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Mini Review

Non-dilated Left Ventricular Cardiomyopathy - inclusion Criteria and Risk Assessment

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Abstract

The newly developed term non-dilated left ventricular cardiomyopathy includes Takotsubo syndrome, hypertrabecularisation cardiomyopathy and arrhythmogenic left ventricular cardiomyopathy. The risk assessment differs a lot depending on left ventricular function is severely or mildly depressed or normal.

Keywords: Non-dilated left ventricular cardiomyopathy, Arrhythmogenic left ventricular cardiomyopathy, Extensive hypertrabecularisation, Takotsubo syndrome, Non-compaction cardiomyopathy

Introduction

Non-dilated left ventricular cardiomyopathy is a newly developed term in the classification and definition of cardiomyopathies [1]. The former term was non-dilated hypokinetic left ventricle [2] characterizing a special form of dilated cardiomyopathy or arrhythmogenic left ventricular cardiomyopathy according to the so-called Padua criteria [3]. A ring-like enhancement of gadolinium of the left ventricle is a typical finding of arrhythmogenic left ventricular cardiomyopathy and has a high risk of severe ventricular arrhythmias and prophylactive ICD Implantation [4].

Otherwise, in the newest publication from *Arbelo, et al.* [1] some terms like non-compaction cardiomyopathy, takotsubo syndrome and arrhythmogenic cardiomyopathy, especially arrhythmogenic left ventricular cardiomyopathy are lacking. Nowadays, these heart diseases are included in non-dilated left ventricular cardiomyopathy. For risk assessment, other definitions are of interest.

Extreme Hypertrabecularisation with Severe Left Ventricular Dysfunction

Hypertrabecularisation with severe left ventricular dysfunction has a very high risk for thromboembolism, sudden cardiac death and heart failure [5]. According to a Scandinavian study, non-compaction left ventricle with severe left ventricular dysfunction has a higher risk for heart transplantation compared to dilated cardiomyopathy [6].

Extreme Hypertrabecularisation with Normal Left Ventricular Function

Hypertrabecularisation with normal left ventricular function has a normal life and is in many cases an incidental finding.

Takotsubo Syndrome

Takotsubo syndrome is generally characterized by a regression of left ventricular function (normalization) within two to three weeks. If left ventricular dysfunction (apical ballooning, mid-ventricular or basal ballooning) lasts longer than 4 weeks or segmental contraction abnormalities with normal ejection fraction remain, the risk of heart failure increase significantly [7]. According to a Japanese study the number of men with takotsubo syndrome increase significantly with frequent mid-ventricular ballooning and

increase risk of heart failure [8]. Takotsubo syndrome can occur with a happy heart syndrome with decreased risk of heart failure according to a German study [9].

Arrhythmogenic Left Ventricular Cardiomyopathy with Severe Left Ventricular Dysfunction

Arrhythmogenic cardiomyopathy includes right dominant, biventricular and left dominant disease. The new term non-dilated left ventricular cardiomyopathy is mainly used instead of arrhythmogenic left ventricular cardiomyopathy. Left dominant arrhythmogenic cardiomyopathy is in 3 – 10% of cases part of arrhythmogenic cardiomyopathy and is characterized by ring-like or severe left ventricular late gadolinium enhancement by cardiac MRI and special appearance of standard ECG. ECG is characterized by low voltage in limb leads and T-wave inversion or flattering in inferolateral leads [3]. LVAD or heart transplantation due to therapy-resistant heart failure and, in minor terms, ventricular arrhythmia events or sudden cardiac death are typical findings of arrhythmogenic left ventricular cardiomyopathy with severe left ventricular dysfunction. According to the above cited Scandinavian study [6] arrhythmogenic biventricular cardiomyopathy and arrhythmogenic left ventricular cardiomyopathy bare a higher risk of heart transplantation compared to dilated cardiomyopathy.

Arrhythmogenic Left Ventricular Cardiomyopathy with Mild Left Ventricular Dysfunction

Arrhythmogenic left ventricular cardiomyopathy with normal or mildly reduced left ventricular function and ring-like late gadolinium enhancement bares a high risk of sudden arrhythmic death [4], even pronounced in Lamin A/C [10] or filamin C gene mutations [11].

Discussion

The newly developed term of non-dilated left ventricular cardiomyopathy includes several forms of cardiomyopathy like non compaction left ventricle, takotsubo syndrome and arrhythmogenic cardiomyopathy not mentioned in the paper of *Arbelo, et al.* [1]. This paper must be critically discussed, as several different cardiomyopathies exist with different risks of heart failure or sudden cardiac death. In particular, arrhythmogenic cardiomyopathy including right dominant, biventricular and left dominant has been defined not long ago in 2020 in the so-called *Pudua* criteria [3]. To define arrhythmogenic right ventricular cardiomyopathy as a distinct cardiomyopathy and not to subdivide biventricular or left dominant arrhythmogenic cardiomyopathy is a mistake per se and a great concern of the newly defined term of non-dilated left ventricular cardiomyopathy [1].

Acknowledgments

None.

Conflicts of Interest

None.

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