

Date: 23.05.2018 - 12:06 (CEST)

Participant: Dr. Monika Poppe

# Project "USZ-HAE-Amyloidosis registry"

DB schema (user): US229 Last DB

modification: 18.05.2018

- 12:12:24 (CEST)

**Type:** • Registry • Part project

Project name: USZ-HAE-Amyloidosis registry

11:51:29 (CEST)

Reason: New user management

Vers. V<sub>1.01</sub> Vers. 2 Last vers. modification: 23.05.2018 -

label: 11:51:29 (CEST)

### **Used catalogues** (0)

### Form families (7)

Displayed order	Form family name	Туре	Hidden
10	- Sub Casenode	Subform	
	Sub Medical History <emnpus229_cd> Sub Concomitant Treatment and Medication <emnpus2< td=""><td>29_cm&gt;</td><td></td></emnpus2<></emnpus229_cd>	29_cm>	
50	- NEU: subforms	Subform	
	sub AL therapy <emnpus229_sub_therapy> sub AA therapy <emnpus229_sub_aa_therapy> sub ATTR / other therapy <emnpus229_sub_attr_thrpy: <emnpus229_sub_compl_al="" complications="" sub=""> coagulation disorders <emnpus229_coagul_dis> please specify <emnpus229_specify> specify other biopsy <emnpus229_spec_biopsy> specify other symptoms leading to diagnosis <emnpus2 <emnpus229_affected_relations="" <emnpus229spec_pr<="" affected="" history="" medical="" other="" past="" relative(s)="" specify="" td=""><td>:29specify_sltd&gt; &gt;</td><td></td></emnpus2></emnpus229_spec_biopsy></emnpus229_specify></emnpus229_coagul_dis></emnpus229_sub_attr_thrpy:></emnpus229_sub_aa_therapy></emnpus229_sub_therapy>	:29specify_sltd> >	
9	- baseline	Visit	
	diagnosis (FILL IN / SAVE FIRST!) <mnpus229_diagno <mnpus229_demographics="" demographics=""> medical history <mnpus229_med_history> organ involvement baseline <mnpus229_organ_involv> bone marrow <mnpus229_bmarrow_st> laboratory values <mnpus229_laboratory></mnpus229_laboratory></mnpus229_bmarrow_st></mnpus229_organ_involv></mnpus229_med_history></mnpus229_diagno>		
20	- follow up	Visit	
	condition of the patient <mnpus229_condition> organ response follow up <mnpus229_resp_criteria> AL: treatment since registry inclusion / last follow up <m <mn="" aa:="" attr="" follo<="" follow-up="" inclusion="" last="" other:="" since="" study="" td="" treatment=""><td>pus229_aa_treatment&gt;</td><td></td></m></mnpus229_resp_criteria></mnpus229_condition>	pus229_aa_treatment>	
20	- Study End	Casenode	
	Study End <mnpus229_end> Premature Study End <mnpus229_end_prem></mnpus229_end_prem></mnpus229_end>		
10	- Subform centre	Centre Subform	

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Sub Principal Investigator <emnpus229\_pi> Sub Participants Setup <emnpus229\_prt\_stp> Sub Participants Productive <emnpus229\_prt\_prd>

10 - Centre Centre

Participants and roles - Setup <mnpus229\_prt\_stp>
Participants and roles - Productive <mnpus229\_prt\_prd>

Date: 23.05.2018 - 12:06 (CEST) Participant: Dr. Monika Poppe Project "USZ-HAE-Amyloidosis registry"

secuTrial<sup>®</sup> 5.3.0.15, 2018

Project	USZ-HAE-Amyloidosis regis	try							
						Treatment ar	m AL follow up		
									AL follow-up
									Created visits: 1
Family	Form	Table(s)		baseline	1. AL follow-up	2. AL follow-up	3. AL follow-up	4. AL follow-up	5. AL follow-up
			Day of visit	0	183	365	548	730	365
			at interval to	entry	baseline	baseline	baseline	baseline	preceding, planned visit
			Туре	flexible	flexible	flexible	flexible	flexible	flexible
			possible deviation in days						
Visit plan			117			Visit p	lan tab		<u> </u>
baseline									
	diagnosis (FILL IN / SAVE	mnpus229_diagnosis,		х					
	FIRST!)	emnpus229_spec_biopsy,							
		emnpus229specify_sltd							
	demographics	mnpus229_demographics		Х					
	medical history	mnpus229_med_history,		Х					
		emnpus229_affected_relat,							
		emnpus229spec_pmh							
	organ involvement baseline	mnpus229_organ_involv		Х					
	bone marrow	mnpus229_bmarrow_st		Х					
	laboratory values	mnpus229_laboratory		Х					
follow up									
	condition of the patient	mnpus229_condition,			Х	Х	Х	Х	х
		emnpus229_sub_compl_al							
	organ response follow up	mnpus229_resp_criteria			Х	Х	Х	Х	Х
	AL: treatment since registry	mnpus229_treatment,			Х	Х	Х	Х	Х
	inclusion / last follow up	emnpus229_sub_therapy							
	AA: treatment since last	mnpus229_aa_treatment,							
	follow-up / study inclusion	emnpus229_sub_aa_therapy							
	ATTR / other: treatment since								
	study inclusion / last follow- up	emnpus229_sub_attr_thrpy							

						Treatment arr	n AA follow up		
									AA follow-up
									Created visits: 1
Family	Form	Table(s)		baseline	1. AA follow-up	2. AA follow-up	3. AA follow-up	4. AA follow-up	5. AA follow-up
			Day of visit	0	183	365	548	730	365
			at interval to	entry	baseline	baseline	baseline	baseline	preceding,
									planned visit
			Туре	flexible	flexible	flexible	flexible	flexible	flexible
			possible						
			deviation in days						
Visit plan						Visit p	lan tab		
baseline									
	diagnosis (FILL IN / SAVE	mnpus229_diagnosis,		Х					
	FIRST!)	emnpus229_spec_biopsy,							
	,	emnpus229specify_sltd							
	demographics	mnpus229 demographics		х					
	medical history	mnpus229 med history,		х					
	,	emnpus229_affected_relat,							
		emnpus229spec_pmh							
	organ involvement baseline	mnpus229 organ involv		Х					
	bone marrow	mnpus229_bmarrow_st		Х					
	laboratory values	mnpus229 laboratory		Х					
follow up	,	<del>,                                      </del>							
	condition of the patient	mnpus229 condition,			х	х	Х	х	х
		emnpus229 sub compl al							
	organ response follow up	mnpus229_resp_criteria			Х	х	Х	Х	Х
	AL: treatment since registry	mnpus229_treatment,							
	inclusion / last follow up	emnpus229 sub therapy							
	AA: treatment since last	mnpus229_aa_treatment,			х	х	Х	Х	х
	follow-up / study inclusion	emnpus229_sub_aa_therapy							
	, , , , , , , , , , , , , , , , , , , ,								
	ATTR / other: treatment since	mnpus229_attr_treatment,							
	study inclusion / last follow-	emnpus229_sub_attr_thrpy							
	up								

						Treatment arm AT	ΓR / other follow u	p	
									ATTR / other
									Created visits: 1
				baseline	1. ATTR / other	2. ATTR / other	3. ATTR / other	4. ATTR / other	5. ATTR / other
Family	Form	Table(s)			follow up	follow-up	follow-up	follow-up	follow-up
			Day of visit	0	183	365	548	730	365
			at interval to	entry	baseline	baseline	baseline	baseline	preceding,
									planned visit
			Type	flexible	flexible	flexible	flexible	flexible	flexible
			possible						
			deviation in days						
Visit plan						Visit p	lan tab		
baseline									
	diagnosis (FILL IN / SAVE	mnpus229_diagnosis,		X					
	FIRST!)	emnpus229_spec_biopsy,							
		emnpus229specify_sltd							
	demographics	mnpus229_demographics		Х					
	medical history	mnpus229_med_history,		X					
		emnpus229_affected_relat,							
		emnpus229spec_pmh							
	organ involvement baseline	mnpus229_organ_involv		Х					
	bone marrow	mnpus229_bmarrow_st		X					
	laboratory values	mnpus229_laboratory		X					
follow up		1	1						
	condition of the patient	mnpus229_condition,			Х	Х	х	Х	х
		emnpus229_sub_compl_al							
	organ response follow up	mnpus229_resp_criteria			Х	Х	х	Х	Х
	AL: treatment since registry	mnpus229_treatment,							
	inclusion / last follow up	emnpus229_sub_therapy							
	AA: treatment since last	mnpus229_aa_treatment,							
	follow-up / study inclusion	emnpus229_sub_aa_therapy							
	ATTR / other: treatment since	manus 220 otto trootmont			Х	х	х	X	х
	study inclusion / last follow-	mnpus229_attr_treatment, emnpus229_sub_attr_thrpy			^	_ ^	^	^	^
	-	emnpus229_sub_attr_thrpy							
	up					ļ		ļ	ļ
Study End						Cason	ode tab		
Study Liid	Study End	mnpus229 end				Casen	Jue lab		
	Premature Study End	mnpus229_end_prem							
	Tremature Study End	mipus225_end_prem	<u> </u>						
Centre						Cent	re tab		
Contro	Participants and roles - Setup	mnpus229_prt_stp,				Cent			
	. s. delpants and roles Setup	emnpus229_pi,							
		emnpus229_prt_stp							
	Participants and roles -	mnpus229_prt_prd,							
	Productive	emnpus229_prt_prd							
L		cbiozzzz_bi t_bio							



Date	
Participant	
Centre	
Б	

Patient	
Visit	
Form family	follow up

Project USZ-HAE-Amyloidosis registry (V1.01) Form AA: treatment since last follow-up / study inclusion

AA: treatment since last follow-up / study inclusion

		o luci lonon up / clau, l					( - /
Follow up pe	eriod: (do not fill in)						
data entry r	ange from:	until:					
	dd.mm.yyyy	dd.mm.yyyy					
ti	reatment						
	reatment since last f	follow up?					
	○Yes ○No	ionow up.					
ι	Jse 'weitere' / 'more'	to state all relevant therapie	es in this period of time				
t	herapy 1						
	○ pharma ○ transpla						
	pharmaco	ological therapy					
		treatment start		dd.mm.	уууу		
		ongoing treatment		OYes ONo			
		treatment end		dd.mm.	vvvv		
	disease m	nodifying drugs			,,,,,		
	dioodoo iii	iounying urugo			please specify		
		1)			picase specify		
	transplant	tation					
		transplanted organ		2)			
		please specify					
		date of transplantation		dd.mm.	yyyy		
		pharmacological immunos	suppression	OYes ONo Onot know			
	More						
;	Signature						
ı	Place, date	Si	ignature				
ı	Possible entries						
F	Please select one of the	he following entries for the corr	responding items marked above.				
1)	colchicine						
	OCIOI IIOII IC						

TNF-alpha blocker

IL-1 blocker

rituximab

tocilizumab

corticosteroids

NSAID

eprodisate

other

2)

heart
kidney
liver
other



Patient	
/isit	
orm family	follow up

Form AL: treatment since registry inclusion / last follow up treatment since registry inclusion / last follow up Follow up period: (do not fill in) data entry range from: until: dd.mm.yyyy dd.mm.yyyy treatment since last follow up? OYes ONo Use 'weitere' / 'more' to state all relevant chemotherapies in this period of time therapy 1 O pharmacological Otransplantation start of treatment dd.mm.yyyy OYes ONo ongoing treatment end of treatment dd.mm.yyyy cycle numbers chemotherapy please specify please specify please specify please specify please specify please specify transplantation transplanted organ please specify date of transplantation dd.mm.yyyy OYes ONo Onot known pharmacological immunosuppression hematologic response after therapy hematologic response during this treatment response criteria: Gertz MA et al. AJoH. 79:319-328 (2005) date of data collection remission status dd.mm.yyyy serum immunofixation Opositive Onegative Onot known dd.mm.yyyy urine immunofixation Opositive Onegative Onot known not done dd.mm.yyyy total free light chain kappa dd.mm.yyyy total free light chain lambda serum M-gradient g/I dd.mm.yyyy bone marrow examination performed yes dd.mm.yyyy bone marrow plasma cells % More hematologic response (no therapy since last follow up) response criteria: Gertz MA et al. AJoH, 79:319-328 (2005) date of data collection dd.mm.yyyy remission status serum immunofixation Opositive Onegative Onot known dd.mm.yyyy eCRF Amyloidosis Registry V1.0.1 / 23.05.2018 Page 8 of 43 urine immunofixation Opositive Onegative Onot known not done total free light chain kappa 

	total free light chain lambda g / I		dd.mm.yyyy		
	serum M-gradient g / I		dd.mm.yyyy		
	bone marrow examination performed  yes		dd.mm.yyyy		
	bone marrow plasma cells %				
	Signature				
	Gignature				
	Place, date Signat	ure			
	Possible entries				
	Please select one of the following entries for the corresp	onding items marked above.			
1)	corticosteroid monotherapy				
	melphalan based				
	thalidomide based				
	lenalidomide based				
	bortezomib based				
	bendamustin based				
	pomalidomide based				
	carfilzomib based				
	high-dose therapy and autologous HSCT				
	other				
2)					
-,	melphalan - prednisone				
	melphalan - dexamethasone other				
	outer				
3)	bortezomib - dexamethasone				
	bortezomib - melphalan - prednisone				
	bortezomib - cyclophosphamide - dexamethasone				
	bortezomib - lenalidomide - dexamethasone				
	other				
4)	thalidomide - dexamethasone				
	other				
5)	lenalidomide - dexamethasone				
,	lenalidomide - dexametrasorie lenalidomide - melphalan - prednisone				
	other				
	out.				
6)	High-dose therapy and autologous HSCT (melphalan 1	00mg/m2)			
	High-dose therapy and autologous HSCT (melphalan 1	20mg/m2)			
	High-dose therapy and autologous HSCT (melphalan 1	40mg/m2)			
	High-dose therapy and autologous HSCT (melphalan 2	00mg/m2)			
	other				
7)	book				
',	heart				
	kidney				
	allogeneic stem cell				
	other				
3)	sCR				
	CR				
	VGPR				
	PR				

not known



Date	
Participant	
Centre	
Destant	LIOZ LIAE Annudelidade na elektro 0.44 0.41

Patient	
Visit	
Form family	follow up

For USZ-HAE-Amyloidosis registry (V1.01) Form ATTR / other: treatment since study inclusion / last follow-up Project ATTR / other: treatment since study inclusion / last follow-up (V1.01) Follow up period: (do not fill in) data entry range from: until: dd.mm.yyyy dd.mm.yyyy treatment treatment since last follow up? OYes ONo Use 'weitere' / 'more' to state all relevant therapies in this period of time therapy 1 O pharmacological O transplantation pharmacological therapy start of treatment dd.mm.yyyy ○Yes ○No ongoing treatment end of treatment dd.mm.yyyy disease modifying drugs TTR please specify transplantation transplanted organ please specify date of transplantation dd.mm.yyyy OYes ONo Onot known pharmacological immunosuppression More Signature Place, date Signature

#### Possible entries

Please select one of the following entries for the corresponding items marked above.

1) doxycycline tafamidis diflunisal siRNA other

2) liver kidney heart other

UniversitätsSpital	Date Participant Centre Project USZ-HAE-Amyloidosis registry (V	1.01)	Patient Visit Form family baseline Form bone marrow	
baseline period: (do not fill in) data entry range until: from:  plasma cells				
plasma cells in bone marrow  percentage percentage date of exar  further examinations  aberrant implasma cell	mination % in biopsy dd.mm.yyyy			
FISH				
FISH performed date of performance		○Yes ○No ○not known dd.mm.yyyy		
ι(11,14)	2) s ONo Onot known O No Onot known O No O not known			

OYes ONo Onot

OYes ONo Onot

OYes ONo Onot

OYes ONo Onot

○Yes ○No ○not

known

known

known

known

t(14;16)

del17p

gain 1q

loss 1p

del 13

eCRF Amyloidosis Registry V1.0.1 / 23.05.2018

(V1.01)

	known  OYes ONo Onot please specify known
	Signature
	Place, date Signature
	Possible entries
	Please select one of the following entries for the corresponding items marked above.
1)	kappa
	lambda
	not known
->	
2)	normal
	abnormal
	not known



Date	
Participant	
Centre	
Destant	LICZ LIAE Amulaidania anniata (A/4 04

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USZ-HAE-Amyloidosis registry (V1.01)

condition of the patient

		,,,,,,,,,,,,,,,,,,,,,,,,,,,,,,,,,,,,,,,		
	condition of the patient			(V1.01)
Follow up p	period: (do not fill in)			
data entry	range from: until:			
	dd.mm.yyyy	dd.mm.yyyy		
	Is patient alive			
	is the patient alive?		date of death	
	○Yes ○No		dd.mm.yyyy	
	info: please fill in, print and	sign the form "study end"		
	cause of death please specify death related to	2)		
	please specify			
	complications leading to ho	ospitalisation		
	complications since last fol	low up		
	○Yes ○No			
	complication 1			
	reason of hospi		please specify	
	More	3)		
	quality of life			
		onnaire "EQ-5D-3L" OYes O	No	
	Signature			
	Place, date	Signature		
	Possible entries			
	Please select one of the follo	wing entries for the corresponding items marked above.		
1)	infection			
	cardiac			
	renal			
	bleeding			
	other			
2)	related to amyloidosis	eCRF Ar	nyloidosis Registry V1.0.1 / 23.05.2018	Page 15 of 43

treatment related

concomittant disease	
other	
nfection	
cardiac	



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rticipant	
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oject	USZ-HAE-Amyloidosis registry (V1.01)

Patient	
Visit	
Form family	baseline
Form	demographics

		-1	, , , , , , , , , , , , , , , , , , , ,			
demogr	raphics					(V1.01)
demogra	aphic data					
date of I	dd.mm.yyyy	Age Score				
<b>gender</b> ○ femal ○ male						
body cha	aracteristics					
baseline period: (c	do not fill in)					
data entry range from:	until:					
Irom:						
	not dor	ne				
height	cm 🗆					
weight	kg 🗆		date of weight measurement	dd.mm.yyyy	Body Mass Index Score	
quality o	of life					
	filled out questionnaire number of points date, questionnaire wa		○Yes ○No ○no			
		o mieu out		dd.mm.yyyy		
Signatu	re					
Place, da	ate	Signatu	re			
				1		



Date	
Participant	
Centre	
Project	US7-HAF-Amyloidosis registry (V1 01)

Patient	
Visit	
Form family	baseline
	discounts (EUL IN CONTESTIDOTI)
Form	diagnosis (FILL IN / SAVE FIRST!)

diagnosis	(V1.01
informed consent	
informed consent signed O Yes O No O not known date informed consent signed dd.	n .mm.yyyy
<u></u>	
type of amyloidosis	
please specify 2) please	e specify
date of diagnosis	
date of diagnosis  dd.mm.yyyy	
confirmed by biopsy  O Yes O No  date of genetic testing	genetic testing / g performed confirmed by szintigraphy (99mTc-DPD)  O Yes O No date of szintigraphy dd.mm.yyyy dd.mm.yyyy
amyloid deposit in tissue biopsy	
subcutaneous fat date of biopsy congo red immunohistochemistry please specify electron microscopy ms-based proteomics	O Yes O no amyloid detected O no biopsy performed  dd.mm.yyyy O positive O negative O not done O kappa O lambda O aa O attr O other O not conclusive O not done  C conclusive O not conclusive O not done O conclusive O not conclusive O not done
heart date of biopsy congo red immunohistochemistry please specify electron microscopy ms-based proteomics	O Yes O no amyloid detected O no biopsy performed  dd.mm.yyyy O positive O negative O not done O kappa O lambda O aa O attr O other O not conclusive O not done O conclusive O not conclusive O not done O conclusive O not conclusive O not done
kidney date of biopsy congo red immunohistochemistry please specify electron microscopy ms-based proteomics	O Yes O no amyloid detected O no biopsy performed  dd.mm.yyyy O positive O negative O not done O kappa O lambda O aa O attr O other O not conclusive O not done O conclusive O not conclusive O not done O conclusive O not conclusive O not done
intestinal date of biopsy congo red	○ Yes ○ no amyloid detected ○ no biopsy performed  dd.mm.yyyy ○ positive ○ negative ○ not done

	immunohistochemistry	kappa lambda aa attr other not conclusive not d	lone
	please specify		
	electron microscopy	O conclusive O not conclusive O not done	
	ms-based proteomics	Oconclusive Onot conclusive Onot done	
	liver	○Yes ○no amyloid detected ○no biopsy performed	
	date of biopsy	dd.mm.yyyy	
	congo red	Opositive Onegative Onot done	
	immunohistochemistry	Okappa Olambda Oaa Oattr Oother Onot conclusive Onot d	lone
	please specify		
	electron microscopy	○ conclusive ○ not conclusive ○ not done	
	ms-based proteomics	Oconclusive Onot conclusive Onot done	
	ms-baseu proteomics	Conclusive Office conclusive Office dolle	
	skin	○ Yes ○ no amyloid detected ○ no biopsy performed	
	date of biopsy	dd.mm.yyyy	
	congo red	Opositive Onegative Onot done	
	immunohistochemistry	Okappa Olambda Oaa Oattr Onot done Oother Onot conclu	sive
	please specify		
	electron microscopy	O conclusive O not conclusive O not done	
	ms-based proteomics	O conclusive O not conclusive O not done	
	nonvo	OVer One amulaid detected One bioness performed	
	nerve date of biopsy	O Yes O no amyloid detected O no biopsy performed	
	• •	dd.mm.yyyy	
	congo red	Opositive Onegative Onot done	
	immunohistochemistry	Okappa Olambda Oaa Oattr Oother Onot conclusive Onot d	lone
	please specify		
	electron microscopy	Oconclusive Onot conclusive Onot done	
	ms-based proteomics	O conclusive O not conclusive O not done	
	lung	○Yes ○no amyloid detected ○no biopsy performed	
	date of biopsy		
		dd.mm.yyyy	
	congo red	Opositive Onegative Onot done Okappa Olambda Oaa Oattr Oother Onot conclusive Onot d	lono
	immunohistochemistry	Okappa Olambua Olaa Olatti Ootner Onot conclusive Onot d	ione
	please specify		
	electron microscopy	Oconclusive Onot conclusive Onot done	
	ms-based proteomics	○ conclusive ○ not conclusive ○ not done	
	bone marrow	○ Yes ○ no amyloid detected ○ no biopsy performed	
	date of biopsy	dd.mm.yyyy	
	congo red	Opositive Onegative Onot done	
	immunohistochemistry	Okappa Olambda Oaa Oattr Oother Onot conclusive Onot d	lone
	please specify	Chappa Chambad Cad Cada Cada Charter Charter	
		Operation Operation Constitution	
	electron microscopy ms-based proteomics	O conclusive O not conclusive O not done O conclusive O not conclusive O not done	
	ma-baseu proteomitos		
	other	○ Yes ○ no amyloid detected ○ no biopsy performed	
specify	y other biopsy 1		
	affected tissue		
	date of biopsy	dd mm yaaay	
		dd.mm.yyyy	
	congo red	Opositive Onegative Onot done	Inot dono
	immunohistochemistry	○ kappa ○ lambda ○ aa ○ attr ○ other ○ not conclusive ○	not done
	please specify		
	electron microscopy	O conclusive O not conclusive O not done	
	ms-based proteomics	○ conclusive ○ not conclusive ○ not done	
More			
sympto	oms leading to diagnosis		
	heart failure	OYes ONo Onot known	
	syncope/-s	OYes ONo Onot known	
	stroke	O Yes O No O not known	
	atrial fibrillation	OYes ONo Onot known	
	edema	○Yes ○No ○not known	
	chronic kidney failure	OYes ONo Onot known	
	(creatinine clearance, nephrotic syndrome	•	Dogo 10 of 42
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	weight loss		○Yes ○No ○not	known			
	weight gain		○Yes ○No ○not	known			
	fatigue		OYes ONo Onot	known			
	•						
	bleeding diathesis		OYes ONo Onot	known			
	anemia		OYes ONo Onot				
	anemia		O res O No O not	KIIOWII			
			a a a .				
	carpal tunnel syndro		OYes ONo Onot				
	peripheral neuropath	ıy	○Yes ○No ○not	known			
	autonomic neuropath	hy	OYes ONo Onot	known			
	gastrointestinal disea	ase	OYes ONo Onot	known			
	(diarrhea, hepatomeg	galy, splenomegaly,					
	etc.)						
	pathologic fracture		OYes ONo Onot	known			
	osteolytic lesions		OYes ONo Onot	known			
	macroglossia		OYes ONo Onot				
		·	OYes ONo Onot				
	periorbital bleeding (	racoon-eyes)					
	other		OYes ONo Onot	known			
	fever		OYes ONo Onot	known			
	fatigue		OYes ONo Onot				
			O Yes O No O not				
	neuropathy						
	cardiac failure		OYes ONo Onot				
	chronic kidney disea	se	OYes ONo Onot				
	other		OYes ONo Onot	known			
other	symptom leading to dia	aanosis 1					
	please specify						
	_	y					
More							
sympt	toms of inflammation						
	abdominal pain / peri		OYes ONo Onot				
	inflammatory joint pa	ain	OYes ONo Onot				
	pleuritis and/or peric	arditis	OYes ONo Onot	known			
	production decision position	ai aitio	O 103 O 110 O 1101	KIIOWII			
	myalgia	ar arrio	OYes ONo Onot				
	•	ur unio		known			
	myalgia		○Yes ○No ○not	known known			
	myalgia rash / skin		OYes ONo Onot OYes ONo Onot	known known			
chron	myalgia rash / skin fever without any of t above		OYes ONo Onot OYes ONo Onot	known known			
chron	myalgia rash / skin fever without any of t		OYes ONo Onot OYes ONo Onot	known known			
	myalgia rash / skin fever without any of t above	the listed symptoms	OYes ONo Onot OYes ONo Onot	known known	date of diagnosis		
prese	myalgia rash / skin fever without any of t above ic infection	the listed symptoms	OYes ONo Onot OYes ONo Onot OYes ONo Onot	known known		dd.mm.yyyy	
prese	myalgia rash / skin fever without any of t above ic infection ence of chronic infection	the listed symptoms	OYes ONo Onot OYes ONo Onot OYes ONo Onot	known known		dd.mm.yyyy	
prese	myalgia rash / skin fever without any of t above ic infection ence of chronic infection	the listed symptoms	OYes ONo Onot OYes ONo Onot OYes ONo Onot	known known		dd.mm.yyyy	
prese O Yes	myalgia rash / skin fever without any of t above  ic infection ence of chronic infection s ○ No ○ not known	the listed symptoms	OYes ONo Onot OYes ONo Onot OYes ONo Onot	known known		dd.mm.yyyy	
prese O Yes	myalgia rash / skin fever without any of t above ic infection ence of chronic infection s ○ No ○ not known	the listed symptoms n pl	O Yes O No O not O Yes O No O not O Yes O No O not	known known known		dd.mm.yyyy	
prese O Yes	myalgia rash / skin fever without any of t above ic infection ence of chronic infection s ○ No ○ not known	the listed symptoms	O Yes O No O not O Yes O No O not O Yes O No O not	known known known		dd.mm.yyyy	
prese O Yes	myalgia rash / skin fever without any of t above  ic infection ence of chronic infection s ○ No ○ not known  ic inflammatory disease presence of chronic in	the listed symptoms  n pl  e  inflammatory disease	O Yes O No O not O Yes O No O not O Yes O No O not	known known known		dd.mm.yyyy	
prese O Yes	myalgia rash / skin fever without any of t above  ic infection ence of chronic infection s ○ No ○ not known  ic inflammatory disease presence of chronic i rheumatological dise	the listed symptoms  n pl  e  inflammatory disease	O Yes O No O not O Yes O No O not O Yes O No O not lease specify O Yes O No O not	known known known		dd.mm.yyyy	
prese O Yes	myalgia rash / skin fever without any of tabove  ic infection ence of chronic infection s	the listed symptoms  n pl  e  inflammatory disease	O Yes O No O not O Yes O No O not O Yes O No O not  lease specify  O Yes O No O not O Yes O No O not	known known known		dd.mm.yyyy	
prese O Yes	myalgia rash / skin fever without any of t above  ic infection ence of chronic infection s ○ No ○ not known  ic inflammatory disease presence of chronic i rheumatological dise	the listed symptoms  n pl  e  inflammatory disease	O Yes O No O not O Yes O No O not O Yes O No O not  lease specify  O Yes O No O not O Yes O No O not	known known known		dd.mm.yyyy	
prese O Yes	myalgia rash / skin fever without any of tabove  ic infection ence of chronic infection s	n pl	O Yes O No O not O Yes O No O not O Yes O No O not  lease specify  O Yes O No O not O Yes O No O not	known known known known known		dd.mm.yyyy	
prese O Yes	myalgia rash / skin fever without any of tabove  ic infection since of chronic infection since of chronic infection fic inflammatory disease presence of chronic infection rheumatological dise please specify date of diagnosis chronic inflammatory	n pl	O Yes O No O not O Yes O No O not O Yes O No O not  lease specify  O Yes O No O not O Yes O No O not	known known known known known		dd.mm.yyyy	
prese O Yes	myalgia rash / skin fever without any of tabove  ic infection ence of chronic infection s	n pl	O Yes O No O not O Yes O No O not O Yes O No O not  lease specify  O Yes O No O not O Yes O No O not O Yes O No O not	known known known known known d.mm.yyyy known		dd.mm.yyyy	
prese O Yes	myalgia rash / skin fever without any of tabove  ic infection since of chronic infection since of chronic infection fic inflammatory disease presence of chronic infection rheumatological dise please specify date of diagnosis chronic inflammatory	n pl	O Yes O No O not O Yes O No O not O Yes O No O not  lease specify  O Yes O No O not	known known known known known		dd.mm.yyyy	
prese O Yes	myalgia rash / skin fever without any of tabove  ic infection ence of chronic infection s	n pl	O Yes O No O not O Yes O No O not O Yes O No O not  lease specify  O Yes O No O not O Yes O No O not O Yes O No O not	known known known known known d.mm.yyyy known		dd.mm.yyyy	
prese O Yes	myalgia rash / skin fever without any of tabove  ic infection ence of chronic infection s	n pl	O Yes O No O not O Yes O No O not O Yes O No O not  lease specify  O Yes O No O not	known known known known known d.mm.yyyy known		dd.mm.yyyy	
prese O Yes	myalgia rash / skin fever without any of tabove  ic infection  ence of chronic infection s   No  ont known  ic inflammatory disease presence of chronic infection rheumatological dise please specify date of diagnosis chronic inflammatory please specify date of diagnosis other please specify	n pl	O Yes O No O not O Yes O No O not O Yes O No O not  lease specify  O Yes O No O not	known known known known known d.mm.yyyy known		dd.mm.yyyy	
prese O Yes	myalgia rash / skin fever without any of tabove  ic infection ence of chronic infection is O No O not known  ic inflammatory disease presence of chronic in rheumatological dise please specify date of diagnosis chronic inflammatory please specify date of diagnosis other	n pl	O Yes O No O not O Yes O No O not O Yes O No O not  lease specify  O Yes O No O not	known known known known known d.mm.yyyy known		dd.mm.yyyy	
prese ○ Yes chron	myalgia rash / skin fever without any of tabove  ic infection  ence of chronic infection s   No  ont known  ic inflammatory disease presence of chronic infection rheumatological dise please specify date of diagnosis chronic inflammatory please specify date of diagnosis other please specify	n pl	O Yes O No O not O Yes O No O not O Yes O No O not  lease specify  O Yes O No O not	known known known known known d.mm.yyyy known		dd.mm.yyyy	
prese O Yes chron	myalgia rash / skin fever without any of ta above  ic infection ence of chronic infection is O No O not known  ic inflammatory disease presence of chronic in rheumatological dise please specify date of diagnosis chronic inflammatory please specify date of diagnosis other please specify date of diagnosis other please specify date of diagnosis	n pl	O Yes O No O not O Yes O No O not O Yes O No O not  Dease specify  O Yes O No O not	known known known known known d.mm.yyyy known d.mm.yyyy			
prese O Yes  chron  hered prese	myalgia rash / skin fever without any of tabove  ic infection ence of chronic infection ic inflammatory disease presence of chronic in rheumatological dise please specify date of diagnosis chronic inflammatory please specify date of diagnosis other please specify date of diagnosis other please specify date of diagnosis itary periodic fever syntace of hereditary	n pl	O Yes O No O not O Yes O No O not O Yes O No O not  lease specify  O Yes O No O not	known known known known known d.mm.yyyy known d.mm.yyyy		number of attacks	
prese O Yes chron hered prese period	myalgia rash / skin fever without any of tabove  ic infection ence of chronic infection ic inflammatory disease presence of chronic in rheumatological dise please specify date of diagnosis chronic inflammatory please specify date of diagnosis other please specify date of diagnosis other please specify date of diagnosis itary periodic fever synthic fever synthic fever syndrome	n pl	O Yes O No O not O Yes O No O not O Yes O No O not  Dease specify  O Yes O No O not	known known known known known d.mm.yyyy known d.mm.yyyy	ate of diagnosis	number of attacks	
prese O Yes chron hered prese period	myalgia rash / skin fever without any of tabove  ic infection  ence of chronic infection ic inflammatory disease presence of chronic infection rheumatological dise please specify date of diagnosis chronic inflammatory please specify date of diagnosis other please specify date of diagnosis itary periodic fever syntemice of hereditary dic fever syndrome is ONO Onot	n pl	O Yes O No O not O Yes O No O not O Yes O No O not  Dease specify  O Yes O No O not	known known known known known d.mm.yyyy known d.mm.yyyy			
prese O Yes chron hered prese period	myalgia rash / skin fever without any of tabove  ic infection ence of chronic infection ic inflammatory disease presence of chronic in rheumatological dise please specify date of diagnosis chronic inflammatory please specify date of diagnosis other please specify date of diagnosis other please specify date of diagnosis itary periodic fever synthic fever synthic fever syndrome	n pl	O Yes O No O not O Yes O No O not O Yes O No O not  Dease specify  O Yes O No O not	known known known known known d.mm.yyyy known d.mm.yyyy	ate of diagnosis	number of attacks	
prese O Yes chron hered prese period	myalgia rash / skin fever without any of tabove  ic infection  ence of chronic infection ic inflammatory disease presence of chronic infection rheumatological dise please specify date of diagnosis chronic inflammatory please specify date of diagnosis other please specify date of diagnosis itary periodic fever syntemice of hereditary dic fever syndrome is ONO Onot	n pl	O Yes O No O not O Yes O No O not O Yes O No O not  Please specify  O Yes O No O not	known known known known known d.mm.yyyy known d.mm.yyyy	ate of diagnosis	number of attacks	
prese O Yes chron hered prese period	myalgia rash / skin fever without any of tabove  ic infection  ence of chronic infection ic inflammatory disease presence of chronic infection rheumatological dise please specify date of diagnosis chronic inflammatory please specify date of diagnosis other please specify date of diagnosis itary periodic fever syntemice of hereditary dic fever syndrome is ONO Onot	n pl	O Yes O No O not O Yes O No O not O Yes O No O not  Dease specify  O Yes O No O not	known known known known known d.mm.yyyy known d.mm.yyyy	ate of diagnosis	number of attacks	

	mutation analyse done p	lease specify mutation	family history of HPFS	please specify: affected family members / type of HPFS
	○ Yes ○ No ○ not known		○Yes ○No ○not known	
	Signature			
	Place, date	Signature		
	Possible entries			
	Please select one of the following entries	s for the corresponding items ma	arked above.	
1)	AL (immunoglobulin light chain amyloid	osis)		
	AA (serum amyloid a amyloidosis)			
	other (ATTR, AApo etc.)			
2)	not known (congo red positive, no further	er conclusive evaluations)		
	Aβ2M (β2-microglobulin, wild type)			
	Aβ2M (β2-microglobulin, variant)			
	ATTR (transthyretin, wild type; SSA)			
	ATTR V30M (transthyretin variant)			
	ATTR I111M (transthyretin variant)			
	ATTR V122I (transthyretin variant)			
	ATTR other mutation (transthyretin varia	ant)		
	AApoAI (Apolipoprotein A I, variants)			
	AApoAII (Apolipoprotein A II, variants)			
	AApoAIV (Apolipoprotein A IV, wild type	)		
	AGel (gelsolin, variants)			
	ALys (lysozyme, variants)			
	ALeuc2 (leukocyte chemotactic factor-2	<u>'.')</u>		
	AFib (fibrinogen α, variants)			
	ACys (cystatin c, variants)			
	ABri (ABriPP, variants)			
	other			
3)	FMF (familial mediterranean fever)			
	HIDS (hyperimmunoglobulinemia D with	n periodic fever syndrome)		
	TRAPS (TNF receptor-associated perio	odic syndrome)		
	MWS (muckle-wells syndrome)			
	FCAS (familial cold autoinflammatory sy	yndrome)		
	CINCA (chronic infantile neurologic cata	aneous articular syndrome)		

other



atient	
isit	
orm family	baseline

USZ-HAE-Amyloidosis registry (V1.01) Form laboratory values laboratory values baseline period: (do not fill in) data entry range from: until: paraprotein paraprotein examination performed? ○Yes ○No affected paraprotein affected paraprotein affected light chain immunofixation Opositive Onegative Onot done serum immunofixation dd.mm.yyyy date of immunofixation urine immunofixation Opositive Onegative Onot done date of immunofixation dd.mm.yyyy immunoglobulins serum m-gradient ○Yes ○No ○not known please specify not done urine bence jones protein Opositive Onegative Onot known please specify not done total IgG not done total IgM not done total IgA not done not done total IgD not done total IgE kU/I date of data collection dd.mm.yyyy total free light chain total free light chain lambda mg/l not done mg/l total free light chain kappa not done date of FLC data collection dd.mm.yyyy other laboratory values (clinical chemistry, immunology, hematology) clinical chemistry serum calcium mmol/l not done corrected serum calcium mmol/l not done CRP not done mg/dl not done SAA (serum amyloid a) eCRF Amyloidosis Registry V1.0.1 / 23.05.2018 date of data collection dd.mm.yyyy Page 22 of 43

immunolog	ıy				
	β-2 microglobulin	mg/l	not done □		
	date of data collection	dd.mm.yyyy			
		,,,,			
hematolog	v				
J	Hb	g/I	not done □		
	leucocytes	G/I	not done		
	platelets	G/I	not done		
	plasma cells	% in peripheral blood	not done □		
	neutrophiles	% in peripheral blood	not done □		
	lymphocytes	% in peripheral blood	not done □		
	date of data collection	dd.mm.yyyy			
		,,,,			
	Signature				
	Place, date	Signature			
	Possible entries				
	Please select one of the following	ng entries for the corresponding items	s marked above.		
1)	IgG				
	IgM				
	IgA				
	IgD				
	IgE				
	light chain only				
	none				
	not known				
2)	lambda				
,	kappa				
	none				
	not known				

	Date		Patient		
	Participant		Visit		
UniversitätsS <sub>I</sub>	pital Centre		Form	baseline	
<b>Zürich</b>	Project	USZ-HAE-Amyloidosis registry (V1.01)	family Form	medical history	
medical history		(* 1.01)	i Oiill	medical filstory	(V1.01)
family history					
	m/2				
amyloidosis in family histo ○ Yes ○ No ○ not known	ı y r				
please specify type of amy	loidosis	please speci	fy		
		1)			
affected relative 1					
please specify					
	2)				
More					
past / actual medical histor	y				
congestive cerebrove peripheral pulmonal deep verice other organ disorders  chronic peripheral pulmonal deep verice peripheral pulmonal deep verification of the patopa gastroint deep verification deep ver	rascular disease  rascular disease  ral arterial disease  ry embolism Oyes  nous thrombosis  oulmonary disease  athy Oyes ONo  restinal disease Oyes  ic disorder Oyes Onour Oyes ONo	Yes ONo Onot known ONo Onot known es ONo Onot known OYes ONo Onot known s ONo Onot known Onot known es ONo Onot known ONO Onot known			
endocrinology diabetes	mellitus O Yes O No	o ○ not known			
infectious disorders viral hep	atitis OYes ONo	○ not known			
human ir	mmunodeficiency viru	OYes ONo Onot			
	s disease ○Yes ○N	KNOWN			

	atologic disorders	
		bleeding diathesis O Yes O No O not known
		MGUS (monocl. gammopathy OYesONoOnot of unknown significance) known
		date of diagnosis dd.mm.yyyy
		multiple myeloma OYes ONo Onot known
		date of diagnosis dd.mm.yyyy
		lymphoplasmocytic lymphoma OYes No Onot known
		date of diagnosis dd.mm.yyyy
		other lymphoma O Yes O No O not known Please specify
		date of diagnosis dd.mm.yyyy
	other	
	○Yes ○No ○no	t known
	other past / actual	
	please spec	
	piodes oper	
	More	
	Signature	
	Place, date	Signature
	Possible entries	
	Please select one	of the following entries for the corresponding items marked above.
1)	Al (incres us a sila but	lin light chain amyloidosis)
''	AL (Immunodiobu	
',		
1)	AA (serum amyloi	d a amyloidosis)
.,		id a amyloidosis) tin amyloidosis)
',	AA (serum amyloi ATTR (transthyret	d a amyloidosis) tin amyloidosis) mic amyloidosis)
',	AA (serum amyloi ATTR (transthyret SSA (senile syste AFib (fibrinogen a	d a amyloidosis) tin amyloidosis) mic amyloidosis)
',	AA (serum amyloi ATTR (transthyret SSA (senile syste AFib (fibrinogen a	id a amyloidosis) tin amyloidosis) mic amyloidosis) imyloidosis)
	AA (serum amyloi ATTR (transthyret SSA (senile syste AFib (fibrinogen a AApoAl (apolipopi other	id a amyloidosis) tin amyloidosis) mic amyloidosis) imyloidosis)
2)	AA (serum amyloi ATTR (transthyret SSA (senile syste AFib (fibrinogen a AApoAl (apolipopi other	id a amyloidosis) tin amyloidosis) mic amyloidosis) imyloidosis)
	AA (serum amyloi ATTR (transthyret SSA (senile syste AFib (fibrinogen a AApoAl (apolipopi other mother father	id a amyloidosis) tin amyloidosis) mic amyloidosis) imyloidosis)
	AA (serum amyloi ATTR (transthyret SSA (senile syste AFib (fibrinogen a AApoAl (apolipopi other mother father sister	id a amyloidosis) tin amyloidosis) mic amyloidosis) imyloidosis)
	AA (serum amyloi ATTR (transthyret SSA (senile syste AFib (fibrinogen a AApoAl (apolipopi other mother father sister brother	id a amyloidosis) tin amyloidosis) mic amyloidosis) imyloidosis)
	AA (serum amyloi ATTR (transthyret SSA (senile syster AFib (fibrinogen a AApoAl (apolipoprother mother father sister brother daughter	id a amyloidosis) tin amyloidosis) mic amyloidosis) imyloidosis)
	AA (serum amyloi ATTR (transthyret SSA (senile syste AFib (fibrinogen a AApoAl (apolipopi other mother father sister brother daughter son	id a amyloidosis) tin amyloidosis) mic amyloidosis) imyloidosis)
	AA (serum amyloi ATTR (transthyret SSA (senile syste AFib (fibrinogen a AApoAl (apolipopi other mother father sister brother daughter son maternal aunt	id a amyloidosis) tin amyloidosis) mic amyloidosis) imyloidosis)
	AA (serum amyloi ATTR (transthyret SSA (senile syster AFib (fibrinogen a AApoAl (apolipopr other  mother father sister brother daughter son maternal aunt paternal aunt	id a amyloidosis) tin amyloidosis) mic amyloidosis) imyloidosis)
	AA (serum amyloi ATTR (transthyret SSA (senile syster AFib (fibrinogen a AApoAl (apolipoprother mother father sister brother daughter son maternal aunt paternal aunt	id a amyloidosis) tin amyloidosis) mic amyloidosis) imyloidosis)
	AA (serum amyloi ATTR (transthyret SSA (senile syster AFib (fibrinogen a AApoAl (apolipopi other  mother father sister brother daughter son maternal aunt paternal aunt maternal uncle	d a amyloidosis) tin amyloidosis) mic amyloidosis) myloidosis) rotein a1 amyloidosis)
	AA (serum amyloi ATTR (transthyret SSA (senile syste) AFib (fibrinogen a AApoAl (apolipop) other  mother father sister brother daughter son maternal aunt paternal aunt maternal uncle paternal uncle maternal grandmo	d a amyloidosis) tin amyloidosis) mic amyloidosis) myloidosis) rotein a1 amyloidosis)  other
	AA (serum amyloi ATTR (transthyret SSA (senile syste) AFib (fibrinogen a AApoAl (apolipop) other  mother father sister brother daughter son maternal aunt paternal aunt maternal uncle paternal uncle maternal grandmo	d a amyloidosis) tin amyloidosis) mic amyloidosis) myloidosis) rotein a1 amyloidosis)  other ther
	AA (serum amyloi ATTR (transthyret SSA (senile syste) AFib (fibrinogen a AApoAl (apolipop) other  mother father sister brother daughter son maternal aunt paternal aunt maternal uncle paternal uncle maternal grandmo	d a amyloidosis) tin amyloidosis) mic amyloidosis) myloidosis) rotein a1 amyloidosis)  other ther ther



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articipant	
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atient	
sit	
orm family	baseline

	Project US	SZ-HAE-Amyloidosis registry (V1.01)	Form	organ involvement baseline	
	organ involvement				(V1.01)
	definition of organ involvement: Gertz MA et	al. American Journal of Hematology 79:319	328 (2005)		
	(link: AJoH, definition of organ involvement)				
	<ul> <li>biopsy of affected organ demonstrates amyloi or</li> </ul>	d			
	biopsy at an alternate site demonstrates amyle	oid plus typical organ involvement			
	baseline period: (do not fill in)				
	data entry range from: until:				
	baseline period: (do not fill in)				
	data entry range from: until:				
	heart				
	heart involvement	NYHA	date of examination		atrial fibrillation
	○ Yes ○ No ○ not known	1)	dd.mm.yyyy		○Yes ○No ○not known
	involvement: mean septum thickness >12mm	(echocardiography), no other cardiac cause; co	nsensus criteria: Gertz et al. AJoH 79:319-328 (20	005) (link above: definition of organ involvem	nent)
		NYHA	date of examination		atrial fibrillation
	○ Yes ○ No ○ not known	1)	dd.mm.yyyy		○Yes ○No ○not known
vit	tal parameters	not done	blood parameters	not done	
	heart rate bpm		2)	ng/l (=pg/ml)	
	ppiii			Hg/I (−pg/III)	
	blood pressure	3)	troponin T	µg/l □	
	standing position: systolic /		date of data collection	dd.mm.yyyy	
	standing position: diastolic mmHg				
	sitting / lying position: systolic /				
	sitting / lying position: diastolic mmHg				
	date of measurement dd.mm.yy	VVV			
	auto or modeli ometri.	,,,			
	transthoracic echocardiogram				
	echocardiography (TTE) performed?	date of examination			
	○Yes ○No ○not known	mm.yyyy			
	left ventricle / left atrium not done	diastolic function	not done right	ventricle not done	
				○ Yes	
	LVEF biplan %	E/e'		ight ventricular O No	
	LVLI DIPIAII 70 LJ			not vypertrophy O not known	Page 26 of 43
				KIIOWIII	

EDVI ml/m²		diastolic dysfunction	4)			ΔPsyst RV/RA	mmHg 🗆	
LVMMI g/m²						0		
rTh						pericardial O Yes effusion O not kr	nown	
septum thickness mm		speckle tracking				O HOUR		
○ Yes left atrial ○ No		apical sparing	○ Yes					
dilatation ⊜ not known		арісаі эраппу	O not known					
	m² □	global peak longitudinal st	rain %	not done				
LAVI ml/m²								
further investigations of	the heart (MRI / right heart	catheter)						
MRI performed?	are neart (man, ngm neart		nathologic late gade	olinium enhancement		date	of performance	
O Yes		(	O Yes				dd.mm.yyyy	
○ No ○ not known			○ No ○ not known					
						01/5		
right-heart catheter performed?	mean wedge pressure	not done	mean pulmo	nal pressure not done		CVP	not done	date of performance
O Yes	mmHg		mmHg	, 🗆		mmHg		dd.mm.yyyy
○ No ○ not known								
kidney								
involvement: 24h urine pr O Yes O No O not knowr	otein > 0.5g/day, predominar า	ntly albumin. Gertz et al. <i>i</i>	AJoH 79:319-328 (20	005). (link above: definition	on of organ involv	vement)		
renal involvement ○ Yes ○ No ○ not knowr	ı							
renal replacement OY therapy ON	o please specify	5)						
On		dd.mm.yyyy						
N.								
serum parameters	not done	mo	rning urine		not done	2	4h urine	not done
serum creatinine µm	ol/l		urine creatinine	mmol/l			24h urine creatinine r	nmol/d
serum urea mmol/l			urine protein	g/l			24h urine protein	g/d 🗆
serum albumin g/l			urine albumin	mg/l			24h urine albumin	mg/d
eGFR ml/min/1	.73m²						collected volume	ml $\square$
6	5)							
data of			doto@fic A		00.05.0040		data of	
date of data collection	dd.mm.yyyy		data collection	losis Registry V1.0.1	23.05.2018		date of data collection	<sub>dd.mm.yyy</sub> Page 27 of 43

r	renal sonography performed? OYes ONo Onot known length diameter right ki	dney cm not done
	date of performance dd.mm.yyyy length diameter left kid	ney cm not done
d	digestive system	
_	gastrointestinal involvement  O Yes O No O not known	
	involvement: total liver span >15cm in the absence of heart failure or alkaline phosphatase >1.5 times institutiona  O Yes O No O not known	upper limit of normal. Gertz et al. AJoH 79:319-328 (2005). (link above: definition of organ involvement)
	symptoms  constipation OYes ONo O not known  diarrhea OYes ONo O not known  malabsorption OYes ONoO not known	
so	sonography	
	Iver size   O normal   max. diameter   cm   ascites	○Yes ○No ○not known
	liver parameters  alkaline phosphatase U/I not done upper limit of normal U/I not known date of data collection dd.mm.yyyy	
n	nerve system	
	nerve system involvement  O Yes O No O not known	
ро ав <b>р</b>	involvement:  peripheral: clinical; symmetric lower extremity sensorimotor peripheral neuropathy  autonomic: gastric-emptying disorder, pseudo-obstruction, voiding dysfunction not related to direct organ infiltration  peripheral sensoric  O Yes O No O not known  O Yes O No O not known	. Gertz et al. AJoH 79:319-328 (2005). (link above: definition of organ involvement)  autonomic  ○ Yes ○ No ○ not known

**ENMG** 

NIS-LL

eCRF Amyloidosis Registry V1.0.1 / 23.05.2018

Bril V, Eur Neurol 41 (Suppl. 1):8–13, 1999 not done

FAP-scale (TTR-amyloidosis)

Planté-Bordeneuve. I Neurol Planté-Bordeneuve, J Neurol (2014) 261: 1227-1233

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Iung  Iung involvement  Yes No not known involvement: interstitial radio Yes No not known pulmonary radiography interstitial radiographic	OVec ONe Onet	muscle power grading total sensory/reflex activity grading date of performance  . AJoH 79:319-328 (2005). (link at	pints mm.yyyy	organ involvement)	FAP-scale  date of performance	mm.yyyy	
function test  FVCex I  FEV1 I  SO2 (pulsoxymetry) date of pulmonary function test	know  dd.mm.yyyy	not done					
soft tissue							
soft tissue involvement O Yes O No O not known							
involvement: if any of the following Yes ONo Onot known	owing listed symptoms are	positive. Gertz et al. AJoH 79:319	0-328 (2005). (link	above: definition of organ involved	rement)		
arthropat claudicat skin myopath lymph no	al bleeding (racoon eyes)	OYesONoOnot known t known t known t known es O No O not known					
coagulation disorders							
coagulation disorders ○ Yes ○ No ○ not known							

	fa	actor VII deficien	cy OYes ONo Onot known						
			s ONo Onot known						
	h	yperfibrinolysis (	O No O not known						
	а	cquired platelet	unction disorder OYesONoOnot known						
	0	ther (	Yes ONo Onot known						
	р	lease specify							
	Signature								
	Place, date		Signature						
	Possible entries								
	Please select one of the fo	llowing entries for	r the corresponding items marked above.						
1)	not known								
-	no dyspnea								
	1								
	II								
	III								
	IV								
2)	NT-pro BNP								
	BNP								
3)	sitting / lying position								
	standing position								
	both performed								
	not done								
4)	normal								
	grade I - relaxation dysfur	nction							
	grade II - pseudonormal								
	grade III - restrictive								
	not known								
5)	hemodialysis								
	peritoneal dialysis								
	high cut-off dialysis								
6)	CDK-EPI								
	MDRD								
7)	axonal degeneration								
	demyelination								
	axonal degeneration + de	myelination							
	conduction block								
	normal		eCRF Amyloidosis Registry V1.0.1 / 23.05.2018	Page 30 of 43					

not known

8)	stage I
	stage II
	stage III
	not known



Patient	
Visit	
Form family	follow up
Form	organ response follow up

	P	roject USZ-HAE	-Amyloidosis registry (V1.01)		Form organ response follow up	
	organ response					(V1.01)
	data entry range any time since last follow up, nea always indicate date of data colle	arest to actual date ection!	e of follow up.			
	Follow up period: (do not fill in)					
	data entry range from:	until:				
	dd.mm.yyyy		dd.mm.yyyy			
	response criteria: Gertz MA et a	ıl. AJoH, 79:319-32	<u>8 (2005)</u>			
	heart					
vita	l parameters	ne	ot done	blood para	ameters not done	
	heart rate bpm		]		1) ng/l (=pg/ml)	
	blood pressure	2)		troponii	n T µg/l □	
	standing position: systolic /			date of	data collection dd.mm.yyyy	
		mHg				
	sitting / lying position: systolic	1		NYHA		
	sitting / lying position: diastolic	mmHg			3)	
	date of data collection	dd.mm.yyyy		date of	examination dd.mm.yyyy	
	transthoracic echocardiogram					
	echocardiography (TTE)					
	performed? O Yes O No O	not known				
	date of examination					
	left ventricle / left atrium	not done	diastolic function	not done	right ventricle	
					○ Yes	
	LVEF biplan %		E/e'		right ventricular O No	
	Evel biplan		2.0		hypertrophy O not known	
	EDVI ml/m²		diastolic dysfunction	4)	ΔPsyst RV/RA mmHg	
	LVMMI g/m²				0	
	rTh				pericardial O Yes effusion O No	
			amaalda		o not known	
	septum thickness mm		speckle tracking			
	left atrial OYesONoOnot		○ Yes	not done		
	dilatation know		apical sparing? ○ No ○ ng†kagwn		V1.0.1 / 23.05.2018	Page 32 of 43
	LA ESDI cm/m²		global peak 0/.		V 1.0.17 23.03.2010	Faye 32 01 43
		_	longitudinal strain /6	<del>-</del>		

LAVI	ml/m²					
kidney						
renal replace therapy	ement O Yes O No O not known start of dialysis	dd.mm.yyyy				
serum para	meters	not done	morning urine	not done	24h urine	not done
	reatinine µmol/l		urine creatinine mmol/l		24h urine creatinine mmol/d	
serum u serum a eGFR			urine protein g/l urine albumin mg/l		24h urine protein g/n 24h urine albumin mg collected volume ml	
	6)	_				_
date of data col	lection dd.mm.yyyy		date of data collection	dd.mm.yyyy	date of data collection	dd.mm.yyyy
perform			th diameter right kidney cm _not do			
liver						
upper li	phosphatase U/I mit of normal U/I mit of normal U/I	not done ☐ not known ☐ not known ☐				
nervous syst	em response and progression					
<b>ENMG</b> perfor	med? ○Yes ○No ○not known	NIS-LL <u>Bril V, Eur Neuro</u>	<u>l 41 (Suppl. 1):8–13, 1999</u> not done	FAP-scale (TTR-am <u>Planté-Borden</u>	nyloidosis) euve, J Neurol (2014) 261: 1227-1233	
result	7)	muscle power grading sensory/reflex activity grading	total points	FAP-scale	8)	
since date o	ges in result glast follow up 9)  of mance mm.yyyy	date of performance	mm.yyyy	date of performance	mm.yyyy	
perior						

○Yes ○No ○not known			
		not done	
immunofixation			
serum immunofixation Opositive C	negative		
urine immunofixation Opositive C	negative		
date of immunofixation	dd.mm.yyyy		
total free light chain			
total free light chain kappa	g / I		
total free light chain lambda	g / I		
date of FLC data collection	dd.mm.yyyy		
m-gradient / bone marrow			
serum M-gradient g / I			
bone marrow examination performed	□ yes		
bone marrow plasma cells %			
date of examination	dd.mm.yyyy		
hematologic data have to be entered in the n	next form "treatment since last follow up"		
laboratory values			
	not done		
Hb g/l			
leucocytes G/I			
platelets G/I			
plasma cells  % in peripheral blood			
neutrophiles  % in peripheral blood			
lymphocytes  % in peripheral blood			
date of dd.mm.yyyy	,		
data collection dd.nin.yyyy			
CRP mg/l			
SAA (Serum amylolu a)    mg/l			
date of			
date of data collection dd.mm.yyyy			
date of	/		
date of data collection dd.mm.yyyy	not done		
date of data collection dd.mm.yyyy  physical examination  weight kg	/		
date of data collection dd.mm.yyyy	not done		

paraprotein examination performed

fever attacks

	○ Yes since last follow up ○ No		
	○ not known number of attacks per year /year		
	symptoms of inflammation		
	symptoms of inflammation		
	abdominal pain / peritonitis		
	joint pain OYes ONo Onot known		
	Plausitia / pasiagaditia O Yes O No O not		
	myalgia OYes ONo Onot known rash / skin OYes ONo Onot known		
	rash / skin O Yes O No O not known other O Yes O No O not known		
	please specify		
	Signature		
	Place, date S	inature	
	Flace, date	naure	
	Possible entries		
	Please select one of the following entries for the cor	esponding items marked above.	
1)	BNP		
	NT-pro BNP		
2)	sitting / lying position		
-,	standing position		
	both performed		
	not done		
•			
3)	not known		
	no dyspnea		
	II		
	III		
	IV		
4)			
٠,	normal grade I - relaxation dysfunction		
	grade II - pseudonormal		
	grade III - restrictive		
	not known		
5)	homodialyais		
٠,	hemodialysis peritoneal dialysis		
	high cut-off dialysis		
6)	CDK-EPI	eCRF Amyloidosis Registry V1 0 1 / 23 05 2018	Page 35 of 43

MDRD

7)	axonal degeneration
	demyelination
	axonal degeneration + demyelination
	conduction block
	normal
	not known
8)	stage I
	stage II
	stage III
	not known
9)	improved
	worsened
	no change

not known

			Date				
			Participant			Patient	
1111/2	Univers	itätsSpital	Centre			Form	Centre
	Zürich	•	Project	USZ-HAE	-Amyloidosis registry	family	
				(V1.01)		Form	Participants and roles - Productive
	Participants ar	d roles					(V1.01)
	Please note: This	form is only filled in t	for the PRO	DUCTIVE	area. Data entry will be	done by th	e CTC DM.
Partic	cipants at center						
No. G	Given name	Surname	Role	- 1	First Access to product	ve With	drawn? Access until
1					dd.mm.y	уууу 🔲	dd.mm.yyyy
More	e						
	Signature						
	Place, date			Signature	<u> </u>		7



Date	
Participant	
Centre	
Project	USZ-HAE-Amyloidosis registry (V1.01)

Patient	
Form family	Centre
Form	Participants and roles - Setup

Participants and roles	
Please note: This form is only filled in for the SETLIP area	Data entry will be don

(V1.01)

Flease hote. This form is only filled in for the SETOF area. Data entry will be done by the CTC Divi.					
Center					
Registered (by CTC)	dd.mm.yyyy				
Hospital / Organisation Department City					
Language menus in secuTrial (standard for all participants) Release for productive = enter study data?	○ Deutsch ○ Englisch ○ Französisch ○ Spanisch ○ Automatisch durch CTC DM ○ Initial durch Sponsor				
Principal Investigator					
Current PI of center Salutation Given name Surname E-Mail PI has changed?	OMr OMs				
Previous PI					
No. Salutation Given name  1 OMr OMs	Surname	PI until dd.mm.yyyy			

#### Participants and Qualification

#### Please note:

secuTrial has two different areas to enter data: Setup = training area, Productive = enter study data.

Befor you get access to Productive area you need to qualify.

Necessary steps of qualification depend on your role and the privileges assigned to this role. For each participant the qualification steps are shown below.

### Training / RUS-Log

The objective is to get to know secuTrial and all relevant functionalities for data entry and validation.

#### Participants at USZ / UZH

- A personal training is mandatory. Please contact us for a training session.
- You will receive a training record for documentation with the TMF.

#### Participants at external centers

- We will send you the manual for secuTrial and provide access to the Setup area.
- You need to confirm self study of the manual using a RUS-Log
- The RUS-Log needs to be signed by you and the PI
- Put the signed form in the TMF
- Send a signed copy to CTC DM

### **Testpatient**

The objective is to get accustomed to the eCRF at hand.

- Enter a test patient in the setup area.
- Enter all visits according to protocol
- Fill in each form at least once; note that there are different index tabs
- Enter an AE and SAE
- Fill in the AE and SAE forms completely
- Enter a follow up for the AE / SAE
- Send the pseudonym of your test patient to the CTC DM

Participant

1 More		
	Signature	
	Place, date Signature	
	Possible entries	
	Please select one of the following entries for the corresponding items marked above.	
1)	No qualification necessary	
	Training / RUS-Log	
	Training / RUS-Log + Test patient	

W	UniversitätsSpita Zürich
	Premature Study End
	Please fill in the form 'Study End'
	Course
	Premature Study End
	Date of premature study en
	Withdrawal of informed consent
	Please note: If the patient has withdrawn consent
	Has the patient withdrawn informed consent?
	Date of withdrawal
	Reason for discontinuation

Origin

Univers	itätsS	pital
Zürich		

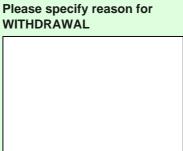
	Date					
	Participant			Patient		
<b>UniversitätsSpital</b>	Centre			Form	Study End	
Zürich	Project		nyloidosis registry	family	-	
		(V1.01)		Form	Premature Study End	04)
nature Study End					(V1	.01)
se fill in the form 'Study End' fir	rst!					
se						
ature Study End						
Date of premature study end			dd.mm.yyyy			
drawal of informed consent			-			
e <u>e note:</u> patient has withdrawn consent th	ne forms sho	ould be locked	d. For this use the it	ems below.		
Has the patient withdrawn th informed consent?	e					
Date of withdrawal			dd.mm.yyyy			
on for discontinuation						
n						
Cause for discontinuation lie		Patient				
with		Study team				
		Authorities other				
If OTHER: Please explain						]
ner explanation for premature to	ermination					7
se explain the reasons for discont			ly)			
	,		Applies			
h of the patient						
ocol violation / inadequate com ocol	npliance wi	th				
ision / Exclusion criteria are no	longer me	et.				
drawal of informed consent son given)						
drawal of informed consent			_			

### Further explanation for premature

Please explain the reasons for disco

Death of the patient	
Protocol violation / inadequate compliance with protocol	
Inclusion / Exclusion criteria are no longer met.	
Withdrawal of informed consent (reason given)	
Withdrawal of informed consent (no reason given)	
Lost to Follow Up	
Other reason	

C	Comment (optional)				
Γ					



Please specify OTHER REASC	)[

# **Exclusion**

Please note: If the checkbox 'Lock patient' is clicked, the forms for this patient will be locked!

The locking can be undone by unselecting the Lock patient, no further data entry	checkox and saving the form. This will be documented in the audit trail.
Signature and Confirmation	
Confirmation of the investigator (Prüfperso With my electronic signature (username and p this CRF.  Date  Name of investigator (Prüfperson)	assword) I confirm the <b>completeness</b> and <b>correctness</b> of the data entered in dd.mm.yyyy
Signature  Date (handwritten after printout)	Signature (handwritten after printout)
Please <b>print out</b> this form, <b>sign</b> it and	d put it in the <b>patient folder</b> .
Signature	
Place, date	Signature



Date			
Participant		Patient	
Centre		Form	Study End
Project	USZ-HAE-Amyloidosis registry	family	
	(V1.01)	Form	Study End

l ladice valdäde Caldal	0 1				
UniversitätsSpital	Centre		Form	Study End	
Zürich	Project	USZ-HAE-Amyloidosis registry		a = .	
- · · - ·		(V1.01)	Form	Study End	
Study End					(V1.01)
Study End					
Please give the date of the last study v With premature study end, please give			ppropriate form	ı <b>.</b>	
Has the patient completed the study	accord	ling to protocol? Date of I	ast study visit	conducted	
○ Yes			dd.mn	1.уууу	
○ No					
Signature and Confirmation					
Confirmation of the investigator (Pri	ifperso	n)			
With my electronic signature (usernam this CRF.	e and pa	assword) I confirm the <b>complete</b>	eness and corr	ectness of the dat	a entered in
Date	Г	dd.mm.yyyy			
Name of investigator (Prüfper	rson)				
Signature					
Date (handwritten after printout)		Signature (ha	ndwritten afte	r printout)	
Date (name miles and printedly		Oignaturo (na		, printedly	
Please print out this form, sign	it and	put it in the patient folde	r.		
Signature					
Place, date		Signature			
1 1000, 0010		5.9.744010			

# **Explanation of items in the Header:**

• Date:

Date the form is filled in [dd.mm.yyyy]

• Participant:

Member of the study team (e.g. study nurse) filling in the form.

• Centre:

Centre where the patient is registered.

Project

Internal name of the project in SecuTrial

• Patient:

Pseudonym of the patient according to Patient Identification Log

• Visit:

Name of the visit for which the form is filled in according to visit plan.

Adverse Event:

For each patient the adverse events will be labeled with a unique number

• Form family:

Designation of the group of forms the current form belongs to.

• Formular:

Title of the form.

# **Explanation of items in the footer:**

• Place, Date:

Where and when was the form filled in.

Signature:

Signature of the participant filling in the form. States correctness and completeness of the data.