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T1 mapping in Fabry Cardiomyopathy

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Fabry disease (FD) is a rare, x-linked lysosomal storage disorder. FD has two types, type-1 is the classical type, with systemic involvement; type -2 is later-onset, has specific target organ involvement. Taiwan had high prevalence of cardiac variant type FD with IVS4 919G>A (FD-IVS4) mutation. T1 mapping is particular useful in cardiac MR, since GB3 storage actually is a kind of lipid; which will result in T1 decrease, and has been used a biomarker for Fabry cardiomyopathy. However, during the disease course, the developing myocardial fibrosis would cause increase of T1 value, therefore, a so-called "pseudo-normalization" of T1 value of Fabry cardiomyopathy. The complexity of the interplays among T1 value, focal scar and hypertrophy are further unpredictable in the type-2 FD than in type-1 FD. Here I will update the current staging of Fabry cardiomyopathy and Taiwanese experience of type-2 FD with IVS4 919G>A mutation. We believe the experience would shed light on Asian cardiac MR community about Fabry cardiomyopathy.