

The 10th Congress of the Asian-Pacific Society of Atherosclerosis and Vascular Diseases (10th APSAVD Congress) / [APSAVD] Sponsored Symposium

## **[APSAVD] Sponsored Symposium 1 Familial Hypercholesterolemia (FH) -High Risk-**

2016年7月14日(木) 8:30-10:30      APSAVD Lecture Room | 南館 4階 錦

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英語

英語セッション

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## **Who has Extreme High Cardiovascular Risk in Patients with Familial Hypercholesterolemia?**

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Familial hypercholesterolemia (FH) is the most severe monogenic hyperlipidemia characterized by tendon xanthoma and premature coronary artery disease. The occurrence of cardiovascular events is predominantly decided by the cumulative exposure to low-density lipoprotein (LDL) cholesterol. Thus, FH represents a very high risk condition of cardiovascular disease. However, traditional coronary risk factors may also affect the threshold of cumulative LDL-cholesterol level for the onset of coronary artery disease, even in FH patients. Homozygous FH (hoFH) patients display an extremely high risk of cardiovascular events including acute coronary syndrome and acute heart failure due to aortic valvular and/or supraaortic stenosis as a result of extreme hyper-LDL-cholesterolemia around 550 mg/dL from birth. Compound or double heterozygous FH patients, who are sometimes misdiagnosed as heterozygous FH (heFH), should be diagnosed promptly. Since hoFH is highly resistant to conventional LDL-cholesterol lowering medications, extraordinary therapies such as LDL-apheresis, microsomal triglyceride transfer protein inhibitor, apoB antisense, or liver transplantation should be considered. The mean level of LDL-cholesterol in heFH is around 260 mg/dL. Although the approved maximum doses of three different oral hypocholesterolemic medications consisting of potent statin, ezetimibe, and bile acid-sequestering resin could reduce LDL-cholesterol levels by 66%, there are still a significant number of insufficiently treated heFH. HeFH patients with coronary artery disease, especially with recurrent or recent acute coronary syndrome are at extreme high risk of additional recurrences. HeFH with extracoronary atherosclerotic disorders, such as peripheral artery disease or cerebrovascular infarction, are also at extreme risk. HeFH carries a high life-long cardiovascular risk, including patients with multiple, premature, poor-controlled risk factors, who are also at extreme high risk. PCSK9 inhibitor effectively reduces LDL-cholesterol levels of heFH as well as hoFH, and should be prescribed for FH patients with extremely high cardiovascular risk.