Percutaneous trans-mitral commissurotomy in juvenile mitral stenosis with aneurysmal left atrium — a technically challenging situation

Przezskórna komisurotomia mitralna u nastolatka ze stenozą mitralną i tętniakiem lewego przedsionka — zabieg trudny technicznie

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Abstract

Percutaneous trans-mitral commissurotomy (PTMC) using an Accura balloon is an effective procedure for the management of patients with rheumatic mitral stenosis. It is not infrequent to encounter an aneurysmal left atrium (LA), especially in the developing world where patients often present very late. This is a technically challenging procedure as the septum often bulges into the right atrial side, making the puncture often difficult. Furthermore, it bulges anteriorly and inferiorly so it becomes even more challenging. Because of the vertical lie of the septum, the needle often dissects the septum rather than puncturing it.

Here, we describe the case of a 17 year-old boy with severe mitral stenosis with a giant, aneurysmal left atrium (LA — 13.5 × 16 cm) where an inter-atrial septal puncture was achieved by probing the fossa ovalis to enter the left atrium instead of a conventional transseptal puncture. PTMC was successfully done, increasing the mitral valve area from 0.8 cm² to 1.9 cm².

Key words: aneurysmal left atrium, probing fossa ovalis, Brockenbrough needle, percutaneous trans-mitral commissurotomy, trans-septal puncture

Introduction

Percutaneous trans-mitral commissurotomy (PTMC), introduced in 1984 by Inoue et al [1], opened a new window for the treatment of patients with symptomatic mitral stenosis (MS) with suitable valve anatomy. In the presence of a left atrial clot, left atrial appendage clot, or calcified valve, the complication rate may rise [2]. Another challenging substrate is an aneurysmal left atrium (LA) where the inter-atrial septum becomes fatter with a more horizontal orientation. The fossa ovalis lies lower down. As LA pressure is high, the atrial septum usually bulges into the right atrium. For these reasons, the needle tends to make a tangential track in the atrial septum. These factors pose a formidable challenge during a trans-septal puncture. The conventional technique requires some modifications when faced with these atypical situations.

Case report

A 17 year-old boy presented with exertional dyspnoea — New York Heart Association (NYHA) class II for the past three
years, progressing to class III for the past eight months. His chest X-ray showed an aneurysmal left atrium (Figure 1). Transthoracic echocardiogram revealed severe mitral stenosis with a mitral valve area of 0.8 cm$^2$ by planimetry, and Wilkin’s score — 8/16 (M2, C2, T2, S2). Mean gradient across the mitral valve was 35 mmHg by pressure half time (PHT). His left atrium was aneurysmally dilated, with the size being 11 × 9 cm on echocardiogram (Figure 2). His height was 163 cm. Transoesophageal echocardiography was performed to look for a left atrial clot, degree of mitral regurgitation (MR), and the suitability of PTM. Pre-procedural consent was obtained. Femoral artery and vein were accessed with a 6 F and a 8 F sheath respectively. A Mullins sheath over a 0.035” guidewire was parked in the left brachiocephalic vein. The wire was removed and a Brockenbrough needle was inserted into the sheath. LA entry was facilitated using the probing fossa ovalis technique after making a gradual descent. A small puff of dye was injected to confirm the left atrial entry. Subsequently, the Mullins sheath was advanced over the needle and contrast was injected through the sheath following removal of the needle, which opacified the left atrium, confirming its aneurysmal dilatation (Figure 3A). Inter-atrial septum was

Figure 1. Chest X-ray postero-anterior view showing aneurysmal left atrium

Figure 2. 2D trans-thoracic echocardiogram in apical four-chamber view showing aneurysmal left atrium (11 × 9 cm)

Figure 3A. Contrast being injected through the sheath opacifying the left atrium (LA); B. Loop LA wire was parked in LA
dilated in a left atrial oblique (LAO) 30° view. Looped LA wire was parked in left atrium (Figure 3B). An Accura balloon (Vascular Concepts, Essex, UK) was sized to 26.5 mm based on Hung’s formula. Preprocedure LA pressure was 49/28 mm Hg (mean = 39 mm Hg). Following septal dilatation, the Accura balloon was placed in the LA. Because of the giant LA, the Accura balloon was pushed over the wire very deep inside the LA (Figure 4). LA wire was removed, and J-shaped metallic stylet was introduced. A gentle anticlockwise twist was applied to the metallic stylet, and the Accura balloon was gradually pulled until a bobbing movement at its tip was noted. Once the bobbing movement of the Accura balloon was established, it was pushed into the left ventricle through the mitral valve. Once in the left ventricle, the J-shaped metallic stylet was exchanged for a 0.035” wire. The balloon was inflated in its distal part and pulled back to anchor it at the mitral valve and inflated further to achieve its full dilatation (Figure 5). The procedure was performed successfully, thereby bringing the LA mean pressure to 6 mm Hg, and achieving a mitral valve area of 1.9 cm$^2$.

**Discussion**

In a developing country like India, where patients with rheumatic mitral stenosis often present very late, it is not uncommon to find an aneurysmal left atrium. In such situations, the anatomy of the interatrial septum is often distorted. Wood et al have proposed a radiological classification of LA enlargement wherein grade-3 denotes where the left atrium bulges conspicuously on both sides of the heart, extending beyond the shadow of the right atrium on that side. This gross enlargement is described as aneurysmal [3].

Due to the high left atrial pressure in mitral stenosis, an atrial septum will usually bulge into the right atrium, where the atrial septum becomes flatter with a more horizontal orientation, which distorts the anatomy of the region of the fossa ovalis as it lies lower down [4]. These factors combined make septal puncture challenging because the needle tends to dissect the septum rather than puncturing it because it tends to track tangentially. Also, if it punctures the muscular part of the septum, sometimes septal catch is encountered, which makes negotiating the balloon across the mitral valve difficult. Moreover, there are potential risks during transseptal puncture in the form of needle tip perforation < 3%, tamponade < 1%, and death < 0.5% [5]. Instead of a conventional transseptal puncture, the fossa ovalis probing technique, as described by Krishnamoorthy et al [6], may be tried, as in our case. Under fluoroscopic guidance, the needle and the sheath as an assembly make a gradual descent into the right atrium. When the Mullins sheath snaps into the fossa ovalis, it is firmly pressed against the atrial septum, keeping the needle tip just inside the sheath (by about 2–3 mm). This gives support to the sheath without risk of injury to the left atrial wall. Proper positioning of the sheath tip prior to probing should be ensured on fluoroscope, and a test injection with contrast material made to identify any tenting of the atrial septum at the site of the fossa. Left atrial entry should
be sought by repeated attempts to probe the fossa using gentle and controlled pressure applied to the sheath tip. Once entry to the left atrium is suspected, with a feeling of giving way, only 2–3 mm of the tip of the sheath should be allowed to enter the chamber. Further advancement of the sheath should be done only after confirming its entry into the left atrium.

These precautions avoid damage to the left atrial wall at a site other than the fossa, and consequent damage to aorta or pericardium. This avoids the production of an iatrogenic atrial septal defect, although the incidence of this is itself much less frequent with a conventional transseptal puncture. One should avoid the risk of a tear outside the fossa ovalis by keeping the needle tip well inside the sheath throughout the procedure. Surgeons should ensure proper positioning of the sheath tip, the application of gentle and controlled pressure allowing only 2–3 mm of the tip of the sheath to enter the left atrium after probing, and final advancement of the sheath only after the confirmation of left atrial entry by a puff injection. One should avoid probing the fossa with force. This will be successful as high left atrial pressure leads to stretching and thinning. This has the additional advantage in certain situations such as a distorted atrial septum, a significant right or left atrial enlargement, for those on anticoagulants, and for pregnant women, because it reduces fluoroscopy time and complications.

If this fails, a transseptal puncture will be the default choice. This requires slight modification in the form of increasing the curvature of the Brockenbrough needle so that it can face more posteriorly to enable the puncture, especially when the LA is aneurysmal. In difficult cases, transthoracic, transoesophageal, or intracardiac ultrasound navigated transseptal puncture may be employed [6].

Usually, the balloon is negotiated in a right anterior oblique view because it profiles the left ventricular long axis. In a situation of aneurysmal LA, a lateral view (90°) may be better sometimes. Other improvisations may include a non-ideal puncture site such as more cephalad or leftward (closer to mitral valve), very low punctures, changing the shape of the stylet, and different techniques of balloon entry such as the over the wire technique [7, 8], and floating a Swan-Ganz catheter into the left ventricle [9].

Our method was a modification of an existing technique that turned a complex situation into a simple one.

Conflict(s) of interest
The authors declare no conflict of interest.

Streszczenie
Przezskórna komisurotomia mitralna (PTMC) przy użyciu balonu Accurato to skuteczny zabieg stosowany w leczeniu chorych ze stenozą mitralną (MS) spowodowaną chorobą reumatyczną. Nierzadko spotyka się u tych chorych tętniak lewego przedsionka (LA), zwłaszcza w krajach rozwijających się, gdzie pacjenci często bardzo późno zgłaszają się do lekarza. Zabieg ten jest trudny technicznie, ponieważ przegroda często wpukła się do prawego przedsionka, utrudniając wykonanie nakłucia. Co więcej, przegroda wybrzusza się w kierunku przednio-dolnym, co dodatkowo zwiększa trudność zabiegu. Ze względu na pionowe położenie przegrody igła często rozdziela przegrodę, zamiast ją przebić. W niniejszej pracy opisano przypadek 17-letniego chłopca z ciężką MS i olbrzymim tętniakiem LA (LA — 13.5 × 16 cm). Aby dostać się do LA, sondowano otwór owalny zamiast konwencjonalnego nakłucia przegrody. Przeprowadzono z powodzeniem zabieg PTMC, zwiększając powierzchnię zastawki mitralnej z 0,8 cm² do 1,9 cm².

Słowa kluczowe: tętniak lewego przedsionka, sondowanie otworu owalnego, igła Brockenbrougha, przeszkórná komisurotomia mitralna, nakłucie przegrody międzyprzedsionkowej

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