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CLINICAL CASE

Heart transplantation for the treatment of isolated left ventricular myocardial noncompaction. First case in Mexico☆

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KEYWORDS
Myocardial noncompaction of the left ventricle;
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Heart transplantation

Abstract
Background: Myocardial noncompaction of the left ventricle is a congenital cardiomyopathy characterised by left ventricular hypertrabeculation and prominent intertrabecular recesses. The incidence ranges from 0.15% to 2.2%. Clinical manifestations include heart failure, arrhythmias, and stroke. Prognosis is fatal in most cases. Heart transplantation is a therapeutic option for this cardiomyopathy, and few had been made worldwide.
Clinical case: The case is presented of a 20 year-old male with noncompacted myocardium of the left ventricle, who had clinical signs of heart failure. His functional class was IV on the New York Heart Association scale. He was successfully transplanted. Its survival to 15 months is optimal in class I New York Heart Association, and endomyocardial biopsies have been reported without evidence of acute rejection.

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Conclusion: It is concluded that heart transplantation modified the natural history and improved survival in patients with this congenital heart disease.

PALABRAS CLAVE
Miocardio no compactado del ventrículo izquierdo;
Cardiopatía congénita;
Insuficiencia cardiaca;
Trasplante cardiaco

Trasplante cardiaco: una opción para tratamiento del miocardio no compactado aislado de ventrículo izquierdo. Primer caso en México

Resumen
Antecedentes: El miocardio no compactado del ventrículo izquierdo es una miocardiopatía congénita caracterizada por hipertrabeculación del VI y prominentes recesos intertrabeculares. La incidencia oscila entre 0.15% a 2.2%. Las manifestaciones clínicas son: insuficiencia cardiaca, arritmias y embolias. Su pronóstico es mortal en la mayoría de los casos. El trasplante cardiaco es una opción terapéutica para esta miocardiopatía, y pocos han sido realizados a nivel mundial.
Caso clínico: Varón de 20 años con miocardio no compactado del ventrículo izquierdo que presentó datos clínicos de insuficiencia cardiaca en clase funcional IV de la Asociación Cardiológica Neoyorquina, y fue trasplantado en forma exitosa. Su sobrevida a los 15 meses es óptima en clase funcional I de la Asociación Cardiológica Neoyorquina y las biopsias endomiocárdicas se han reportado sin datos de rechazo agudo.
Conclusión: El trasplante cardiaco es una opción terapéutica que modifica la sobrevida para este tipo de casos.

Background

Myocardial noncompaction of the left ventricle (MNCLV) is a congenital and genetic myocardial disease characterised by hypertrabeculation of the left ventricle (usually the smooth endocardium), prominent intertrabecular recesses that receive blood flow directly from the left ventricular cavity rather than the coronary arteries, and coronary anomalies.[1,2] It is believed that it is caused by noncompaction of the left ventricle during the 5th to 8th weeks of gestation.[3]

The incidence ranges from 0.15% to 2.2%, although in recent series with better diagnostic technology it has increased to 18%, especially if familial.[4] MNCLV can be isolated, exclusive to the left ventricle, combined with the right ventricle or other cyanogen congenital heart diseases, or left or right ventricle outlet obstruction.[1] In 1934, Bellet reported a case of congenital heart disease similar to MNCLV; Engberding published a similar case in 1984,[5] and the first broad series of 8 cases was brought to the scientific world by Chin in 1990.[6] It is, therefore, a recent disease categorised by the World Health Organisation since 1996 as an unclassified cardiomyopathy.[7] Since 2006, the American Heart Association (AHA) has categorised the condition as a congenital heart disease.[8,9]

Clinical manifestations are principally heart failure (53%), arrhythmias that can be fatal (41%) and stroke (24%).[10] Up to 82% of cases are accompanied by neuromuscular symptoms or manifestations.[2] Manifestations can start from the second decade of life; principally heart failure due to dilation of the left ventricle. The natural progression of the disease can be death due to heart failure or fatal arrhythmias.[10,11]

Diagnosis is made by echocardiography, using the criteria of Chin[6] and Jenni (2001).[12]

Because this diagnosis has a poor prognosis and there is a risk of sudden death and heart failure, one of the treatments indicated is placement of an automatic implantable defibrillator[13] and heart transplantation.[14,15]

There are few cases in the literature on treatment of MNCLV by heart transplantation; the majority are for severe heart failure due to dilation of the left ventricular cavity.

On this occasion, we report a case of MNCLV after heart transplantation and their current progress.

Clinical case

A 20-year-old male student with no relevant history and neurologically intact. The patient presented with symptoms of severe heart failure from August 2013, manifested by fluid retention, leg oedema, progressive dyspnoea on little exertion, orthopnoea and paroxysmal nocturnal dyspnoea, and heart arrhythmias. Treatment was started with amiodarone, diuretics and angiotensin-converting-enzyme Inhibitors. ECG showed hypertrabaculations on the lateral, apical and inferior surface of the left ventricle, relating to non-compacted myocardium with compacted epicardium higher than 2, occupying 21% of the ventricular surface