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Atrioventricular Block and Atrial Septal Defect, A Rare but Serious Association: about a Case

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ABSTRACT

The medical management of several diseases including congenital heart disease has been changed by the CoVid 19 pandemic. We report through this observation the case of a child with complete atrioventricular block associated with atrial septal defect.

KEYWORDS: complete atrioventricular block, atrial septal defect

I. INTRODUCTION

Atrial septal defect is the second cardiac malformation in terms of frequency after aortic bicuspidism, it represents approximately 6 to 8% of congenital cardiac malformations in childhood.

The vast majority of atrial septal defect occurs sporadically and is isolated, or at least no causal factor can be identified [1].

However, there are certain familial forms where the mutation of a gene can lead to CIA or a polymalformative syndrome associating structural deformations and developmental defects of the cardiac conduction system which significantly increases the risk of mortality [2].

II. CASE REPORT

We report in this medical observation, the case of the child DG aged 08 years old with no significant personal or family history, born of a pregnancy carried to term with normal growth and psychomotor development, which has reported for 1 year the occurrence of repeated lipothymic discomfort at the rate of 2 to 3 episodes per week which prompted the patient to consult.

A child in good general condition at the initial physical examination has been found with a BMI at 15.56 kg / m² with a blood pressure at 100/64 mmHg. A systolic murmur with a weak pulmonary focus, followed by a doubling of B2 have been revealed by the cardiovascular examination.

A complete atrioventricular block with ventricular rate at 50 cycles / min and fine QRS has been recorded by the electrocardiogram

A septal atrial defect of the ostium secundum type mesuring 14 mm of large diameter with hyperechoic edges "image 1" and with color doppler a left right shunt "image 2" and moderate dilation of the right ventricle has been diagnosed by transthoracic echocardiography.

Given the emergency context of the degree of the conductive disorder, a temporary cardiac stimulation has been underwent before the implantation of a single-chamber pacemaker with simple operating suites.

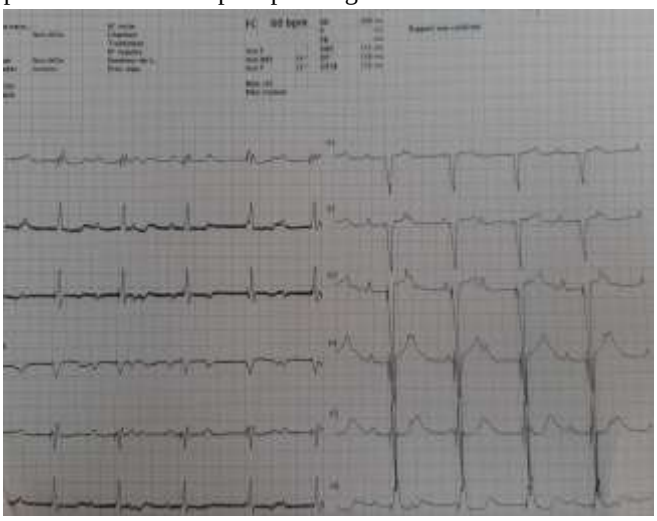

Figure 1 : Electrocardiogram showing the rhythm trained by the pacemaker.

In a particular work context where activity in our service is greatly reduced because of the situation of a global pandemic due to the coronavirus, the closure of the atrial septal defect by interventional cardiac catheterization has been deferred as well as the rest of the assessment and in particular the genetic exploration in search of the NKX2.5 mutation.

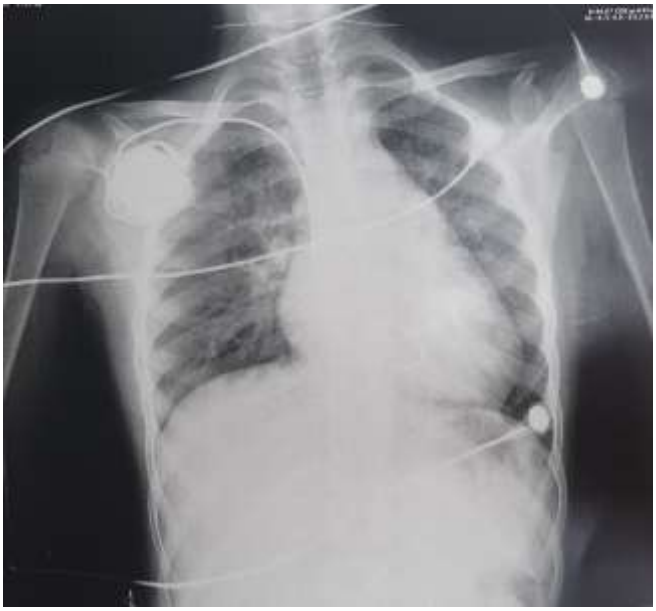

Figure 2: Chest x-ray after pacemaker placement

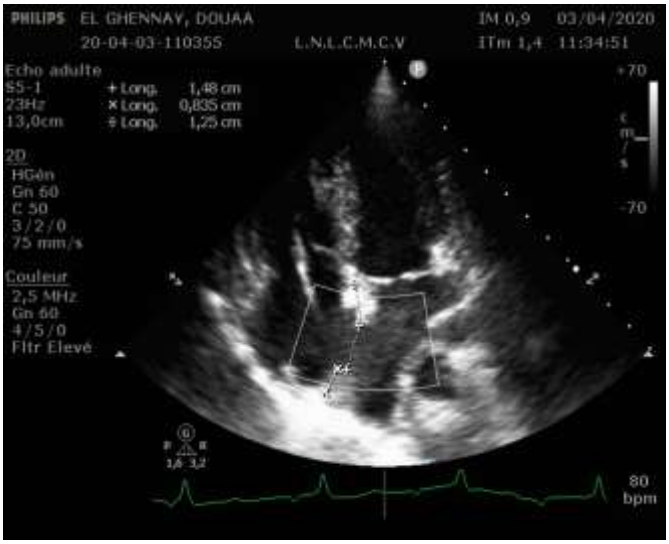

Figure 3: Visualization of the pacemaker probe and interauricular communication in apical four chambers view

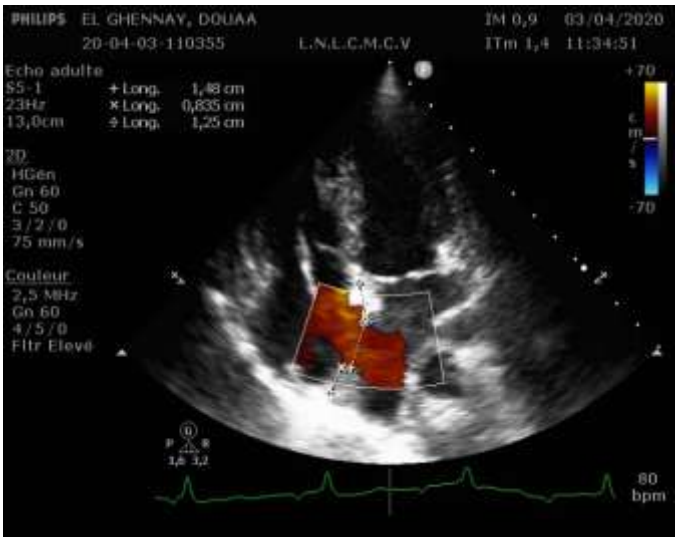

Figure 4: View of the pacemaker probe and the left shunt right in apical with color doppler

III. DISCUSSION

The association of congenital complete heart block and atrial septal defect can be seen in ostium primum type , but considered very rare in the ostium secondum type.

The importance of recognizing this association, however rare it may be, lies in its poor prognosis with an increased risk of sudden death [1].

This could be confirmed by the study carried out by Schott et al on four families carrying the NKX2-5 mutation associating atrioventricular block and cardiac malformation (ASD and Fallot tetralogy), conductive disorders have been presented by their 33 affected individuals and implantation of a pacemaker have been decided for 14 among them. Unfortunately six of them, after surgical repair of ASD and without implantation of pacemaker have died suddenly following a sudden cardiac death [3]. The relentless care of our young patient has been justified by this study, with the setup of a provisional cardiac stimulation when the diagnosis of complete atrioventricular block and septal atrial defect has been revealed followed by the implantation of a single-chamber pacemaker with simple operating suites.

Therefore, when septal atrial defect is associated with a conduction disorder, a family history should be sought and a careful clinical examination of all those related to the patient should be performed [4]. In our case a significant family history has not been found.

The complete clinical examination of our patients is also of great importance, in particular the muscular and osteo-articular examination, in search of malformations in the upper limbs, in particular in the thumb, which is often associated with septal atrial defect and a complete atrioventricular block in Holt-Oram syndrom [5,6].

In our young patient, the atrial septal defect lends itself perfectly to a closure by interventional catheterization but given the constraints of the pandemic it was decided to set up a single chamber pacemaker in order to be able to subsequently propose an interventional closure.

IV. CONCLUSION

Hereditary forms should encourage us to deepen our family investigation in search for a history of sudden cardiac death, To make a complete clinical examination and evoke the association ASD AVB which is certainly rare, but dreadful by its poor prognosis.

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