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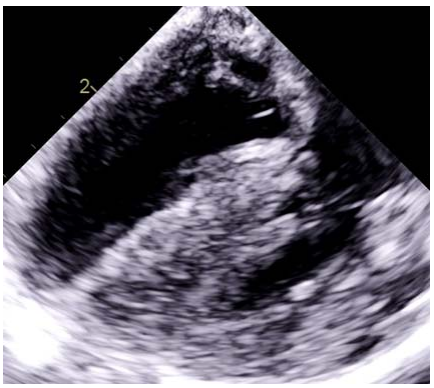
Suspected protamine-induced lethal myocardial ischemia after congenital heart surgery in an infant with congenital disorder of glycosylation



Panagiotou E.(1), Ostermayer S.H.U.(1), Gruszka A.(1), Koester S.(1), Hanten J.(1), Maizza A.F.(2), Vazquez J.(2), Buding B.(3), Kerst G.(1)
Department of Pediatric Cardiology, RWTH Aachen University Hospital, Germany (1); Department of Pediatric and Congenital Heart Surgery, RWTH Aachen University Hospital, Germany (2); Department of Anesthesiology, RWTH University Hospital Aachen, Germany (3)

Introduction:
Congenital heart disease (CHD) can be associated with the heterogenous group of congenital disorders of glycosylation (CDG). Affected children are exposed to risks of both thrombosis and bleeding. We report on lethal myocardial ischemia associated with heparin neutralization by protamine after cardiopulmonary bypass (CPB) for biventricular repair in an infant with CDG and Double-Outlet Right Ventricle (DORV).

Methods:
A 10 month old infant with CDG type Ia and DORV with a severely hypoplastic pulmonary valve and hypoplastic pulmonary arteries was admitted for surgery. Previously, a 2,5mm modified BT-Shunt had been implanted at 6 weeks of age because of increasing cyanosis. To further promote pulmonary artery growth, balloon angioplasty of the shunt and the hypoplastic pulmonary valve had been performed at 6 months of age. After shunt implantation the infant received acetylsalicylic acid at 3mg/kg/d despite congenital thrombocytopenia. Coagulation tests were normal. There never was evidence for bleeding or thrombosis. Biventricular repair consisted of VSD-closure, transannular pulmonary patch-plasty and implantation of a monocusp contegra conduit. Transesophageal echocardiography (TOE) showed a good biventricular function with no residual defects and no pulmonary regurgitation. Weaning from CPB was uneventful. Protamine was given for heparin neutralization, immediately followed by cardiopulmonary reanimation an intensified catecholamine support. TOE showed new-onset massive cardiac edema with severely impaired biventricular function accompanied by ST-elevations on ECG. We decided to put the patient on extracorporeal membrane oxygenation (ECMO) using heparin for anticoagulation. Neither excessive bleeding nor thrombosis occurred. The myocardial edema initially regressed only to recur upon reduction of ECMO-flow. Given the severe neurodevelopmental delay and the limited prognosis in CDG, parental consent was given to discontinue ECMO-support on the 6th postoperative day. The patient died 24 hours later in catecholamine-resistant heart failure.

Results:
Administration of protamine in an infant with CHD and CDG resulted in massive myocardial edema leading to severely impaired biventricular function and death.

Conclusions:
Protamine as a polycationic protein might interact with abnormally glycosylated membranes in CDG and lead to thrombotic myocardial microangiopathy, myocardial ischemia and terminal heart failure. It deems pruden to avoid protamine during CPB in patients with CHD and CDG.