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Prenatal lesions of the myocardium in patients with congenital single-ventricle heart disease

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Today congenital heart disease with functionally only one ventricle can be remediated successfully by surgery.

We report two cases with univentricular heart. One showed a hypoplastic left ventricle (HLHS), atresia of the aortic valve and restriction of the foramen ovale (case I). The other showed a hypoplastic right ventricle (HRHS), atresia of the pulmonary valve, an intact ventricular septum and a large fistula between the left anterior descending artery and the right ventricular outflow tract (case II).

In case I, a modified Norwood-I operation (Blalock-Taussig anastomosis, reconstruction of the aortic arch and atrioseptectomy) was performed at the age of 4 days. This child died unexpectedly at the age of 9 days, presumably due to a thrombosis of the anastomosis.

Also in case II, a Blalock-Taussig anastomosis was constructed. The coronary fistula was thereby disconnected temporarily. This prematurely born twin died in tabula at the age of 15 days.

Using light-optical microscopy, the myocardium of both children showed severe lesions, in the case of HLHS: on the right, in the case of HRHS: on the left side. Histologically, signs of necrosis of different ages, partly presenting scars and calcification, could be observed. This necrosis had to be older than the time since delivery.

No cardiotropic virus could be detected in samples obtained from myocardium using nested-PCR on paraffin-embedded tissue.

We conclude the already intrauterine myocardial load and hypoxaemia lead to irreversible damage of the myocardium, thus limiting the prognosis of these children.