walled and kinking coronary arteries were observed, especially multiple ventriculocoronary arterial connections and thick-walled and kinking coronary arteries were observed, especially in those infants with patent left ventricular inflow and obstructed left outflow [3].

Over the past two decades, we have continuously progressed in our ability to treat these patients pre-, peri- and postoperatively. We have learned that the success is deeply rooted in novel surgical techniques in conjunction with excellent intensive care therapy, particularly regarding the principle of a well-balanced relationship between pulmonary and systemic flow.

Thanks to staged Norwood operations nowadays, the most children with HLHS achieve palliation to Hemi-Fontan and modified...
Fontan circulation. However, this remains one of the most challenging high-risk procedures in cardiac surgery, with mortality rates over 10–15%, followed by additional inter-stage mortality and still unknown long-term results [4].

Hansen et al. [5] present new scientific findings in this issue, confirming the early post-mortem series with the high rate of severe coronary disorders in HLHS. But this confirmation is not an autopsy series, but rather the first large, intravitam angiographic evaluation of the coronary artery anatomy in 84 survivors with HLHS with a median age of 1.8 years. This is a new population that would not have survived two decades ago. Hansen et al. describe the epicardial course and coronary artery anomalies, as well as the dimensions of the native aorta, pointing out potential risk factors regarding the long-term course of these patients. Surprisingly, half of the patients presented collaterals between coronaries and extracardiac vessels, either resulting from postoperative adhesions (as 70% of them had undergone two or more extensive surgical procedures), because vessel growth might have been endogenously triggered by cyanosis and the additional need for cardiac blood supply. One should keep in mind that these vessels may cause coronary steal phenomenon in later life.

Another important revelation from the innovative paper of Hansen et al. that has the potential to affect the long-term treatment of these children is based on the diameter of the aorta: in contrast to the subgroup with aortic atresia and severe hypoplastic aorta ascendens, the early surgical Norwood procedure seems to be easier in children with antegrade flow via aortic stenosis and a moderate ascending aorta diameter, but we need to remember that due to competitive antegrade and retrograde flow in the aortic root, resulting in stasis, these children are at high risk for thrombembolic events, which thus requires more intensive lifelong anticoagulation.

Both the diameter of the aorta ascendens and the structure and anomalies of the coronary arteries are risk factors in the treatment of HLHS. It is key that we keep the early postmortem examinations in mind and compare them with our current coronary angiography study.

In echocardiographic examinations, Vida et al. [6] found 23% with the subtype mitral stenosis and aortic atresia (MS-AA) to represent the high-risk group (mortality 29 vs. 8% in the other subtypes). Fifty per cent of this MS-AA subtype showed ventriculocoronary connections, and in these cases early mortality rose to 50%. This is unfortunately a key limitation, namely that the non-survivors of Norwood-I—who might have the worst coronary anomalies—could not be considered for the analysis by Hansen et al. Especially, the larger ventriculocoronary connections causing steal phenomenon of blood from the left coronary artery via sinusoids into the rudimentary left ventricle and across mitral regurgitation into the atrium may be under-represented. This steal phenomenon is well known in pulmonary atresia with an intact ventricular septum and may be underrepresented in HLHS survivors. It seems possible that these are children whose reasons for early peri- and postoperative death during the Norwood-I procedure seemed to be unclear in the past.

The findings of Hansen et al. help us learn more about coronary arteries as risk factors for dismal outcome in HLHS. With a better understanding of risk factors, such as ventriculocoronary arterial connections, and thick-walled and kinking coronary arteries, more timely therapy decisions can be made, leading to better total outcome results in the future of these challenging patients.

Yet, we also know that new scientific findings lead us to pose better questions, followed by new solutions. This coronary analysis represents an important step further toward stretching the borders and achieving better results in predicting prognoses and in treating children with hypoplastic left heart syndrome.

**REFERENCES**


