Congenital Heart Disease Prognosis Still a Mystery

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TORONTO — Experts continue to look for the best way to assess the risk for sudden death in adults with congenital heart disease.

Adults with congenital heart disease face a threat from sudden cardiac death that is far higher than that of the general population, with a mortality risk of 0.9 events per 1,000 patients per year in patients who are younger than age 20 years. But as of now, there is no good way to distinguish patients with the highest risk from those congenital heart disease patients who have a substantially lower risk.

"No single hemodynamic or electrophysiologic risk factor appears to be sufficiently predictive," Dr. Louise Harris said at the 14th World Congress on Heart Disease. Overall, she said, about a quarter of the deaths that occur in adults with congenital heart disease are sudden cardiac deaths.

Researchers may eventually develop a scoring system that takes into account several risk factors, she said. Recent findings have established that patients with congenital heart disease may have abnormalities in several organ systems, and risk assessment needs to take all these variables into account. Renal function, for example, is impaired in a significant percentage of patients with congenital heart disease, and was linked with an increased risk for death (Circulation 2008;117:2320-8).

"Sudden cardiac death is not one physiologic entity, but a manifestation of a number of disorders," said Dr. Harris, who is a cardiologist in the congenital cardiac center for adults at Toronto General Hospital.

The risk for sudden cardiac death is highest in patients who survived any of four types of congenital heart diseases: tetralogy of Fallot, transposition of the great arteries, coarctation, and aortic stenosis. Among these patient groups, the most studied have been patients treated for tetralogy of Fallot.

Most sudden cardiac deaths in congenital heart disease survivors involve an electrophysiologic disorder, such as an arrhythmia. These can be ventricular tachycardia, ventricular fibrillation, an atrial arrhythmia, or asystole. But sudden death can also occur secondary to a vascular catastrophe, including pulmonary embolism or an aneurysm rupture. Hemodynamic abnormalities causing both mechanical and structural issues, such as acute heart failure, also cause sudden death.

So far, limited data exist to prove that effective interventions are available to help high-risk patients with congenital heart disease.

In patients who survived tetralogy of Fallot, a QRS duration of more than 180 msec is a reliable predictor of ventricular tachycardia and sudden death. Patients with a QRS duration this long have had a greater-than-twofold increased risk of sudden death, compared with patients who had a shorter QRS interval, Dr. Harris said at the congress, which was sponsored by the International Academy of Cardiology.

Indicators of hemodynamic abnormalities—such as increased right ventricular dimension, decreased right ventricular function, or a left ventricular ejection fraction of less than 40%—boost the risk for sudden death when they are coincident with prolonged QRS in patients who had tetralogy of Fallot.

In addition, inducible sustained monomorphic or polymorphic ventricular tachycardia during an electrophysiology study predicted sudden death with a sensitivity of 77% and specificity of 79%. Other noninvasive electrophysiologic measures—such as a signal-average ECG, T-wave alternans, and ambulatory ECG monitoring—have been less reliable for predicting risk.

A significant limitation on risk stratification of congenital heart disease patients is that this strategy presumes that identifying a high-risk patient is to be followed by an intervention that improves the patient's outcome. So far, limited data exist to prove that effective interventions are available to help these patients, Dr. Harris said. ■