

**Role of late gadolinium enhancement magnetic resonance imaging in the management of patients with primary cardiomyopathy.**

*Yamazawa H., Takeda A., Takei K., Furukawa T., Izumi G., Abe J.  
Hokkaido University Hospital, Sapporo, Japan*

**Introduction:**

Late gadolinium enhancement magnetic resonance imaging (LGE-MRI) has been widely accepted for detection of myocardial fibrosis, which has been implicated as a factor in cardiovascular events including sudden cardiac death. We introduce some cases of primary cardiomyopathy that MRI study including LGE-MRI was useful for management.

**Methods:**

We performed MRI study for five patients with primary cardiomyopathies. Two siblings with hypertrophic cardiomyopathy (HCM) caused by an alpha-tropomyosin1 (TPM1) mutation. Twin sisters and a girl with overlap of restrictive and hypertrophic cardiomyopathy (RCM/HCM) caused by a cardiac troponin-I (TNNI3) mutation and a myosin light chain (MYL2) mutation, respectively. Elder brother with HCM (patient 1) was diagnosed at 13 years old, and younger brother (patient 2) was diagnosed at 6 years old. Twin sisters with RCM/HCM (patient 3 and patient 4) were diagnosed at 11 years old. A girl with RCM/HCM (patient 5) patient was diagnosed at 7 years old. The extent of LGE was expressed as the percentage of the total left ventricular mass (the % of LV mass).

**Results:**

The extent of myocardial fibrosis by LGE-MRI (age at examination) in patients 1 to 5 was 8% (18), 1% (10), 18% (11), 7% (11), and 6% (18) of LV mass, respectively. Life-threatening events were found in patient 1 (syncope due to ventricular tachyarrhythmias) at 18 years old and patient 3 (sudden death) at 11 years old. Patient 2, 4, and 5 have been uneventful. The extent of LGE increased to 16% of LV mass by re-examination two years later in patient 2.

**Discussion:**

Implantable cardioverter defibrillator (ICD) was implanted for secondary prevention of sudden death in patient 1. Furthermore, we decided to place a primary prophylactic ICD in patient 2 because the extent of LGE in patient 2 was larger than that of the patient 1. In the case of RCM/HCM, we arrange heart transplantation because myocardial fibrosis might be in progress with patient 4 as in the case of patient 3.

**Conclusions:**

LGE-MRI could be useful to manage patients with cardiomyopathy and to estimate their prognosis.