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The Coronary Arteries in Patients with Hypoplastic Left Heart Syndrome – An Angiographic Study and its Clinical Implications

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Introduction: Coronary anatomy in hypoplastic left heart syndrome (HLHS) has attracted increasing attention. This is the first angiographic study of coronary artery anatomy in HLHS while prior descriptions mostly based on post-mortem examinations.
Methods: Coronary angiograms obtained by selective native aortic root injection were reviewed in 84 patients (mitral atresia / aortic atresia, n=39; mitral stenosis / aortic stenosis, n=25; mitral stenosis / aortic atresia, n=13; mitral atresia / aortic stenosis, n=7). Origin and epicardial course, anomalies, coronary dominance and native ascending aorta dimensions were analyzed.
Results: Right dominance was present in 43 (51.2%), left in 31 (36.9%) and balanced type in 10 (11.9%) patients. Coronary dominance was unrelated to anatomic subtypes (p=0.163), but left dominance and balanced type was more common in patients with absent left ventricular cavity (p=0.011). Coronary artery fistulas were found in 15 (17.9%) and tortuosity of the coronary arteries, mostly affecting the left anterior descending artery, in 28 (33.3%) patients. Both occurred more frequently with mitral stenosis / aortic atresia (Tortousity 12 of 13 patients, p≤0.001; Coronary artery fistulas 6 of 13 patients, p=0.001). Collaterals to extra-cardiac vessels were visualized in 41 patients. The native ascending aorta was smaller with aortic atresia (44.4 ±20.5 mm^2/m^2 vs. 127.8 ±71.8 mm^2/m^2, p≤0.001). In 18 (21.4%) patients with relatively large native ascending aorta (113.8 ±69.2 mm^2/m^2 vs. 65.9 ±56.2 mm^2/m^2) retention of contrast media in the aortic root identified areas of low blood flow. One of these patients experienced myocardial infarct and cerebral stroke 7 years after catheterization and thrombus formation was detected within the aortic root.
Conclusions: Left dominance was more prevalent compared to the normal population. Observed anomalies were tortuosity, coronary artery fistulas and collateral vessels. The latter could be interpreted as a result of repeated surgical interventions. The long-term effect of these findings is unclear. Most coronary artery fistulas are small and coronary perfusion seems not to be altered. Native ascending aorta dimension remained larger in patients with aortic stenosis. This might bear a risk for thrombus formation in the aortic root. Anticoagulation might be warrantable.