

Cantrell's Syndrome

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Cantrell's syndrome is characterized by:

- 1) Omphalocele
- 2) Sternal cleft
- 3) Anterior diaphragmatic hernia
- 4) Ectopia cordis
- 5) Intracardiac defects

With an incidence of 5 to 8 cases per million of newborn babies, this syndrome is caused by mesodermal defects in early stages of the embryonic period. There are two ways of presentation: complete and incomplete.

The incomplete way is when the heart is covered by skin, pericardium or both of them. In the complete way, the sternum is absent or it has a wide defect. There is no parietal pericardium and the heart is totally out of the thorax with its apex upwards and there is an important reduction in the size of the thoracic cavity. Generally, with omphalocele. 80% of the cases with incomplete way present congenital heart defect and in the complete way this percentage is 100%. Truncocoanal abnormalities are the most usual.

The studied case had interventricular communication (IVC) and the emergence of the great vessels was difficult to observe.

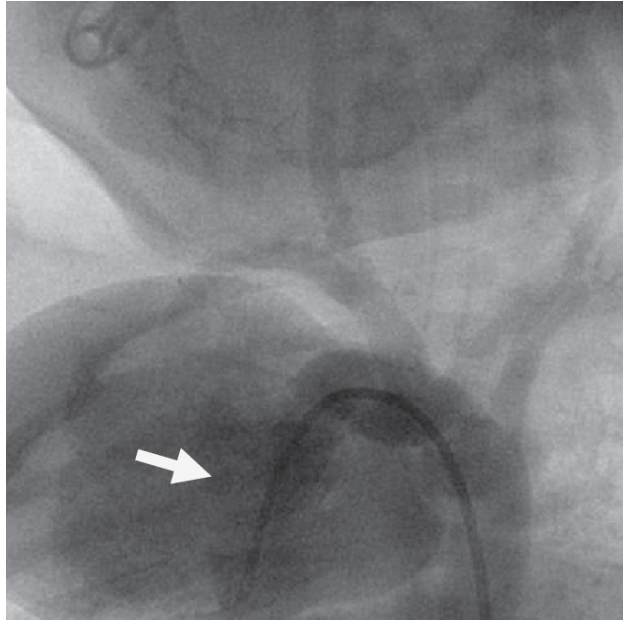


Fig. 2. Catheter in the LV. Anteroposterior position. Concordant ventriculoarterial connection. A muscular IVC is observed.



Fig. 1. Catheter in the RV. Anteroposterior position. Concordant ventriculoarterial connection.

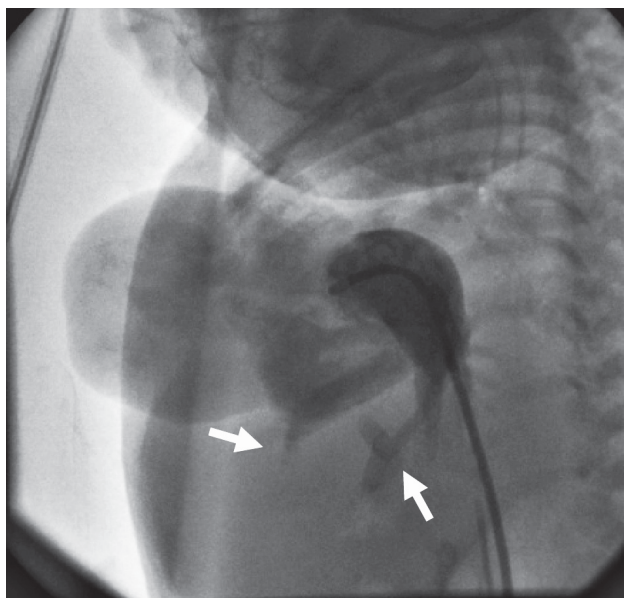


Fig. 3. Catheter in the RA. Left profile position. Concordant AV connection. A dual system of suprahepatic veins entering the RA is observed.

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Giant Left Atrial Appendage with Atrial Tachycardia

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BACKGROUND

The real anatomical knowledge is important in electrophysiology for the success of the curative treatment, not only of complex arrhythmias but also of those that may be associated with structural alterations that could complicate the procedure. Nowadays, the possibility to carry out a magnetic resonance imaging (MRI) or a multislice computed tomography is very important when facing the suspicion of left arrhythmias. These procedures allow placing structures and orientating the treatment. The association to navigation systems allows avoiding complications when doing movements in unknown structures.

We present the case of a 53-year-old woman with background of focal atrial fibrillation. Isolation of pulmonary veins with a conventional technique at other center was performed. The patient is referred to the specialist due to episodes of regular tachyarrhythmia which was not registered in the electrocardiogram. A two-dimensional echocardiogram, which reported normal left ventricular diameters with preserved ventricular function, normal left atrial size and typical myxomatous mitral valve prolapse, was carried out.

Begin the treatment with atenolol 25 mg daily and anticoagulation with acenocoumarol. Thyroid hormone values are normal. A 24-hour holter monitor is carried out. We may observe in it sinus rhythm, a heart rate of 66 bpm, less frequent ventricular extrasystoles (194 in 24 hrs) and frequent supraventricular extrasystoles (1311 in 24 hrs).

The patient continues with episodes of palpitations which are of short duration. As a result, she is admitted into the emergency department with an atrial tachycardia (110 bpm), which due to its P wave morphology that is negative in aVl with superior axis, allows the suspicion of a left location. We decide to ask for a MRI to assess the anatomy when facing the

possibility to perform a new ablation of such focus and also, to assess the present state of pulmonary veins post-ablation. In the posteroanterior slice, we may observe a severe dilation at the level of the left atrial appendage close to the mouth of the left superior pulmonary vein, with growth backwards and upwards (Figure 1).

An electrophysiological study is carried out, and atrial tachycardia with the same morphology that the one already observed in the echocardiogram during the clinical episode is induced (Figure 2).

Due to the possible left and superior origin of atrial tachycardia, transseptal puncture is performed and an automatic focus, which is originated within the left atrial appendage, is checked. In relation to the

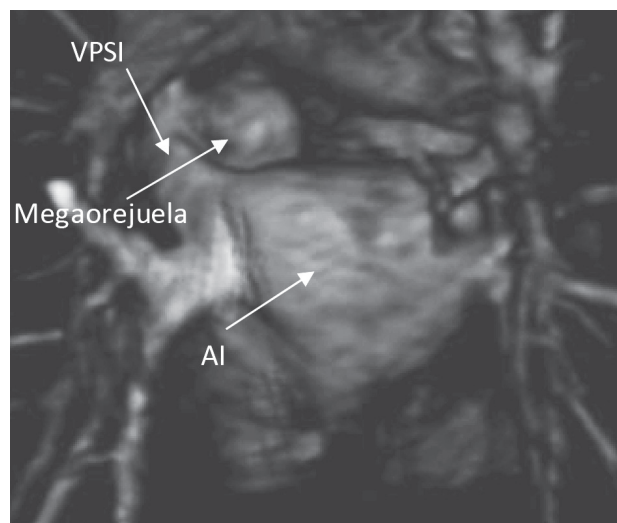


Fig. 1. Falta la traducción

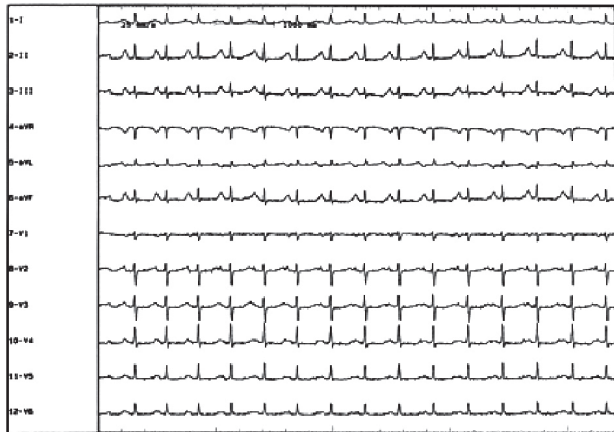


Fig. 1. Falta la traducción

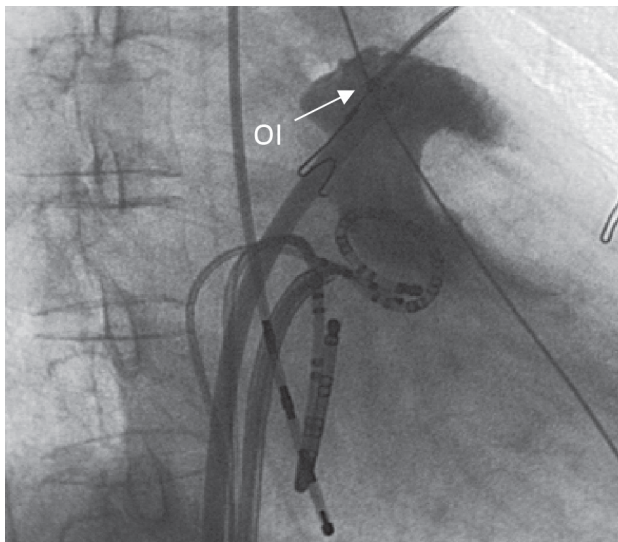


Fig. 1. Falta la traducción

anatomy described in the MRI, a contrast injection in the left appendage is carried out. It is performed to place the mapping in relation to its special anatomical

characteristic (Figure 3). Guided by a loop catheter, the ablation of the focus is successfully performed and with no complications. The patient evolves without symptoms and with no new register of arrhythmias.

Discussion: the alterations in the usual morphology of the left atrial appendage may be the origin of ectopic focuses. The surface echocardiogram approaches us to their location, but it does not give us any specific information.^{1,2} The use of new imaging diagnosis methods such as MRI^(3,4) or multislice CT,⁵ allows us to know the real anatomy of the atrium in its entirety and besides these methods give us information to guide radiofrequency in a precise way. MRI allows us to relate the different mediastinal structures with the heart and it also makes possible a more detailed analysis of anatomical alterations of the atrium.⁶ MRI constitutes an element of great importance in the assessment of arrhythmias of unusual location.

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