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Guidewire-driven Left Ventricular Pacing During Transcatheter Aortic Valve Implantation

Estimulación ventricular izquierda a través de la guía del implante percutáneo de válvula aórtica

To the Editor,

Cardiac pacing at 180 to 200 bpm is an effective means to stabilize the balloon during aortic valvuloplasty and transcatheter aortic valve implantation (TAVI). Classic temporary pacing requires femoral or jugular puncture for placement of the active fixation electrode in the right ventricle (RV). However, effective cardiac pacing can also be achieved using a unipolar electrode in the left ventricle (LV).[1] We describe a prospective and consecutive series of 25 patients (Table) who underwent femoral TAVI. Because evaluation by the cardiology team responsible for patient selection revealed that the patients were at high or moderate risk[2,3] from conventional valve replacement, they were selected for TAVI, preferably using a minimalist approach (sedation, without systematic transesophageal echocardiography, without previous aortic valvuloplasty, percutaneous femoral occlusion). In addition, implantation of an active fixation electrode in the RV was omitted because, in our first 105 TAVI procedures with expandable balloon (EB) valves, RV perforation occurred in 2 patients and rapid LV pacing was induced through a 0.035" super-stiff guidewire used for the TAVI.

All patients had severe aortic stenosis with elective or emergent indication for TAVI or dysfunction of a valve prosthesis previously implanted via aortic valve replacement surgery. Once the aortic valve was crossed with a 5-Fr Amplatz AL1 catheter, the guidewires used were a 0.035" super-stiff wire (Cook; Bloomington, Indiana, United States) precurved in the shape of a pig's tail and placed within the LV or a Safari guidewire (Boston Scientific), which is already spiral-shaped and did not require manipulation. The extreme end of the guidewire was connected to the negative electrode (cathode) of a Medtronic 5348 temporary pacemaker using an alligator clamp while the positive electrode (anode) was connected to the skin of the right inferior extremity using an intramuscular or curved needle and similarly connected by an alligator clamp to the external pacemaker (Figure A). Pacing was performed at between 180 and 240 bpm, with a maximal output and reduced sensitivity. We waited until the blood pressure was reduced to 40 mmHg with a pulse wave of about 10 mmHg (Figure B) before implanting the Edwards-SAPIEN 3 and Edwards-SAPIEN XT EB-type valves (Figure C), with a 0 to 5 mmHg reduction in the systolic gradient of the LV and aorta and no regurgitation (Figure D and video of the supplementary material). In our series, there was 1 incident of complete atrioventricular block that required pacing via the 0.035" guidewire during the time required to implant a temporary pacemaker through a jugular approach to the RV; the patient subsequently required a permanent pacemaker.

We believe that rapid pacing should not be used when there are predictors of atrioventricular block after TAVI, self-expanding valves are used, or the center has limited experience.[5] We have evidence from only 2 publications[4,6] that used TAVI guidewire-driven rapid LV pacing. Faurie et al.[4] did not report the number of TAVIs with EBs and self-expanding valves but reported an intraprocedural temporary pacing rate due to post-TAVI blocks of 13.8%, estimating a predominance of the self-expanding valve, which does not necessarily involve rapid pacing. In the series by Hilling-Smith et al.,[6] permanent pacemaker implantation was required after TAVI in 21.2% of patients, and an EB was used in only 6% of patients. In our prospective and consecutive series, all prostheses used had EBs, and the rapid pacing mode allowed the implantation of valves with EBs, elimination of potential complications due to the need for additional venous punctures, and

Table
Patients' Characteristics, Surgical Risk, and 30-day Results

Patients, n	25
Age, y	79.2 ± 4.6
EuroSCORE II, %	5.35 ± 3.9
STS, %	5.81 ± 4.2
Previous pacemaker	1 (4)
Previous percutaneous coronary intervention	6 (24)
NYHA class III/IV	11 (44)
Porcelain aorta	1 (4)
Chronic kidney disease stage IV/V	5 (20)
Edwards-SAPIEN 3	19 (76)
Edwards-SAPIEN XT	6 (24)
Direct implantation	23 (92)
Dysfunctional surgical aortic valve prosthesis	7 (28)
Pacing at 180 bpm	18 (72)
Pacing at 200 bpm	3 (12)
Pacing at 220 bpm	3 (12)
Pacing at 240 bpm	1 (4)
Pacing failure	0
Pacemaker 30 d after the TAVI	2 (8)
Tamponade	0
Vascular complication	1 (4)
Stroke	1 (4)
Periprocedural myocardial infarction	0
Death	1 (4)
TAVI procedure success	25 (100)
TAVI success with complications	24 (96)

EuroSCORE II, European system for cardiac operative risk evaluation; NYHA, New York Heart Association functional class; STS, cardiac surgery mortality risk score of the Society of Thoracic Surgeons; TAVI, transcatheter aortic valve implantation. Unless otherwise indicated, the data represent No. (%).

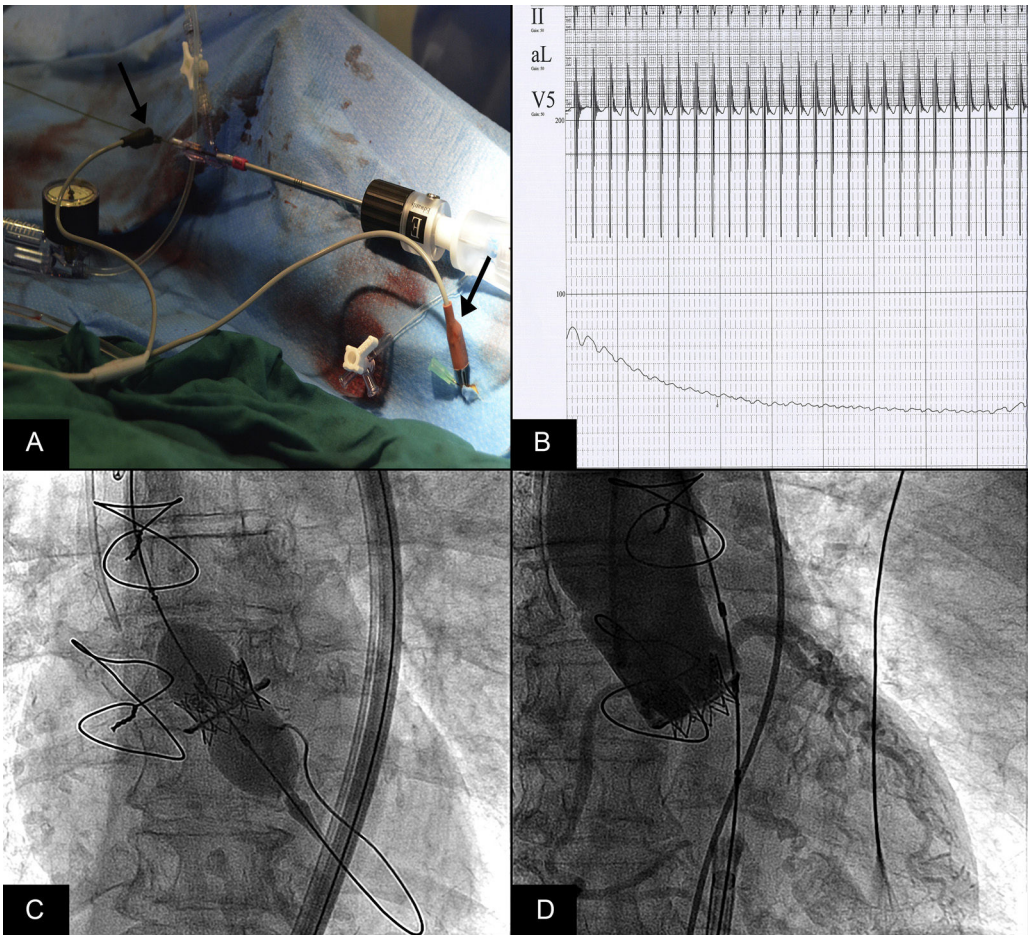

Figure. A: The left arrow indicates the negative electrode (cathode) connected to the guidewire and the right arrow indicates the positive electrode (anode) connected to an intramuscular needle in the skin. B: Reduction in the aortic pressure curve during rapid left ventricular pacing. C: Expansion of the SAPIEN XT prosthesis with rapid pacing through the guidewire contacting the left ventricle. D: Immediate result after prosthesis implantation.

avoidance of additional problems, costs, and minor risks associated with the creation of arteriovenous fistulas, as well as complications from RV perforation with the active fixation electrode of the temporary pacemaker required for rapid pacing at the time of Edwards-SAPIEN 3 and XT valve implantation. Effective pacing of the LV requires good isolation of the guidewire with the prosthesis insertion device, adequate contact between the guidewire and the LV endocardium, and correct assembly with the pacing system, where the guidewire is used as the cathode and the electrode in the skin as the anode connected to an intramuscular needle or wire in an anesthetized area.

Our series of patients who underwent TAVI with EB supports our continued use of high-support guidewire-driven pacing of the LV as a safe and effective alternative to conventional right ventricular pacing in TAVI.

SUPPLEMENTARY MATERIAL

Supplementary material associated with this article can be found in the online version available at http://dx.doi.org/10.1016/j.rec.2017.08.009.

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The p.Arg118Cys Variant in the *GLA* Gene Does not Cause Fabry Disease. More Evidence

La variante p.Arg118Cys en el gen GLA *no causa enfermedad de Fabry. Más evidencias*

To the Editor,

Fabry disease (FD) is an X-linked disorder caused by mutations in the *GLA* gene. This leads to deficiency in α-galactosidase A, the enzyme responsible for degrading certain glycosphingolipids. The enzyme deficiency leads to accumulation of these compounds, in turn resulting in dysfunction of vital organs (mainly the kidney and heart) and premature death. Both sexes are affected in similar proportions, but the disease is usually less serious and presents later in life in women. FD is classed as a rare disease, with a frequency of between 1:40 000-1:120 000.[1]

Diagnosis is challenging and cardiologists are often the first to suspect the disease after patients are referred for investigation of hypertrophic cardiomyopathy (HCM). The diagnosis is confirmed by genetic study, which can prompt a family study; however, there are genetic variants of doubtful pathogenicity.[1] Pathogenicity is, however, a crucial consideration as enzyme replacement therapy (ERT) is available but is expensive and not free of complications.

We present the case of a family with FD in which the p.Arg118Cys variant was demonstrated not to cause the disease, thus highlighting the need to be alert to other similar variants.

A 45-year-old man with proteinuria since he was 36 years old and whose renal function was deteriorating underwent a kidney biopsy. Electron microscopy showed zebra bodies indicative of FD (Figure A). In view of these findings, the patient was referred to the hereditary heart disease unit to complete the study. Echocardiography showed severe left ventricular hypertrophy (LVH) and electrocardiography revealed short PR interval and LVH (Figure B and C). A genetic study was conducted by next-generation sequencing of a panel of 16 genes associated with HCM (including *GLA*). The p.Arg118Cys variant was detected, but deletion of exons 3 and 4 (p.Val124_Lys213del) was also present.

The family study (Figure D) found that the patient's mother also had HCM and had died of kidney failure at age 64 years. Some family members were living in another region of Spain, and we therefore

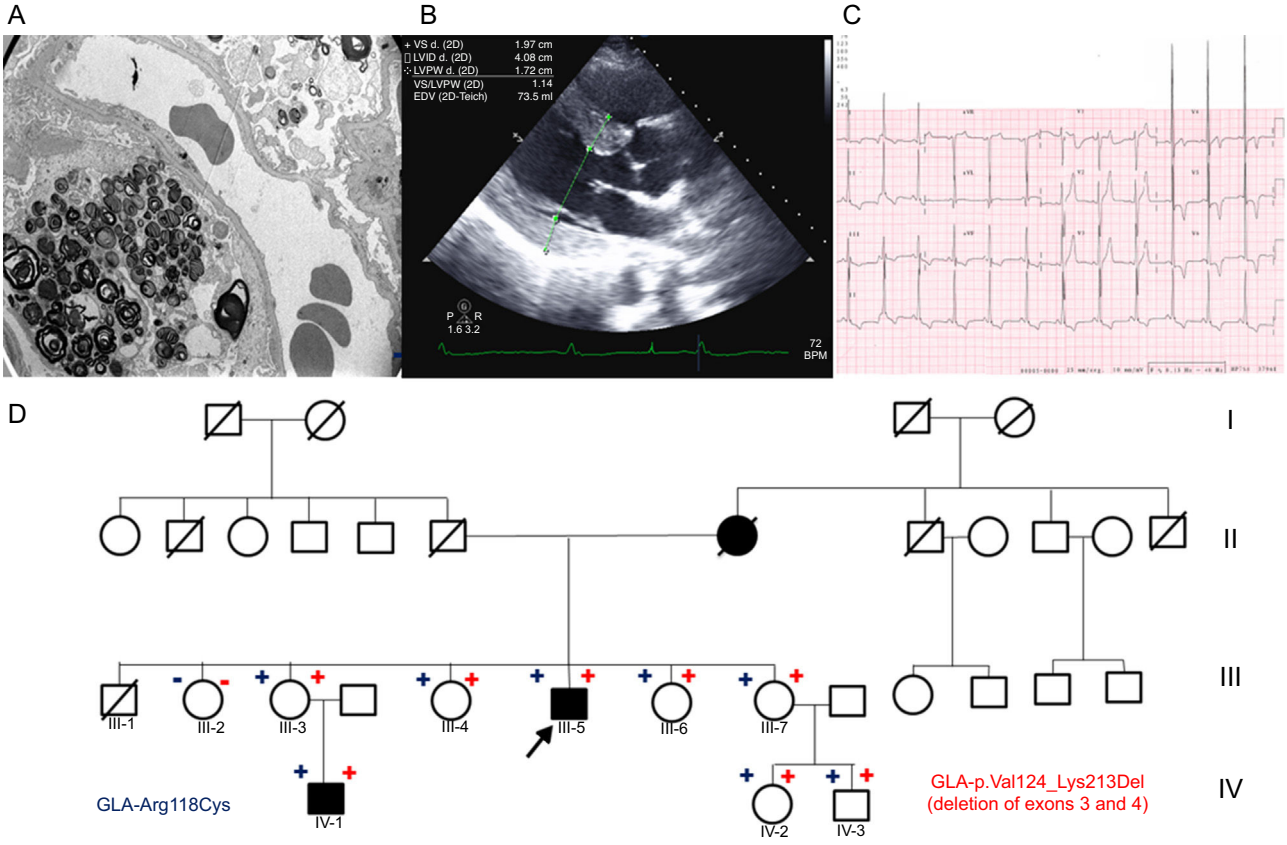

Figure. A: Electron microscopy of kidney biopsy with zebra bodies. B: Echocardiogram of the index case with LVH. C: Electrocardiogram of index case with LVH and short PR interval. D: Family tree; square, male; circle, female; in boldface, phenotype involvement; arrow, proband; diagonal, dead; + red, carrier of exon 3 and 4 deletion; -red, noncarrier of exon 3 and 4 deletion; + blue, carrier of p.Arg118Cys variant; blue, noncarrier of the p.Arg118Cys variant. LVH, left ventricular hypertrophy.