What to do for a 50 Year Old Woman with Inoperable PA VSD and MAPCA Complaining of Recurrent NSVT?

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Adult patients with congenital heart disease (CHD) are at high risk for sudden cardiac death caused by ventricular tachyarrhythmia. In the documented absence of heart disease, spontaneous non-sustained ventricular tachycardia (NSVT) does not carry any adverse prognostic significance, especially in patients with preserved left ventricular function. However, the prognostic value of NSVT in adult patients with unrepaired CHD is not known.

Until now, NSVT in CHD patients has mainly been investigated in tetralogy of Fallot (TOF) patients. Symptomatic NSVT predicted implantable cardioverter defibrillator (ICD) shocks for primary prevention in these patient groups.

Recent study demonstrated that CHD patients with documented NSVT, normal to moderate prolonged QRS duration and at least a moderate cardiac function rarely developed sustained VT or VF during a follow-up period of 5 years. They suggest that in CHD patients with a moderate to good ventricular function, normal or limited conduction delay and NSVT, a wait-and see treatment strategy seems justified. Therefore, in adult patient with inoperable PA with VSD, antiarrhythmic pharmacotherapy (Class III agents: sotalol, amiodarone ± beta blocker) may be helpful in reducing the symptom of NSVT in moderate to normal ventricular function. In contrast to, ICD therapy for primary prevention can be reasonable in selected adults patients with inoperable PA with VSD and multiple risk factors for sudden cardiac death, such as left ventricular systolic (LVEF< 35%) or diastolic dysfunction, higher burden of NSVT, QRS duration more than 180 ms, extensive right ventricular scarring on cardiac MRI, or inducible sustained VT at electrophysiologic study.

Catheter ablation can be applicable as adjunctive therapy to an ICD in adults with CHD and recurrent monomorphic VT, a VT storm, or multiple appropriate shocks that are not manageable by device reprogramming or drug therapy. Further prospective study focusing on the prognostic role of NSVT in adult CHD patients might give more insight for development of sustained VT or