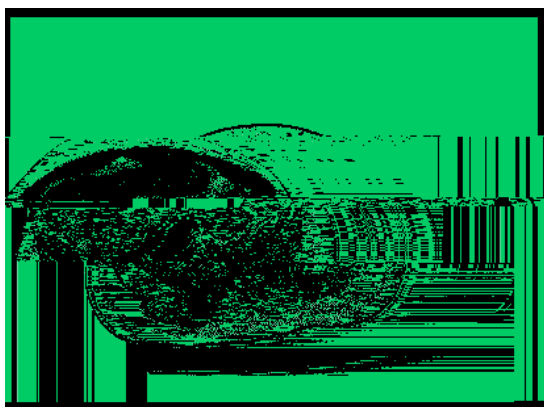




due to the introduction of nonsteroidal anti-inflammatory drugs in the treatment of chronic hemophilic arthropathy [4]. Bleeding on the intestinal wall is rare [5]. According to the literature, and extending the review carried out by Katsumi et al. in 2006 [6], there are 33 described cases of intramural hematoma of the gastrointestinal tract from 1964



**Figure 1:** Edema and dilatation of small intestine loops with gas levels.



**Figure 2:** Parietal thickening of a wide segment of the jejunum with homogeneous enhancement after the administration of intravenous contrast. It loses its normal layered structure and shows hypodensity of the submucosa, associated with dilatation of the proximal segments, compatible with submucosal hemorrhage.

The patient was hospitalized with digestive rest and nasogastric decompression. Also, plasma factor VIII with high concentration of von Willebrand factor was administered intravenously every 12 hours for seven days, with monitoring of FVIII:C > 80% and a gradual descent of the need to administer FVIII. In addition, no anemia was observed and the patient did not require support with a transfusion or iron. The pain disappeared progressively and previously the patient started oral diet with good tolerance. A control CT scan performed 7 days after admission shows that the hematoma had resolved completely and the patient was discharged with no recurrences so far.

## Discussion

It is known that the most common cause of intramural spontaneous hematoma in the small intestine is the intake of oral anticoagulants [1]. Gastrointestinal bleeding in hemophilic patients may appear in between 10% and 25% of the cases, and up to 85% of them are due to peptic ulcers [3]. These cases have been growing over the last years

APCC, bMBA Age	Sex	Diagnosis (activity of Factor VIII)	FVIII level (%)	Inhibitor (BU/mL)	titer	Location in the gastrointestinal tract	Treatment	Reference
1	20	Hemophilia A	-	-	-	Jejunum	FVIII	This report
2	7	Severe hemophilia A	>10	-	-	sigma	APCC, rVIIa	[8]
3	37	Hemophilia A	-	-	-	Distal ileum	FVIII	[7]
4	17	Severe hemophilia A (<1%)	A	1048	-	Jejunum	APCC, rVIIa	[6]
5	34	Severe hemophilia A	-	High	-	Jejunum	rVIIa	[5]
6	74	Mild hemophilia A	-	27	-	Small intestine	APCC, rVIIa	[10]
7	78	Acquired FVIII inhibitor	-	>15	-	Esophagus	Intravenous immunoglobulin, prednisolone, and cyclophosphamide	human oral [22]
8	n.s	Hemophilia A	-	-	-	Esophagus	FVIII	[23]
9	14	Severe hemophilia A (<1%)	A	-	-	Jejunum	FVIII	[11]
10	47	Severe hemophilia A	-	0.9	-	Small intestine	FVIII	[12]
11	31	Hemophilia A (4.5%)	-	-	-	Esophagus	Cryoprecipitate and antituberculous treatment	[24]
12	Child	Hemophilia	-	-	-	Duodenum	Deferred surgery	[26]
13	Child	Hemophilia	-	-	-	Duodenum	Deferred surgery	[26]
14	Child	Hemophilia	-	-	-	Duodenum	Conservative therapy	[26]
15	Child	Hemophilia	-	-	-	Duodenum	Conservative therapy	[26]
16	31	Hemophilia A	-	-	-	Small intestine	Laparotomy	[13]
17	40	Hemophilia A	-	-	-	Jejunum	FVIII	[13]
18	8	Hemophilia A	-	-	-	Proximal jejunum, jejuno-jejunal intussusception	FVIII, laparotomy	[28]
19	9	Severe hemophilia A	-	4~8	-	Stomach	FVIII	[17]
20	34	Hemophilia A	-	Positive	-	Small intestine	FVIII	[14]
21	19	Hemophilia A	-	-	-	Small intestine	FVIII	[14]
22	47	Hemophilia A	-	-	-	Jejunum	FVIII	[14]
23	16	Hemophilia B	-	-	-	Ileocolic intussusception	FIX	[14]
24	23	Severe hemophilia A (<1%)	A	-	-	Stomach	Cryoprecipitate	[19]
25	13	Severe hemophilia A (<1%)	A	-	-	Ileocolic intussusception	Cryoprecipitate, enema reduction	barium [27]
26	38	Hemophilia A	-	-	-	Esophagus	FVIII	[23]
27	22	Hemophilia A	-	-	-	Stomach	Cryoprecipitate	[18]
28	16	Hemophilia A	-	-	-	Small intestine	Fresh blood	[15]
29	8	Hemophilia A	-	-	-	Stomach	Anti-hemophilic globulin	[20]
30	15	Hemophilia A	-	-	-	Ileum	Anti-hemophilic globulin	[16]



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35. Collins PW, Palmer BP, Chalmers EA, Hart DP, Liesner R5, et al. (2014) Factor VIII brand and the incidence of factor VIII inhibitors in previously untreated UK children with severe hemophilia A, 2000-2011. *Blood* 124: 3389-3397.
  36. Kouides PA, Fogarty PF (2010) How do we treat: upper gastrointestinal bleeding in adults with haemophilia. *Haemophilia* 16: 360-362.
  37. Srivastava A, Brewer AK, Mauser-Bunschoten EP, Key NS, Kitchen S, et al. (2013) Guidelines for the management of hemophilia. *Haemophilia* 19: e1-47.
  38. Verbruggen B, Novakova I, Wessels H, Boezeman J, van den Berg M, et al. (1995) Comparison of the Bethesda assay for factor VIII:C inhibitors: improved accuracy and reliability. *Thromb Haemostasis* 73: 247-251.
  39. Hay CR, DiMichele DM; International Immune Tolerance Study (2012) Principal results of the International Immune Tolerance Study: a randomized dose comparison. *Blood* 119: 1335-1344.
  40. Coppola A, Di Minno MN, Santagostino E (2010) Optimizing management of immune tolerance induction in patients with severe haemophilia A and inhibitors: towards evidence-based approaches. *Br J Haematol* 150: 515-528.