## PB1370 | Acquired FXIII Inhibitor: Patient Characteristics and Treatment Outcome, a Case Series

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**Background**: Acquired FXIII inhibitor, or autoimmune hemophilia-like disease due to anti-FXIII antibody, is a rare but possibly under-diagnosed bleeding disorder. To date, reported cases have been less than a hundred but the number increased rapidly in recent years, especially in Japan. With normal result in most screening coagulation assays, this disease is easily overlooked. Most clinics still don't have FXIII activity assay or even FXIII screening assay, such as urea solubility test. Hence true incidence could be higher than reported. Because of rarity, there was no treatment guideline available. In some reports, physicians treated it similarly as acquired hemophilia A.

**Aims**: To gain more understanding of acquired FXIII inhibitor.

**Methods**: From Jan 2015 to Dec 2018, we collected consecutive patients diagnosed with acquired FXIII inhibitor. FXIII activity and inhibitor were measured by an amine incorporation assay and Bethesda method, respectively.

**Results**: We found 5 patients with detectable FXIII inhibitor, including 1 male and 4 females. Four of them were older than 70. Two had autoimmune disease and none had cancer or used attributable medications. All patients presented with ecchymosis and intramuscular hematoma. No life-threatening bleeding was observed. Delayed diagnosis was common with varied time period to achieve a correct diagnosis. (table 1)

All bleedings were treated and improved by cryoprecipitate with or without tranexamic acid. Steroid was given to all patients and cyclophosphamide, rituximab, and other immune suppressants were also used. FXIII inhibitor was totally resolved in two, partially resolved in one (0.1-0.5 BU), and persisted in two patients with minimal bleeding. (table 2)

**Conclusions**: We presented 5 patients with acquired FXIII inhibitor. The most common presentations were ecchymosis and intramuscular hematomas. Screening FXIII assay was positive in all patients. Cryoprecipitate was effective in controlling most bleeds. Steroid, cyclophosphamide and rituximab were effective to eradicate inhibitor in some patients.

**TABLE 1** Patient characteristics

| No. | Age | Gender | Autoimmune diseases | Other diseases | Bleeding site | Time to diagnosis (month) |
|---|---|---|---|---|---|---|
| 1 | 41 | F | Yes, SLE | None | SC, IM | 25.8 |
| 2 | 85 | F | Yes, SLE | None | SC, IM | 23.9 |
| 3 | 72 | F | None | Renal stone | SC, IM, HU | 3.0 |
| 4 | 72 | M | None | CKD | SC, IM | 34.3 |
| 5 | 79 | F | None | DM | SC, IM, HA | 0.5 |

SLE: systemic lupus erythematosus CKD: chronic kidney disease DM: diabetes mellitus SC: subcutaneous/ecchymosis IM: intramuscular hematoma HA: hemarthrosis HU: hematuria

**TABLE 2** Diagnosis, treatment and outcome

| No. | Residual FXIII activity (%) | Peak FXIII inhibitor (BU) | Hemostatic treatment | Immune suppressants | Time to remission (month) | outcome |
|---|---|---|---|---|---|---|
| 1 | <5 | 1.89 | CRYO, TA | S, PS, CP, R, HQ | 1.0 | CR |
| 2 | <5 | 1.47 | CRYO, TA | S, CP, R, HQ | 1.4 | CR |
| 3 | <5 | 40.00 | CRYO | S | 12.8 | PR |
| 4 | 9.0 | 2.21 | CRYO, TA | S, CP, MF, R | - | NR |
| 5 | 6.4 | 2.37 | CRYO, TA | S, PS, R, CP | - | NR |

CRYO: cryoprecipitate TA: tranexamic acid, R: rituximab, S: steroid, PS: pulse steroid CP: Cyclophosphamide, HQ: hydroxychloroquine, MF: mycophenolate mofetil, CR: complete remission, PR: partial remission, NR: no remission

## PB1371 | Venous Thromboembolism Trends and Impact in Patients with Cirrhosis: Analysis of The National Inpatient Sample Database 2003-2014

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**Background**: Despite the decreased synthesis of coagulation factors and high bleeding risk in patients with cirrhosis, they are not free of venous thromboembolism (**VTE**) risk. The impact of VTE in patients with cirrhosis is not well understood, even less so the impact of acute VTE during hospitalization.

**Aims**: Establish the impact of inpatient VTE occurrence in mortality, bleeding, length and cost of hospitalization in patients with cirrhosis.

**Methods**: We identified individuals ≥ 18 years old from the 2003-2014 National Inpatient Sample database with diagnosis of cirrhosis using ICD-9-CM codes. Cirrhosis, acute deep vein thrombosis of extremities and acute pulmonary embolism diagnoses were allocated if present among the top 10 admission diagnoses. Demographics and inpatient outcomes of patients with VTE were compared to those without VTE. We used SPSS 25 to calculated odds ratios (**OR**) with confidence intervals (**CI**), and trend analysis.

**Results**: A total of 1,165,369 cirrhotic patients were included (weighted cases= 5,594,565). 11,049 (0.9%) VTE cases were diagnosed. Patients with VTE were older (61.9 vs 58.4 years, p< 0.0001), and less likely females (36.8% vs 39%, p< 0.0001). Patients with VTE were more likely to die during the hospitalization (**OR** 1.6 [CI 95% 1.5-1.71, p< 0.0001]), to have intracranial bleeding (**OR** 1.3 [CI 95% 1.03-1.64, p=0.02]), retroperitoneal bleeding (**OR** 2.36 [CI 95% 1.68-3.3, p< 0.0001]), had longer inpatient stay (9.6 vs 6.0 days, p< 0.0001), and higher costs (72,434 vs 41,670 USD, p< 0.0001). There was an increasing trend of inpatient VTE per year (0.27% 2003 vs 1.16% 2014, p< 0.0001) but a decreasing trend in inpatient mortality per year in cirrhotic patients with VTE (17.6% 2003 vs 9.2% 2014, p= 0.0001).

**Conclusions**: VTE occurrence is associated with increased inpatient mortality, intracranial bleed, retroperitoneal bleed, length and cost of hospitalization. More thorough VTE prophylaxis strategies may need to be implemented in patients with cirrhosis.

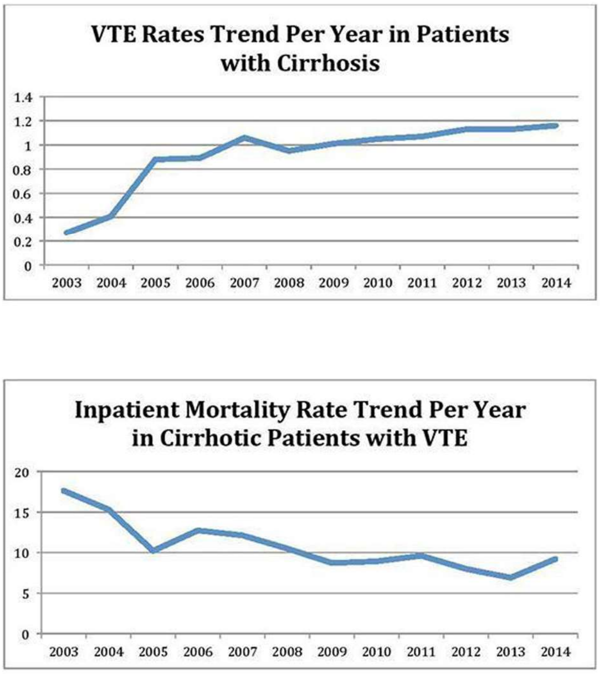


**FIGURE 1** VTE Rates Trend Per Year and Inpatient Mortality Rate Trend Per Year in Cirrhotic Patients with VTE

## PB1372 | Implication of Deranged Coagulation Parameters on Clinical Management of Patients with Dengue Fever

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**Background**: Dengue is the second most prevalent mosquito- borne infection after malaria, with over 40 million victims.Dengue fever is endemic in Pakistan with seasonal rise in cases. Morbidities and mortalities are proportionately reported to be increasing in number and associated with disseminated intravascular coagulation resulting in hemorrhagic or thrombotic manifestations in patients having deranged coagulation profiles.

**TABLE 1** VTE-Associated Comorbidities

| Comorbidity | VTE (%) | No VTE (%) | OR (95% CI) | P value |
|---|---|---|---|---|
| Heart Failure | 1,679 (15.2) | 118,099 (10.2) | 1.57 (1.49 - 1.65) | p<0.0001 |
| Metastatic Cancer | 680 (6.2) | 22,245 (1.9) | 3.33 (3.08 - 3.61) | p<0.0001 |
| Non-metastatic Cancer | 780 (7.0) | 47,583 (4.1) | 1.76 (1.64 - 1.90) | p<0.0001 |
| Liver Cancer (Primary) | 633 (5.7) | 47,600 (4.1) | 1.41 (1.30 - 1.53) | p<0.0001 |
| Valvular Heart Disease | 578 (5.2) | 45,952 (4.0) | 1.33 (1.22 - 1.44) | p<0.0001 |
| COPD | 1,171 (10.6) | 109,220 (9.5) | 1.13 (1.06 - 1.20) | p<0.0001 |