primitive atrium, through the sinoatrial orifice. It is the smooth portion of the atrium and does not present pectinate muscles. The defect results from incomplete migration of sinoatrial orifice to the
right, impairing secundum septal formation thus creating a defect that is high in interatrial septum. Sinus venosus atrial septal defects exist outside the confines of the OF and account for 2% to 14% of all ASDs. Its most striking feature is overriding of superior caval vein over the atrial septum, above the OF. Sinus venosus atrial septal defect of inferior type is particularly rare.

In most typical cases of SVASD, SCV orifice overrides the defect whose inferior margin is the superior border of the OF. The posterior wall of SVC and anterior wall of RUPV are absent, but the posterior wall persists connected to LA. This creates anomalous drainage of the RUPV, through the defect and, therefore, SVC has bialtrial drainage.

Right inferior pulmonary vein is normally connected to LA, middle lobe vein connects to the area of the defect and RUPV usually connects directly to SVC. Left pulmonary vein drainage is a rare finding, present in as much as 10% of cases.

Due to SVC overriding over interatrial septum and partial anomalous pulmonary venous drainage, SVASDs are generally corrected by surgery. Lack of superior margin with overriding SVC and partial anomalous venous drainage either prevent or difficult implantation of regular ASDs dedicated nitinol mesh devices. Despite that, surgery is not free from significant complications, specially arrhythmias, due to sinus node lesion, obstruction of SVC-AD junction and, mainly, pulmonary vein obstruction.

Sinus venosus atrial septal defect transcatheter occlusion was first reported by Meier et al. and Garg et al. in 2014. Meier managed to implant a regular nitinol ASD device and plugged the RUPV. Garg’s patient had no innominate vein and SVC was thinner than normal. A premounted covered Advanta stent was used successfully. In 2015, Crystal et al. presented with a solution called “wedding cake”, using a self-expanding covered stent. These documents were the pillars of the idea that transcatheter occlusion of SVASD using covered stents was feasible. Nowadays, other authors have opted for this type of SVASD correction.

There is no way to determine by TEE, in advance, which patient will or will not be suitable for the procedure. The only way to determine suitability is by performing simultaneous RUPV angiography with SVC balloon occlusion. Probably the use of more modern imaging methods, such as computed tomography angiography or magnetic resonance imaging, may determine with certainty the position of RUPV, and indicate which cases will be suitable for transcatheter occlusion.

In this small case-series, the age of patients stands out, all over the third decade of life, which is unusual in ASDs series. Those were cases that escaped detection at TTEs, in which this diagnosis is rather tricky, demanding an experienced specialist on congenital heart defects. All our cases were diagnosed by TEE only.

The use of different types of occluding balloons was due to material availability. Usually, an exchange guide wire entering the right femoral vein, being captured and retrieved from the right jugular vein is used to obtain trackability for the implant of venous stents. In our cases, a Super Stiff™ long guidewire introduced in the femoral vein was positioned high in SVC, and proven sufficient to provide all support needed to implant the stents.

The major issue, in our cases, was the slightly short length of stents (45mm) hindering SVC adhesion and total coverage of the defect. In both cases, a very short extension of the stents was expanded in SVC. It is important to emphasize the role of TEE orienting stent placement, to cover the inferior border of the defect. Unfortunately larger and longer stents, utilized by others, were not available at the time of the procedures.

Possible complications of the procedure are similar to surgery, mainly sinus node lesion and RUPV stenosis.

CONCLUSION

Transcatheter occlusion of sinus venosus atrial septal defect is an appealing alternative to surgery. The small number of cases and short-term follow-up are significant limitations of this report. Longer follow-up and more numerous case-series are warranted to establish the efficacy and safety of this procedure.

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None.

CONFICTS OF INTEREST

The authors declare there are no conflicts of interest.

CONTRIBUTION OF AUTHORS

Conception and design of the study: FC; data collection: FC, DP, RT, JCT and MOL; data interpretation: FC, DP and RT; text writing: FC, DP and RT; approval of the final version to be published: FC, DP e RT.

REFERENCES