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MANAGING DUAL RISKS: PERCUTANEOUS CORONARY REVASCULARIZATION IN CONGENITAL FACTOR VII DEFICIENCY

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УПРАВЛЕНИЕ НА ДВОЙНИЯ РИСК: ПЕРКУТАННА КОРОНАРНА РЕВАСКУЛАРИЗАЦИЯ ПРИ ВРОДЕН ДЕФИЦИТ НА ФАКТОР VII

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Abstract. Factor VII deficiency is a rare inherited coagulation disorder characterized by decreased activity of factor VII, leading to variable bleeding tendencies that may not correlate with measured FVII levels. While many patients remain asymptomatic, others can experience severe spontaneous hemorrhages. The condition poses significant challenges during surgical or invasive procedures due to the potential for uncontrollable bleeding. Data regarding percutaneous coronary intervention (PCI) in such patients are extremely limited, mostly confined to isolated case reports. PCI requires anticoagulation during the procedure and dual antiplatelet therapy afterward, both of which elevate bleeding risk. Conversely, administering FVII concentrate may increase the chance of thromboembolic events. Therefore, individualized planning and a multidisciplinary approach are crucial to balance these opposing risks. We present the case of a 75-year-old Bulgarian woman with congenital FVII deficiency and severe three-vessel coronary artery disease who underwent successful transradial PCI without FVII replacement. Drug-eluting stents were implanted in the left anterior descending and circumflex arteries, enabling short-term dual antiplatelet therapy. No bleeding complications occurred peri-procedurally, and only one minor episode of epistaxis was observed during follow-up. This case illustrates that, with meticulous preparation and procedural care, PCI can be a safe and effective revascularization strategy in patients with congenital FVII deficiency.

Key words: percutaneous coronary intervention; factor VII deficiency; short-duration dual antiplatelet therapy; case report

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Резюме. Дефицитът на фактор VII е рядко наследствено нарушение на коагулацията, характеризиращо се с намалена активност на фактор VII, което води до различна степен на кървене, не винаги съответстваща на измерените нива на фактора. Докато част от пациентите остават безсимптомни, при други се наблюдават тежки спонтанни хеморагии. Състоянието представлява сериозно предизвикателство при хирургични и инвазивни процедури поради риска от неконтролируемо кървене. Данните относно перкутанната коронарна интервенция (PCI) при такива пациенти са изключително ограничени и се базират основно на единични клинични съобщения. Провеждането на PCI изисква интраоперативна антикоагулация и последваща двойна антиагрегантна терапия, които повишават риска от кървене, докато приложението на концентрат от фактор VII може да увеличи вероятността от тромбоемболични усложнения. Поради това, индивидуализираното планиране и мултидисциплинарният подход са от решаващо значение за балансиране на тези противоположни рискове. Представяме случай на 75-годишна българка с вроден дефицит на фактор VII и тежка триклонова коронарна болест, при която успешно е проведена трансрадиална

PCI без заместителна терапия с фактор VII. Имплантирани са медикамент-отделящи стентове в лявата предна низходяща и лявата циркумфлексна артерия, което позволява краткотрайна двойна антиагрегантна терапия. Не са наблюдавани кръвоизливни усложнения в перипроцедурния период, а по време на проследяването е регистриран един лек епизод на епистаксис, овладян консервативно. Този случай показва, че при внимателна подготовка и прецизно провеждане на процедурата PCI може да бъде безопасна и ефективна стратегия за реваскуларизация при пациенти с вроден дефицит на фактор VII.

Ключови думи: перкутанна коронарна интервенция; дефицит на фактор VII; краткотрайна двойна антиагрегантна терапия; клиничен случай

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Introduction

Factor VII deficiency is an uncommon autosomal recessive coagulation disorder resulting in decreased activity of factor VII (FVII) within the coagulation cascade [1]. The clinical spectrum is highly variable and often does not correlate with measured FVII activity, ranging from asymptomatic individuals to cases of spontaneous bleeding in vital organs [1-4]. This condition may cause severe, uncontrolled bleeding in patients undergoing surgical interventions [5, 6], yet data concerning percutaneous coronary intervention (PCI) remain extremely limited, mostly restricted to single case reports [7-9]. On one hand, intraprocedural anticoagulation and subsequent dual antiplatelet therapy significantly elevate the bleeding risk; on the other, FVII supplementation in the form of recombinant factor VII, plasma, or prothrombin complex concentrate may predispose to thromboembolic complications during the procedure [5, 8]. Therefore, such clinical scenarios necessitate a multidisciplinary approach, meticulous pre-procedural planning, and individualized management strategies. We present a case of a patient with chronic coronary syndrome, three-vessel coronary artery disease, and congenital FVII deficiency who successfully underwent percutaneous coronary revascularization with drug-eluting stent implantation.

Case presentation

A 75-year-old white Caucasian Bulgarian woman with a history of hypertension, type 2 diabetes mellitus, and dyslipidemia presented with substernal chest pain provoked by mild exertion. The symptoms had begun approximately four months before presentation, lasted several minutes, and resolved with rest. The patient had been diagnosed with congenital factor VII deficiency in early childhood following recurrent spontaneous epistaxis. Apart from occasional nosebleeds and mild perimenstrual bleeding, there was no history of other spontaneous or provoked hemorrhagic events. One month before admission, she underwent surgical extraction of a biliary concrement, during which recombinant factor VII was administered perioperatively.

Physical examination revealed no pathological findings. Careful inspection of the skin and visible mucosa showed no petechiae, bruising, or active bleeding. Vital signs were within normal limits. A resting electrocardiogram (ECG) demonstrated normal sinus rhythm with pathological Q waves and inverted T waves in leads II, III, and aVF, consistent with an old inferior myocardial infarction. Transthoracic echocardiography showed mild left atrial enlargement (LA diameter 4.0 cm), left ventricular hypertrophy (interventricular septum 1.4 cm; posterior wall 1.51 cm), preserved left ventricular ejection fraction (51%), and mild mitral regurgitation. Laboratory investigations revealed coagulation abnormalities typical of FVII deficiency (Table 1).

Coronary angiography demonstrated three-vessel coronary disease with critical stenosis in the mid segment of the left anterior descending artery (LAD) **(Figure 1)**, high-grade significant stenosis of proximal left circumflex artery (LCX) **(Figure 2)**, and a chronic total occlusion of the right coronary artery.

The Heart Team – consisting of a cardiologist, interventional cardiologist, cardiothoracic surgeon, and hematologist – evaluated the case and concluded that the patient was an appropriate candidate for coronary artery bypass grafting. However, the patient declined surgical treatment, and percutaneous coronary revascularization was subsequently undertaken. The procedure was performed via a transradial approach using a 5 French introducer sheath. The intervention on the LAD artery involved balloon pre-dilatation followed by implantation of a drug-eluting stent (DES – BioFreedom, Biosensors) at the site of the mid-LAD stenosis (**Figure 3**). The lesion in the LCX artery was similarly managed with balloon pre-dilatation and DES (BioFreedom, Biosensors) implantation (**Figure 4**). Final angiography demonstrated thrombolysis in myocardial infarction grade III flow and excellent immediate procedural results.

Table 1. The patient's laboratory test results at initial presentation, at the time of PCI, at admission for epistaxis during follow-up. Units are given in brackets after the name of the laboratory assay

Laboratory assay	At initial presentation (17 days prior to PCI)	At the time of PCI	At admission for epistaxis (17 days after PCI)
Haemoglobin (g/L)	136.0	133.0	88.0
RBC (1012/l)	4.1	4.2	2.96
HCT (L/L)	0.407	0.407	0.274
PLT (109/l]	187	185	226.0
Prothrombin time (%)	No coagulation	No coagulation	No coagulation
Activated partial thromboplastin time (s)	24.2	24.4	23.4
Factor VII plasma activity (%)	0%	-	-
Troponin I (ng/ml)	0.04	0.01	0.01
Creatin kinase MB (U/L)	12	14	33

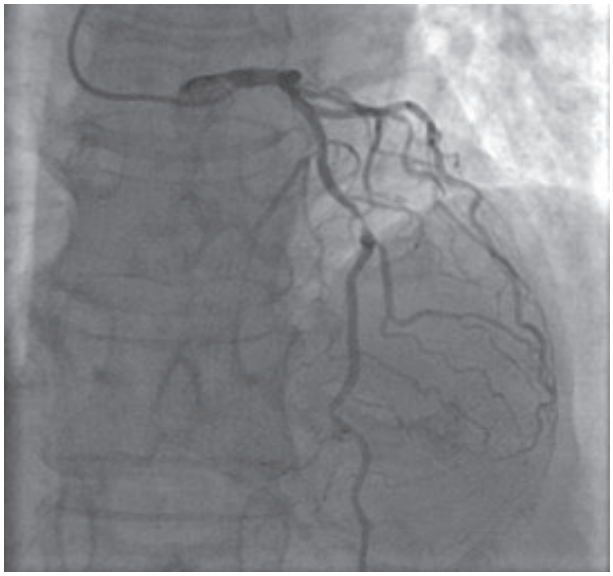

Figure 1

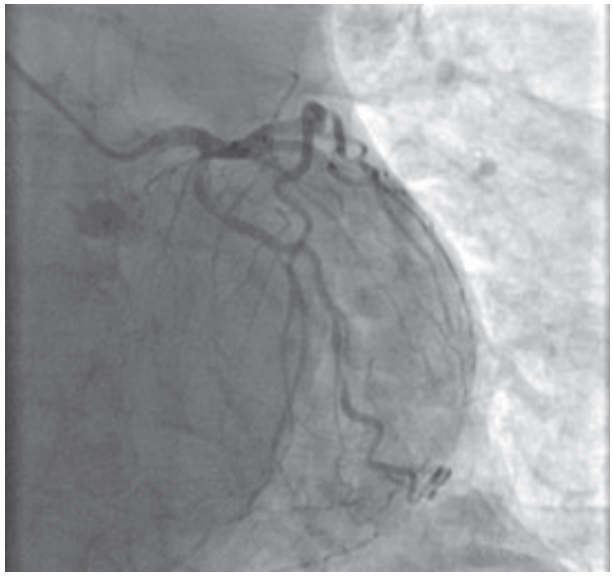

Figure 2

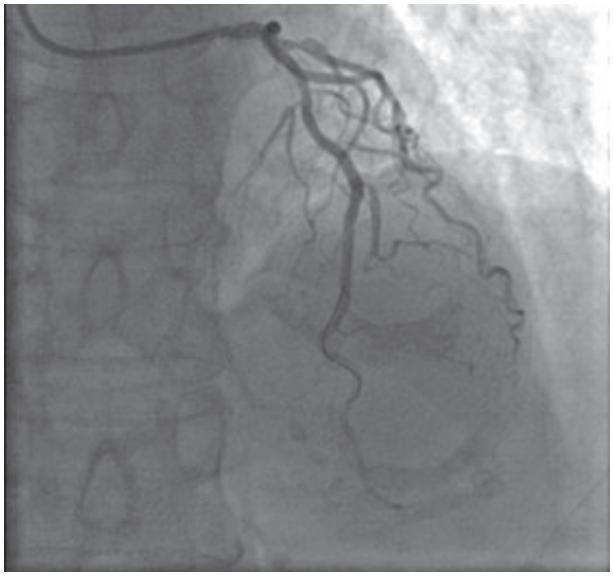

Figure 3

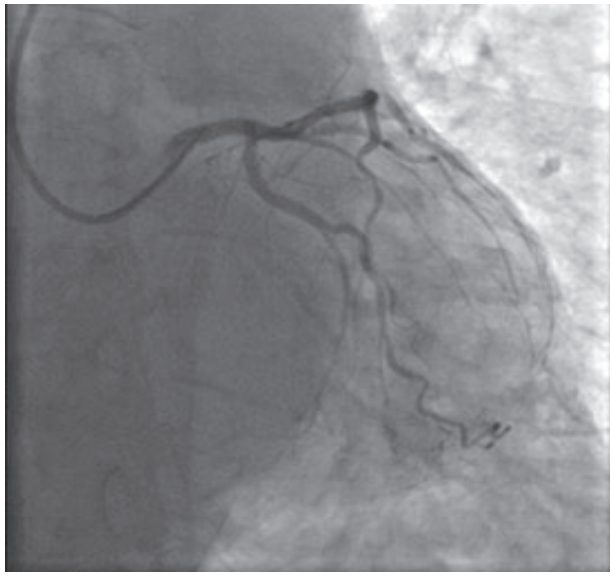

Figure 4

During the intervention, unfractionated heparin was administered at a dose of 100 U/kg. Activated clotting time (ACT) was measured – 372. Local haemostasis at the puncture site was successfully achieved through bandage compression. Recombinant factor VII and fresh frozen plasma were kept available for immediate use if required, but neither was ultimately administered. No bleeding complications occurred during hospitalization.

At discharge, the patient was prescribed dual antiplatelet therapy (DAPT) for 28 days – aspirin 75 mg once daily and clopidogrel 75 mg once daily. In addition, a statin, ezetimibe, angiotensin receptor blocker, diuretic, and beta-blocker were initiated as part of secondary prevention.

Seventeen days post-discharge, the patient was readmitted with persistent epistaxis that had continued for several days without prior treatment, resulting in a decrease in haemoglobin levels (Table 1). Management included transfusion of two units of plasma and two units of packed red blood cells, along with local haemostatic measures, which successfully controlled the bleeding. The patient was subsequently discharged on single antiplatelet therapy with clopidogrel 75 mg daily for one week.

No further bleeding events have been documented to date. At six-month follow-up, the patient remains clinically stable and free of symptoms.

Discussion and conclusions

We describe the case of a patient with congenital factor VII deficiency and severe three-vessel coronary artery disease who underwent successful percutaneous coronary revascularization. To our knowledge, only a few similar cases have been reported in the literature to date, and none involved patients with FVII activity below 3%, a level considered to confer a high risk of bleeding complications [7-9].

Due to the rarity of this condition, there are currently no evidence-based recommendations regarding the optimal management of such patients, making an individualized, multidisciplinary approach essential.

Several challenges arose in determining the therapeutic strategy. Firstly, percutaneous coronary intervention, unlike surgical revascularization, carries a dual risk—coronary and arterial thrombosis in addition to the inherent bleeding tendency associated with coagulopathies [8]. To minimize the risk of local bleeding and hematoma formation, a transradial approach was selected. Similar reasoning guided other published reports, where radial access was favoured for its safety [9], or vascular closure devices were employed following femoral access [7].

Unfractionated heparin was used as the intraprocedural anticoagulant because its anticoagulant effect can be monitored via activated clotting time (ACT) and promptly reversed if necessary. Heparin was administered at the standard dose of 100 U/kg. Given that recombinant FVII replacement carries a potential thrombotic risk [2, 8], periprocedural supplementation was withheld. The intervention was thus performed at 0% factor VII activity, with recombinant FVII and plasma readily available in case of bleeding. In previously published cases, peri-procedural factor replacement was administered without reported thromboembolic events [7, 9].

Another consideration in patients with coagulation disorders undergoing PCI is the requirement for DAPT. To reduce DAPT duration, both LAD and LCX lesions were treated within a single procedure – representing, to our knowledge, the first such report in a patient with congenital FVII deficiency. To reduce the bleeding risk, new-generation drug-eluting stents permitting short-term DAPT (only 28 days) were implanted, and the patient was discharged on aspirin 75 mg and clopidogrel 75 mg daily for one month. Earlier case reports describe the use of bare-metal stents with DAPT durations of at least one month [7, 9].

Since the long-term prognosis in such patients depends primarily on the progression of ischemic heart disease, ongoing management should focus on optimal medical therapy for ischemia, excluding prolonged antiplatelet or anticoagulant use where possible.

Although one bleeding episode occurred during follow-up, the need for transfusion was likely due to persistent bleeding and delayed haemostatic intervention. Following in-hospital management, haemostasis was achieved, and no further adverse events were recorded during the six-month post-procedural period.

This report has several important limitations that should be considered when interpreting the findings. It describes a single clinical case, which limits the generalizability of the findings. Genetic characterization of factor VII deficiency and global coagulation assessment using viscoelastic methods were not performed, which may have restricted a more detailed evaluation of the patient's hemostatic profile. In addition, the relatively short follow-up period does not allow firm conclusions regarding late thrombotic or hemorrhagic complications.

No conflict of interest was declared

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