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Cardiovascular magnetic resonance (CMR) implementation in a newly-established cardiomyopathies unit.

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INTRODUCTION: Cardiomyopathies are a group of often inherit diseases characterised by abnormalities and associated dysfunction of myocardium. Knowledge of the etiology of a cardiomyopathy is important for diagnosis, therapy and prognosis. CMR provides a noninvasive gold-starndard measure of cardiac chamber size and structure, both left and right function and tissue composition. We present our experience with Cardiac-MRI in a newly-established cardiomyopathies unit.

METHODS: Since January 2017 until October 2018, 283 patients were evaluated in the unit and CMR were ordered in 83 of them. Suspected diagnosis were mainly: Hypertrophic cardiomyopathy (38,6%) Left ventricular non-compaction (20,5%) Dilated cardiomyopathy (16,9%). Arrhythmogenic ventricular cardiomyopathy (7,2%), marfan syndrome and other aortic disorders (6%) amyloidosis (3,6%) and restrictive cardiomyopathy (1,2%). Steady state free precesion (SSFP) was used for for determination of myocardial mass and volumes. Basal perfusion and Late Gadolinium Enhancement (LGE) imaging were performed in all cases too.

RESULTS: In general, 72,3% were males, 68,7% proband, 31,3% family members. Suspected diagnosis was confirmed in 83,6% cases, mainly: Hypertrophic cardiomyopathy (28,4%) Noncompaction Cardiomyopathy(14,9% Dilated cardiomyopathy (13,4%). Arrhythmogenic ventricular cardiomyopathy (7,5%) and amyloidosis (4,5%). On the other hand, CMR study was normal in 13,4%, letting a 10% final discharge from the Cardiomyopathy Unit, most frequently due to: Non-significant left ventricular hypertrabeculation, suspected arrhythmogenic ventricular cardiomyopathy with no diagnostic criteria and slightly increased LV-mass cases with any HCM-hallmark. LGE was observed in 30% and in 3% that was determinent in implantable cardioverter defibrillators (ICT) decision-making. Also, LGE patients held a significantly lower LV eyeccion fraction (47 to 55%, p = 0,014) Finally, 87,7% of patients with a pathological CMR-study showed a pathological genetic test too. The most frequent mutations detected were those involving genes encoding for the cardiac sarcomere: MYBPC3 (Arg502Gln 8,8%, Arg495Gln 1,8%, Glu542Gln 1,8%) y TTN (Cys3529Ser 7%) (Figure 1)

CONCLUSION: Cardiomyopathies include a variety of myocardial disorders that manifest with various structural and functional phenotypes and are frequently genetic. Their assessment still relies primarily upon echocardiographic evaluation, but, in somes cases, cardiomyopahties can be particulary challenging. In this regard, CMR imaging is clearly helpful, providing valuable information that are complementary to other aspects of both clinical and echocardiography evaluation. Our experience is in line with that current trend of CMR emerging as a powerful tool in cardiomyopathies assessment.

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