

## MP2-15

### **Stereological analysis of mitochondria in hypertrophic cardiomyopathy - A proposal for pathological diagnostic criteria of mitochondrial cardiomyopathy**

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**Introduction:** Mitochondrial density in myocardium is highly consistent within each species according to the energy supply-demand balance. Thus, increased mitochondrial density may indicate excessive myocardial demand or adaptation to the reduced mitochondrial respiratory capacity. It is not well known whether mitochondrial density differ depending on the two status.

**Methods:** Eight patients with hypertrophic cardiomyopathy (HCM) were studied. Respiratory chain (RC) enzymatic assay and electron microscopy were performed using endomyocardial biopsy samples. Four patients were diagnosed as mitochondrial cardiomyopathy (MCM) with reduced RC enzymatic activity. In each electron microscope images from endomyocardial biopsy samples, 18-25 sampling areas per each patient were studied. In each sampling area, mitochondrial volume density ( $V_{vmit}$ ), myofibrillar volume density ( $V_{vmyo}$ ), and mitochondrial surface area density ( $S_{vmit}$ ) were calculated using multipurpose test system for stereological analysis. Thus, mitochondrial volume density to myofibrillar and mitochondrial volume density ratio ( $V_{vmit}/(V_{vmyo}+V_{vmit})$ ) and mitochondrial surface density to volume density ratio ( $S_{vmit}/V_{vmit}$ ) were calculated.

**Results:** In MCM group, all the patients showed reduced activity of complex I. Two patients also had reduced activity of complex IV.  $V_{vmit}$ ,  $S_{vmit}$  and  $V_{vmit}/(V_{vmyo}+V_{vmit})$  were significantly higher than non-MCM group (0.57 vs 0.31, 2.91 vs 1.81, 0.63 vs 0.37, respectively).  $S_{vmit}/V_{vmit}$  was significantly higher in non-MCM group (5.21 vs 5.90).

**Discussion:** MCM group had significantly increased density and larger in size of mitochondria in myocardium. These data suggest that stereological quantification of mitochondria could be a pathological diagnostic criteria for mitochondrial cardiomyopathy. In addition, increased mitochondrial density in non-MCM group have a great impact on excessive energy consuming, which could be a prognostic factor.

**Conclusions:** Stereological analysis of mitochondria in hypertrophic cardiomyopathy could be a pathological diagnostic criteria of mitochondrial cardiomyopathy