



GYNAECOLOGICAL AND OBSTETRICAL BLEEDING IN WOMEN WITH FACTOR XI DEFICIENCY- a systematic review



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BACKGROUND

Individuals with factor XI (FXI) deficiency are prone to bleed excessively during and/or after surgery and particularly in areas of high fibrinolysis such as the mouth and genitourinary system. Pregnancy, menstrual bleeding and delivery present an intrinsic haemostatic challenge to women with bleeding disorders such as FXI deficiency.

OBJECTIVE

To provide a systemic overview of studies in which gynaecological and obstetrical bleeding problems in women with FXI deficiency are discussed.

MATERIAL & METHODS

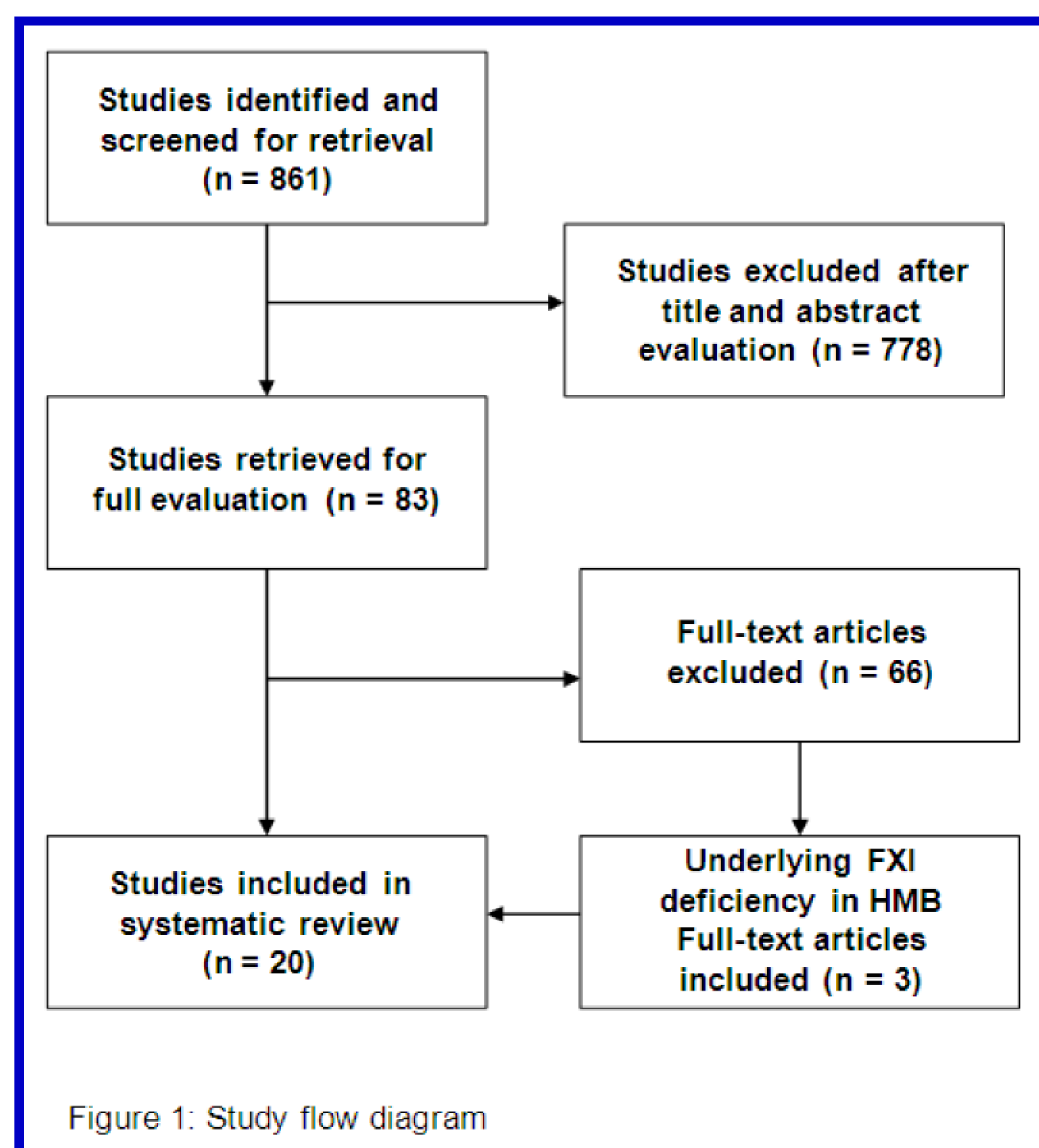
We searched MEDLINE, EMBASE and the Cochrane library to identify studies that focus on the incidence, the levels and treatment options in FXI deficient women with gynaecological and obstetrical bleeding. Papers were eligible if they presented original data in adults. We excluded case reports and case series with <5 cases of FXI deficient patients. The articles were read independently by two reviewers.

REFERENCES

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- Kadir RA, et al. Pregnancy in women with von Willebrand's disease or factor XI deficiency. BJOG 1998.
- Salomon O, Steinberg DM, Tamarin I, et al. Plasma replacement therapy during labor is not mandatory for women with severe FXI deficiency. Blood Coagul Fibrinolysis 2005.
- Santoro R, et al. FXI deficiency: a description of 34 cases and literature review. Blood Coagul Fibrinolysis 2011.
- Singh A, et al. FXI deficiency and obstetrical anesthesia. Anesth Analg 2009

RESULTS

We identified 20 studies (figure 1), with a total of 300 women with FXI deficiency (factor XI < 70 U/dL). More than half of the patients had a mild deficiency (FXI \geq 20 U/dL). In most studies bleeding phenotype was not described, heavy menstrual bleeding (HMB) was reported in 29-67% (range within studies) of the women with FXI deficiency. In women who underwent gynaecological procedures, 7 out of 18 had a bleeding complication. Seven hysterectomies were performed, two women had bleeding complications.



CONCLUSION

Women with factor XI deficiency have a clearly increased risk of HMB, and bleeding complications after miscarriage, TOP and delivery. Despite high bleeding risks, in the majority of reported surgeries and deliveries, no prophylactic treatment was given.

RESULTS (cont.)

During pregnancy FXI levels do not increase. 2.8-20% of reported pregnancies ended in a miscarriage; of these miscarriages 0-25% (4 of 24) were complicated by a bleeding. Four out of 11 of the terminations of pregnancies (TOP) were complicated by bleeding. In 83 out of 424 deliveries, post partum haemorrhage (PPH) was reported, ranging from 0-40%. In only 21% of the deliveries the patient received prophylaxis, 11% of those had PPH (table 1). In the 44 times epidural analgesia was performed, with or without prophylaxis, no complications were reported.

Table 1: Prophylactic treatment at deliveries in patients with factor XI deficiency

Authors	Prophylaxis ante-partum	Sort of prophylaxis	# of patients with PPH	Treatment for PPH
Chi ¹	19/47 deliveries	12 TA 7 FXIc	1 TA had primary PPH 2 FXIc had secondary PPH	TA FXIc then TA
Kadir ²	5/25 deliveries	3 FXIc 2 FFP	None	
Salomon ³	20/164 deliveries	20 FFP	2 FFP	?
Santoro ⁴	4/24 deliveries	4 FFP	None	
Singh ⁵	8/13 deliveries	8 FFP	1 FFP	RBC and FFP
Total	56/273 deliveries	12 TA 10 FXIc 34 FFP	6/56 (1/12 TA; 2/10 FXIc; 3/34 FFP)	

#: number; PPH: post-partum haemorrhage; TA: tranexamic acid; FXIc: Factor XI concentrate; FFP: Fresh Frozen Plasma; RBC: Red Blood Cells.

We have no conflicts of interest to disclose.

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