

Evaluation of Coronary Artery Abnormalities and Sudden Death Risk in Williams Syndrome Patients Using Myocardial Perfusion Scintigraphy and CT Angiography

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Introduction: Sudden death risk in Williams syndrome (WS) patients has been shown to be 25-100 times higher than in the general population. This study aims to detect coronary artery anomalies and myocardial perfusion defects in WS patients using noninvasive diagnostic methods.

Patients and Methods: This study features 38 patients diagnosed with WS. In addition to physical examination, electrocardiography, and echocardiography, computed tomography (CT) angiography and rest/dipyridamole stress technetium-99m sestamibi (99mTc-sestamibi) single photon emission computed tomography (SPECT) myocardial perfusion scintigraphy (MPS) were performed.

Results: Twenty-one patients (55%) were male; 17 (45%) were female. The average patient age was 12±5 years (2.5-26 years); the average follow-up period was 7.2±4.2 years (6 months-18 years). Cardiovascular abnormalities were found in 89% of patients, the most common one being supraaortic stenosis (SVAS). CT angiography revealed coronary anomalies in 10 patients (26%), the most common ones being ectasia of the left main coronary artery and proximal right coronary artery as well as myocardial bridging. SVAS was present in 80% of patients with coronary artery anomalies. 99mTcsestamibi SPECT MPS revealed findings possibly consistent with myocardial ischemia in 29% of patients, and ischemia in 7 out of 10 patients (70%) with coronary anomalies shown on CT angiography (p=0.03).

Conclusion: Coronary artery abnormalities are relatively common in WS patients and are often accompanied by SVAS. CT angiography and dipyridamole 99mTc-sestamibi SPECT MPS seem to be less invasive methods of detecting coronary artery anomalies and myocardial perfusion defects in WS patients.