The coronary arteries in patients with Hypoplastic Left Heart Syndrome – An angiographic study and its clinical implications

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Introduction

Coronary artery anatomy in hypoplastic left heart syndrome (HLHS) was predominantly assessed in post-mortem analysis and focused on coronary anomalies and morphologic changes. The impact of the coronary circulation on the outcome of HLHS patients is unclear. Evaluation of coronary anatomy in survivors of staged palliation for HLHS might be capable to identify anomalies and morphologic changes of the coronary arteries which have further implications for survival.

Methods

We reviewed coronary angiograms obtained by native aortic root injection of 84 patients. Epicardial course, anomalies, coronary dominance and native ascending aorta dimensions were analyzed.

Results

Angiography was performed prior to Hemifontan operation in 24 (28.6%) and before or after completion of the Fontan circulation in 47 (55.9%) and 13 (15.5%) patients, respectively. Median age at catheterization was 1.8 (0.1-9.9) years.

Results of native aortic root and coronary angiography

	MA/AA (n=39)	MS/AS (n=25)	MS/AA (n=13)	MA/AS (n=7)	Total (n=84)	p-value
LAD diagonals	39 (100.0%)	25 (100.0%)	13 (100.0%)	7 (100.0%)	**84 (100.0%)**	1.000
LAD septals	28 (71.8%)	16 (64.0%)	11 (84.6%)	7 (100.0%)	**62 (73.8%)**	0.368
Circumflex artery	39 (100.0%)	24 (96.0%)	13 (100.0%)	7 (100.0%)	**83 (98.8%)**	0.536
Coronary dominance						0.163
right	18 (48.7%)	14 (56.0%)	10 (84.6%)	1 (14.3%)	**43 (51.2%)**	
left	16 (41.0%)	9 (36.0%)	2 (15.4%)	4 (57.1%)	**31 (36.9%)**	
balanced	5 (12.8%)	2 (8.0%)	1 (7.7%)	2 (28.6%)	**10 (11.9%)**	
Origin of sinus node artery						0.704
RCA	12 (30.8%)	10 (40.0%)	6 (46.2%)	2 (28.6%)	**30 (35.7%)**	
CX	22 (56.4%)	10 (40.0%)	7 (53.8%)	4 (57.1%)	**43 (51.2%)**	
LCA	0 (0.0%)	1 (4.0%)	0 (0.0%)	1 (14.3%)	**2 (2.4%)**	
unknown	5 (12.8%)	4 (16.0%)	0 (0.0%)	0 (0.0%)	**9 (10.7%)**	
Coronary tortuosity						
Overall*	11 (28.2%)	6 (24.0%)	10 (76.9%)	1 (14.3%)	**28 (33.3%)**	0.005
LAD	9 (23.1%)	6 (24.0%)	10 (76.9%)	1 (14.3%)	**26 (31.0%)**	
CX	4 (10.3%)	0 (0.0%)	2 (15.4%)	0 (0.0%)	**6 (7.1%)**	
RCA	1 (2.6%)	0 (0.0%)	0 (0.0%)	0 (0.0%)	**1 (1.2%)**	
VCCs	7 (17.9%)	2 (8.0 %)	6 (46.2%)	0 (0.0%)	**15 (17.9%)**	0.026
Collateral arteries	24 (61.5%)	10 (40.0%)	5 (38.5%)	2 (28.6%)	**41 (48.8%)**	0.174

MA/AA: Mitral atresia/aortic atresia; MS/AS: Mitral stenosis/aortic stenosis; MS/AA: Mitral stenosis/ aortic atresia; MA/AS: Mitral atresia/aortic stenosis; LAD: left anterior descending; RCA: right coronary artery; CX: circumflex artery; LCA: left coronary artery; VCCs: ventriculocoronary connections. Asc. ao.: ascending aorta. P-values refer to global tests, footnotes indicate results of post-hoc tests. *MS/AA > MA/AA, MS/AA > MS/AS

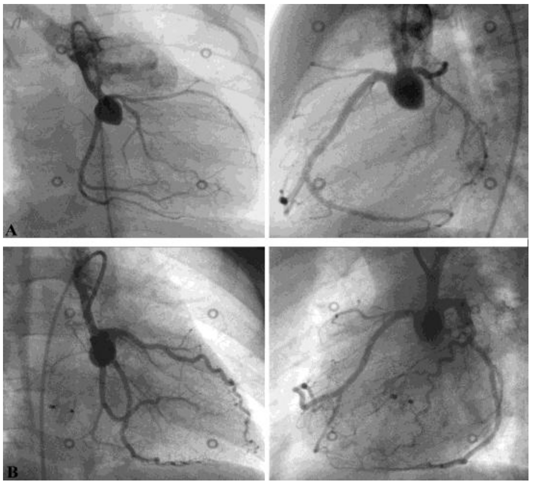

Figure 1. Native aortic root angiography in posterior-anterior and lateral view in a patient with mitral atresia/aortic atresia. Right dominance with the posterior descending artery arising from the right coronary artery. Origin of the sinus node artery from the right coronary artery (**A**). Native aortic root angiography in right and left anterior oblique view in a patient with mitral atresia / aortic atresia. Left dominance with the posterior descending artery arising from the circumflex artery. Tortuosity of the left anterior descending artery (**B**).

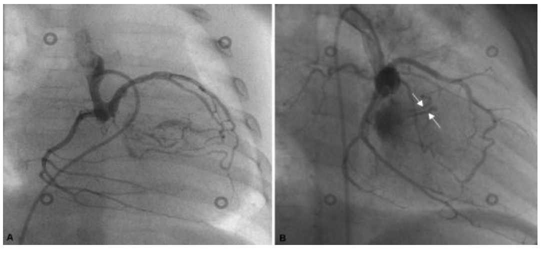

Figure 2. Ventriculocoronary connections were detected in 15 patients and more common in patients with mitral stenosis/aortic atresia. Native aortic root angiography in two different patients with mitral stenosis/aortic atresia shows multiple small ventriculo-coronary connections between the left anterior descending and circumflex artery and the residual left ventricular cavity in the first patient (**A**). Two ventriculocoronary connections (white arrows) between the left anterior descending artery and the residual left ventricular cavity are displayed in the second patient. Opacification of the residual left ventricular cavity is noted. (**B**)

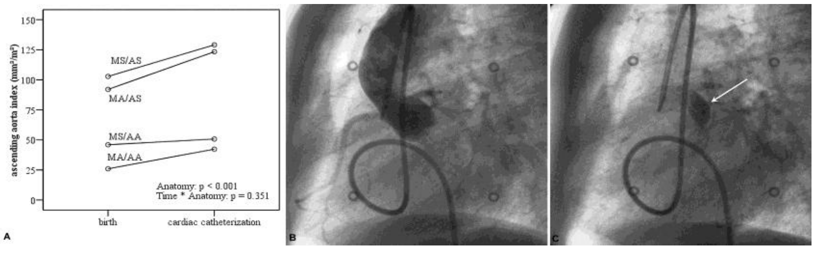

Figure 3. Estimated marginal means of the native ascending aorta index at birth and at cardiac catheterization showed a significant difference between anatomic subgroups but not for the interaction between anatomic subtype and time of measurement. This indicates that the native aorta did not had a significantly better growth potential in patients with aortic stenosis compared to those with aortic atresia (**A**). Native aortic root angiography did not show stenosis or thrombus formation in any patient. In 18 (21.4%) patients with relatively large native ascending aorta (113.8 ±69.2 mm²/m² vs. 65.9 ±56.2 mm²/m²) retention of contrast media in the aortic root identified areas of low blood flow. Native aortic root angiography in lateral view in a patient with mitral stenosis / aortic stenosis shows the native aortic root during contrast application (**B**) and after injection (**C**). Retention of contrast in the aortic root after the injection is noted (white arrow).

Conclusion

The epicardial course of the coronary arteries and the separation into the main branches were basically similar to normal hearts in the majority of patients. Left coronary dominance was more common with HLHS than in the normal population. Observed anomalies of the coronary arteries were tortuosity, ventriculocoronary connections (VCCs) and collateral vessels. The latter could be interpreted as a result of repeated surgical interventions. None of these findings seems to have any hemodynamic adverse effects. VCCs and tortuosity of the coronary arteries were more frequently found with MS/AA. VCCs were usually small and did not cause relevant diastolic runoff. They were never associated with coronary stenosis or interruption, which could impair myocardial perfusion. The native ascending aorta did not have a significantly better growth potential in patients with aortic stenosis compared to those with aortic atresia. A potential risk for thrombus formation in the native aortic root might exist for patients with a dilated native aortic root. Anticoagulation should be considered in this subset of patients to prevent coronary obstruction or systemic embolism.