

Ceredih Activity report

2025-01-08

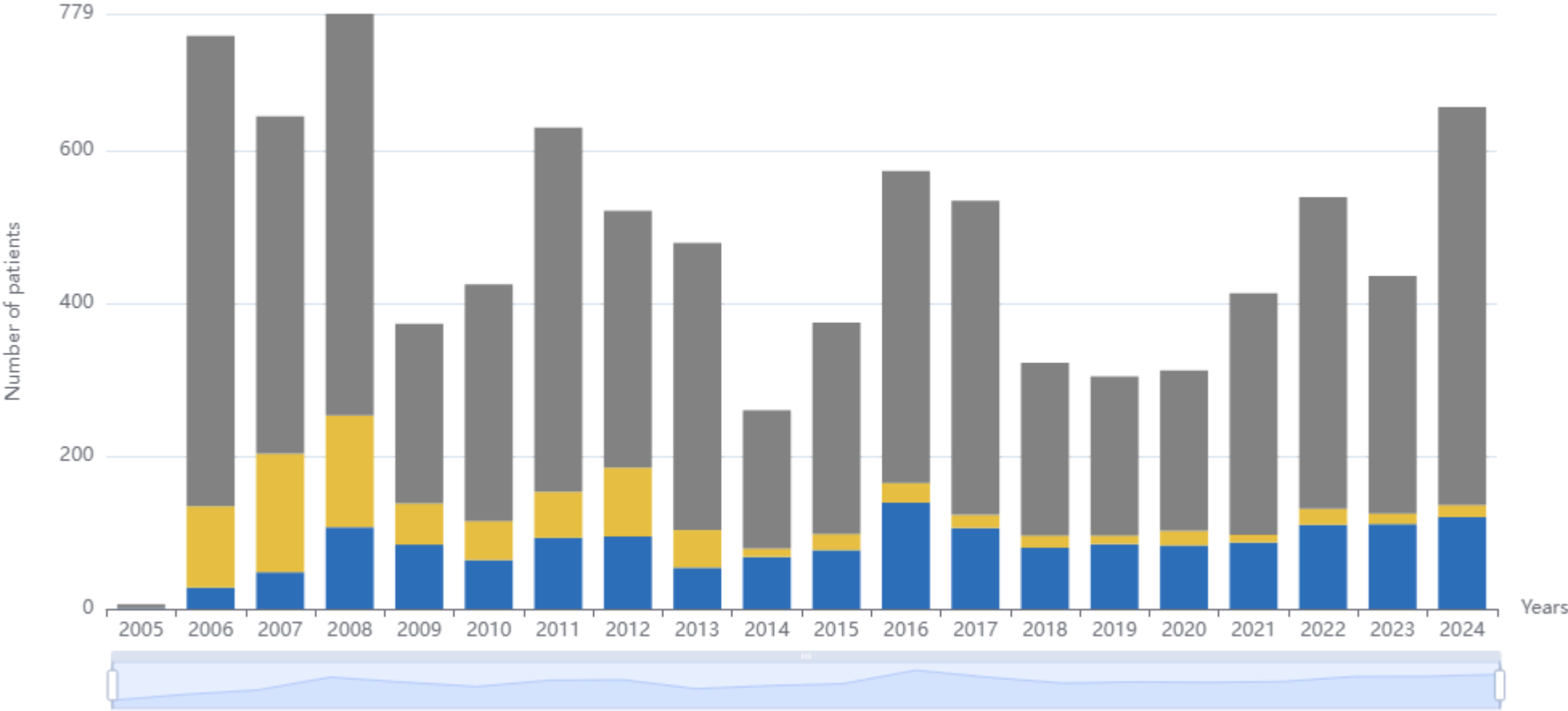
Registry description as of 2024, Nov. 11th

Inclusion

Number of inclusion in the CEREDIH registry

(incident cases were defined as included the year after diagnosis)

Incident cases Included deceased Prevalent cases

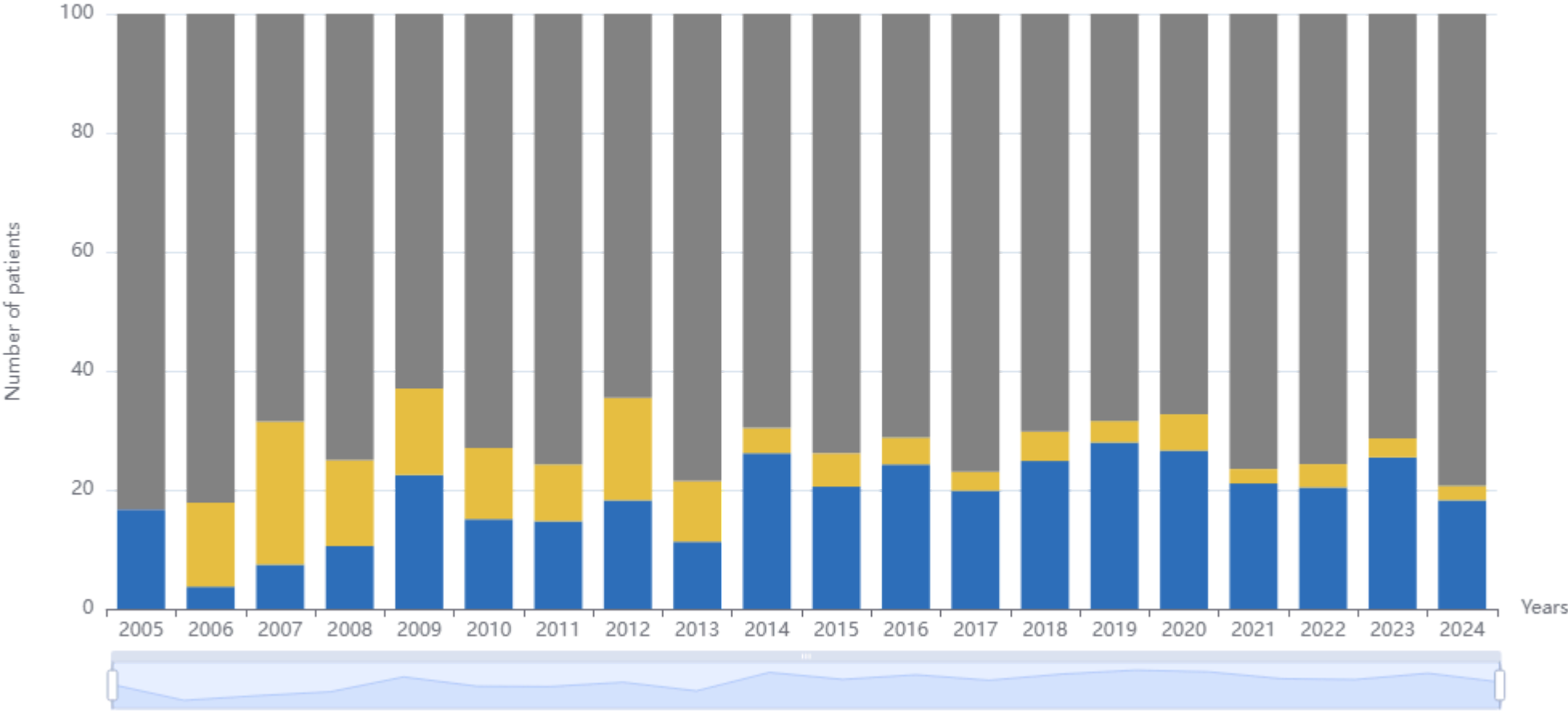


Inclusion

Percentage of inclusion in the CEREDIH registry

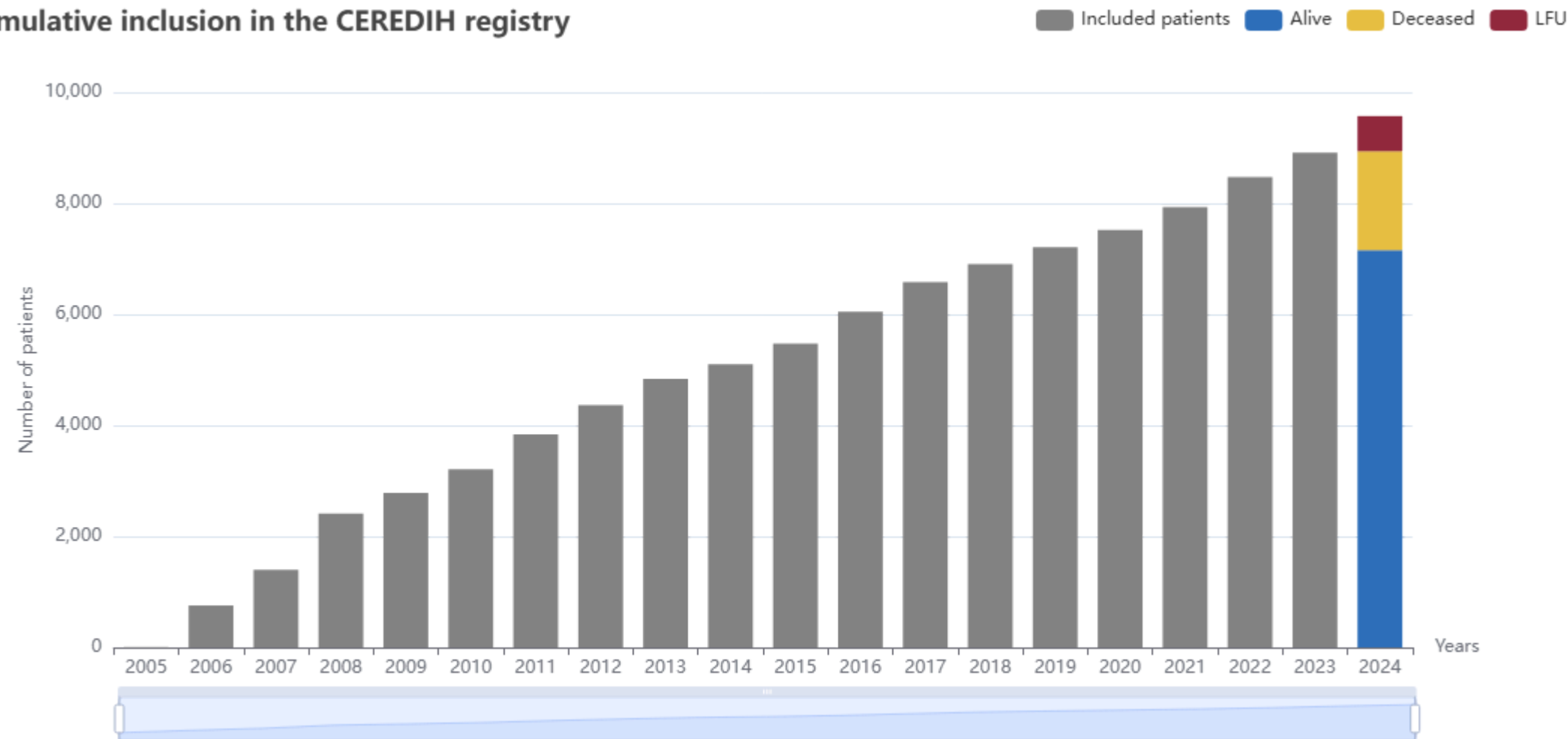
(incident cases were defined as included the year after diagnosis)

Incident cases Included deceased Prevalent cases



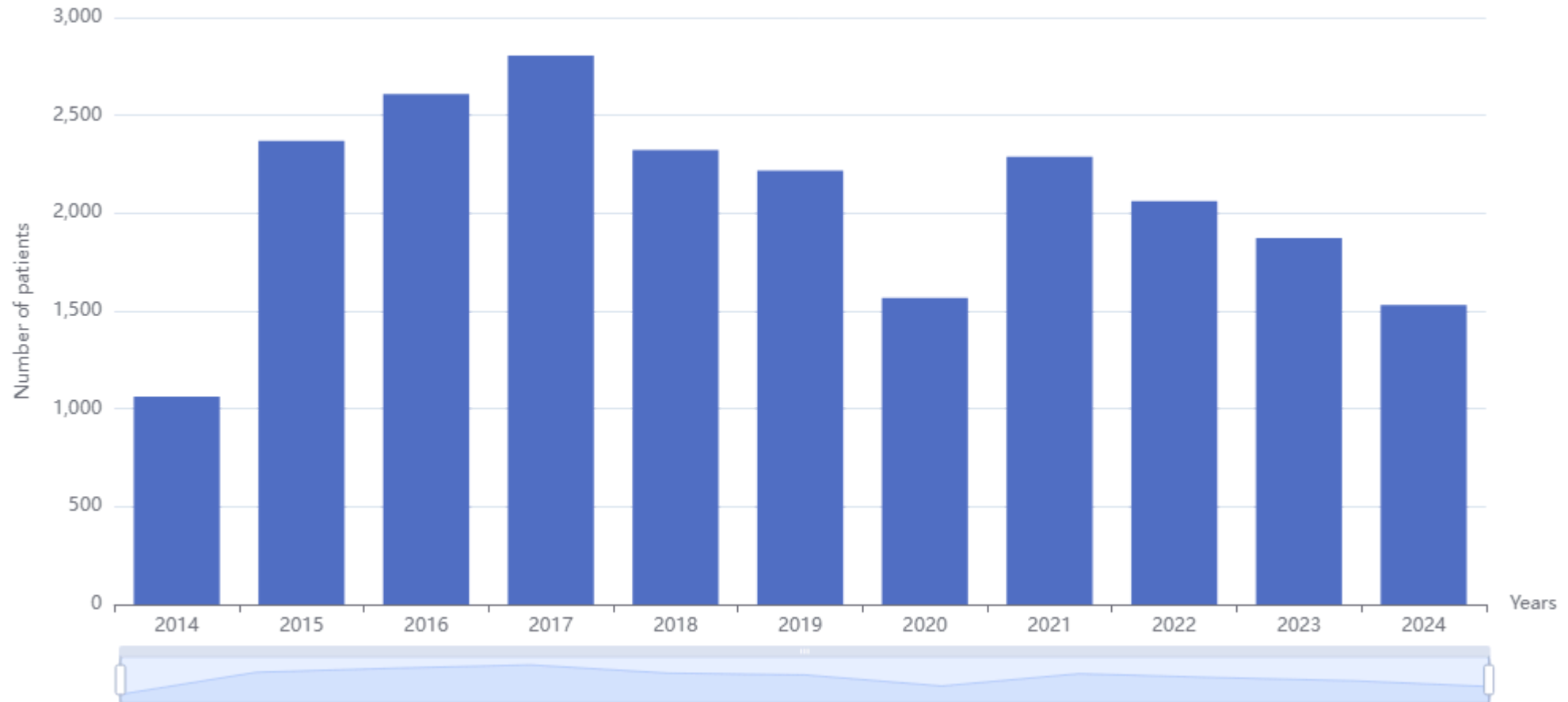
Inclusion (n = 9571)

Cumulative inclusion in the CEREDIH registry



Follow-up (total = 22707)

Evolution of number of Fup since May. 2014



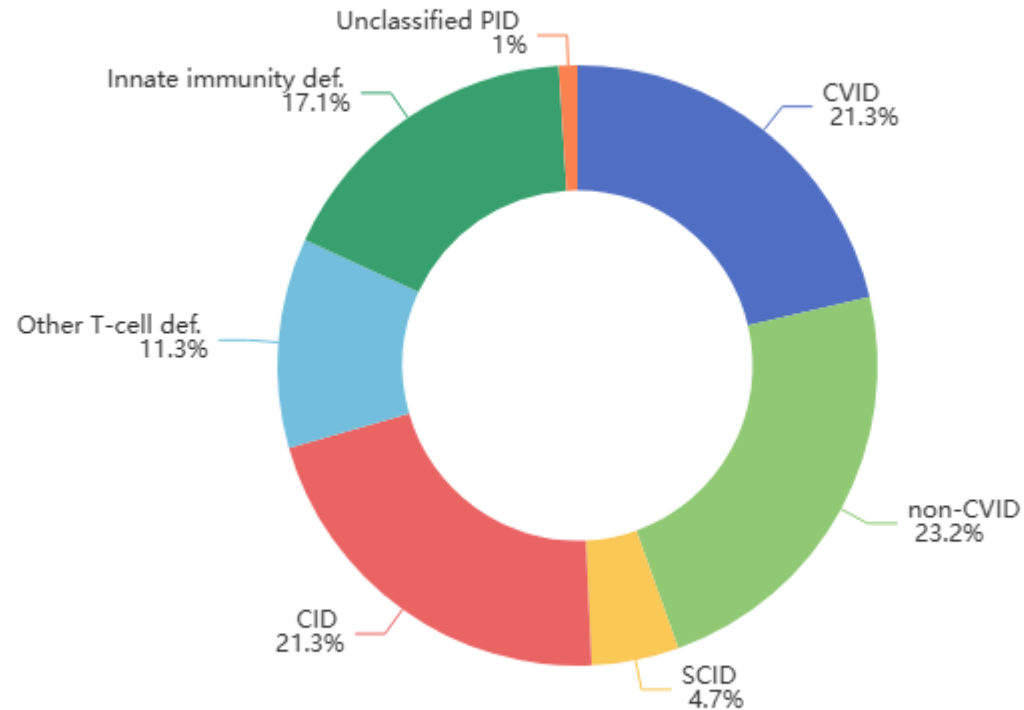
Registry (n = 9571)

Covariates	Pop.	N	Percentage
Males	9571	5213	54.5
Adults	9546	5766	60.4
Consanguinity	9033	1239	13.7
Familial case	9210	2605	28.3
Born in France	8191	7356	89.8
Living in France	9551	9367	98.1
Alive	9571	7157	74.8
Deceased	9571	1781	18.6
Lost	9571	633	6.6

Main PID category

Repartition of main PID category in the CEREDIH registry

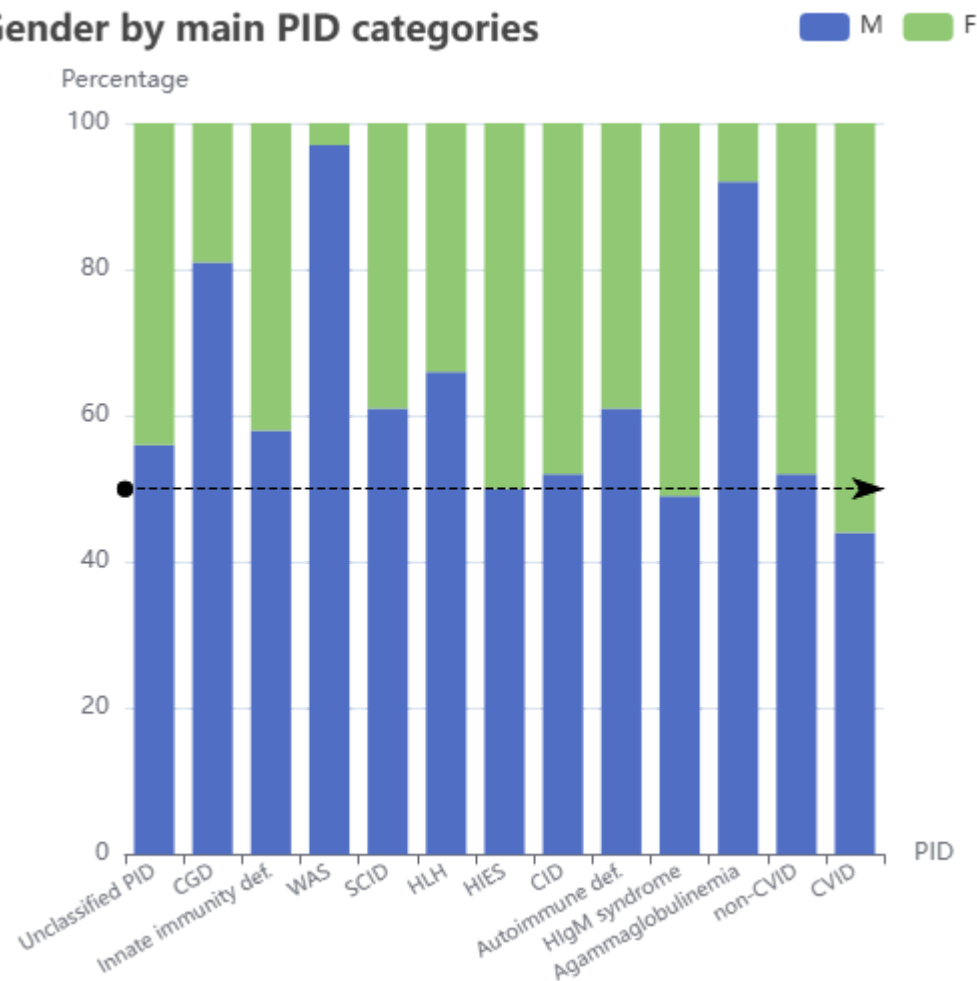
■ CVID ■ non-CVID ■ SCID ■ CID ■ Other T-cell def. ■ Innate immunity def. ■ Unclassified PID



Main categories of PID

Categories	N	Percentage
B-cell deficiency	4262	45
CVID	2038	21
Non-CVID	2224	23
Agammaglobulinemia	375	4
HIgM syndrome	41	0.43
T-cell deficiency	3575	37
Autoimmune	471	5
CID	1725	18
HIES	157	2
HLH	454	5
SCID	454	5
WAS	314	3
Innate immunity deficiencies	1638	17
CGD	330	3
Unclassified	96	1

Gender by main PID categories



Evolution of main PID categories

	2009	2024
Categories	N (%)	N (%)
B-cell deficiency	950 (35)	4262 (45)
CVID	469 (17)	2038 (21)
Non-CVID	481 (18)	2224 (23)
Agammaglobulinemia	375 (14)	375 (4)
HIgM syndrome	41 (2)	41 (0.43)
T-cell deficiency	755 (28)	3575 (37)
Autoimmune	68 (3)	471 (5)
CID	602 (22)	1725 (18)
HIES	40 (1)	157 (2)
HLH	152 (6)	454 (5)
SCID	231 (9)	454 (5)
WAS	136 (5)	314 (3)
Innate immunity deficiencies	522 (19)	1638 (17)
CGD	156 (6)	330 (3)
Unclassified	16 (1)	96 (1)

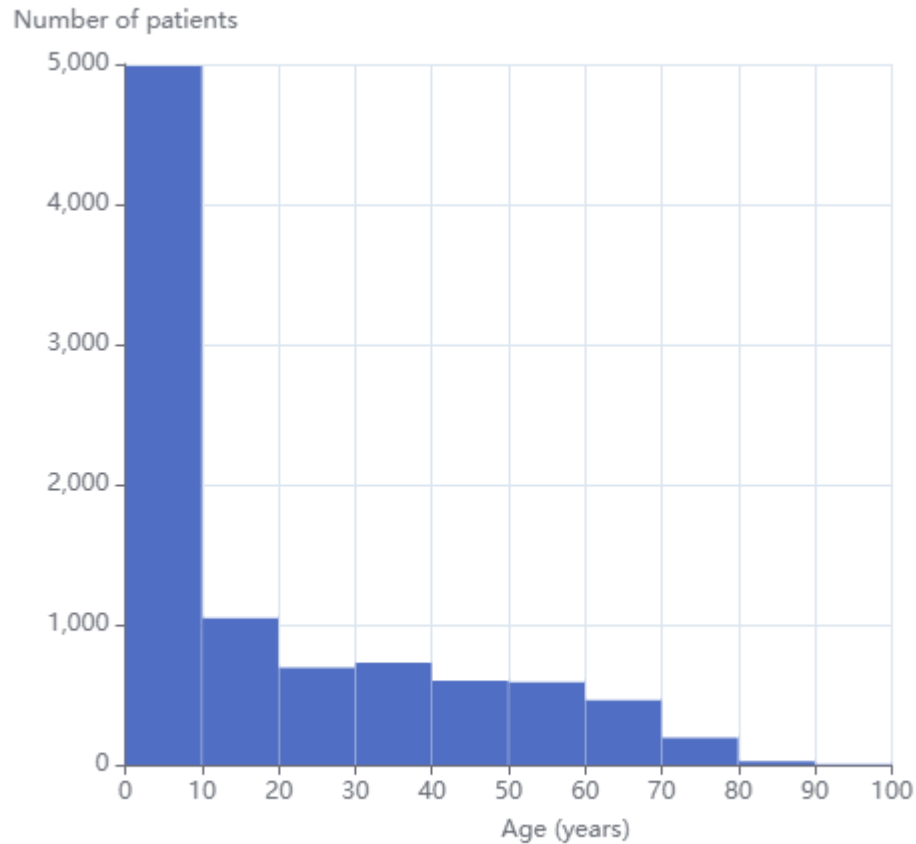
Timeline

Clinical diagnosis (CD)

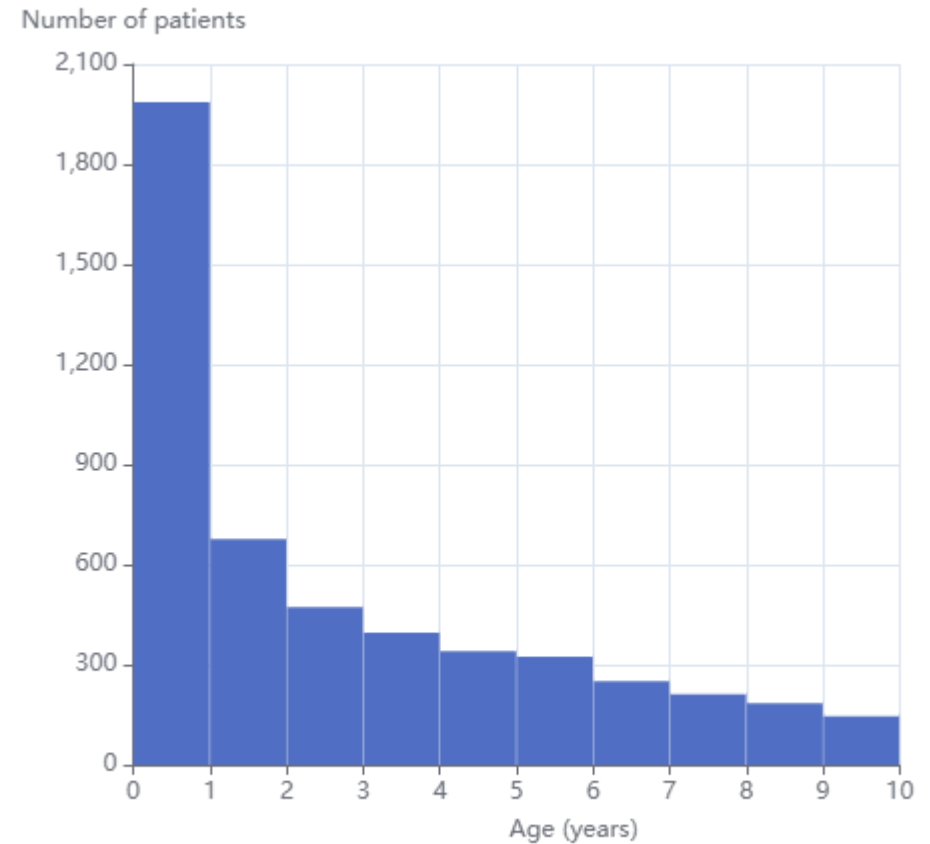
Covariates	Median
Age at CD, years (n=9172)	7.57
Delay between onset and CD, years (n=7976)	1.45
CVID	5.01
non-CVID	2.5
SCID	0.08
CID	1
Other T-cell def.	0.52
Innate	0.5

Clinical diagnosis (CD)

Age at CD

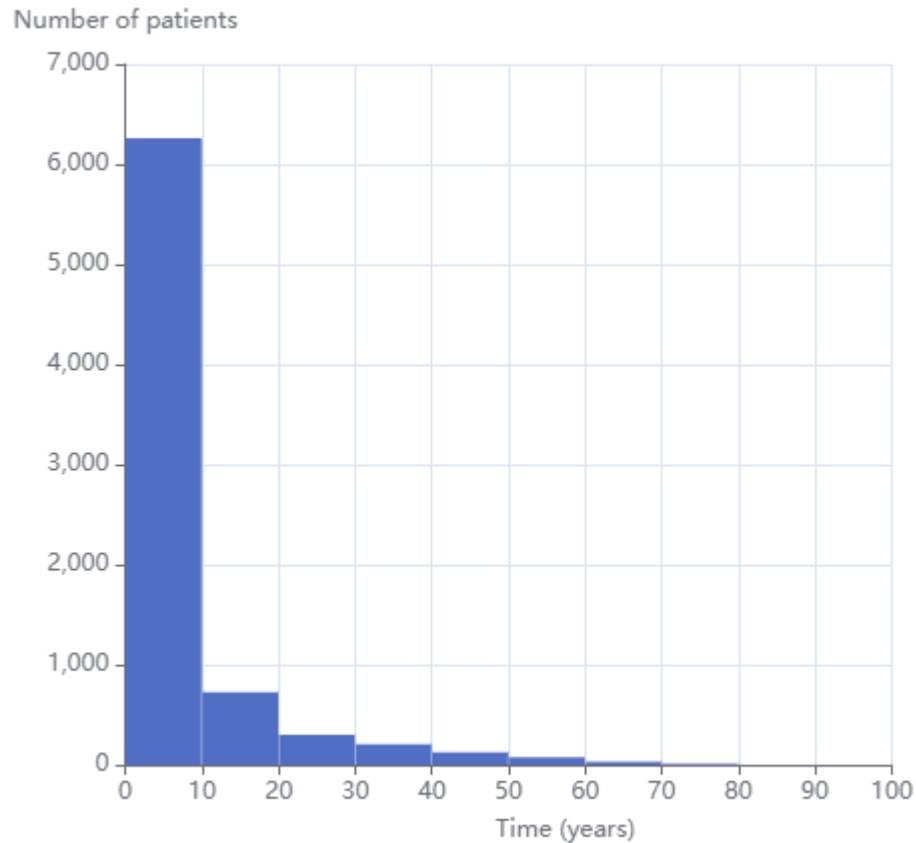


Age at CD (0 - 10 years)

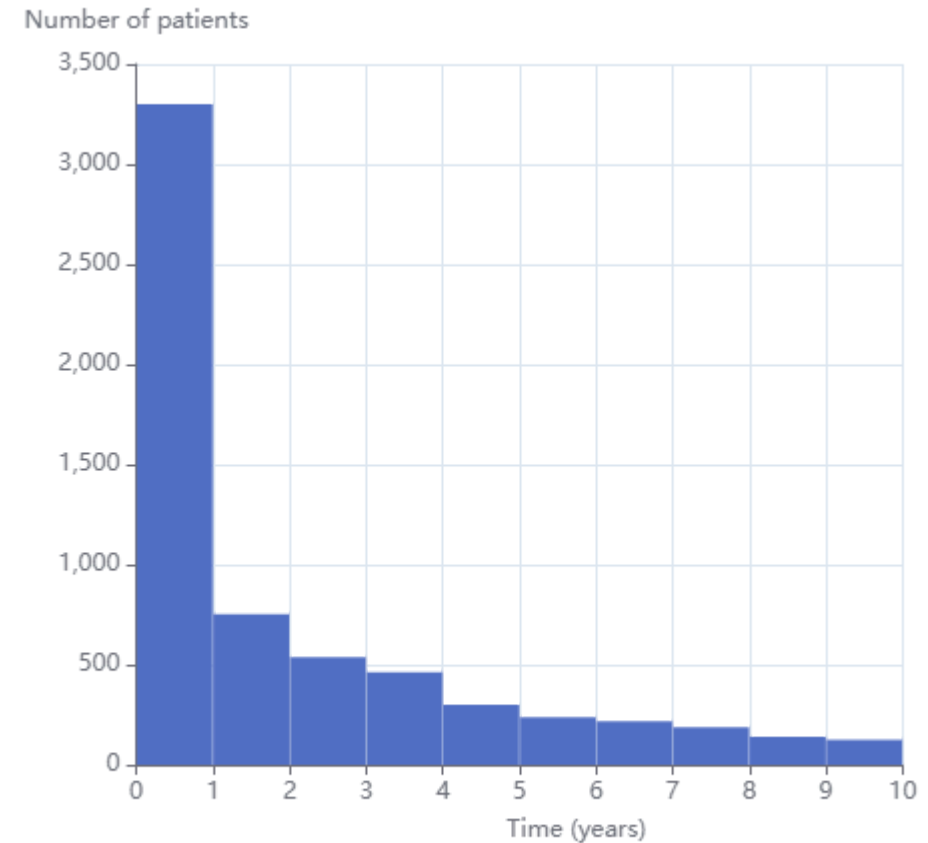


Clinical diagnosis (CD)

Delay between onset and CD



Delay between onset and CD (0 - 10 years)



Genetic diagnosis (GD)

Covariates	N	Percentage
Genetic mutation found (n=9364)	4516	48
CVID	159	8
non-CVID	347	16
SCID	1605	79
CID	388	85
Other T-cell def.	883	82
Innate	1211	74
Additional gene found	483	5

Genetic diagnosis (GD)

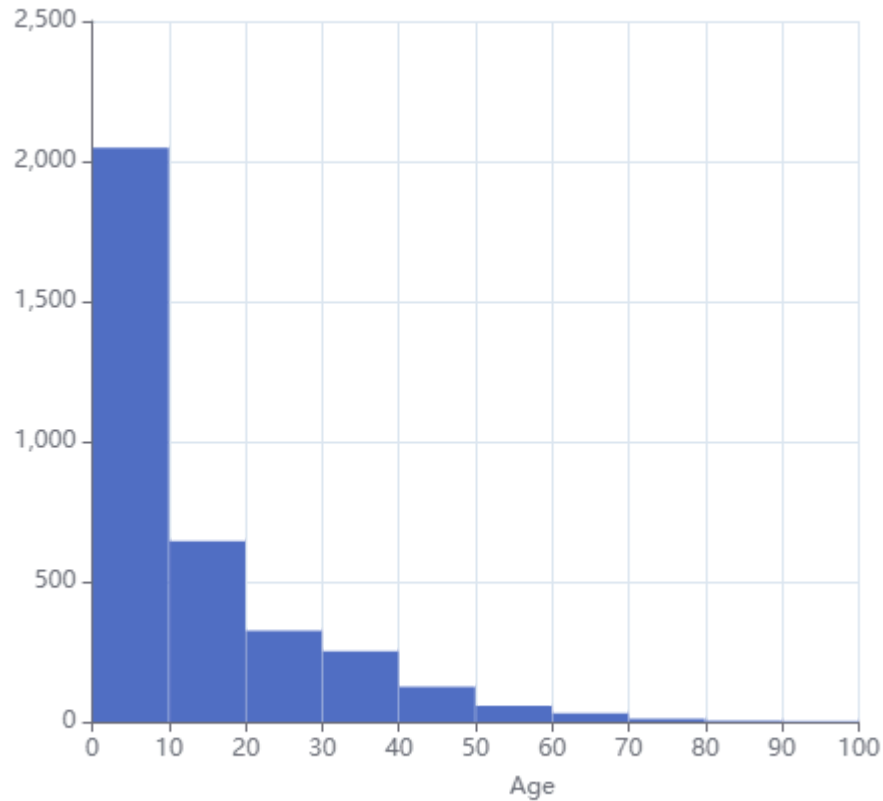
Covariates	Median
Age at GD, years (n=3484)	6.8
CVID	31.23
non-CVID	5.83
SCID	0.47
CID	0.47
Other T-cell def.	8.29
Innate	7.97

Genetic diagnosis (GD)

Covariates	Median
Delay between CD and GD, years (n=3459)	0.41
CVID	4.01
non-CVID	1.13
SCID	0.05
CID	0.05
Other T-cell def.	0.18
Innate	0.37

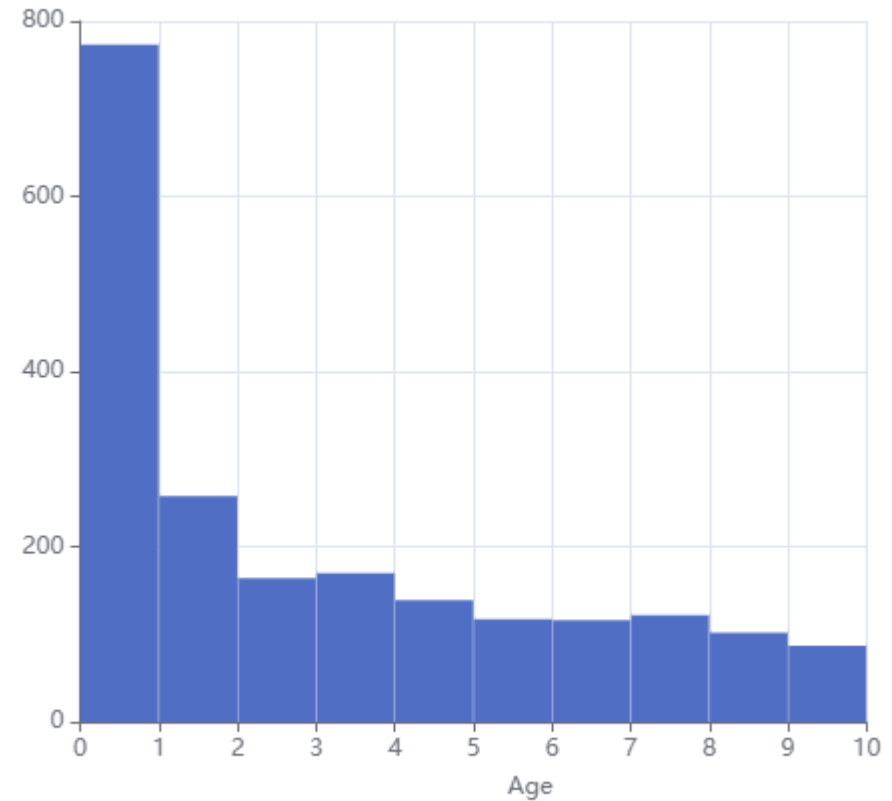
Genetic diagnosis (GD)

Distribution of age at genetic diagnosis



Distribution of. age at genetic diagnosis

(0 - 10 years)

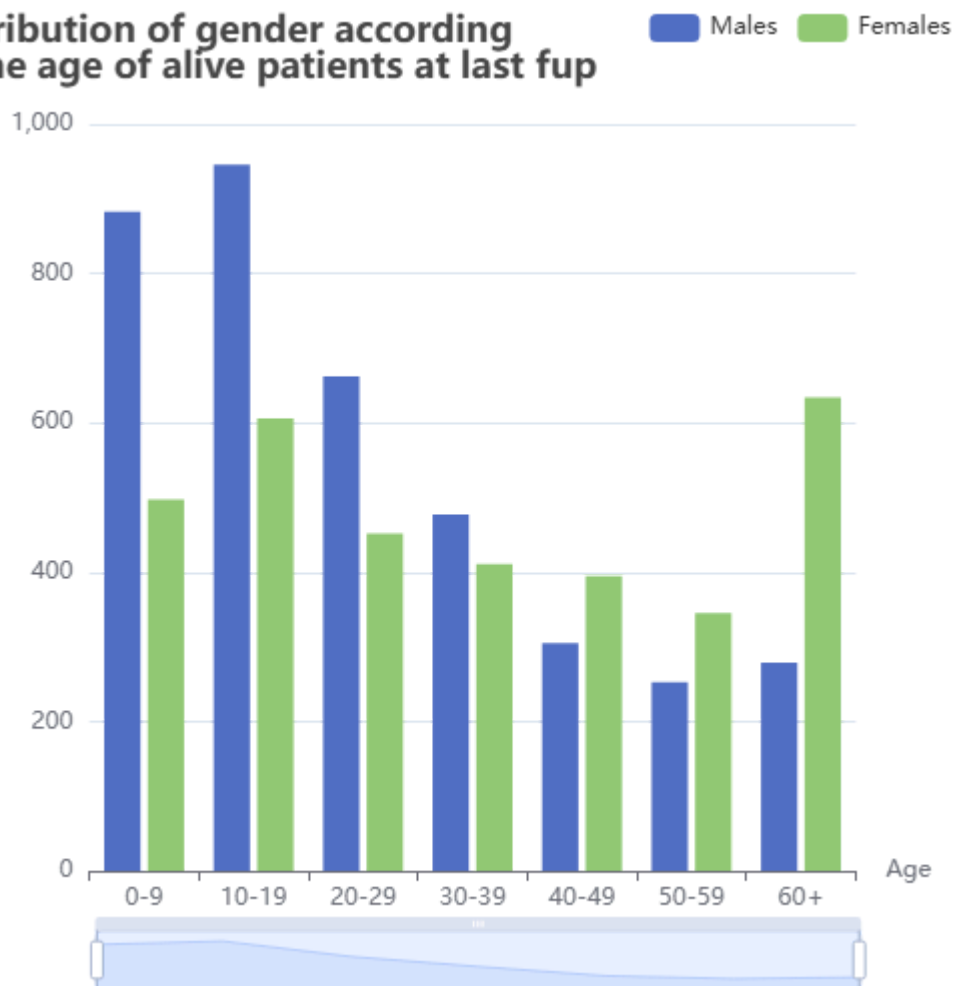


Last follow-up

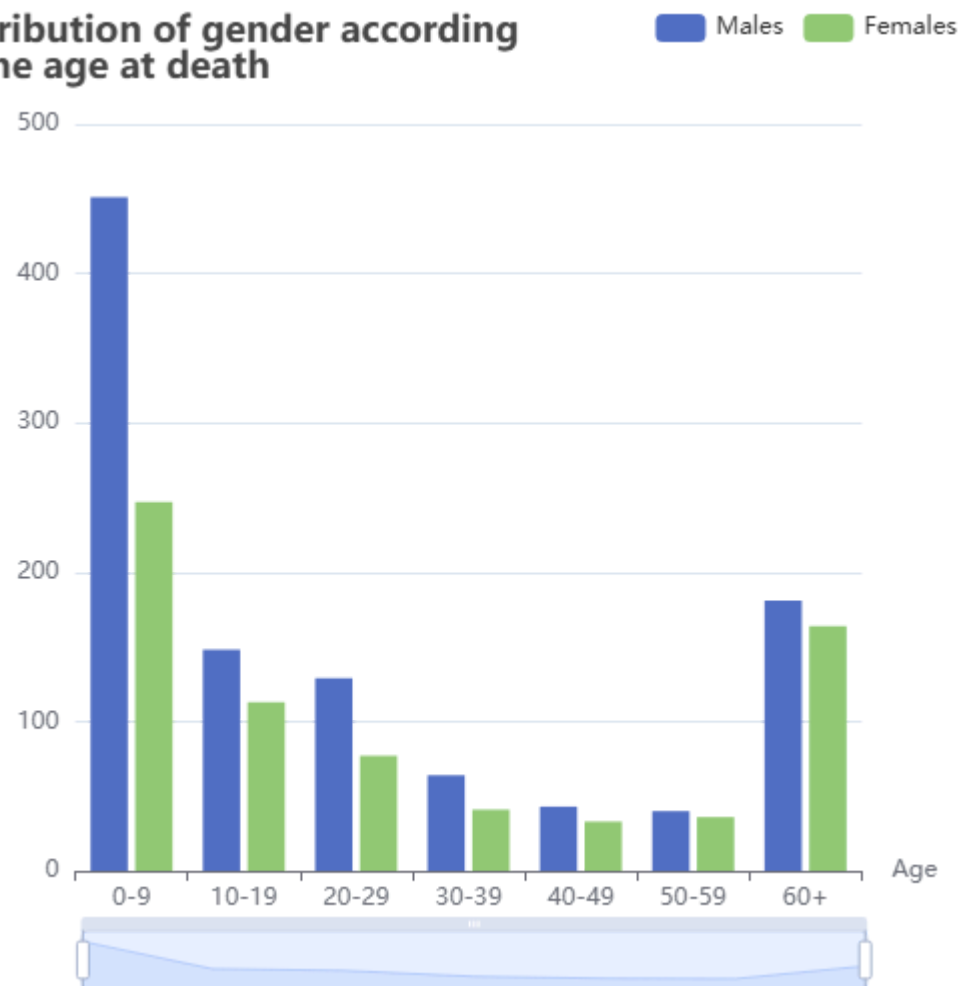
Covariates	Pop.	Median
Age at last news (alive and lost to follow-up patients), years	7779	24.82
Alive	7157	25.07
Lost to fup	633	22.7
Age at death, years	1760	17.07

Last follow-up

Distribution of gender according to the age of alive patients at last fup



Distribution of gender according to the age at death

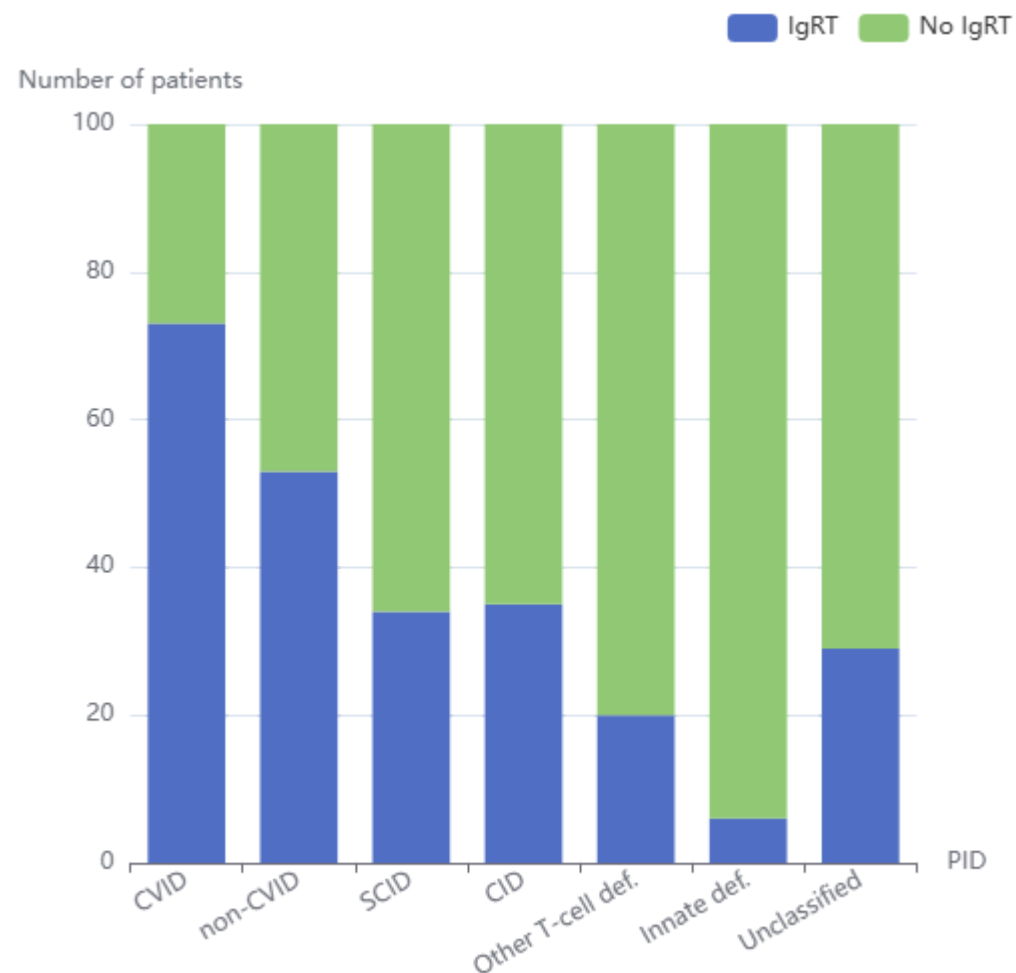
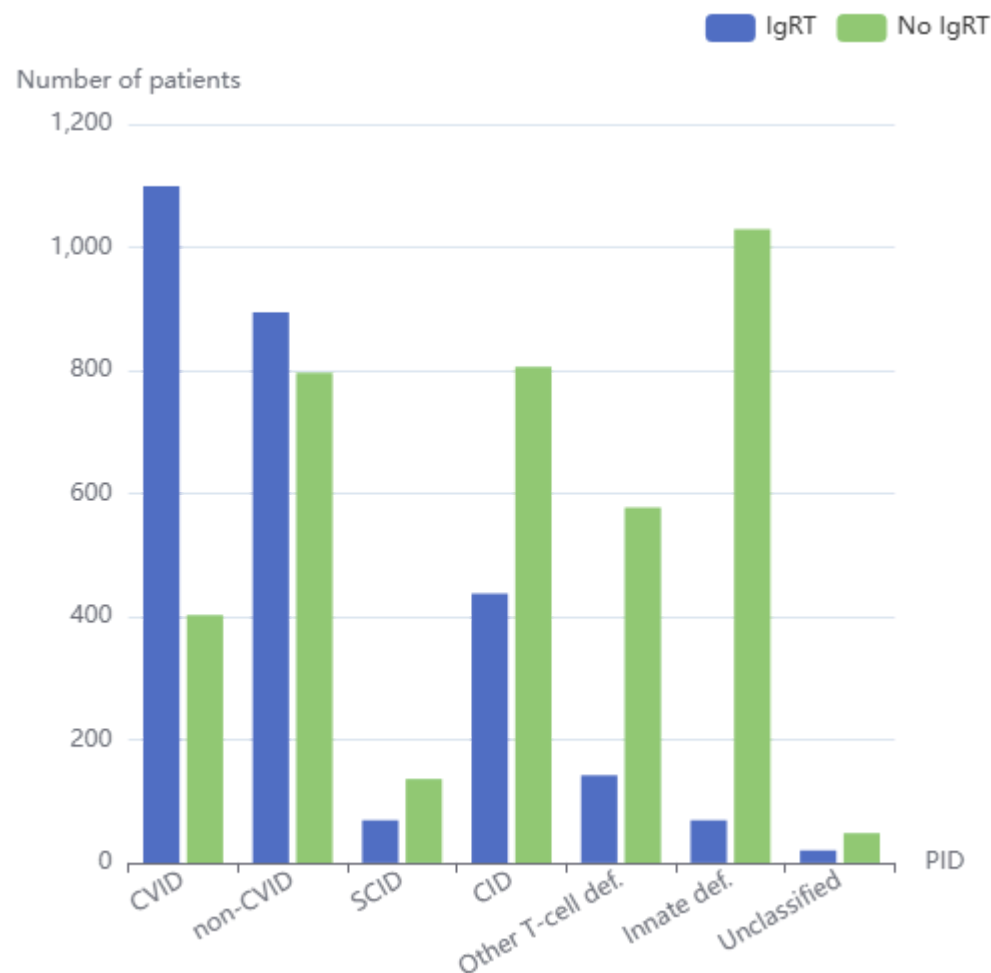


Treatments

Description of IgRT (on currently replaced population)

Covariates	Pop.	Median or N	Percentage
Ig replacement	6524	2730	42
Type of Ig replacement (IV)	2669	1265	47
Place of Ig replacement (home)	2643	1535	58
IV at home	2643	212	8
Age at first Ig replacement, years	3575	17.04	
Delay between onset of symptoms and Ig replacement	3276	4.42	
Delay between clinical diagnosis and Ig replacement	3554	0.26	

Current IgRT status by main PID categories



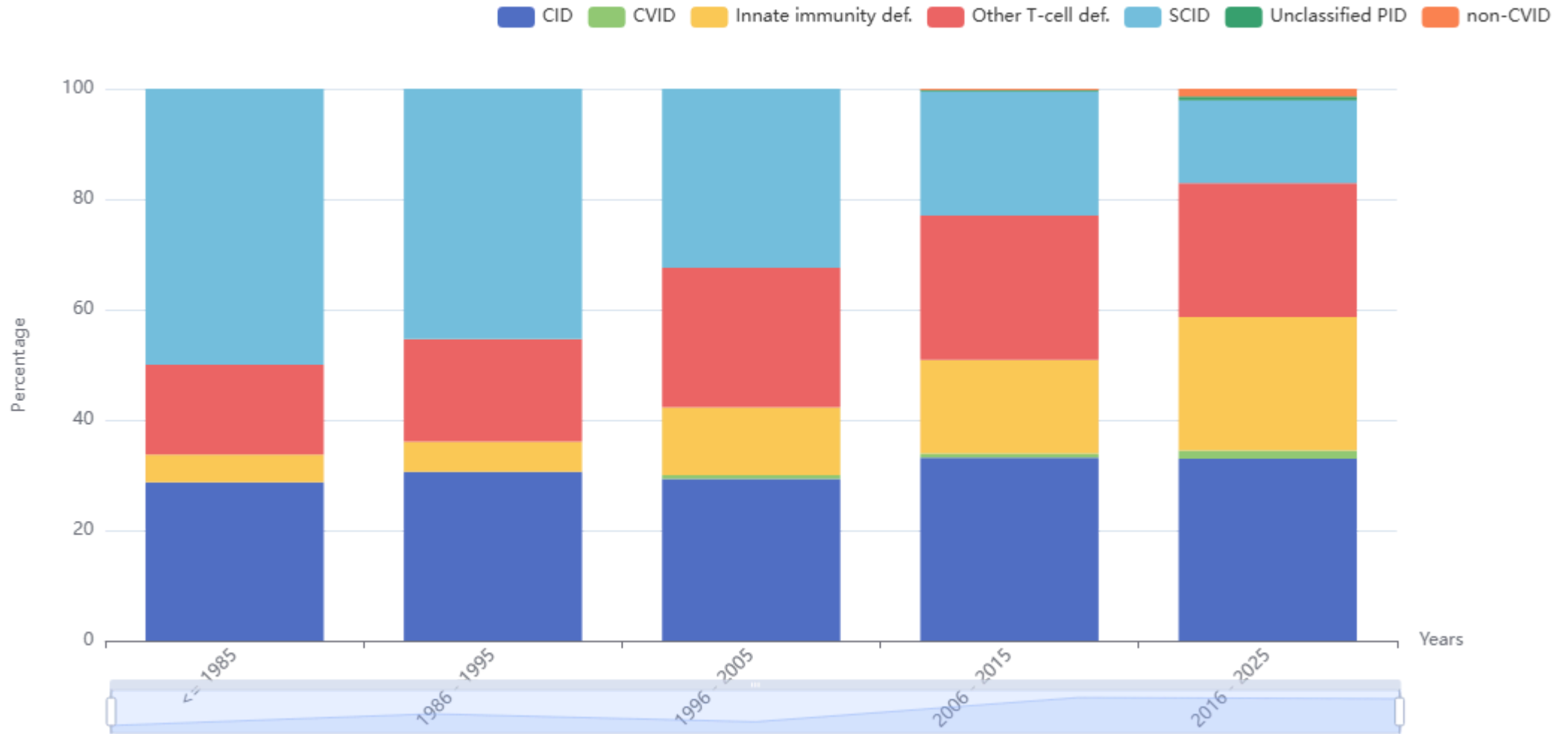
Description of alloHSCT

Covariates	Pop.	Median or N	Percentage
alloHSCT	9364	1490	16
Age at first alloHSCT, years	1448	1.67	
Delay between onset of symptoms and HSCT	1247	0.53	
Delay between clinical diagnosis and HSCT	1435	0.53	

Description of PID categories by decade of first alloHSCT



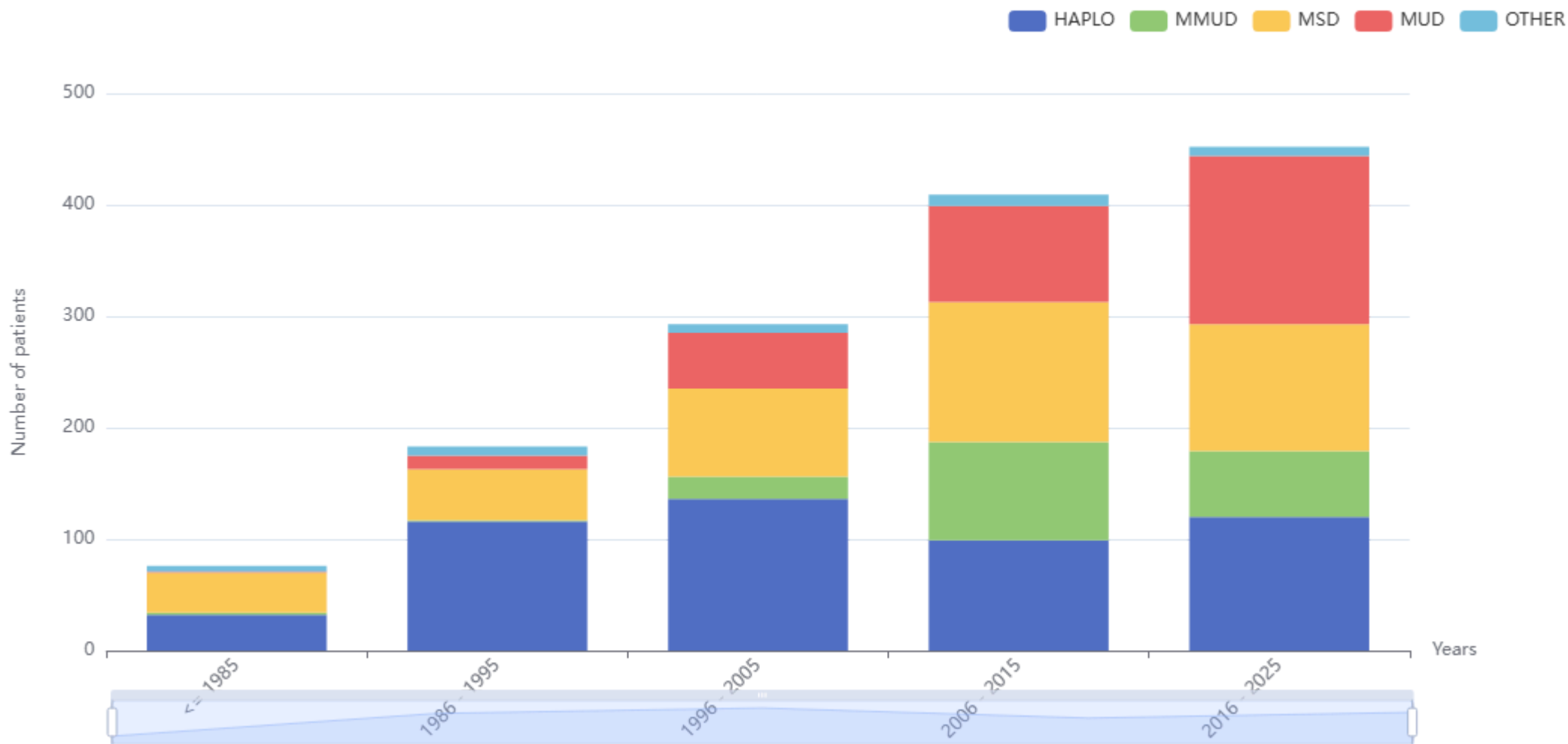
Description of main PID categories by decade of first alloHSCT



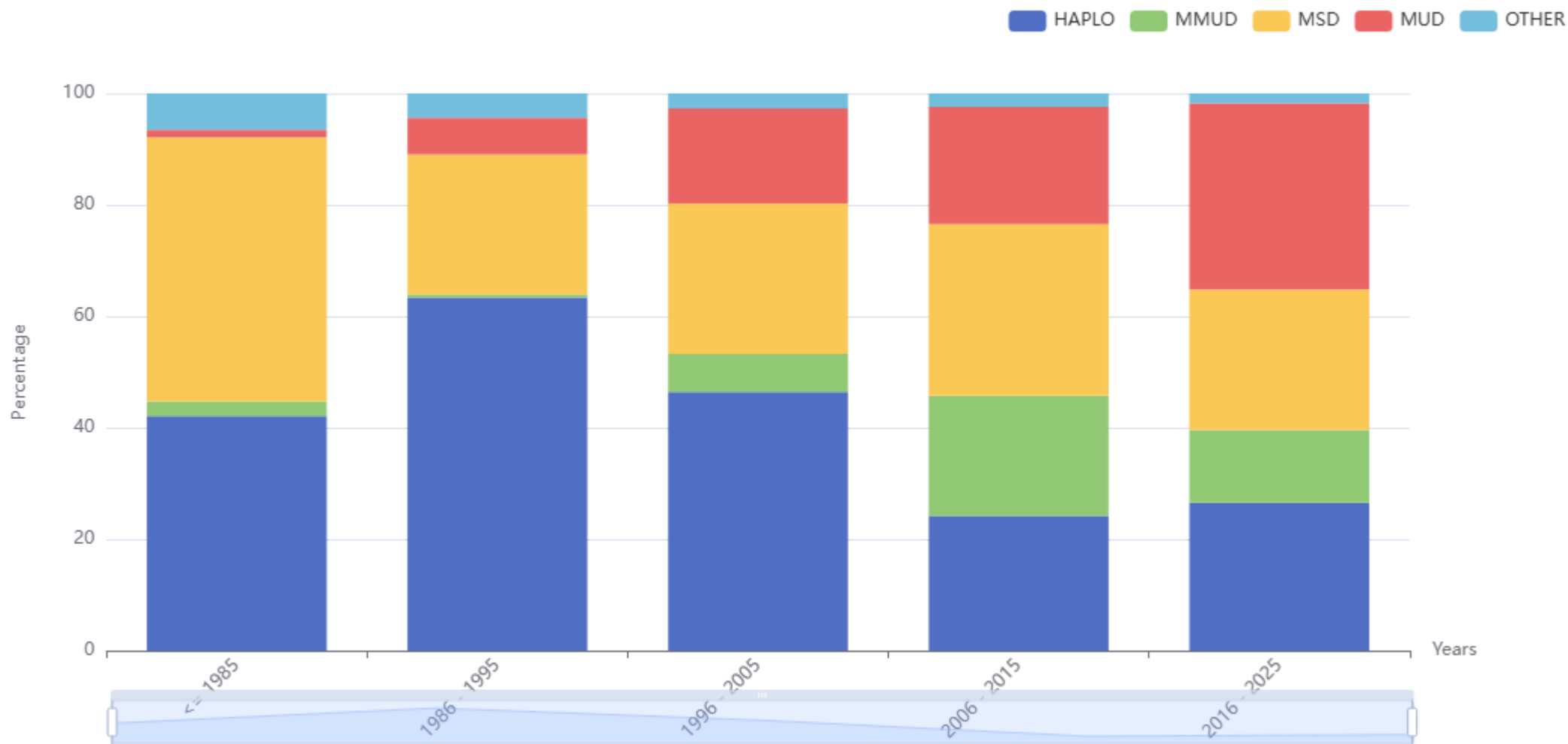
First alloH SCT donor (n = 1448)

Type of donor	N	Percentage
HAPLO	503	35.6
MMUD	170	12.0
MSD	401	28.4
MUD	300	21.2
OTHER	39	2.8

Description of donor by decade of first alloH SCT



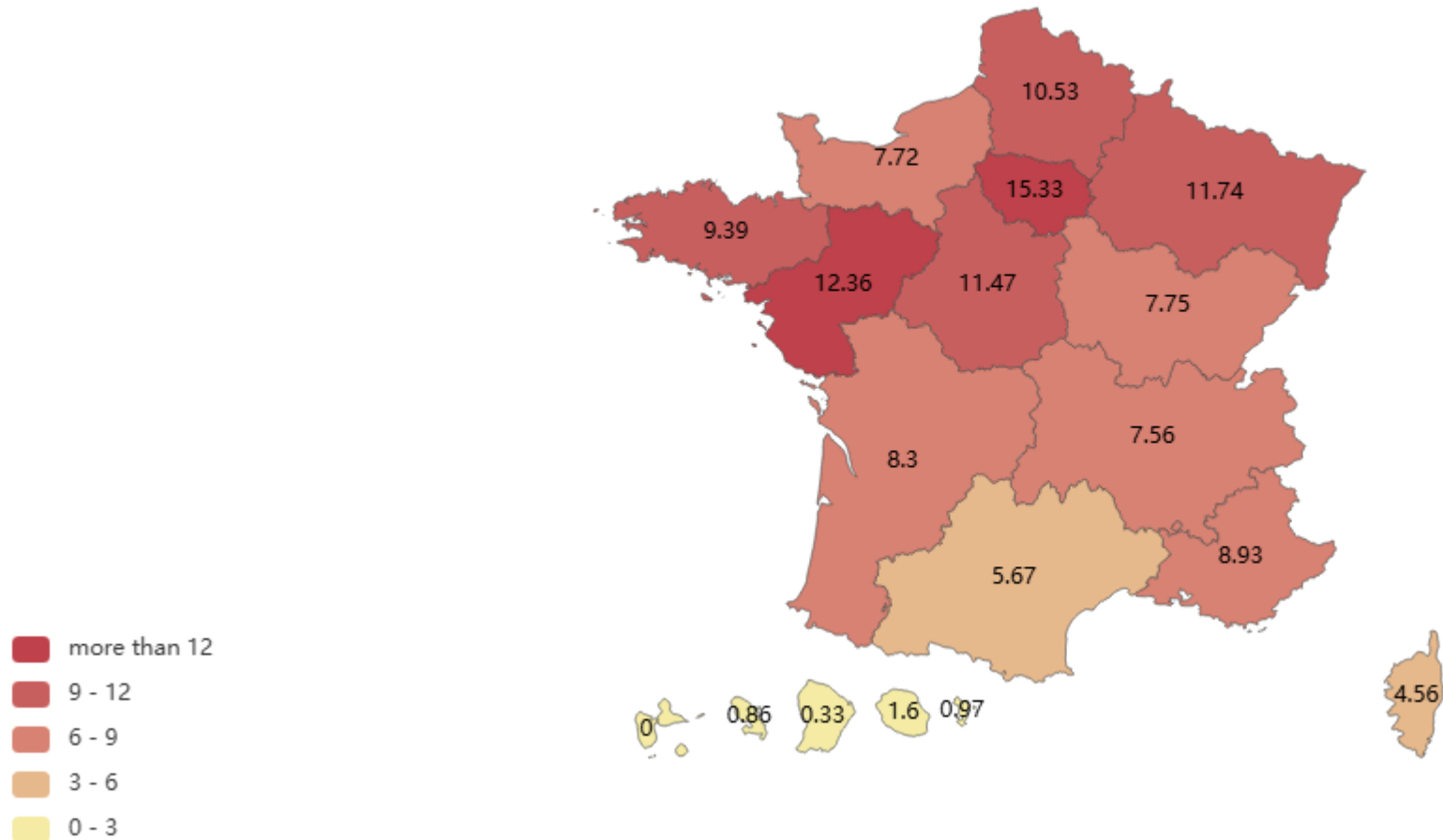
Description of donor by decade of first alloH SCT



Epidemiology

275,259 patient-years

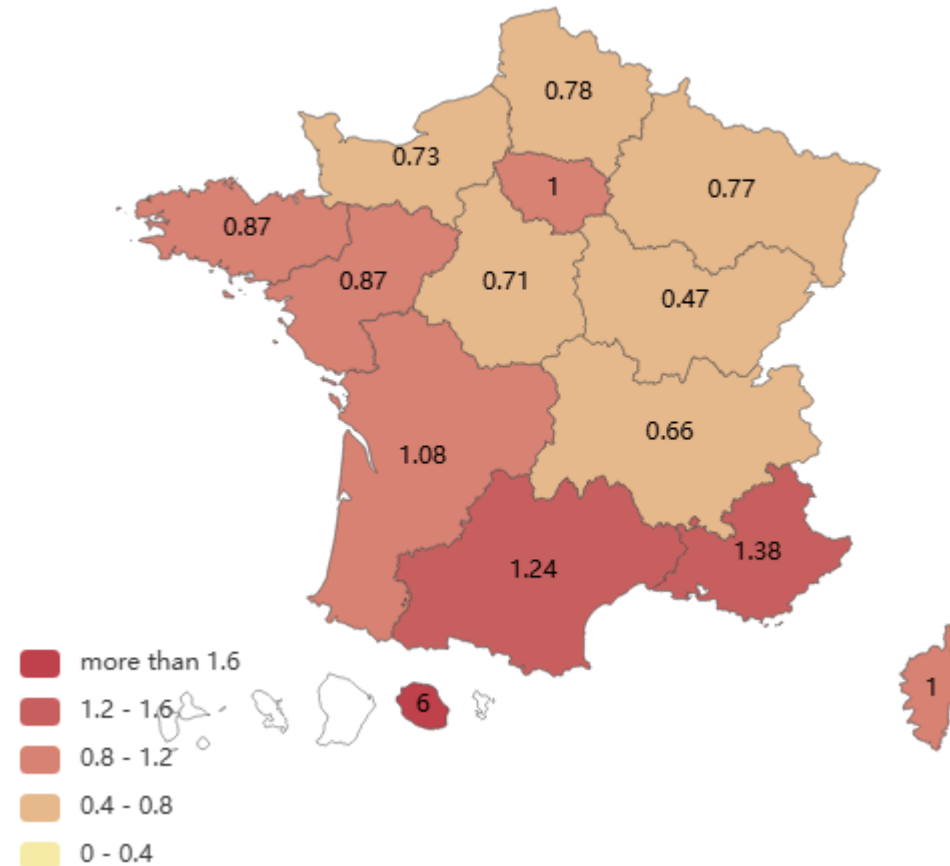
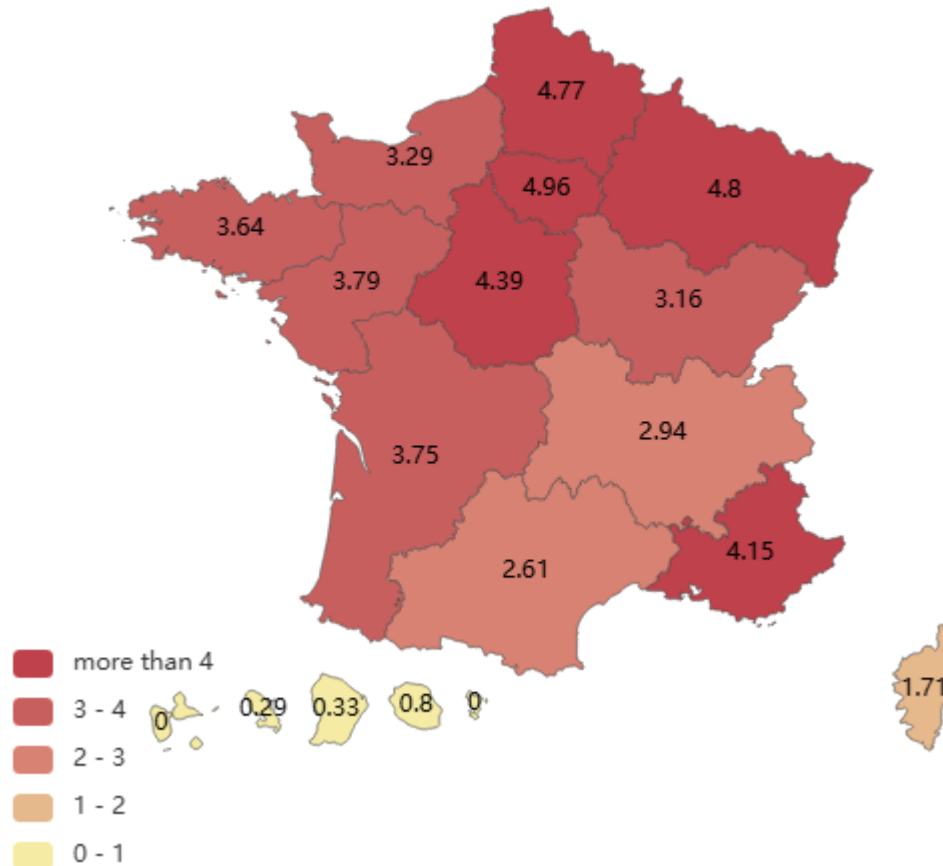
Prevalence of PID in France : 10.27 for 100 000 inhabitants



Immunoglobulin replacement therapy (overall prevalence : 4.03 for 100 000 inhabitants)

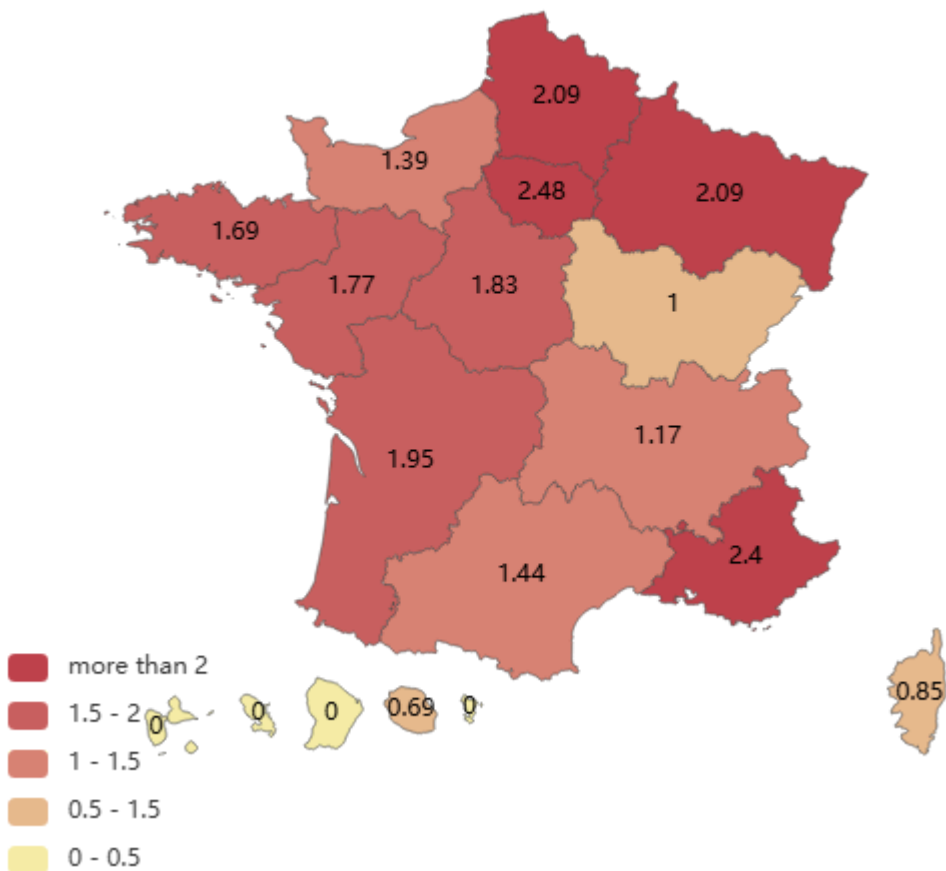
IV and SC

IV/SC ratio

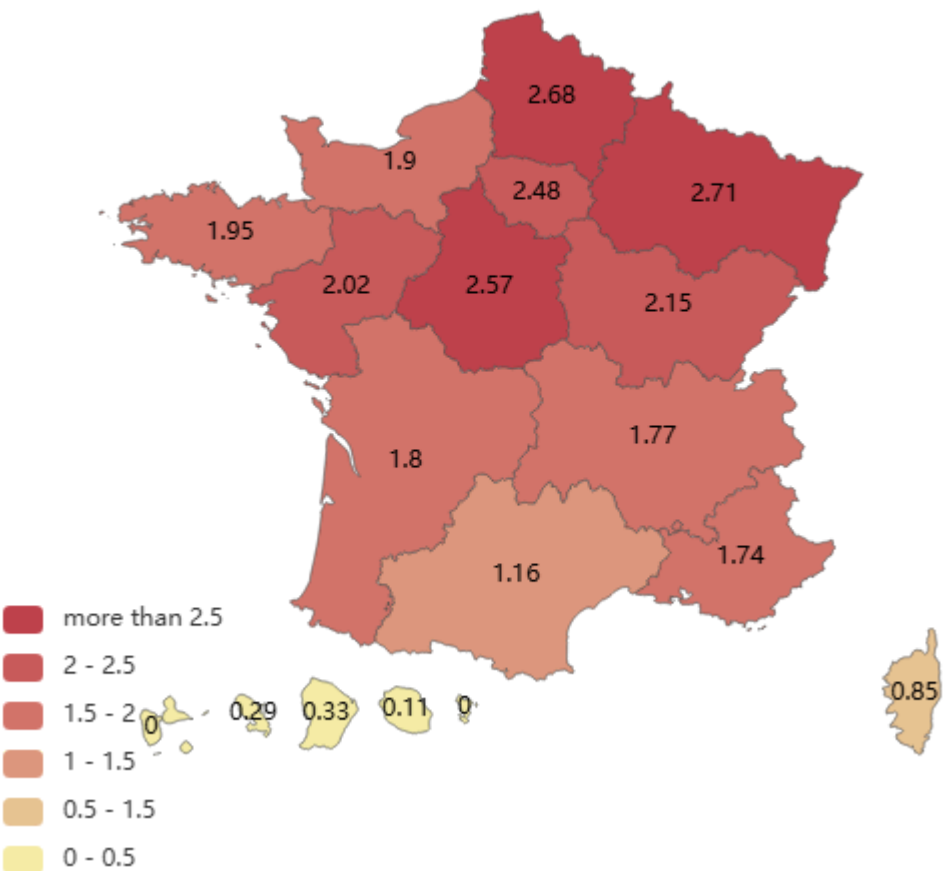


Immunoglobulin replacement therapy

IV



SC

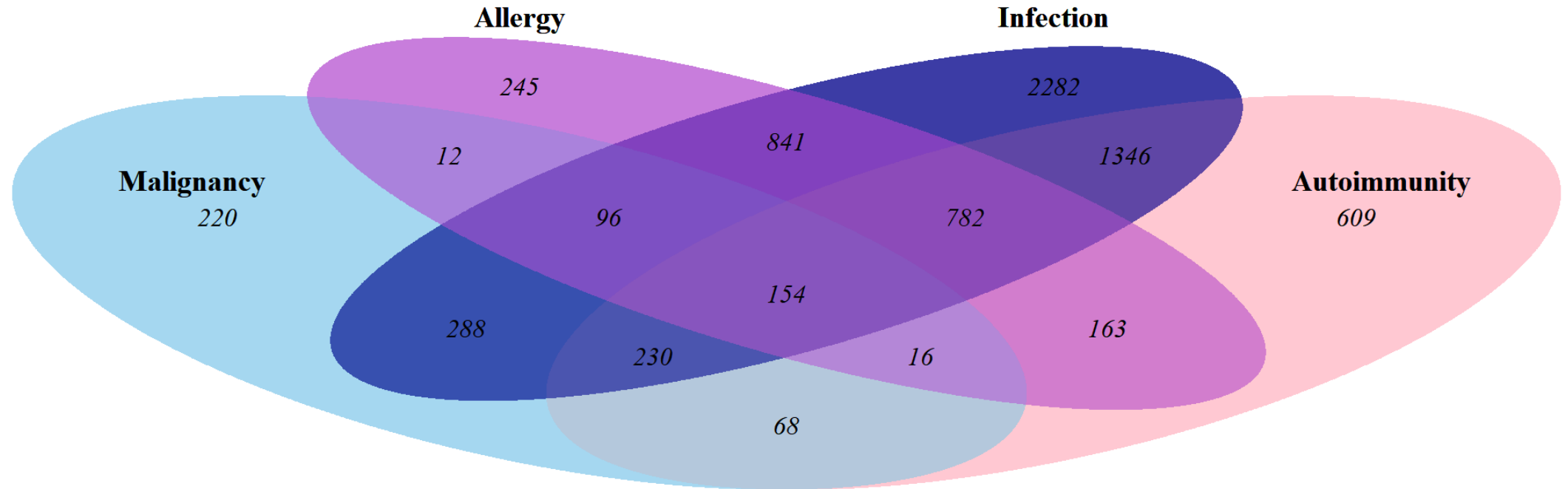


Description of clinical manifestations

Clinical manifestations

Covariates	Pop.	Median or N	Percentage
Age at first clinical manifestation, years	5733	7.1	
Severe allergy	8213	2309	28.1
Age at first severe allergy, years	1518	5.12	
Autoimmunity	8280	3368	40.7
Age at first autoimmunity, years	2919	12.47	
Malignancy	9211	1084	11.8
Age at first malignancy, years	1031	38.61	
Infection	6786	6019	88.7
Age at first infection, years	3687	6.93	

Clinical manifestations



Acknowledgment

Acknowledgment

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This presentation was created by Merlin Simsen Baratault