Journal of the Saudi Heart Association

Volume 33 | Issue 1 Article 13

2021

Heyde's syndrome: An enigma

Recommended Citation

Khazi, Fayaz Mohammed; Siddiqi, Nayyer R.; Karaly, Yehia Mohamed; Aljassim, Obaid; and Al-Halees, Zohair Y. (2021) "Heyde's syndrome: An enigma," *Journal of the Saudi Heart Association*: Vol. 33 : Iss. 1 , Article 13.
Available at: https://doi.org/10.37616/2212-5043.1237

CASE REPORT

Heyde's Syndrome – An Enigma

Fayaz Mohammed Khazi [a,*], Nayyer R. Siddiqi [a], Yehia Mohamed Karaly [a], Obaid Aljassim [b], Zohair Y. Al-Halees [c]

[a] Cardiothoracic Anaesthesia, Dubai Hospital, Dubai Health Authority, Dubai, United Arab Emirates
[b] Cardiothoracic Surgery, Dubai Hospital, Dubai Health Authority, Dubai, United Arab Emirates
[c] Cardiothoracic Surgeon, King Faisal Specialist Hospital and Research Centre, Riyadh, Saudi Arabia

Abstract

Heyde's syndrome (HS) is described as the association between recurrent bleeding from angiodysplasia of the gastrointestinal tract and aortic stenosis. Aortic valve replacement has been reported to stop the bleeding. In unfit patients, the options available are interventional or conservative management. We hereby report an elderly obese patient with severe comorbidity with complicated HS involving a narrow aortic root. She underwent left ventricular outlet myomectomy and aortic root replacement to promote better forward flow and prevent restenosis and recurrence of symptoms. She was discharged home symptom-free despite being on coumadin anticoagulants.

Keywords: Aortic stenosis, Gastrointestinal bleeding, von willebrand, Angiodysplasia, Heyde's syndrome, Cardiac surgery

Introduction

Heyde's syndrome (HS) is a triad of aortic stenosis (AS), acquired coagulopathy (Von Willebrand syndrome -2A, (VWS-2A)) and anaemia due to bleeding from intestinal angiodysplasia. Replacement of the aortic valve has been reported to stop the bleeding. Recently, transcatheter aortic valve implantation (TAVI) or other conservative strategies have been recommended as treatments of choice for high-risk surgical patients. In addition, TAVI may not be technically suitable for a patient with a narrow aortic root, and haemostatic abnormalities may recur when a mismatch is found between the patient and prosthesis size.

Case presentation

A 66-year-old woman was admitted with chest discomfort and transient loss of consciousness. Her past medical history included anaemia, hypertension, type 2 diabetes and morbid obesity with a BMI of 42 and obstructive sleep apnoea. Blood investigations revealed a haemoglobin of 8.4 g/dl (12–15 g/dl), a platelet count of 169/mm^3 (150–410/mm^3) a troponin T of 16 ng/L (<14 ng/L) with normal vitamin B12 and folate levels, but iron levels were consistent with iron-deficiency anaemia with serum iron 44 μg/dL (37–145 μg/dL), Iron saturation 11.8% (15–45%) and serum ferritin 12.5 ng/mL (25–300 ng/mL), transferrin and iron binding capacity 474.98 μg/dL (250–435 μg/dL). Faecal occult blood was positive, and esophagogastroduodenoscopy was normal. However, colonoscopy revealed multiple angiodysplasia in the splenic flexure and rectum. She underwent clipping and argon plasma coagulation at multiple sites (Fig. 1). AS was detected first time during this admission with transthoracic echocardiogram that showed severe AS, valve peak gradient of 96 and mean 63 mm Hg, Aortic valve area 0.8 cm^2, left ventricular outlet diameter 1.9 cms and left ventricular ejection fraction of 62% (Fig. 2). Her subsequent investigations including molecular

Disclosure: Authors has nothing to disclose with regard to commercial support.

Received 14 December 2020; revised 17 January 2021; accepted 19 January 2021.
Available online 26 April 2021

* Corresponding author at: Department of Cardiothoracic Surgery, Dubai Hospital, Dubai Health Authority, Al Baraha, Dubai, United Arab Emirates.
E-mail address: mkhazi@dha.gov.ae (F.M. Khazi).

https://doi.org/10.37616/2212-5043.1237

genetic analysis for Von Willebrand factor (VWF) gene was negative. The Coomb's test was negative and the Gliadin antibody tests was performed were negative that excluded the diagnosis of haemolysis and celiac disease respectively. The underlying malignancy or other structural diseases of gastrointestinal tract were also excluded.

She was clinically diagnosed with HS with a narrow aortic root complicated by obesity. She continued to suffer from anaemia and required multiple transfusions to improve her haemoglobin. Considering her comorbidities with surgical EuroSCORE II of 16.8%, the initial choice of treatment was decided to be TAVI rather than surgical AVR

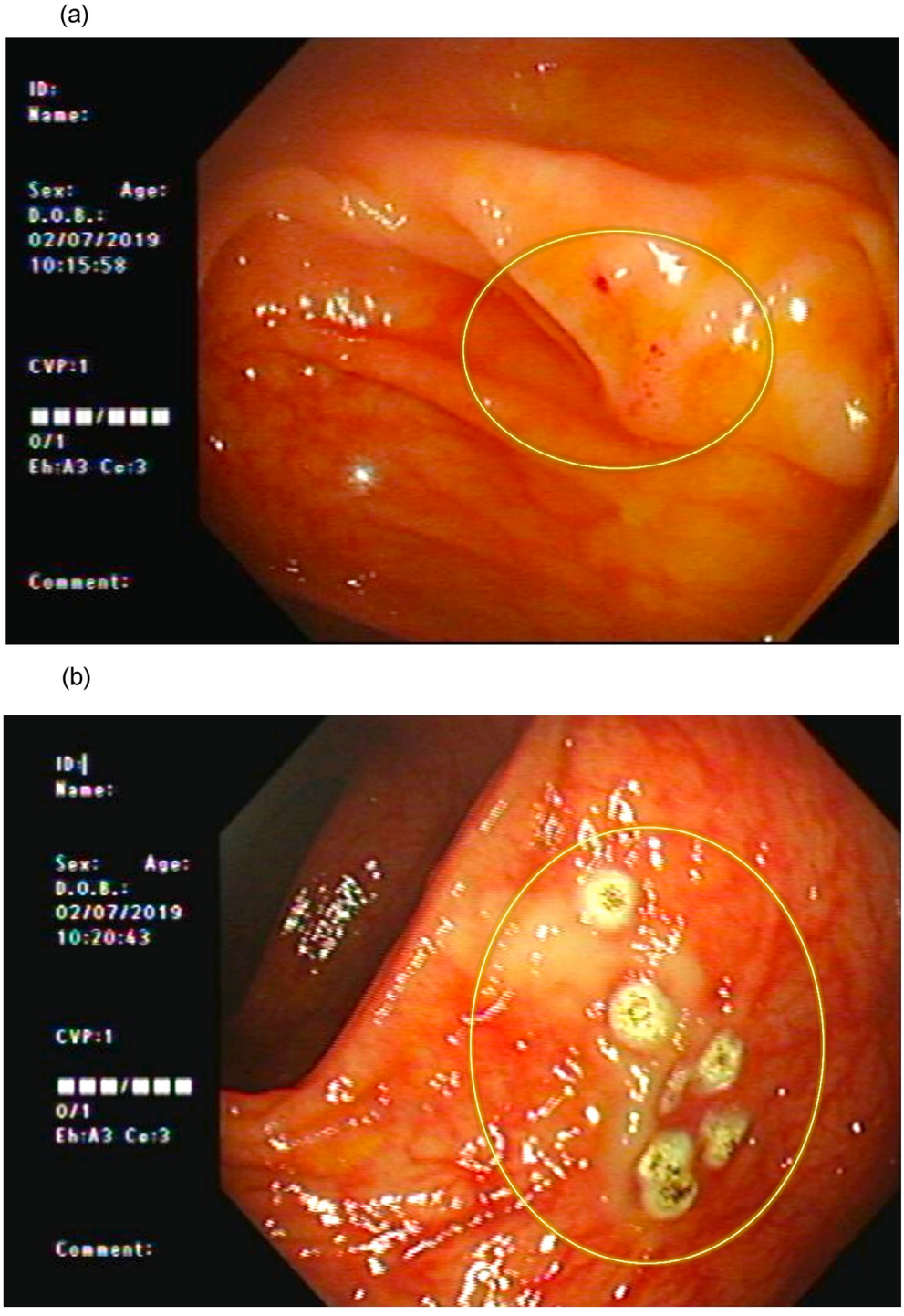


Fig. 1. Endoscopic pictures showing angiodysplasia (yellow circle) in (a) and multiple argon coagulation treatment spots in (b).

(SAVR) during the multidisciplinary (MDT) meeting. However, in view of her calcified narrow aortic outlet in relation to a large body surface area, the decision was finally changed to Bentall procedure, as it will allow us to implant a larger biological valve to avoid turbulence, provide better forward flow and avoid a possible mismatch that can occur with surgical AVR. Patient underwent aortic outlet myomectomy and Bentall procedure with Medtronic freestyle 25 size porcine valve under cardiopulmonary bypass (see Fig. 3). The postoperative course was complicated by bleeding compounded by atrial fibrillation and hypoventilation, resulting in a few unsuccessful weaning trials. Tracheostomy was performed on the 11th postoperative day. She needed several days of high dependency and rehabilitation care. Her tracheostomy was decannulated on the 17th day and discharged home on the 23rd day with a haemoglobin of 10.6 g/dl. She did not show any more signs of intestinal bleeding during her hospital stay or follow-up for twenty months. She was treated with coumadin for 3 months and continued on aspirin.

Discussion

The evidence that AS is the root cause of coagulopathy in HS is compelling, with 5%–20% of these patients manifesting recurrent bleeding [1] with severity depending on the gradients across a stenotic AV [2]. VWS-2A arises from degradation of VWF multimers by shear stress across the AV, resulting in coagulopathy. These multimers are required to maintain haemostasis in high-flow conditions, such as in intestinal arteriovenous malformations in patients with AS [3]. The pathogenesis of angiodysplasia could be due to low-grade chronic

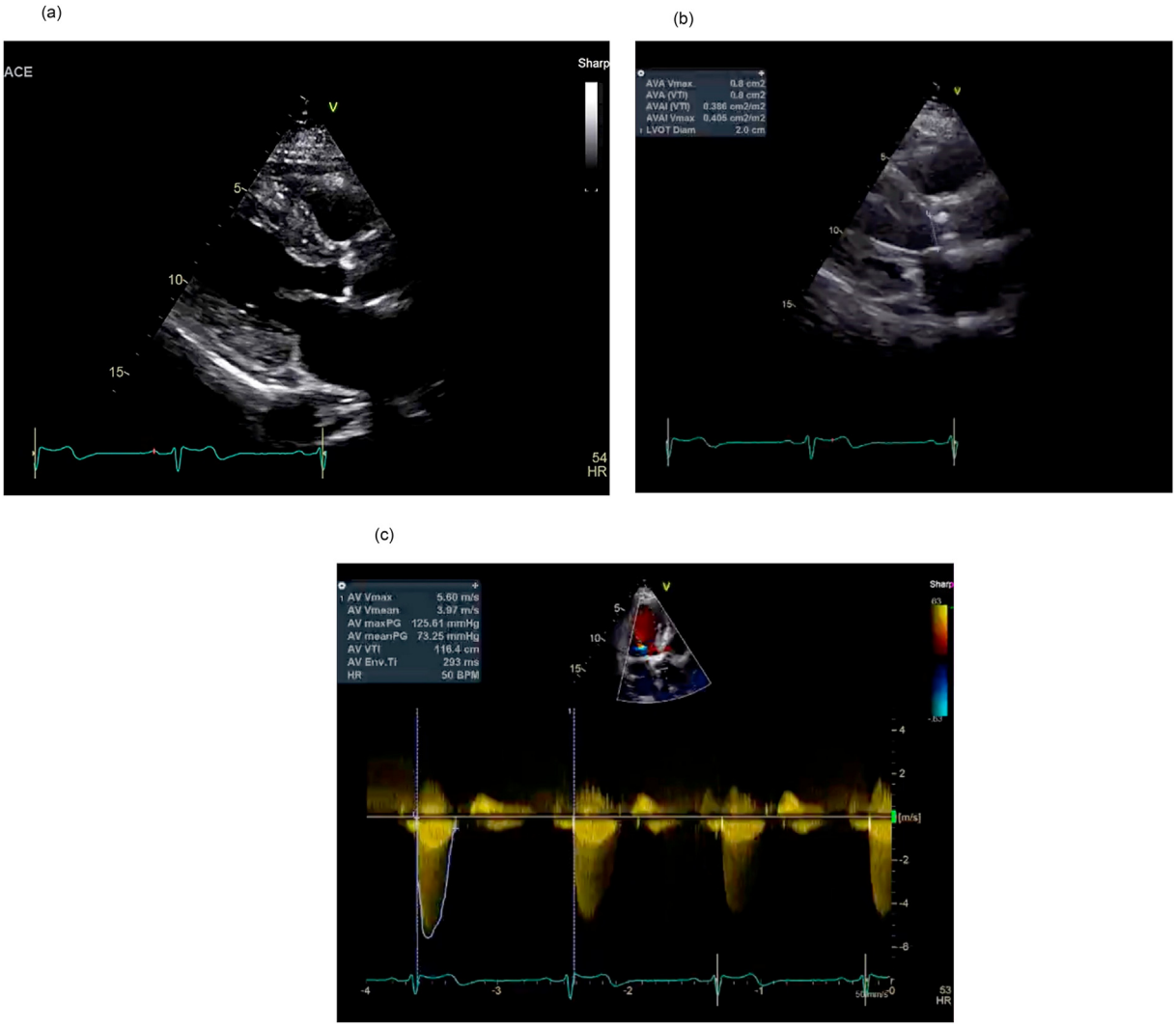
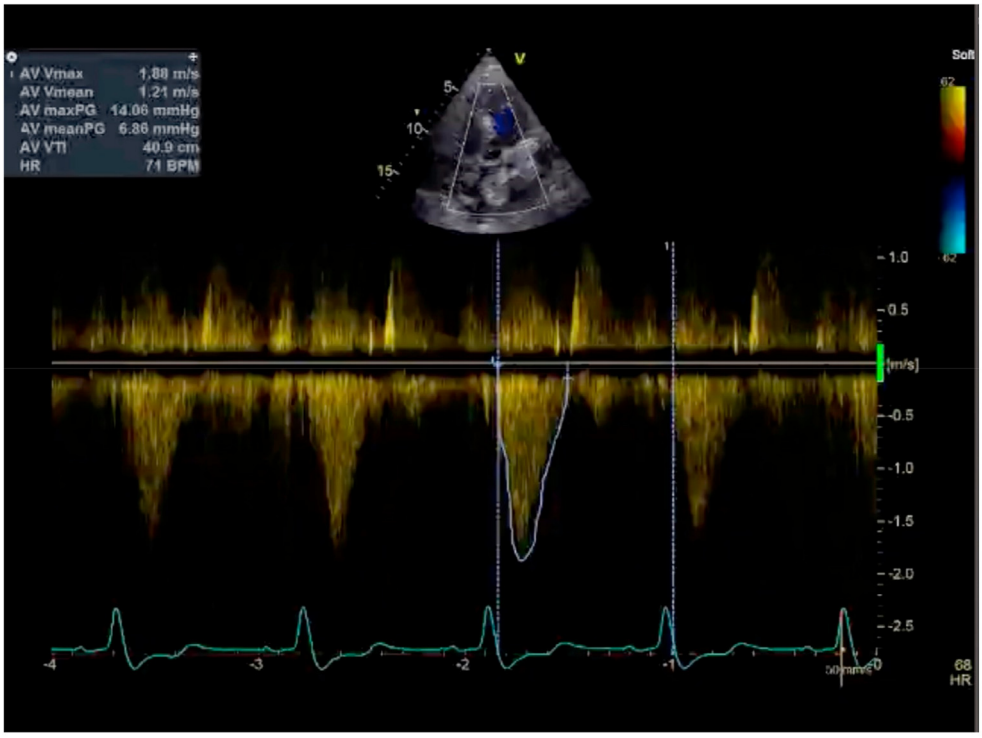


Fig. 2. Preoperative Transthoracic Echocardiography showing severe calcified aortic valve in (a), a narrow LVOT gradient and AV area in (b) and narrow Increased AV gradient in (c).

(a)



(b)

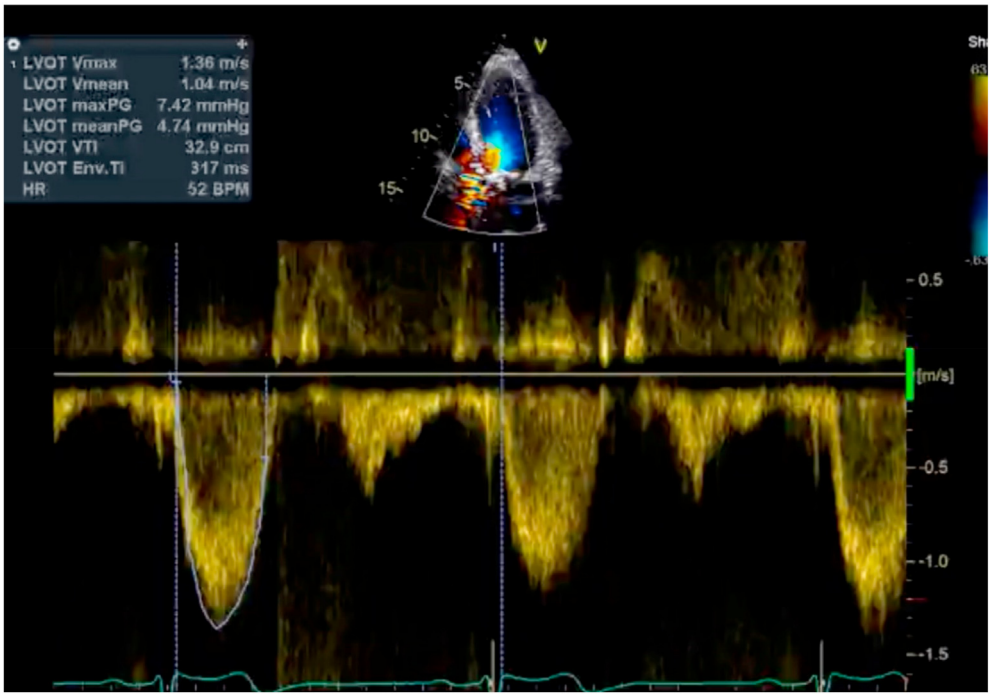


Fig. 3. Postoperative transthoracic Echocardiography showing Decline in postop LVOT gradient in (a) and decline in AV gradient in (b).

hypoxia that stimulates sympathetic vasodilation reflex progressing to fixed smooth muscle relaxation and true ectasia [4]. These could also be due to mucosal hypoxia caused by cholesterol emboli or by the altered pulse waveform secondary to AS. Most of them remain undiagnosed and are found as an incidental finding in 15%–25% of patients with repeated bleeding [5]. Unfortunately, Von Willebrand factor test could not be performed in our patient due to financial constraints and the diagnosis was made on clinical grounds. VWS-2A also occurs in other similar high-stress conditions such as left ventricular assist device implantation, hypertrophic obstructive cardiomyopathy, ventricular septal defect and patent ductus arteriosus.

Even though AV replacement eliminates the risk of bleeding [1], the procedure itself may cause serious bleeding complications. Other issues such as type of procedure (open vs interventional), prosthesis (mechanical vs biological) and perioperative anticoagulation are always debatable. More recently, TAVI has emerged as a feasible option with bioprosthetic valve [7] and postoperative low-dose aspirin. In the presence of prosthesis mismatch, GI bleeding has been found to recur because of the persistence of a pathophysiological mechanism. We opted for Bentall procedure, as it allowed us to place a larger valve with less gradient and better haemodynamics. Unlike surgery, endoscopic cauterisation alone was only temporarily successful and was associated with a high recurrence rate [6]. Our patient could even tolerate warfarin anticoagulation without bleeding recurrence following corrective surgery for three months. We opted 3 months of coumadin anticoagulation followed by aspirin as per recent guidelins [9]. However, we fully agree that the associated morbidity was not negligible, with significant postoperative complications and increased length of stay. Sleep-disordered breathing is highly prevalent in obese patients [8] with associated upper airway muscle weakness, vascular endothelial dysfunction, cardiac arrhythmia, alveolar hypoventilation and sensitivity to anaesthetic medications leading to sudden cardiovascular death, myocardial infarction, stroke, worsening overall postoperative recovery and higher pacemaker use [10].

HS management requires a multidisciplinary approach. Treatment options include octreotide infusion, endoscopic interventions, colon surgery and AV replacement. Surgical excision of angiodysplasia is a high risk in the presence of AS, and studies have shown that bleeding recurrence remains with underlying AS pathophysiology [11]. If AVR cannot stop bleeding, subsequent anaesthesia for bowel resection would be less risky. In our patient, improvement in anaemia was possibly due to the resolution of angiodysplasia with improved cardiac output and recovery of VWF multimers. This case report suggests that more radical procedures offered to these high-risk complicated HS patients, such as Bentall procedure, can still be associated with favourable outcomes. However, a careful understanding of pathophysiology, preoperative optimisation including a multidisciplinary approach, endoscopic treatment, blood transfusion, perioperative echocardiography with appropriate postoperative ventilatory and rehabilitation strategy will be key to successful results.

To the best of our knowledge, this is the first patient who successfully underwent Bentall procedure in complicated HS with complete resolution of anaemia despite coumadin anticoagulation. Larger prospective studies are required to revise AVR guidelines to include anaemia and GI bleeding as criteria of the type, timing and type of surgery and anticoagulation regimen.

Author contribution

Fayaz Mohammed Khazi: Conception and design of Study. **Fayaz Mohammed Khazi, Nayyer R Siddiqi, Yehia Mohamed Karaly, Obaid Aljassim, Zohair Y. Al-Halees:** Literature review, Acquisition of data, Analysis and interpretation of data, Research investigation and analysis, Data collection, Drafting of manuscript, Revising and editing the manuscript critically for important intellectual contents, Data preparation and presentation.

References

[1] Massyn MW, Khan SA. Heyde syndrome: a common diagnosis in older patients with severe aortic stenosis. Age Ageing 2009;38:267–70. https://doi.org/10.1093/ageing/afp019.

[2] Vincentelli A, Susen S, Le Tourneau T, Six I, Fabre O, Juthier F, et al. Acquired von Willebrand syndrome in aortic stenosis. N Engl J Med 2003;349(4):343–9. https://doi.org/10.1056/NEJMoa022831.

[3] Garcia LR, Garzesi AM, Tripoli G, Campos NLK, Martins AS, Felicio ML. Heyde syndrome treated by conventional aortic valve replacement. Braz J Cardiovasc Surg 2019;34(5):630–2. https://doi.org/10.21470/1678-9741-2019-0004.

[4] Godino C, Lauretta L, Pavon AG, Mangieri A, Chieffo A, Galaverna S, et al. Heyde's syndrome incidence and outcome in patients undergoing transcatheter aortic valve implantation. J Am Coll Cardiol 2013;61(6):686–91. https://doi.org/10.1016/j.jacc.2012.10.041.

[5] Schwaiger JP, Ludwiczek O, Graziadei I, Grander W. A vicious circle: heyde syndrome in mild aortic stenosis. CASE: Cardiovasc Imaging Case Rep 2019;3(4):171–6. https://doi.org/10.1016/j.case.2019.04.005.

[6] Pate GE, Chandavimol M, Naiman SC, Webb JG. Heyde's syndrome: a review. J Heart Valve Dis 2004;13:701–12.
[7] Desai R, Parekh T, Singh S, Patel U, Fong HK, Kumar G, et al. Alarming increasing trends in hospitalizations and mortality with heyde's syndrome: a nationwide inpatient perspective (2007 to 2014). Am J Cardiol 2019 Apr 1;123(7): 1149–55. https://doi.org/10.1016/j.amjcard.2018.12.043.
[8] Abedinov F, Bakalova N, Krastev P, Petrov L, Marinova R, Tsaryanski G. Survival and quality of life of patients with a prolonged stay in the intensive care unit after cardiac surgeries-Remote results. C R Acad Bulg Sci 2019;72(8):1130–6. https://doi.org/10.7546/CRABS.2019.08.16.
[9] Cremer P, Barzilai Benico B. Anticoagulation strategies after bioprosthetic valve replacement: what should we do? J Am Coll Cardiol Dec 19: 2016.
[10] Ding N, Ni BQ, Wang, Ding WX, Xue R, Lin W, et al. Obstructive sleep apnea increases the perioperative risk of cardiac valve replacement surgery: a prospective single-center study. J Clin Sleep Med 2016;12(10):1331–7. https://doi.org/10.5664/jcsm.6182.
[11] Michot JM, Treton X, Brink C, Fabiani JN, Bouhnik Y. Severe gastro-intestinal angiodysplasia in context of Heyde's syndrome durably cured after aortic valve replacement. Presse Med 2012;41:763–6. https://doi.org/10.5551/jat.49239.