ST-segment elevation myocardial infarction (STEMI) due to severe anemia resulting from transient erythroblastopenia of childhood in a 2-year-old boy after arterial switch operation in the absence of coronary artery stenosis

R. Bindermann1, D. Vilser1, E. Limpert1, K. Kentouche1, B. Gruhn1, P. Flosdorff2, B. Hennig1

1 Pediatric Clinic, University Hospital Jena, Germany
2 Pediatric Cardiology, Leipzig Heart Center, Germany

Contact: robert.bindermann@meduni-jena.de

Background
Myocardial ischemia or infarction occurs when the oxygen demand of the myocardium exceeds its supply. In severe anemia the cardiac output is increased whereas the oxygen supply of the myocardium is reduced. This can result in an oxygen supply-demand mismatch and relevant myocardial hypoxemia (1). Both in children and adults a hemoglobin level above 4.3 mmol/l (~ 7 g/dl) is considered safe in terms of complications and in no need of blood transfusion (2, 3). Below 3.1 mmol/l (~ 5 g/dl) even otherwise healthy individuals start to show ST-segment alterations in ECG (1). To our knowledge a myocardial infarction or ischemia due to severe anemia (except for sickle-cell anemia) has not been described in children so far. We report on a myocardial infarction due to anemia in a nearly two-year-old boy after transposition of the great arteries.

Case report
Arterial switch operation (ASO) was performed on a 9-day-old boy with transposition of the great arteries. The postoperative course was uneventful except for a moderate supraventricular pulmonary stenosis and consecutive right ventricular hypertrophy. Aged 1 year 9 months he presented with a history of fatigue of several days. On examination he was very pale with tachycardia and tachypnoea. A loud systolic murmur and upper venous congestion were noted. Diagnostics revealed a severe normochromic normocytic anemia. Hemoglobin was 1.7 mmol/l = 2.7 g/dl, hematocrit was 8%. Cardiac troponin I peaked at 0.98 ng/ml (normal <0.04 ng/ml), CK-MB at 10.5 ng/ml (normal <6.6 ng/ml) and BNP was maximally 4990 pg/ml (normal <20 pg/ml). The electrocardiogram showed ST elevation in leads aVR and V1, and ST depression in leads I, II and V4, V5 and V6. We diagnosed an acute right ventricular myocardial infarction. Apart from a left ventricular shortening fraction at the lower range echocardiography was unremarkable. A hepatopathy with significant elevation of ALT, AST, GDH and a coagulation dysfunction (INR 1.7) was found. The patient responded well to erythrocyte transfusion. His physical condition and all laboratory values normalized, the recovery was uneventful. He received Captopril over six weeks. Neither an echocardiographic dysfunction nor clinical sequelae were observed during follow-up. Further diagnostics revealed transient erythroblastopenia of childhood to be the cause of the anemia. No other hematological abnormalities were found. Coronary angiography showed smooth coronary arteries without stenosis. A moderate supravalvular pulmonary stenosis with a pressure gradient of 45 mmHg was confirmed during cardiac catheterisation.

Transient erythroblastopenia of childhood (TEC)
TEC is a transient, normochromic normocytic anemia due to a temporary stop of erythropoiesis of unknown origin. Previous viral infection might play a role. Affected patients typically are 1 to 4 years old and present with a moderate anemia and hemoglobin levels between 3.5 and 5.0 mmol/l, but even more severe anemia can be seen. However, cardiac involvement has not been described so far. Patients normally recover spontaneously within weeks, initial blood transfusions are occasionally necessary. Diamond-Blackfan anemia is the most important differential diagnosis (4, 5).

Discussion
In our 2-year-old patient after ASO severe anemia led to a relevant oxygen supply-demand mismatch and hypoxemia of the myocardium. Due to this condition heart failure and hepatopathy developed and a ST-segment elevation myocardial infarction (STEMI) was observed. The patient responded well to erythrocyte transfusion. Diagnostics revealed transient erythroblastopenia of childhood as cause of the anemia. Coronary artery stenosis, which occurs in patients who underwent arterial switch operation in about 7% in the further course (6), was ruled out by coronary angiography. The pre-existing moderate supravalvular pulmonary stenosis and the cardiac impairment resulting from previous heart surgery possibly contributed to the extend of the myocardial damage. To our knowledge a myocardial infarction due to anemia had not been described in children so far.

References:
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