Aortic stenosis and concomitant hypertrophic obstructive cardiomyopathy: the bad and the worse

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With the broadening of indications and the consequent massive increase in the number of transcatheter aortic valve implantations (TAVI) some former niche subgroups have gained real clinical relevance. And such severe aortic stenosis (AS) and concomitant obstructive left ventricular hypertrophy is found more and more frequently, causing important diagnostic challenges and treatment dilemmas.

Proper assessment of sequential stenoses and their individual relevance is per se challenging due to the hemodynamic crosstalk. This becomes even more complicated, when this occurs with non-laminar flow of a non-Newtonian fluid in a non-tubular setting, where the surrounding structures have their own, flow- and pressure-dependent dynamic. And this is exactly the case, when AS and left ventricular outflow tract (LVOT) obstruction coexist.

Left ventricular hypertrophy is the common consequence of AS due to the chronic pressure overload. While it is often described in the echocardiography, its magnitude shows marked variation and accordingly its true clinical relevance can be easily overlooked. This occurs mainly because the cardiac contractility is restricted due to the massive afterload and the blood flow is limited over the LVOT, due to the more distal obstruction, namely the AS itself. Accordingly, in this setting the echocardiographic impression and ventricle-to-aorta gradient are dominated by the stenotic aortic valve, masking the true hemodynamic relevance of the LVOT obstruction.

However, once the patient undergoes TAVI, the hemodynamics change quite abruptly: the obstruction by the stenotic aortic valve becomes eliminated so that the afterload falls to almost normal within minutes and the ventricular contractile mechanisms show improvement within hours. This results in consequent exaggeration of all, the septal inotropy, the flow acceleration and the hydrodynamic drag and pull by Venturi effect, leading to dynamic narrowing of the muscular LVOT and to a pronounced systolic anterior motion of the mitral valve, manifesting in ‘unexpected’ hemodynamic deterioration (1, 2).

While concomitant septal myectomy is an optimal solution for patients undergoing surgical valve replacement, for obvious reasons this option cannot be really considered in the cohort, candidate for TAVI. In most of the cases conservative therapy with strict heart rate control and maintenance of filling pressure can be sufficient to stabilize the patient. But in case of failure of conservative management bail-out strategies might be necessary, as reported in the literature. Endo et al. described a case with marked improvement in the hemodynamics with right ventricular apical pacing. (3) As mechanism of its beneficial effect the altered systolic contraction of the basal segments is suggested. Leya et al. has published a case report, where LVOT obstruction was managed with a deep implanted valve-in-valve (4). Despite the success of that case, we believe its application should be considered with extreme caution: on one hand LVOT obstruction is rarely localized purely in the very distal LVOT, but more often even in the mid-ventricular sep-
Another interesting approach was reported by Bode et al., implanting “preventively” a percutaneous mitral edge-to-edge repair system to prevent systolic anterior motion of the mitral valve after the TAVI, which can be definitely considered in certain mitral anatomies, where LVOT obstruction is expected predominantly from the systolic anterior motion of the mitral cusp (5) Verheyen et al. published a case with “conventional” transcatheter alcohol septal ablation as bail-out solution in a patient with cardiogenic shock after transcatheter aortic valve implant due to extreme LVOT obstruction with a gradient, reaching 145 mmHg in rest (more than 240 mmHg after Valsalva maneuver!), which was reduced to 27 mmHg after alcohol septal ablation (6). Transcatheter alcohol septal ablation is definitely an elegant and minimally invasive approach, however its safe and successful performance, especially in a post-TAVI setting, requires a team with extensive experience due to its delicate technical challenges. On the other hand transcatheter alcohol septal ablation in patients with untreated AS (i.e. prior TAVI) has potentially increased risk of septal rupture due to the massive pressure overload and therefore it should be discouraged.

While having potential bail-out strategies in the therapeutic arsenal is crucial, proper diagnosis prior aortic valve replacement is even more important in order to define the best possible therapeutic approach in advance. As described above the proper assessment of individual functional impact is cumbersome or even impossible when AS and LVOT obstruction coexists. Still, there are couple echocardiographic parameters, which can be considered as surrogate characteristics, raising attention for the potential hemodynamic importance of LVOT obstruction. Asymmetric left ventricular hypertrophy with a septal-to-posterior wall thickness ratio of more than 1.3 has been indicated in the literature as marker of ‘latent’ LVOT obstruction. Besides that a certain late systolic peak is often observed in the continuous wave Doppler, caused by the systolic anterior motion of the mitral valve, even if it is not as pronounced as in pure hypertrophic obstructive cardiomyopathy. Finally the morphology of the mitral apparatus has to be also carefully evaluated, since elongated mitral cusps tend to protrude more into the LVOT, causing marked gradient increase. Particularly the extent of residual leaflet length appears to play a crucial role (7). In case of clinical or echocardiographic suspicion invasive evaluation has to be considered as well. With pullback maneuver of a dedicated pigtail catheter from the apex, over the mid-ventricular space, the LVOT up to the aorta during Valsalva and simultaneous measurement of the aortic pressure, more information can be gained about the functional and anatomical characterization of the pressure gradient.

Presence of concomitant AS and LVOT obstruction has to be seriously taken into account during heart team decision and threshold for surgical valve replacement with myectomy should be lower. If careful assessment and multidisciplinary decision finally still indicates TAVI, then it should be performed in a center with sufficient expertise to perform transcatheter alcohol septal ablation and/or edge-to-edge mitral repair even in a post-TAVI setting in case hemodynamic status indicates.

Declaration of interest
The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

References