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Chest pain and syncope in Turner"s syndrome: going beyond the obvious to not miss the critical diagnosis. Role of multimodality imaging approach

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Clinical Presentation: a 18-year-old woman with Turner's syndrome (TS), with history of hypothyroidism treated with L-thyroxin, asymptomatic moderately stenotic bicuspid aortic valve (AV) and without any known cardiovascular risk factor, was admitted to our emergency department (ED) because of syncope and typical chest pain after dinner associated with dyspnea. Chest pain lasted for an hour with spontaneous regression. In the ED the patient (pt) was normotensive. An ECG showed sinus rhythm (88 bpm), nonspecific repolarization anomalies (T wave inversion) in the inferior and anterior leads. Myocardial necrosis biomarkers were negative. A 3D transthoracic echocardiography showed normal biventricular systolic function with left ventricular hypertrophy, dilatation of the ascending aorta, unicuspid AV with severe aortic stenosis (peak/mean gradient 110/61 mmHg, aortic valve area 0,88 cm2-0,62 cm2/m2), mild pericardial effusion (Figure Panel A, B, C). Five days after, the pt had a new episode of typical chest pain without ECG changes. A computerized tomography (CT) was performed to rule out the hypothesis of aortic dissection and showed a dilation of the ascending aorta and pericardial effusion localized in the diaphragmatic wall, no signs of dissection or aortic hematoma. However, CT was of suboptimal quality because of sinus tachycardia (120 bpm) and so the pt underwent a coronary angiography and aortography that ruled out coronary disease, confirmed the dilatation of ascending aorta (50 mm) and showed images of penetrating atherosclerotic ulcer of the ascending aorta (Figure panel D). The pt underwent urgent transesophageal echocardiography (TOE) that confirmed the severely stenotic unicuspid AV and showed a localized type A aortic dissection (Figure Panel E, F, G). The pt underwent urgent AV and ascending aorta replacement (Figure Panel H).

Learning points: Chest pain and syncope are challenging symptoms in pts presenting in ED. AV pathology and aortic dissection should be always suspected and ruled out. TS is associated with multiple congenital cardiovascular abnormalities and is the most common established cause of aortic dissection in young women. 30% of Turner's pts have congenitally AV abnormalities, and dilation of the ascending aorta is frequently associated. However, unicuspid AV is a very rare anomaly, usually stenotic at birth and requiring replacement. The presence of pericardial effusion in a pt with chest pain and syncope should raise the suspicion of aortic dissection, even if those symptoms usually accompany severe aortic stenosis. Even if CT is the gold standard imaging technique to rule out aortic dissection, the accuracy of a test is critically related to the image quality. When the suspicion of dissection is high and the reliability of the reference test is low, it's reasonable to perform a different test to rule out the pathology. Aortography and TOE were pivotal to identify the limited dissection of the ascending aorta.

Abstract P190 Figure.

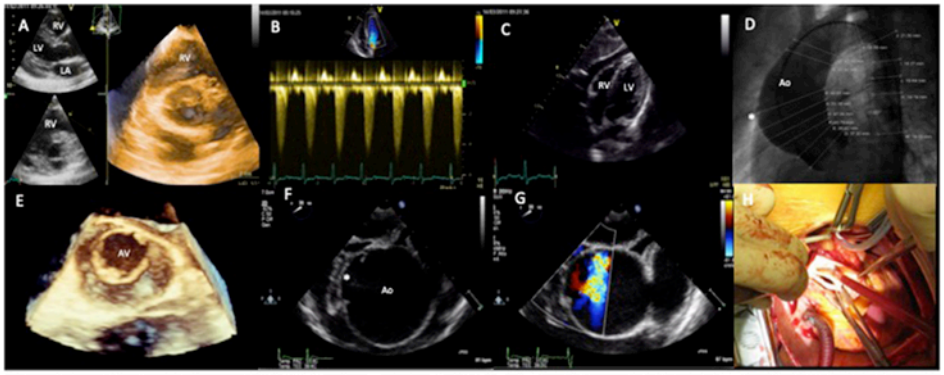

Figure. Panel A: a transthoracic 3 D single beat acquisition and 3 D rendering of the unicuspid aortic valve. **Panel B**: a Continuous Wave Doppler on the Aortic valve shows increased gradients. **Panel C:** a subcostal transthoracic view, white arrow points at the mild pericardial effusion. **Panel D:** aortography showing a dilatation of ascending aorta. **Panel E:** a transesophageal 3D rendering of the unicuspid aortic valve. **Panel F and G:** transesophageal aortic views of the aortic dissection. **Panel H:** unicuspid aortic valve surgery view.
Abbreviations: Ao: Aorta; AV: aortic valve; LA: left atrium; LV: left ventricle; RV: right ventricle. *: aortic dissection.