**ILLUSTRATIVE CASE**

**Demographic and past medical history**

* Female. Date of birth: 15/12/1998
* Situs solitus, levocardia, atrioventricular and ventriculo-arterial discordance, ventricular septal defect.
* Pulmonary artery banding on 17/02/2000
* Complete repair ( Arterial switch, Mustard procedure, ventricular septal defect closure) on 23/3/2003. Coronary sinus drainage was left to the pulmonary atrium.
* Evidence of superior vena cava systemic atrium junction stenosis treated with balloon dilatation on December 2007. In the same year she had undergone endocardial atrial pacemaker implantation due to sinus node dysfunction.
* The patient has always reported normal effort tolerance. NYHA class I.
* She was practicing equitation at competitive level.

**Repeated CPETs evaluations and diagnostic workup.**

The patient underwent serial exercise metabolic tests evaluations as part of the routine assessment at the sport medicine department. CPETs performed in 2012, 2015 and 2018 showed progressive decline in maximal VO2, O2 saturation during effort and increase of VE/VCO2 slope (Table 1 suppl, Figure 1 suppl). At the latest follow up in 2019 significant de-saturation (around 90%) was noted at rest.

These findings prompted second level imaging with cardiac computerized tomography (CT), although transthoracic echo was unremarkable. CT demonstrated occlusion of superior systemic baffle at the level of atrial junction, a six millimeters leak between the inferior systemic baffle and the pulmonary venous atrium, a moderate stenosis beyond the leak and diversion of the superior vena cava flow toward the inferior vena cava through the azygos vein and multiples mediastinal and para-vertebral venous vessels. (Figure 2 suppl)

**Pathophysiologic interpretation**

The clinical picture was interpreted as follow: the progressive stenosis and final occlusion of the superior systemic venous pathway, together with the inferior pathway stenosis caused an ongoing increase of the blood diverted to the inferior baffle and, thus, forced through the leak, eventually triggering a growing magnitude of right to left shunt and worsening de-saturation. Trans-baffle leak is a well known complication after atrial switch. It might remain undetected until hemodynamic changes, like in this case, make the expected left to right shunt to revert into right to left. It is well known that VE/VCO2 slope dramatically rises in cyanotic patients due to a disproportionate ventilation rate (VE) increase and, in this case, turned out as an early and sensitive diagnostic marker.