Table 1. Clinical studies and case series of a- hypo- and dysfibrinogenemia patients where thrombosis was reported

Authors and year of publication	Country	Study population	Methods	Thrombotic events
Lak et al. 1999 [87]	Iran	55 patients (28 F 27 M) with afibrinogenemia. Age range 2–73 years	Retrospective study of bleeding symptoms in afibrinogenemia vs. hemophilia patients	2 patients (4%) with afibrinogenemia developed thrombotic symptoms
Kreuz et al. 2005 [88]	Germany	12 patients (8 F, 4 M) with a-hypo- or dysfibrinogenemia. Median age: 11.5, range 1 day to 29 years	Retrospective study investigating the effects of fibrinogen therapy for bleeding episodes	One patient with afibrinogenemia developed venous thrombosis and non-fatal lung embolism during the treatment. May have been related to fibrinogen
Santacrose et al. 2006 [89]	Italy	18 patients (10 F, 8 M) with a-hypo- or dysfibrinogenemia. Age range: 1–53 years	Retrospective study of patients treated in the clinic over a 10 year time period	An 11 year old boy with cerebral venous thrombosis of the left sagittal sinus was the only case presenting with thrombosis
Casini et al. 2015 [7]	Switzerland	101 patients (68 F, 33 M) with dysfibrinogenemia. Mean age at clinical diagnosis: 29.2 years, standard deviation: 16.8 years	Multicenter study of 101 patients with congenital dysfibrinogenemia to characterize the incidence of hemorrhagic and thrombotic events as well as complications of pregnancy and surgery	There were 28 first thrombotic events, including 20 venous events (11 deep venous thromboses, 3 PEs, 2 superficial vein thrombophlebitis events, and 4 thromboses at unusual sites) and 8 arterial events (4 strokes, 2 acute myocardial infarctions, 1 peripheral artery occlusion, and 1 mesenteric artery thrombosis. Fourteen of these events occurred at time of diagnosis, and 14 during follow-up
Miesbach et al. 2010 [90]	Germany	37 patients with hereditary dysfibrinogenemia. 63% F, 37% M. Age: median 58 years, range: 11–86 years	Laboratory and clinical evidence of hemostatic abnormalities were assessed	19% of probands (9/37, all above age of 50 years), had experienced at least one episode of arterial or venous thrombosis. Among these, were two (7%) with deep venous thrombosis, seven with arterial thrombosis, and five (14%) had experienced both

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Table 1. Continued

Authors and year of publication	Country	Study population	Methods	Thrombotic events
Nguyen et al. 1998 [91]	France	12 patients (4 F, 8 M) with dysfibrinogenemia, 4 with thrombosis, 8 without. Age range: 66–24 years	Retrospective analysis of patients with dysfibrinogenemia presenting with or without thrombotic episodes	4 patients with dysfibrinogenemia had various types of thrombotic episodes
Ramanathan et al. 2013 [92]	Denmark	Family (N = 14) with dysfibrinogenemia (FGA mutation)	Case description of a family affected by dysfibrinogenemia and thrombosis	Deep venous/arterial thrombosis or pulmonary embolism reported in 4 family members with dysfibrinogenemia
Shapiro et al. 2013 [93]	UK	35 patients with heritable dysfibrinogenemia. 20 F, 15 M. Age: median 38 years, range: 11–78 years	Historical symptoms of thrombosis were determined by patient interviews and inspection of hospital records	Thrombosis at any site: 3 (9%) Pulmonary embolus: 2 (6%) Thrombotic stroke: 1 (3%)
Zhou et al. 2015 [11]	China	102 patients with congenital dysfibrinogenemia. 49 F, 53 M. Age: median 35 years, range: 3–76 years	Patients' previous clinical manifestations were recorded and quantified using consensus ISTH bleeding assessment tool	Thrombosis at any site: 4 (3.9%) Lower extremity venous thrombosis: 2 (2.0%) Pulmonary embolism: 1 (1.0%) Portal vein thrombosis: 1 (1.0%) Dorsal vein of foot thrombosis: 1 (1.0%)