

シンポジウム

## シンポジウム19

### 成人先天性心疾患の不整脈

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## [III-S19-01] Sudden Cardiac Death in Adults with Congenital Heart Disease

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Sudden cardiac death (SCD) is a major cause of late mortality (15-26%) in adults with congenital heart disease (CHD). Predicting SCD is challenging due to the anatomic heterogeneity of CHD and the infrequency of sudden arrhythmic events among these patients. However, the relatively low annual SCD incidence rate in the ACHD population varying from 0.09% to 0.26% is still manifold higher than in age-matched controls. The annual incidence of SCD differs among the various types of CHD. SCD in CHD has been studied in Tetralogy of Fallot (TOF) and Mustard or Senning repair of transposition of the great arteries (TGA), and several potential risk factors for SCD have been identified. However, the predictive value of these risk factors in terms of absolute risk of SCD remains unclear. Moreover, some risk factors may or may not be relevant to different types of CHD. By combining three large databases from Toronto (Canada), Leuven (Belgium) and the Netherlands, a large cohort of more than 25000 adults with CHD could be studied. From a total of 1189 deaths, SCD occurred in 171 patients. The majority of these patients had severe cardiac lesions such as Eisenmenger syndrome and TGA. However, arrhythmic death also occurred in mild cardiac lesions, such as septal defects. Documented SVTs, increased QRS duration, and impaired systemic or subpulmonary ventricular function were independent predictors of SCD. These parameters are similar to those in acquired heart disease. Based on these parameters an easily applicable risk score could be developed and validated for prediction of the absolute risk of SCD among adults with various forms of CHD. The model might be useful guiding clinicians in decision making for ICD implantation in CHD. However, prospective data from further studies are required to validate the risk score model.