Acquired deficit of Type IIa von Willebrand factor, characterized by heterogeneity.

Coronary syndrome (ACS) and sudden death. It has traditionally been performed with implantation of a drug-eluting stent, the anterior descending artery of a young woman, we decided to perform angioplasty with implantation of a drug-eluting stent, by loss of the largest VWF multimers, although this causal relationship cannot always be demonstrated, as in this case.

Von Willebrand factor is a high-molecular-weight multimeric protein secreted by endothelial cells that stimulates platelet adhesion and aggregation when there is vascular damage. These multimers are cleared by plasma proteases that are especially active in turbulent blood flow situations.

In aortic stenosis, fragmentation of VWF multimers is increased, which reduces their number and predisposes bleeding. Studies have reported that these coagulation anomalies are directly related to the severity of aortic stenosis and are reversible after valve replacement if successful, so that recurrence of bleeding could be an indication of persistent stenosis. Therefore, although in some cases of extensive bleeding, such as this one, intestinal resection is necessary, many authors have shown that gastrointestinal bleeding ceases after valve replacement, being even more likely to prevent recurrences than intestinal resection.

Heyde’s Syndrome is an entity to be kept in mind, even more nowadays with an aging population, when assessing patients with a history of bleeding or anemia, especially when the bleeding site is not found on initial examination. Based on these data, we propose the hypothesis that this association could be a new indication for valve replacement; however, gastrointestinal bleeding is not used as an indicator in current clinical practice guides.

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Spontaneous Coronary Dissection: Role of Intravascular Ultrasound
Disección coronaria espontánea: papel del ultrasonido intravascular

To the Editor,

A 49-year-old woman with no cardiovascular risk factors was admitted to our department with high-risk non-ST-elevation acute coronary syndrome. Coronary angiography revealed severe stenosis in a segment located in the middle portion of the anterior descending artery, with no evidence of other lesions (Fig. 1). Given the ambiguity of the angiogram and the absence of risk factors, we decided to perform intravascular ultrasound, which revealed localized dissection that had not been visible in the angiogram (Fig. 1). There was no apparent intimal rupture but there was evidence of hematoma within the intima. Given the lack of information on the course of the lesion, and since it was located in the anterior descending artery of a young woman, we decided to perform angioplasty with implantation of a drug-eluting stent, with excellent angiographic results (Fig. 2).

Spontaneous coronary artery dissection is a rare cause of acute coronary syndrome (ACS) and sudden death. It has traditionally been described in 3 groups of patients: patients with coronary artery disease, women during the peripartum period, and a heterogeneous group of patients with idiopathic disease, comprising individuals without cardiovascular risk factors, those with connective tissue disorders (Marfan syndrome, Ehlers–Danlos syndrome, etc.), and cases of intoxication.

There is a clear predominance of women (80% of cases), with a mean age at presentation of 40 years and 25% of cases occurring in the peripartum period. The overall incidence ranges from 0.1% to 1.1%. The most commonly affected artery in women is the anterior descending, whereas in men the right coronary artery is more often affected. Cases of multivessel dissection and of left main coronary artery dissection have also been described.

The pathophysiology involves hemorrhage between the medial layer and the external elastic lamina, leading to separation of the layers, expansion of a false lumen, and compression of the native lumen that results in ischemia. Rupture and hemorrhage of the vasa vasorum has been suggested as a potential mechanism. Eosinophilic infiltrates have also been observed in the dissected region, suggesting a possible inflammatory or vasculitic mechanism, although some authors maintain that the presence of inflammatory cells is a consequence rather than a cause of the dissection. The association with the peripartum period and the use of oral contraceptives supports the hypothesis that the condition occurs as a result of connective tissue changes induced by hormones (endogenous or exogenous) and the hemodynamic changes characteristic of pregnancy. Numerous cases have also been reported in association with acute changes in intrathoracic pressure. Nevertheless, in many patients, no clear trigger is observed and it is likely that there are multiple etiologies.

Diagnosis is based on observation of a radiolucent intimal flap in the coronary angiogram. Diagnosis using angiography alone is often difficult when there is no apparent intimal rupture, with no
observable flap and only vessel stenosis visible. Consequently, the use of intravascular ultrasound is important in young patients with no cardiovascular risk factors in whom spontaneous coronary dissection is suspected, since this allows diagnosis and can differentiate between the false and true lumens.

Consensus is lacking on the most appropriate treatment options, which depend on factors such as the number of affected vessels, whether the left main coronary artery is affected, distal coronary flow, and clinical stability. Thus, patients who are clinically unstable or have poor distal flow should be treated by percutaneous transluminal coronary angioplasty (PTCA) in localized dissections, whereas surgical revascularization would be a more appropriate option in patients with multivessel dissections, long lesions, or lesions of the left main coronary artery. In patients who have ACS with localized dissection and who remain asymptomatic, hemodynamically stable, and have good distal coronary flow, conservative treatment can be employed with double antiplatelet therapy and β-blockers, with angiographic follow-up in a few months, since in many cases the dissection is found to completely reseal.\(^2,3\) Other authors favor early, aggressive treatment with revascularization, since the natural course of this process is poorly understood.

In the case of PTCA, intravascular ultrasound should be used to ensure that the guidewire is in the native lumen and that stent placement does not cause propagation of the dissection or protrusion of a thrombus at one end.\(^3\)

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Acute Heart Failure in an Adult Patient With 2:1 Atrial Flutter: Zebras or Horses?

Insuficiencia cardiaca aguda en un adulto con flutter auricular 2:1: «¿cebras o caballos?»

To the Editor,

We present here the case of a 47-year-old patient with no past medical history of interest who went to the Accident and Emergency Department suffering from palpitations, progressive dyspnoea and orthopnoea. Examination revealed a II/IV systolic murmur; bibasal crackles; preserved and symmetrical pulses, albeit weak; blood pressure of 100/70 mmHg and cardiomegaly on chest X-ray. An electrocardiogram revealed 2:1 atrial flutter with a ventricular rate of 150 bpm. The emergency room, based on an initial diagnosis of suspected “tachycardiomyopathy”, administered digoxin, beta blockers, diuretics, and supplemental oxygen, thereby controlling the ventricular rate (75–100 bpm) with rapid clinical improvement. The next day, an echocardiography was performed that revealed left ventricular dysfunction with ejection fraction of 35%, dilated aortic root, moderate aortic insufficiency, and an abnormal mitral subvalvular apparatus with very elongated chords with a myxoid appearance with moderate mitral regurgitation (Fig. 1).

Given the new data, a multislice CT was performed, which showed a 49 mm aortic root, severe aortic coarctation (Fig. 1) and left atrial thrombus with no coronary abnormalities. Reviewing the basic diagnostic tests, the chest X-ray showed small costal notches that had gone unnoticed. The final diagnosis was left ventricular dysfunction by severe aortic coarctation with associated lesions of aortic and mitral insufficiencies. The rhythm became atrial fibrillation and cardioversion was not indicated by the left atrial thrombus. Cardiac catheterization was performed in an attempt to interrupt the aortic arch gradient of 40 mmHg. A radiofrequency catheter was used to get past the coarctation, allowing subsequent dilation and stenting (Fig. 2) with a disappearance of the gradient. At six months, the patient was asymptomatic, with an ejection fraction of 45% and the patient remained in sinus rhythm and asymptomatic with slight mitral regurgitation.

“Zebras or horses?” This case clearly illustrates one of the basic axioms of medicine: “when you hear hoofbeats behind you, it is almost certainly the sound of horses and not zebras.” This can be translated as “what is most frequent is most likely.” The presence of 2:1 atrial flutter with severe ventricular dysfunction in a young man without any past medical history made us think of the most obvious: dilated cardiomyopathy secondary to tachycardia. However, the echocardiogram revealed valvular disorders (pronounced myxoid degeneration of the mitral chordae and aortic annuloectasia). A subsequent multislice CT revealed another previously undetected disorder (severe aortic coarctation). Reviewing the medical history, blood pressure was normal (which can be due to heart failure), and femoral pulses were present and symmetrical in the arms (possibly due to collateral circulation). Only after a careful review of the X-rays were the small costal notches observed. Coarctation of the aorta constitutes 6% of congenital heart disease in childhood and 15% in adulthood. Its clinical manifestations depend on severity: in mild cases the manifestations do not appear until adulthood, usually with the discovery of hypertension. In our patient, however, there were no disorders until the onset of

Figure 1. Left: echocardiography showing linear images within the left ventricular cavity corresponding to very elongated mitral chordae (1). Right: aortic CT with striking collateral circulation (2), an image of aortic interruption (3), and dilation of the aortic root (4).