Background:
Management of bleeding in congenital cardiac surgery involving cardiopulmonary bypass is challenging, but is especially complicated in patients with other endogenous coagulation deficiencies. In acquired coagulopathies the clinical management based on routine parameters, e.g. PT/INR, PTT, platelet count, and fibrinogen etc., can be difficult. This has lead to an increasing interest in point-of-care testing devices such as rotational thrombelastometry (ROTEM®), which may better reflect hemostasis in such complex coagulopathies. EXTEM is one of the ROTEM essays that works by activation of coagulation through added tissue factor and is used for assessment of coagulation factor deficiencies of the extrinsic pathway. We report a case of a severely coagulopathic neonate scheduled for repair of complex congenital heart disease with preoperatively diagnosed factor VII deficiency, which was undetectable by ROTEM® EXTEM analysis.

Case Report:
The patient was born at 36 weeks and 2.6 kg body weight with pulmonary atresia/intact ventricular septum and Ebsteins anomaly of the tricuspid valve. He was noted to be coagulopathic shortly after birth during his atrial septostomy in the cardiac catheterization suite with a baseline ACT of 203 seconds and PT of 26 seconds. Prior to his cardiac surgery on day three of life (right modified Blalock-Taussig shunt with transannular patch and pulmonary homograft and division of his patent ductus arteriosus) a hematological workup was performed which was most consistent with factor VII deficiency (factor VII level of 18%). In order to optimize his status before surgery and to aid the diagnostic workup, a baseline blood sample was analyzed using ROTEM and unexpectedly resulted in a normal EXTEM study. In view of his low factor VII level, two doses of activated factor VII (90 mcg/kg) were given preoperatively without a change in his ROTEM analysis. In the postoperative period the patient was clinically bleeding despite normal routine coagulation studies as well as normal ROTEM analysis, even after multiple doses of factor VIIa as well as multiple blood products.

Discussion:
The surprising part about this case is that the baseline EXTEM clotting time was not prolonged despite a low factor VII level of 18% and a prolonged PT. In the EXTEM essay, coagulation is activated with a supraphysiologic dose of tissue factor (TF). We speculate that the amount of factor VII present in the patient sample was sufficient to generate a small amount of thrombin when activated by a supraphysiologic dose of TF. The initial thrombin generated in the closed environment of the testing cup subsequently allowed normal amplification of coagulation via thrombin-mediated activation of factors XI, VIII, and V (1). Thus, the supraphysiologic level of TF overcame the low factor VII level of the patient, thereby producing a falsely normal test result.

Conclusion:
ROTEM® EXTEM analysis may not be a suitable test for single coagulation factor deficiencies.

References: