A 19-year-old athlete with sudden onset dyspnoea and progressive exercise intolerance presented to our Heart Centre for assessment of his cardiopulmonary function. A grade 3/6 continuous murmur best heard along the lower left sternal boarder was the only pathological finding upon physical examination. While electrocardiogram depicted a sinus tachycardia and an incomplete right bundle branch block, laboratory testings were within normal range. Transesophageal echocardiography (TEE) demonstrated an oval-shaped echolucent structure originating from the right coronary sinus and protruding into the right atrium (A). Doppler examination revealed permanent shunting from the aorta into the right atrium via a perforation located at the tip of the lesion (B). The tentative diagnosis of a ruptured sinus of valsalva aneurysm was confirmed by contrast-enhanced computed tomography (C) and magnetic resonance imaging (D–E), two diagnostic tools that had infrequently been applied in the diagnosis of this entity. Non-invasive imaging clearly visualized the congenital lesion and contributed to anatomical and functional assessment prior to surgery. The results of non-invasive diagnostics corresponded perfectly to the intra-operative findings. A perforated aneurysmatic sac with a diameter of 26 mm was visualized after right atrial incision (F) followed by an oblique aortotomy (G). The aneurysm was resected from atrial access, followed by a direct continuous suture reinforced with pledgeted mattress sutures. There was no distortion of the right coronary ostium. Final TEE demonstrated excellent coaptation of the aortic cusps and disappearance of the shunt. The patient recovered without periprocedural complications and has resumed his sporting activities 1 year after surgery.