

# Acquired Factor X Deficiency

Deeksha Sarma, MD,<sup>1</sup> Rama Bhagavatula, MD,<sup>2</sup> Prerna Mewawalla, MD<sup>2</sup>

<sup>1</sup> Department of Medicine; <sup>2</sup> Division of Hematology and Oncology, Allegheny Health Network, Pittsburgh, PA



## Introduction

Acquired Factor X (FX) deficiency is a rare clinical entity, presenting with a wide spectrum of hemorrhagic manifestations. FX, a factor in the common pathway of the coagulation cascade, is integral to the formation of fibrin clot and generation of thrombin. The FX inhibitor itself is thought to be due to either a specific FX antibody or lupus anticoagulant, however limited research has been performed in this area.

## Case Presentation

- A 64 year old male presented to an outside facility after experiencing gross hematuria at home, associated with a new onset petechial rash across his body and intermittent fevers as high as 103 F.
- On presentation, labs were notable for prolonged prothrombin time (PT), activated partial thromboplastin time (aPTT), and international normalized ratio (INR) (Table 1). Mixing studies revealed a partial correction of PT and aPTT indicating presence of an inhibitor to FX (Table 2).
- During his hospital stay, he developed acute kidney injury; rheumatologic workup for glomerulonephritis was begun. Laboratory analysis revealed hypocomplementemia, a positive ANA of 1:180, and positive lupus anticoagulant.
- Workup for amyloidosis was also undertaken, including tissue biopsy of his rash, bone marrow biopsy, and abdominal fat pad biopsy, all of which proved negative for amyloidosis.
- Despite multiple units of FFP and FEIBA administration, FX levels did not improve until empiric steroid therapy for FX inhibitor disease was started. At the time of discharge, FX levels continued to improve, and he was discharged home on prolonged steroid taper. Since discharge, he has maintained normal levels of FX off of steroids (Table 1).

## Discussion

- Acquired FX deficiency can stem from a number of pathologies, most often associated with systemic amyloidosis, however also seen with respiratory pathology, malignancy, or as an idiopathic finding.
- Lupus anticoagulant is one identified cause of acquired FX deficiency, and anti-FXa antibodies have been shown to have a high affinity to FX in the setting of antiphospholipid antibodies.
- Treatment is often directed towards the underlying cause of FX inhibitor. Therapies may include steroids, antibiotics, immunosuppressive agents, or even plasmapheresis.

**Table 1.** Coagulation studies at various time points, including at presentation to our facility, few days after beginning empiric steroid therapy, at the time of discharge, and one year later in the outpatient setting.

	At presentation	3 days after starting steroids	Discharge	One year later
PT (sec)	82.4	28.9	17.8	10.3
INR	10.6	2.8	1.5	0.88
PTT (sec)	71	42	28	
FX (U/mL)	<0.12	0.17	0.35	1.16

## Conclusion

Though rare, acquired FX inhibitor is an important diagnosis, given the manifestations and significant morbidity conferred. Diagnosis may also uncover underlying etiologies including amyloidosis, malignancy, or lupus anticoagulant.

## References

1. Uprichard J, Perry DJ. Factor X deficiency. *Blood Reviews*. 2002;16(2):97-110. doi:[10.1054/blre.2002.0191](https://doi.org/10.1054/blre.2002.0191)
2. Ashrani AA, Aysola A, Al-Khatib H, Nichols WL, Key NS. Lupus anticoagulant associated with transient severe factor X deficiency: a report of two patients presenting with major bleeding complications: *Short Report. British Journal of Haematology*. 2003;121(4):639-642. doi:[10.1046/j.1365-2141.2003.04325.x](https://doi.org/10.1046/j.1365-2141.2003.04325.x)
3. Hsia CC, Keeney M, Bosco AA, Xenocostas A. Treatment of acquired factor X inhibitor by plasma exchange with concomitant intravenous immunoglobulin and corticosteroids. *American Journal of Hematology*. 2008;83:318-320.
4. Brown DL, Kouides PA. Diagnosis and treatment of inherited factor X deficiency: FACTOR X DEFICIENCY. *Haemophilia*. 2008;14(6):1176-1182. doi:[10.1111/j.1365-2516.2008.01856.x](https://doi.org/10.1111/j.1365-2516.2008.01856.x)

**Table 2.** Mixing studies of PT and PTT, revealing partial correction of both parameters.

PT (Ref: 11.8-14.3 sec)	69.0
PT Mix	18.1
aPTT (Ref: 23-34 sec)	71
PTT Mix	37