

# Elective major surgery in haemophiliacs: Istanbul experience

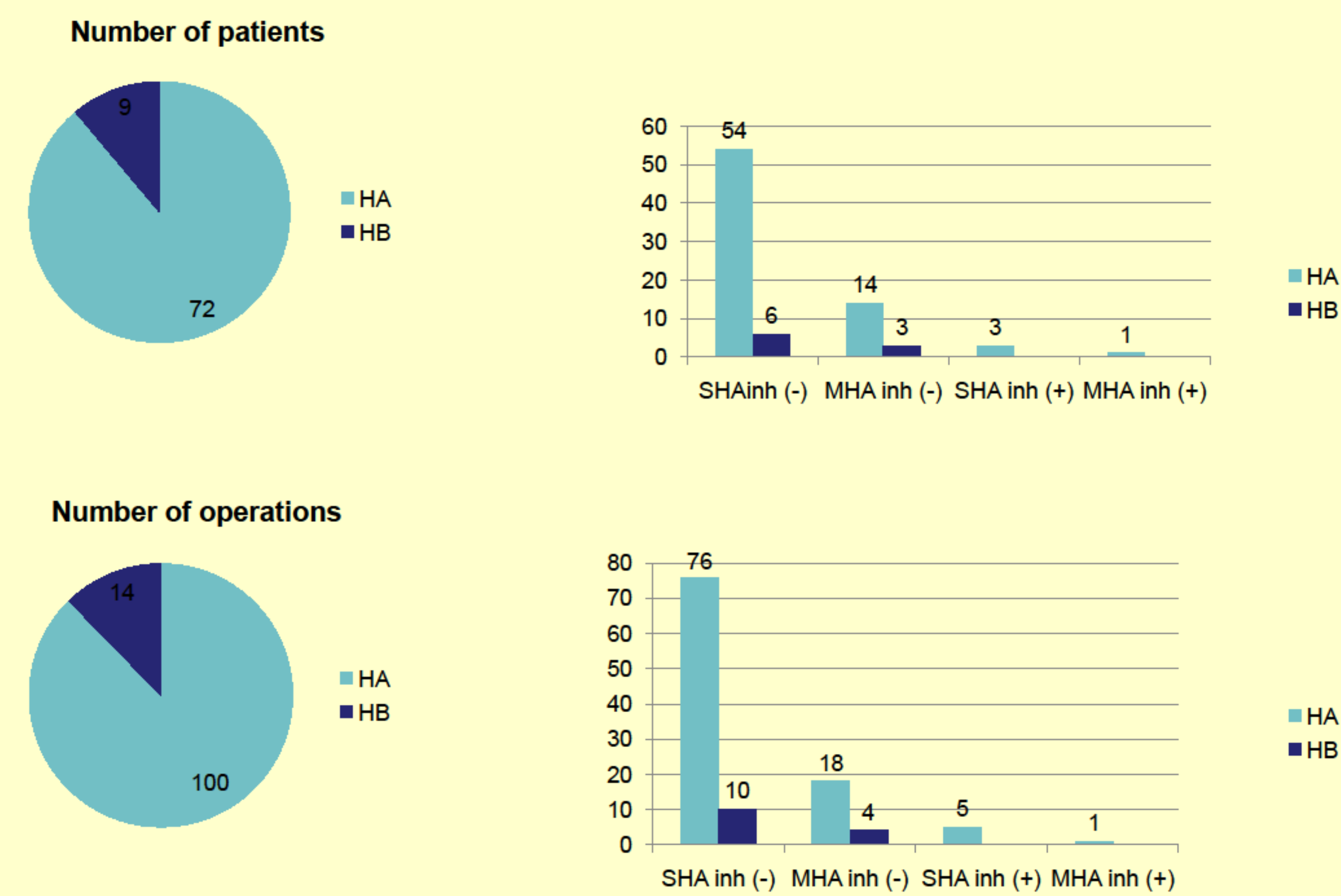
Gulen Tuysuz<sup>1</sup>, Bulent Zulfikar<sup>2</sup>, Onder Kilicoglu<sup>3</sup>, Fatih Dikici<sup>3</sup>, Basak Koc<sup>4</sup>, Fikret Bezgal<sup>5</sup>, Necdet Aras<sup>6</sup>, Recep Güloğlu<sup>7</sup>, Omer Taser<sup>3</sup>

- 1.) Istanbul University, Cerrahpaşa Medical Faculty, Pediatric Hematology Dept, Istanbul, Turkey
- 2.) Istanbul University, Cerrahpaşa Medical Faculty and Oncology Institute, Istanbul and , The Hemophilia Society of Turkey, Turkey
- 3.) Istanbul University, Istanbul Medical Faculty, Department of Orthopedics and Traumatology, Istanbul, Turkey
- 4.) Istanbul University, Cerrahpaşa Medical Faculty and Oncology Institute, Istanbul, Turkey
- 5.) The Hemophilia Society of Turkey
- 6.) Istanbul University, Department of Urology, Istanbul, Turkey
- 7.) Istanbul University, Department of Emergency, Istanbul Turkey

## OBJECTIVES

To review the perioperative management and outcome of elective major surgical procedures in severe and moderate type haemophiliacs.

## MATERIAL METHOD-I



Type of Operations	Number of operations	Haemophilia A	Haemophilia B
Orthopedic	98 (85.97)	85	13
General Surgery	8 (7.02)	8	0
Nose throat ear	3 (2.63)	2	1
Urologic	3 (2.63)	3	0
Neurosurgical	2 (1.75)	2	0
<b>TOTAL</b>	<b>114 (% 100)</b>	<b>100</b>	<b>14</b>

ORTHOPEDIC SURGERIES	1 surgical area	2 surgical area	3 surgical area
Arthroscopic synovectomy	26	2	0
Arthroscopic synovectomy+ others*	0	1	2
Total endoprosthesis	17	7	1
Total endoprosthesis+ others**	0	2	2
Bone resection†	14	0	0
Bone fixation‡	10	0	0
Muscle release*	5	0	0
Removal of hemophilic pseudotumor	2	0	0
Removal of hemophilic pseudotumor+ others§	0	1	1
Other orthopedic procedures ¶	5	0	0
<b>TOTAL</b>	<b>79</b>	<b>13</b>	<b>6</b>

Arthroscopic synovectomy+ others\*= Arthroscopic synovectomy+ radial head resection (1), bilateral arthroscopic synovectomy+ myotomy (1), arthroscopic synovectomy+ arthrodesis+ joint revision (1)  
 Total endoprosthesis+ others\*\*= Total endoprosthesis+ bilateral achilloplasty (2), total endoprosthesis+ joint revision (1), total endoprosthesis+ unilateral total endoprosthesis (1)  
 Muscle release= Achilles lengthening (4), myotomy (1)  
 Bone resection†= Osteotomy (7), radial head resection (3), hemipelvectomy (2), amputation of an extremity (1), open bone biopsy (3)  
 Bone fixation‡= Internal fixation of bone fracture (5), fixation of fractures with circular external fixator (5)  
 Removal of hemophilic pseudotumor + others§= Removal of hemophilic pseudotumor+ osteotomy (1), removal of hemophilic pseudotumor+ Achilles lengthening + myotomy (1)  
 Other orthopedic procedures ¶= Arthrodesis (2), revision of contracture (1), open joint debridement (1), bone cementing (1)

## CONCLUSION

We achieved good haemostasis in our major surgeries by timely replacement therapy and close follow-up of patients after surgery. We conclude that the presence of inhibitors and operation at more than one surgical site for orthopedic surgeries are risk factors for the development of complications. For these patients, close follow-up, more intensive factor replacement therapy, gentle surgical techniques and monitorization of factor levels if available should be considered to lead to better results.

## METHODS

Data pertaining to major surgeries from 1996 to 2013 at our center were retrospectively analysed. All operations were elective and a plan for management of hemostasis was prepared for each patient.

## MATERIAL METHOD- II

### Haemostasis Plan for Inhibitor negative patients

	Severe Haemophilia A	Moderate Haemophilia A	Severe Haemophilia B	Moderate Haemophilia B
12 hours prior to surgery-post-op 10th day	Tranexamic acid 25-40 mg/kg/day: 3-4 dose			
2 hours prior to surgery	40 IU/kg/dose		80 IU/kg/dose	
3-72th hours	60 IU/kg/day (q 3-8 hr)		100 IU/kg/day (q 6-8 hr)	
4-7th days	40 IU/kg/day (q 4-8 hr)	25 IU/kg/day (q 6-8 hr)	70 IU/kg/day (q 8-12 hr)	40 IU/kg/day (q 12 hr)
8-14/21th days	25 IU/kg/day (q 8-12 hr)	10 IU/kg/day (q 12 hr)	40 IU/kg/gün (q 12-24 hr)	20 IU/kg/day (q 24 hr)

### Haemostasis Plan for Inhibitor negative patients

	Prior to surgery	0-24th hours	24-72th hours	4-14th days	14-21th days
rFVIIa	90-120 µg/kg/dose	90-120 µg/kg/dose (12 times/day)	90-120 µg/kg/dose (8 times/ day)	90 µg/kg/dose (6 times/ day)	90 µg/kg/dose (4 times/day)
	Tranexamic acid 25-40 mg/kg/day: 3-4 dose 12 hours prior to surgery-post-op7 days				
aPCC	100 IU/kg/dose	150 IU/kg/day (q 8 hr)		100 IU/kg/day (q 12 hr)	

## RESULTS

- Complications occurred in 12 of 114 procedures (10.5 %).
- All complications occurred in haemophilia A patients;
  - 11 after orthopedic surgeries ;
  - 4 inhibitor positive operations with multiple surgical sites,
  - 4 inhibitor negative operations with multiple surgical sites,
  - 3 inhibitor negative operations with one single site)
  - 1 complication occurred after an open nephrolithotomy operation.

Types of complications;

- ❖ Bleeding (n:6)
- ❖ inhibitor development (n:3)
- ❖ periprosthetic infection (n:2)
- ❖ traumatic fracture (n:1)
- No death or life threatening complication occurred at any of the operations
- Presence of inhibitors and operating multiple orthopedic sites in one procedure were significant predictors for the development of complications

### Statistical analysis of complications

Predisposing factors	No complication (n)	Complication occurred (n)	P
Type of hemophilia; HA/HB	88/14	12/-	0.19
Severity of disease; Severe/moderate	81/21	10/2	0.549
Patients age; <11/11-18/>18	15/17/70	1/3/8	0.69
Infusion type; bolus/continuous	93/9	11/1	0.955
Date of operation; 1996-2001/2002-2007/2007-2013	31/40/31	2/6/4	0.595
Type of surgery; orthopedics/others	87/15	11/1	0.472
Number of surgical site (orthopedic operations); 1/2/3	75/8/4	4/5/2	0.00
Existence of a minor surgical operation (orthopedic surgeries); no/existing	82/5	10/1	0.52
Presence of inhibitors in Hemophilia A patients ; no/existing	86/2	8/4	0.002

