Medical and Research Publications
International Open Access

Case Report **Journal of MAR Cardiology (Volume 3 Issue 5)**

Clinical Cases Implantation of PM and ICD in Thriven of an Abnormality of the left Superior Vena Cava and the Review of the Literature.

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Received Date: October 18, 2021

Published date: November 01, 2021

Introduction:

We proceeded to the implantation of 5 patients: 2 pacemaker **(**PM**)** and 3 defibrillators **(**ICD**)** with abnormality of the left superior vena cava.

Even if it is a rare anomaly, it is interesting to descripting to ovoid failure of implantation. Because there is a little more lead implantation failure.

Population	Age	Sex	HTA	CM	TRT
Patient 1	63	M	+	No cardiomyopathy	ACE inhibitor+ Ca bloquer
Patient 2	65	M	+	Hypertrophic Cardiomyopathy	ACE inhibitor + Ca bloquer
Patient 3	62	M	+	Dilate cardiomyopathy	ACE inhibitor + Ca bloquer+ ADO
Patient 4	64	M	-	Ischemic cardiomyopathy	+ ACE inhibitor BB + AC + Diu
Patient 5	59	M	-	Congenital cardiomyopathy	Ca bloquer

Table: Study population

First patient:

A 63-year-old man, presented to the emergency room for dyspnea that started 2 days ago. He was hospitalized in intensive care unit **(**ICU**)** because the Electrocardiogram **(**ECG**)** shows a complete atrioventricular block **(**CAVB**)** with narrow QRS escape and heart rate **(**HR**)** at 34 beats per minute **(**bpm**)**.

Clinical examination found 130/70mmHg of arterial pressure, a HR at 40 bpm and a systolic murmur 2/6 at the apex. No signs of heart failure were found.

The analysis of the ECG described a CAVB with HR at 34 bpm associated to a right bundle brunch block **(**BBD**)**. The chest X-ray and biological assessment were normal leading to the indication for implantation of a pacemaker **(**PM**)** for degenerative CAVB.

We approached the right deltopectoral groove as usual for PM installation. The introduction of the right ventricular **(**RV**)** lead into the cephalic vein had an unusual progression along the clavicle and then the left edge of the sternum. The probe was removed; and we performed an angiogram which showed a type C left superior vena cava **(**LSVC**)**.

The procedure was particularly difficult, because it was necessary to carry out a preformation of the mandrels to be able to stabilize probes at the apex of the RV and the lateral wall of the right atrium **(**RA**)**. The large size of the coronary sinus **(**CS**)** exposed to several displacements of the probes before fixation.

Probes were actively binding to avoid secondary displacement. The measurements of potentials and thresholds were suitable **(**atrium: potential = 4.8mV, threshold = 0.5V, Impedance = 430 Ohms. Ventricle: potential = 5.7mV, threshold 0.75 V, Impedance = 560 Ohms**)**.

The postoperative chest X-ray as follows illustrates the path and the position of probes.

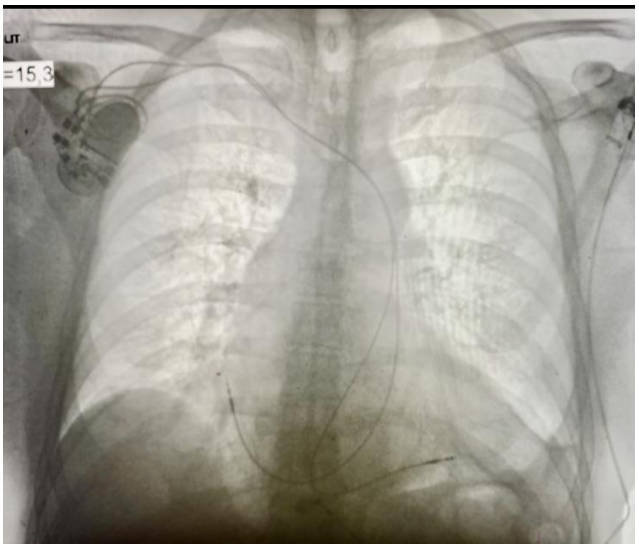


Figure 1: Final X ray

Patient 5 :

A 59-year-old man referred by his cardiologist for outbreak of heart failure.

This patient had as CIA ATCD operated in twist **(**in 70 & 83**)** with aortic bicuspid having undergone a commissurotomy. Clinical examination was BP 110/70 mmHg, RAC systolic breath and congestive heart failure

ECG: shows in sinus rhythm with left bundle branche block Cardiac ultrasound: showed moderately aortic narrowing area at 1cm² and LVEF at 30% The measurements of potential and thresholds were suitable **(**Atrium: potential = 2 mV, Threshold = 0,8 V, Impedance =560 Ω, RV: potential = 9,6mV , Threshold = 0,5 V , Impedance =630 Ω , LV : potential = 7,8mV , Threshold = 1,5 V , Impedance =1130 Ω **)**

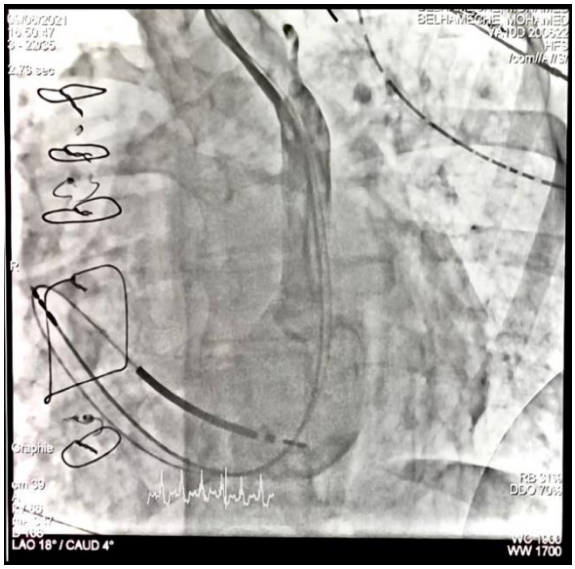


Figure 2: angiographic anatomy of the coronary sinus

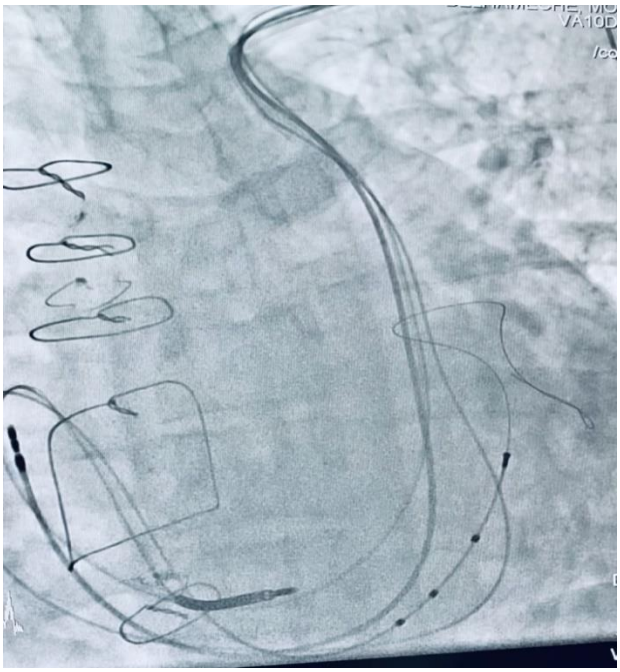


Figure 3: Leads position of atrium a right ventricular

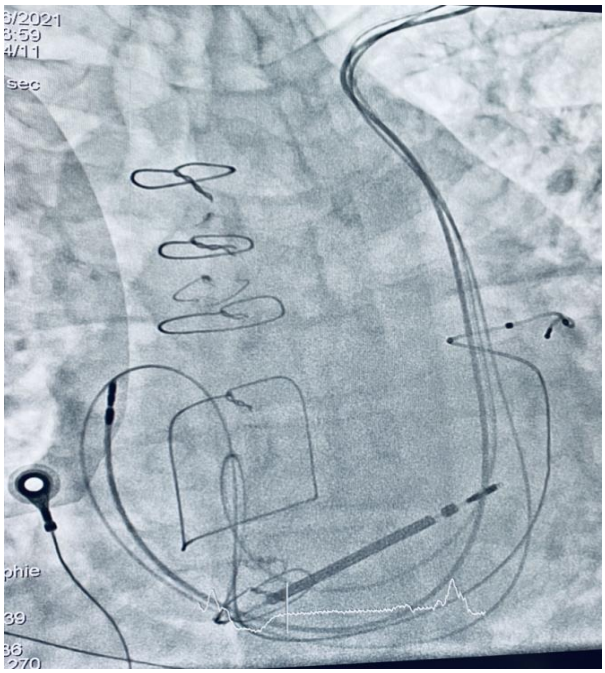


Figure 4: Leads position of atrium a right ventricular and left ventricular (Finale X ray picture)

Patient 3:

This patient was referred about heart failure , he had an arythmic cardiomyopathy treated about AF ablation 3 years ago.

ECG showed: sinus rhythm and left bundle branche bloc.

Ultrasound cardiac found a dilate cardiomyopathy and 35% LVEF.

The measurements of potential and thresholds were suitable **(**Atrium: potential = 3,5mV, Threshold = 0,5V, Impedance =653 Ω, RV: potential = 11,8mV, Threshold = 0,5 V , Impedance = 783 Ω , LV : potential = 22,6mV , Threshold = 2,5 V , Impedance =1340 Ω **)**

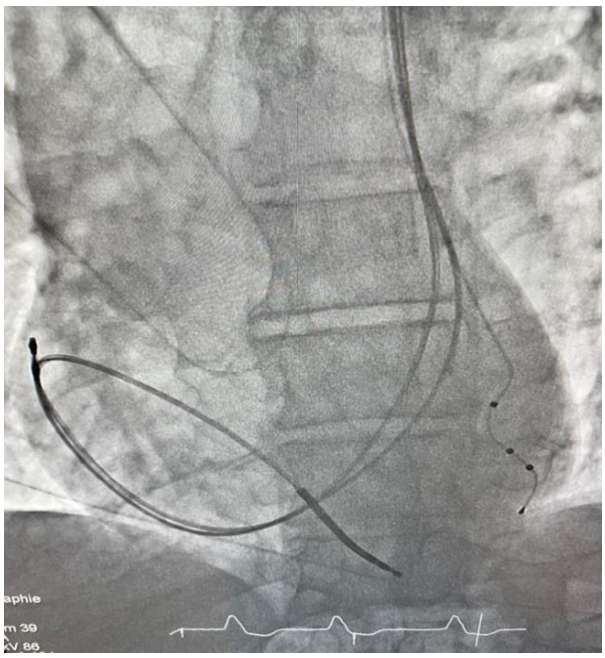


Figure 5: 3 Leads position. Final X ray picture

These two procedures were too difficult, because it was necessary to use long and rigid mandrels to fix and to stabilize the RV probe. The large size of the coronary sinus **(**CS**)** exposed to several displacements of the probes before fixation. And for the second patient we must made a loop in the RA to have a support and put the probe in the lateral vein of the coronary sinus.

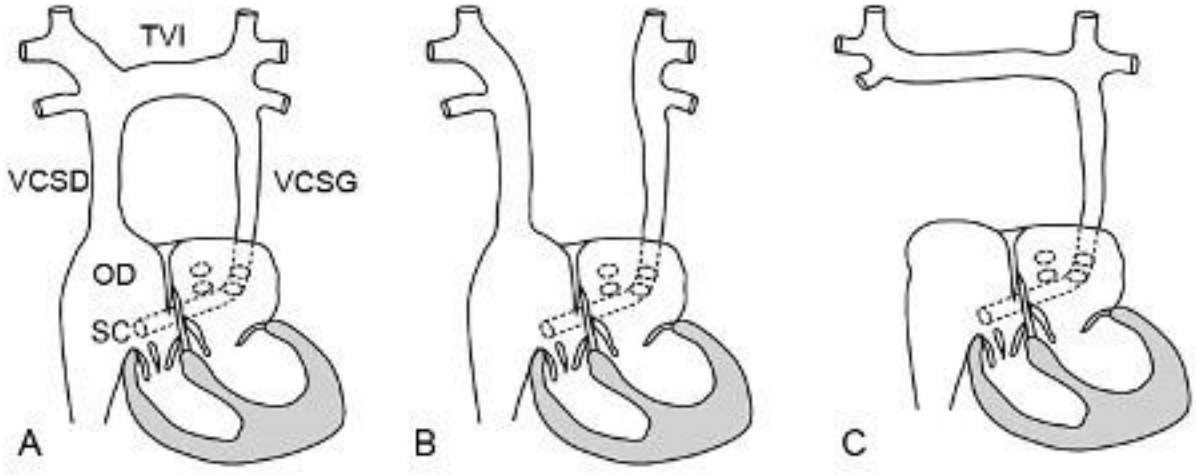
Discussion:

The first interest

This anomaly is rare and is often discovered incidentally. Indeed, the frequency of this congenital anomaly is between 0.3 and 0.5% **(**1**)** and incidence 3% in autopsies **(**12/13**)** The prevalence is 2.8%/year if this anomaly is isolated and 4.3%/year if it is associated with other malformations. In term of embryological development, drainage of the brain is already present since the 8th week. Currently there are three venous segments: a straight segment **(**upper right cardinal vein**)**, a transverse segment and a left segment **(**upper left cardinal vein**)**. The right segment will progress and becomes the vena cava and the right atrium. The transverse segment evolves into the left atrium, coronary sinus, and Marshall's ligament. While the left segment involutes and gives rise to the intercostal veins.

There are 3 types of malformation and several classifications:

Classification according to the existence or not of associated malformation **(**2**)**.



Group I: 2 VCS not associated to a congenital anomaly.

Group II: 2 VCS associated with congenital anomalies:

- Situs inversus.
- Dextrocardia
- Atresia of the tricuspid.
- interatrial and interventricular communication **(**IAC, IVC**)**.

- Pulmonary stenosis.
- Single ventricle.
- Transposition of the great vessels.
- Fallot tetralogy.

Group III: There is a LSVC.

Recent classification derives from MRI and radio data: Schummer (3).

Type A (IIIa): VCS (VCSD and VCSG) connected by an innominate venous trunk.

Type B (IIIb): 2 VCS (VCSD and VCSG) without innominate venous trunk.

Type C (II): The rarest one, there is only a left superior vena cava.

The second interest

Is the association with conductive disorders. Several cases and isolated observations report an association of conductive disorders (sinus dysfunction, atrioventricular block, and intraventricular conductive disorders). Mauro Biffi described 6 patients out of a cohort of 1127 PM implantations where he found 3 sinus dysfunctions, 1 AVB and 2 VT on CMD. On the autopsy data it was noted that the development of the sinus and of the AV node and of the HIs is influenced by the regression of the left segment of the cardinal vein and that in case of VCSG, the sinus node is small with a poverty in myocardial fibers and that at the AV node there are large venous gaps with fibrosis reaching the His; the connection with the myocardium is made through a few fibers (4).

Zerbe described 4 cases out of 661 implantations where he found 3 AVB and 1 sinus dysfunction (5).

The discovery of a VCSG accidentally when an examination for an extracardiac pathology therefore merits cardiological monitoring to prevent the risk of sudden death as has been described (6), (7).

The third interest

Is the technical difficulty of implanting a pacemaker (PM).

In the literature, implantation failure has been noted and lead to epicardial or hybrid implantations. But also changing the implantation site to the right to use an upper right vena cava in type A and B if the first implantation site is on the left as perform many teams. These failures were mainly related to the difficulty of getting into the RV and RA due to the large size of the CS and the fact that the probes were

not stable at the implantation site, especially if there is an associated malformation **(**8,9,10**)**. In our population, all patients have type C anomaly.

We believe that an angiogram should be done to see if there is a RSVC and the possibility of going through that vein into the heart. If it is a type C as in our observation it is then necessary to walk along the left edge of the sternum in the LSVC then to arrive in the coronary sinus then RA and finally RV, for this it is necessary to perform the mandrels and use probes with active fixation to avoid displacement. The shape of the mandrels will depend on the side where we want to implant the PM, and the type of malformation to be able to pass from the coronary sinus which is very wide in the RV. In our observation we preformed the ventricular mandrel in Z, and the atrial mandrel in L **(**11**)**.

It is also necessary to use profile incidence **(**ROA**)** to ensure that the atrial probe is properly fixed in the atrium and not in the CS.

So, preform the mandrels to go to the right atrium and cross the tricuspid to go to the apex on the one hand and use screw probes seems necessary to us to reduce the time of the procedure and to avoid the secondary displacement of probes.

The long-term course is quite good in terms of stimulation apart from the normal course to battery wear **(**12**)**.

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