

**Supplementary table 1.** Antiphospholipid antibodies of aPL patients according to the FXI:C levels.

	<b>FXI:C &lt;70% (N= 13)</b>	<b>FXI:C &gt;150% (N= 18)</b>	<b>p</b>
<b>LA N (%)</b>	10 (76.9)	13 (72.2)	1.00
<b>LA only N(%)</b>	6 (46.2)	6 (33.3)	0.470
<b>aCL</b>			
IgM N (%)	2 (15.4)	3 (16.7)	0.924
IgG N (%)	3 (23.1)	3 (16.7)	0.656
IgM and/or IgG N(%)	4 (30.8)	7 (38.9)	0.641
IgM and IgG N(%)	1 (7.7)	0 (0)	0.232
<b>aCL only N(%)</b>	0 (0)	1 (5.6)	0.388
<b>aβ2GP1</b>			
IgM N(%)	3 (25)	1 (5.6)	0.32
IgG N(%)	4 (30.8)	6 (33.3)	1.00
IgM and/or IgG N (%)	4 (30.8)	6 (33.3)	1.00
IgM and IgG N(%)	2 (16.7)	1 (5.6)	1.00
<b>aβ2GP1 only N(%)</b>	0 (0)	2 (11.1)	0.214
<b>N° aPL</b>			
<b>1 N (%)</b>	6 (46.2)	8 (44.4)	1.00
<b>2 N (%)</b>	0 (0)	3 (16.7)	0.35
<b>3 N (%)</b>	4 (30.8)	4 (22.2)	0.90
<b>Thrombotic aPL profile</b>			
<b>Low N (%)</b>	3 (23.1)	5 (27.8)	1.00
<b>High N (%)</b>	10 (76.9)	13 (72.2)	1.00

Abbreviations: antiphospholipid autoantibodies (aPL); anti β2 glycoprotein B1 (aβ2GP1); anti cardiolipin (aCL); Lupus anticoagulant (LA).

**Supplementary table 2.** Correlation analysis between coagulant activity of FXI and anticardiolipin and anti $\beta$ 2GP1 antibody titre.

	FXI:C (SS)		FXI:C (SFX)	
	Correlation coefficient	p	Correlation coefficient	p
a $\beta$ 2GP1 IgM	-0.23	<b>0.002</b>	-0.21	<b>0.005</b>
a $\beta$ 2GP1 IgG	-0.22	<b>0.002</b>	-0.16	<b>0.031</b>
aCL IgM	-0.15	<b>0.034</b>	-0.14	0.057
aCL IgG	-0.197	<b>0.006</b>	-0.13	0.081

Abbreviations: anti  $\beta$ 2 glycoprotein B1 (a $\beta$ 2GP1); anti cardiolipin (aCL).

**Supplementary table 3.** Genetic defects, FXI:C and type of FXI deficiency detected in 4 cases with congenital FXI deficiency identified in this study.

Patient	Group	cDNA variant	Protein variant	FXI:C	Type of deficiency	Reference HGMD
P1	APS	c.403G>T	p.Glu135Ter	50%	CRM-	CM890042
P2	AaPL	c.403G>T	p.Glu135Ter	49%	CRM-	CM890042
P3	AaPL	c.802C>T	p.Arg268Cys	44%	CRM-	CM035499
P4	HC	c.1327C>T	p.Arg443Cys	23%	CRM-	CM062624

Abbreviations: Asymptomatic carriers of antiphospholipid antibodies (AaPL); Healthy controls (HC); Primary antiphospholipid syndrome (APS).