Device Fracture and Severe Tricuspid Regurgitation After Percutaneous Closure of Perimembranous Ventricular Septal Defect: A Case Report

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We report a case in which fracture of an Amplatzer perimembranous ventricular septal defect (VSD) device occurred within 1 month after placement. This was associated with severe tricuspid regurgitation. Surgical removal of the device and repair of the tricuspid valve was performed with reasonable outcome. We propose a hypothesis on how this complication might have occurred and how it could be prevented.

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INTRODUCTION

During the last few years, percutaneous closure of perimembranous ventricular septal defects has been introduced as an alternative to surgical treatment in selected patients. Especially the introduction of the asymmetrical Amplatzer perimembranous septal occluder has resulted in high closure rates with a low mortality and acceptable morbidity [1–5].

Complications that have been described are as follows: early and late occurrence of total atrioventricular block [6], aortic regurgitation, tricuspid regurgitation, tricuspid stenosis [5,7], and hemolysis [8]. We report a case of device fracture and severe damage to the tricuspid valve within 1 month of percutaneous treatment.

CASE REPORT

A 3-year-old girl was followed-up with a moderately large perimembranous ventricular septal defect. Prior to admission for cardiac catheterization, she was asymptomatic. She had a grade 4/6 holosystolic murmur at the left parasternal border. The electrocardiogram showed signs of left ventricular hypertrophy. The echocardiographic examination showed a 5 mm perimembranous ventricular septal defect with a large left-to-right shunt. The pulmonary-to-systemic blood flow ratio was estimated to be \( > 1.5/1 \). There was a gradient of 85 mm Hg across the defect, excluding the presence of pulmonary hypertension. There was left ventricular dilatation with a left ventricular end-diastolic diameter of 38 mm (z-score 2.2). There was mild grade [1/4] mitral regurgitation and trivial tricuspid regurgitation. On the basis of these findings, a cardiac catheterization was performed with the purpose of closing the perimembranous defect.

Under general anesthesia, a 4 and 6 Fr sheath were placed in the right femoral artery and vein respectively. Angiography as well as transesophageal echocardiography (TEE) were used to define the size and location of the ventricular septal defect (VSD). The presence a moderately large perimembranous defect was confirmed both on angiography as well as on transesophageal echocardiography. The size of the defect was estimated to be around 5–6 mm. There was partial covering of the defect by tissue coming from the tricuspid valve. There was a rim between the aortic valve and the device of 4–5 mm. The pulmonary-to-
systemic blood flow ratio, measured by oximetry was 1.9/1. Mean pressure in the pulmonary artery was 16 mm Hg. On the basis of the angiographic as well as the transesophageal images, a 6-mm Amplatzer perimembranous VSD occluder (AGA®, MN) was implanted according to the technique recommended by the company. Briefly, the VSD was crossed from the left ventricle using a 2.5 Judkins right coronary catheter and a 0.035 Glide wire (Terumo®, Haasrode, Belgium). The glide wire was exchanged for a 0.035 Noodlewire (AGA, MN), and a continuous arteriovenous circuit was formed. The Amplatzer delivery sheath could easily be advanced from the right femoral vein through the VSD and was positioned towards the left ventricular apex. The 6-mm Amplatzer VSD occluder was deployed during a procedure, which was described as uneventful. Angiographic and transesophageal images before and after release of the device showed a nice position within the VSD with a small residual shunt centrally through the device. There was no aortic regurgitation after release of the device. The fluoroscopy time of the whole procedure was 32 min. The procedure was performed after anticoagulation with heparin (1,500 IU intravenously) and administration of intravenous antibiotics. Cine post release of the device showed a pronounced motion of the right ventricular disc and microscrew. This was interpreted as caused by contraction of the septum and pressure transmitted from the LV through the device.

One day after the procedure, a transthoracic echocardiography was performed. The device was normally positioned within the VSD without a residual left-to-right shunt. On color Doppler, turbulence on the tricuspid inflow was noted and the opening of the tricuspid valve was felt to be mildly restricted (Fig. 1). There was a mean gradient on the tricuspid inflow signal of 4 mm Hg. Mild tricuspid regurgitation and trivial mitral regurgitation was noted. These findings did not cause any concern at that time. On ECG, there was normal sinus rhythm and normal AV-conductance. The patient was discharged from the hospital.

One month later she was seen again in our outpatient clinic. The parents did not report any special event. She was clinically doing very well. At clinical examination, a grade 2–3/4 systolic murmur could be detected at the lower left sternal border. There was also a grade 2/4 diastolic inflow murmur. There was mild hepatomegaly with the liver palpable 2 cm below the ribs. The ECG showed normal sinus rhythm with normal PR-interval. The echocardiography showed the device positioned within the VSD (Fig. 2). The most striking finding was a severe grade 4/4 tricuspid regurgitation. Additionally, a small residual ventricular septal defect was noted, which was not remarked on the previous echo. On closer inspection, the device was found to have an abnormal configuration, and a second echodense structure was noted in the right ventricular cavity within the tricuspid apparatus. This structure was thought to be part of the device. The mechanism of the tricuspid regurgitation was difficult to describe, but at that time it was thought to be caused by non-coaptation between the anterior and septal leaflet caused by a ruptured chord. To check the position and integrity of the device, fluoroscopy was performed. On

Fig. 1. Echocardiography 24 h after the procedure. (A) The close relationship between the right ventricular disc and the tricuspid valve can be noted. (B) With color-flow Doppler, some turbulence on the tricuspid inflow can be noted with a Nyquist limit of 0.72 m/s. This indicates some degree of inflow obstruction.
screening, the device was found to be broken into two different pieces with the screw and part of the wires being detached from the rest of the device (the two discs), which was still positioned within the ventricular septal defect (Fig. 3A). Secondary to the severe tricuspid regurgitation there was a mild to moderate right atrial and right ventricular enlargement. Right ventricular function was judged to be normal.

On the basis of these findings, surgery was performed. This confirmed that the microscrew was indeed detached from the device (Fig. 3B) and was entangled within the chordal attachments of the anterior leaflet of the tricuspid valve. There were some wires attached to the microscrew. The rest of the device (the two discs) was positioned within the VSD, and a small residual defect could be detected just at the site where the conduction tissue is normally located. The anterior leaflet of the tricuspid valve was severely damaged as a large traumatic cleft could be detected (Fig. 3C). The device and screw were removed from the heart taking care not to damage the conduction system and the ventricular septal defect.

Fig. 2. Echocardiography 1 month after the procedure. (A) The unusual appearance of the device can be noted. It seems to protrude further into the right ventricle compared to after immediate placement. An echodense structure can be detected in the tricuspid chordae (arrow). (B) On color Doppler the severe tricuspid regurgitation can be noted as well as the residual VSD that was not there at the end of the procedure.

Fig. 3. Device breakage on fluoroscopy and after removal. (A) On fluoroscopy the microscrew is clearly detached from the device. (B) The device after surgical removal. The microscrew with some wire have come loose from the right ventricular disc. (C) The “cleft” anterior leaflet can be seen.
was surgically closed. The anterior leaflet was repaired by closing the cleft. Postoperatively the child did clinically well. The VSD was completely closed, and echocardiographically a moderate grade 2/4 residual tricuspid regurgitation could be observed. The ECG showed normal sinus rhythm with normal conduction. The device was technically evaluated by the manufacturer and no structural deficiencies could be detected explaining the breakage.

**DISCUSSION**

The Amplatzer perimembranous VSD occluder has been reported to be an efficient and safe device for treatment of perimembranous VSDs in children and adults. Complications reported so far are mainly related to the conductance system with the occurrence of total AV-block of up to 5% of the treated patients [4]. The close relationship of both the aortic valve as well as the tricuspid valve to a perimembranous defect has been a concern when developing the device and delivery technique [9]. In a recent report, trivial to mild aortic regurgitation was reported in up to 53% of all patients who underwent device closure [2]. In the same report, damage to the tricuspid valve apparatus was described in one patient when trying to retrieve a device that caused moderate aortic regurgitation after placement.

In our patient, we noted a previously unreported problem. Within 1 month after placement, fracture of the device occurred with the microscrew coming lose from the right atrial disc. Additionally, severe damage was caused to the tricuspid valve; the microscrew with some of the wires got entangled within the chordae of the tricuspid valve. The anterior leaflet was severely damaged with a deep traumatic cleft within the leaflet [2].

We hypothesize the following unfortunate series of events happened in this patient. First, during placement, when passing with the glide wire through the defect, chordae from the tricuspid leaflet got entangled in the wire. This was not noted and the rest of the procedure of the device placement went very smoothly. Apparently, this entanglement did not cause any problem during passage of the sheath from right to left ventricle and no abnormal tricuspid regurgitation was observed throughout the procedure. When the device was unfolded from the sheath tricuspid valve chordae, possibly already entangled by the guidewire, got caught behind the microscrew. When configuring the RV disc, the disc and screw are unprotected by the sheath and at this moment chordae surrounding the device, can get caught behind the microscrew. The design of the device with its relatively long untapered microscrew allows the chordae to be retained by it after release from the delivery cable. In our patient, the chordae pulled on the microscrew with each cardiac contraction. This could be noted in retrospect on the fluoroscopic images obtained immediately after placement. There was a pronounced motion of the right atrial disc, suggesting entanglement. Since there was no more than trivial tricuspid regurgitation after placement, this was not causing any concern. The day after the procedure, the motion of the tricuspid valve was thought to be somewhat restrictive with some flow acceleration on color Doppler. This was felt to be mild and did not cause any concern. Because of the stress on the microscrew with every heartbeat, metal fatigue in the wires occurred and finally the microscrew got detached from the device. As the screw got entangled into the chordae of the anterior tricuspid leaflet, the sharp microscrew and wires probably caused a tear in the anterior valve leaflet. During surgery the microscrew could be removed from the chordae and the device from the VSD. Care was taken in the removal of the device not to damage the conduction tissue. The anterior leaflet could be repaired by closing the “cleft”.

Different strategies could be envisaged to prevent this complication in future cases. First the diagnosis of tricuspid tissue entanglement should be improved during device implantation. The standard method generally applied is to straighten the wire after placement of the arteriovenous loop and look echocardiographically for an increase in tricuspid regurgitation. One of the problems associated with this method is that pulling on the wire will nearly always result in some increase in tricuspid regurgitation. It takes some experience to exactly define what considered to be a significant increase in tricuspid regurgitation. Another hint pointing towards entanglement of chordal tissue from the tricuspid valve is kinking of the wire at the level of the tricuspid annulus when straightening it. In this patient this could not be observed. A third sign that suggests potential entanglement is difficulties encountered when advancing the delivery sheath across the VSD as entangled chordal tissue on the guide wire could interfere with smooth passage of the sheath across the VSD. In our patient the VSD could easily be crossed with the sheath and the sheath could easily be bended towards the left ventricular apex. Perhaps better observation of the tricuspid valve when crossing with the sheath looking for increased tricuspid insufficiency might be helpful. At this stage of the procedure the echocardiographer often focuses on the aortic valve when guiding the interventional cardiologist. Careful attention should be given to the tricuspid valve throughout the whole procedure. A second strategy to prevent the complication could be directed at the tech-
nique used to cross the VSD. In our case the VSD was crossed with a right Judkins catheter and a Terumo glidewire from the left ventricle. Maybe passage with a balloon catheter either from left to right or in the opposite direction could prevent entanglement, but using these catheters is technically more challenging. A guidewire with a curved tip could also make it less likely to entrap part of the tricuspid valve. Finally the microscrew design could possibly be changed. The current device has a long microscrew with untapered edges. Making the screw shorter, smoothened, and tapered, would make it less likely for chordae to get caught by the screw, or they could easily slip from it with cardiac motion.

This case illustrates that during and after the procedure, careful assessment of tricuspid valve motion is extremely important. The right atrial disc had a close relationship with the tricuspid valve and there was some acceleration across the tricuspid valve, which was not present before the procedure. These signs should be considered as indicating entrapment. This case also shows that when the device is thought to be caught by the tricuspid valve it should be immediately removed surgically. Once released and part of the tricuspid chordal apparatus is entangled, no attempts should be made to recapture the device with a snare, as this can cause severe damage to the valve. This was illustrated by one reported case [2]. During surgical removal care should also be taken not to damage the conduction tissue. In this patient we were lucky that the residual VSD was located in the area of the conduction tissue, which facilitated device removal. Maybe future changes in the device design (especially screw design) could prevent this complication.

REFERENCES