UniversitätsSpital Zürich

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

Version: Released **Release:** 23.05.2018 - 11:51:29 (CEST) **By:** Poppe, Monika, Dr. <popmo_us2>
Reason: New user management

Vers. label: V1.01 **Vers. no.:** 2 **Last vers. modification:** 23.05.2018 - 11:51:29 (CEST)

Used catalogues (0)

Form families (7)

Displayed order	Form family name	Type	Hidden
10	- Sub Casenode	Subform	
	Sub Medical History <emnpus229_cd> Sub Concomitant Treatment and Medication <emnpus229_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> sub complications <emnpus229_sub_compl_al> coagulation disorders <emnpus229_coagul_dis> please specify <emnpus229_specify> specify other biopsy <emnpus229_spec_biopsy> specify other symptoms leading to diagnosis <emnpus229specify_sltd> specify affected relative(s) <emnpus229_affected_relat> specify other past medical history <emnpus229spec_pmh>		
9	- baseline	Visit	
	diagnosis (FILL IN / SAVE FIRST!) <mnpus229_diagnosis> demographics <mnpus229_demographics> medical history <mnpus229_med_history> organ involvement baseline <mnpus229_organ_involv> bone marrow <mnpus229_bmarrow_st> laboratory values <mnpus229_laboratory>		
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 <mnpus229_treatment> AA: treatment since last follow-up / study inclusion <mnpus229_aa_treatment> ATTR / other: treatment since study inclusion / last follow-up <mnpus229_attr_treatment>		
20	- Study End	Casenode	
	Study End <mnpus229_end> Premature Study End <mnpus229_end_prem>		
10	- Subform centre	Centre Subform	

		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>	

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secuTrial® 5.3.0.15, 2018

Project	USZ-HAE-Amyloidosis registry								
				Treatment arm 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
			Type	flexible	flexible	flexible	flexible	flexible	flexible
			possible deviation in days						
Visit plan				**Visit plan tab**					
baseline									
	diagnosis (FILL IN / SAVE FIRST!)	**mnpus229_diagnosis, emnpus229_spec_biopsy, emnpus229specify_sltd**		**X**					
	demographics	**mnpus229_demographics**		**X**					
	medical history	**mnpus229_med_history, emnpus229_affected_relat, emnpus229spec_pmh**		**X**					
	organ involvement baseline	**mnpus229_organ_involv**		**X**					
	bone marrow	**mnpus229_bmarrow_st**		**X**					
	laboratory values	**mnpus229_laboratory**		**X**					
follow up									
	condition of the patient	**mnpus229_condition, emnpus229_sub_compl_al**			**X**	**X**	**X**	**X**	**X**
	organ response follow up	**mnpus229_resp_criteria**			**X**	**X**	**X**	**X**	**X**
	AL: treatment since registry inclusion / last follow up	**mnpus229_treatment, emnpus229_sub_therapy**			**X**	**X**	**X**	**X**	**X**
	AA: treatment since last follow-up / study inclusion	**mnpus229_aa_treatment, emnpus229_sub_aa_therapy**							
	ATTR / other: treatment since study inclusion / last follow-up	**mnpus229_attr_treatment, emnpus229_sub_attr_thrpy**							

Family	Form	Table(s)		Treatment arm AA follow up					
									AA follow-up
									Created visits: 1
				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
			Type	flexible	flexible	flexible	flexible	flexible	flexible
			possible deviation in days						
Visit plan				**Visit plan tab**					
baseline									
	diagnosis (FILL IN / SAVE FIRST!)	**mnpus229_diagnosis, emnpus229_spec_biopsy, emnpus229specify_sltd**		**X**					
	demographics	**mnpus229_demographics**		**X**					
	medical history	**mnpus229_med_history, emnpus229_affected_relat, emnpus229spec_pmh**		**X**					
	organ involvement baseline	**mnpus229_organ_involv**		**X**					
	bone marrow	**mnpus229_bmarrow_st**		**X**					
	laboratory values	**mnpus229_laboratory**		**X**					
follow up									
	condition of the patient	**mnpus229_condition, emnpus229_sub_compl_al**			**X**	**X**	**X**	**X**	**X**
	organ response follow up	**mnpus229_resp_criteria**			**X**	**X**	**X**	**X**	**X**
	AL: treatment since registry inclusion / last follow up	**mnpus229_treatment, emnpus229_sub_therapy**							
	AA: treatment since last follow-up / study inclusion	**mnpus229_aa_treatment, emnpus229_sub_aa_therapy**			**X**	**X**	**X**	**X**	**X**
	ATTR / other: treatment since study inclusion / last follow-up	**mnpus229_attr_treatment, emnpus229_sub_attr_thrpy**							

Family	Form	Table(s)		Treatment arm ATTR / other follow up					
									ATTR / other
									Created visits: 1
				baseline	1. ATTR / other follow up	2. ATTR / other follow-up	3. ATTR / other follow-up	4. ATTR / other follow-up	5. ATTR / other 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 plan tab**					
baseline									
	diagnosis (FILL IN / SAVE FIRST!)	**mnpus229_diagnosis, emnpus229_spec_biopsy, emnpus229specify_sltd**		**X**					
	demographics	**mnpus229_demographics**		**X**					
	medical history	**mnpus229_med_history, emnpus229_affected_relat, emnpus229spec_pmh**		**X**					
	organ involvement baseline	**mnpus229_organ_involv**		**X**					
	bone marrow	**mnpus229_bmarrow_st**		**X**					
	laboratory values	**mnpus229_laboratory**		**X**					
follow up									
	condition of the patient	**mnpus229_condition, emnpus229_sub_compl_al**			**X**	**X**	**X**	**X**	**X**
	organ response follow up	**mnpus229_resp_criteria**			**X**	**X**	**X**	**X**	**X**
	AL: treatment since registry inclusion / last follow up	**mnpus229_treatment, emnpus229_sub_therapy**							
	AA: treatment since last follow-up / study inclusion	**mnpus229_aa_treatment, emnpus229_sub_aa_therapy**							
	ATTR / other: treatment since study inclusion / last follow-up	**mnpus229_attr_treatment, emnpus229_sub_attr_thrpy**			**X**	**X**	**X**	**X**	**X**

Study End			**Casenode tab**
	Study End	**mnpus229_end**	
	Premature Study End	**mnpus229_end_prem**	

Centre			**Centre tab**
	Participants and roles - Setup	**mnpus229_prt_stp, emnpus229_pi, emnpus229_prt_stp**	
	Participants and roles - Productive	**mnpus229_prt_prd, emnpus229_prt_prd**	

AA: treatment since last follow-up / study inclusion (V1.01)

Follow up period: (do not fill in)

data entry range from: dd.mm.yyyy **until:** dd.mm.yyyy

treatment

treatment since last follow up?
○ Yes ○ No

Use 'weitere' / 'more' to state all relevant therapies in this period of time

therapy 1

○ pharmacological
○ transplantation

pharmacological therapy

treatment start dd.mm.yyyy
ongoing treatment ○ Yes ○ No
treatment end dd.mm.yyyy

disease modifying drugs

please specify

1)

transplantation

transplanted organ 2)
please specify
date of transplantation dd.mm.yyyy
pharmacological immunosuppression ○ Yes ○ No ○ not known

More

Signature

Place, date Signature

Possible entries

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

1)

colchicine
TNF-alpha blocker
IL-1 blocker
rituximab
tocilizumab
corticosteroids
NSAID
eprodisate
other

2)

heart
kidney
liver
other

treatment since registry inclusion / last follow up (V1.01)

Follow up period: (do not fill in)

data entry range from: dd.mm.yyyy **until:** dd.mm.yyyy

treatment

treatment since last follow up?
○ Yes ○ No

Use 'weitere' / 'more' to state all relevant chemotherapies in this period of time

therapy 1

○ pharmacological
○ transplantation

start of treatment dd.mm.yyyy
ongoing treatment ○ Yes ○ No
end of treatment dd.mm.yyyy
cycle numbers

chemotherapy		please specify		please specify		please specify		please specify		please specify	
1)	2)		3)		4)		5)		6)		

transplantation

transplanted organ 7)
please specify
date of transplantation dd.mm.yyyy
pharmacological immunosuppression ○ Yes ○ No ○ not known

hematologic response after therapy

hematologic response during this treatment

response criteria: Gertz MA et al. AJoH, 79:319-328 (2005)

		not done	date of data collection
remission status	8)		dd.mm.yyyy
serum immunofixation ○ positive ○ negative ○ not known			dd.mm.yyyy
urine immunofixation ○ positive ○ negative ○ not known			dd.mm.yyyy
total free light chain kappa	g / l	☐	dd.mm.yyyy
total free light chain lambda	g / l	☐	
serum M-gradient	g / l	☐	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)

		not done	date of data collection
remission status	8)		dd.mm.yyyy
serum immunofixation ○ positive ○ negative ○ not known			dd.mm.yyyy
urine immunofixation ○ positive ○ negative ○ not known			dd.mm.yyyy
total free light chain kappa	g / l	☐	

total free light chain lambda [ ] g / l ☐ [ ] dd.mm.yyyy

serum M-gradient [ ] g / l ☐ [ ] dd.mm.yyyy

bone marrow examination performed ☐ yes [ ] dd.mm.yyyy

bone marrow plasma cells [ ] %

Signature

Place, date [ ] Signature [ ]

Possible entries

Please select one of the following entries for the corresponding 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

3)

bortezomib - dexamethasone
bortezomib - melphalan - prednisone
bortezomib - cyclophosphamide - dexamethasone
bortezomib - lenalidomide - dexamethasone
other

4)

thalidomide - dexamethasone
other

5)

lenalidomide - dexamethasone
lenalidomide - melphalan - prednisone
other

6)

High-dose therapy and autologous HSCT (melphalan 100mg/m2)
High-dose therapy and autologous HSCT (melphalan 120mg/m2)
High-dose therapy and autologous HSCT (melphalan 140mg/m2)
High-dose therapy and autologous HSCT (melphalan 200mg/m2)
other

7)

heart
kidney
liver
allogeneic stem cell
other

8)

sCR
CR
VGPR
PR
SD
progression
not known

ATTR / other: treatment since study inclusion / last follow-up (V1.01)

Follow up period: (do not fill in)

data entry range from: dd.mm.yyyy **until:** dd.mm.yyyy

treatment

treatment since last follow up?
○ Yes ○ No

Use 'weitere' / 'more' to state all relevant therapies in this period of time

therapy 1

○ pharmacological
○ transplantation

pharmacological therapy

start of treatment dd.mm.yyyy
ongoing treatment ○ Yes ○ No
end of treatment dd.mm.yyyy

disease modifying drugs TTR

please specify

1)

transplantation

transplanted organ 2)
please specify
date of transplantation dd.mm.yyyy
pharmacological immunosuppression ○ Yes ○ No ○ not known

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

bone marrow (V1.01)

baseline period: (do not fill in)

data entry range from: **until:**

plasma cells

plasma cells in bone marrow

percentage in aspirate [ ] % in aspirate not done ☐

percentage in biopsy [ ] % in biopsy not done ☐

date of examination [ ] dd.mm.yyyy

further examinations

aberrant immunophenotype ○ Yes ○ No ○ not known

plasma cell clonality ○ Yes ○ No ○ not known [ ] 1)

FISH

FISH performed ○ Yes ○ No ○ not known

date of performance [ ] dd.mm.yyyy

FISH results

the result was [ ] 2)

hyperdiploid ○ Yes ○ No ○ not known

t(11;14) ○ Yes ○ No ○ not known

t(4;14) ○ Yes ○ No ○ not known

t(14;16) ○ Yes ○ No ○ not known

del17p ○ Yes ○ No ○ not known

gain 1q ○ Yes ○ No ○ not known

loss 1p ○ Yes ○ No ○ not known

del 13 ○ Yes ○ No ○ not

known

other ○ Yes ○ No ○ not known please specify

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

condition of the patient (V1.01)

Follow up period: (do not fill in)

data entry range from: dd.mm.yyyy **until:** dd.mm.yyyy

Is patient alive

is the patient alive? ○ Yes ○ No

date of death dd.mm.yyyy

info: please fill in, print and sign the form "study end"

cause of death 1)

please specify

death related to... 2)

please specify

complications leading to hospitalisation

complications since last follow up ○ Yes ○ No

complication 1

reason of hospitalisation 3)

please specify

More

quality of life

filled out questionnaire *"EQ-5D-3L"* ○ Yes ○ No

number of points

Signature

Place, date Signature

Possible entries

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

1)

infection
cardiac
renal
bleeding
other

2)

related to amyloidosis
treatment related

concomittant disease
other

3)

infection
cardiac
stroke
renal
bleeding
bone disease
other

demographics (V1.01)

demographic data

date of birth dd.mm.yyyy

Age Score

gender

- ○ female
- ○ male

body characteristics

baseline period: (do not fill in)

data entry range from: **until:**

not done

height cm ☐

weight kg ☐ date of weight measurement dd.mm.yyyy

Body Mass Index Score

quality of life

filled out questionnaire *"EQ-5D-3L"* ○ Yes ○ No ○ not known

number of points

date, questionnaire was filled out dd.mm.yyyy

Signature

Place, date Signature

diagnosis (V1.01)

informed consent

informed consent signed ○ Yes ○ No ○ not known

date informed consent signed dd.mm.yyyy

type of amyloidosis

type of amyloidosis 1)

please specify 2) please specify

date of diagnosis

date of diagnosis

dd.mm.yyyy

confirmed by biopsy

○ Yes ○ No

confirmed by genetic testing / genetic testing performed

○ Yes ○ No

date of genetic testing dd.mm.yyyy

confirmed by szintigraphy (99mTc-DPD)

○ Yes ○ No

date of szintigraphy dd.mm.yyyy

amyloid deposit in tissue biopsy

subcutaneous fat ○ Yes ○ no amyloid detected ○ no biopsy performed

date of biopsy dd.mm.yyyy

congo red ○ positive ○ negative ○ not done

immunohistochemistry ○ kappa ○ lambda ○ aa ○ attr ○ other ○ not conclusive ○ not done

please specify

electron microscopy ○ conclusive ○ not conclusive ○ not done

ms-based proteomics ○ conclusive ○ not conclusive ○ not done

heart ○ Yes ○ no amyloid detected ○ no biopsy performed

date of biopsy dd.mm.yyyy

congo red ○ positive ○ negative ○ not done

immunohistochemistry ○ kappa ○ lambda ○ aa ○ attr ○ other ○ not conclusive ○ not done

please specify

electron microscopy ○ conclusive ○ not conclusive ○ not done

ms-based proteomics ○ conclusive ○ not conclusive ○ not done

kidney ○ Yes ○ no amyloid detected ○ no biopsy performed

date of biopsy dd.mm.yyyy

congo red ○ positive ○ negative ○ not done

immunohistochemistry ○ kappa ○ lambda ○ aa ○ attr ○ other ○ not conclusive ○ not done

please specify

electron microscopy ○ conclusive ○ not conclusive ○ not done

ms-based proteomics ○ conclusive ○ not conclusive ○ not done

intestinal ○ Yes ○ no amyloid detected ○ no biopsy performed

date of biopsy dd.mm.yyyy

congo red ○ positive ○ negative ○ not done

immunohistochemistry kappa lambda aa attr other not conclusive not done

please specify

electron microscopy ○conclusive ○not conclusive ○not done

ms-based proteomics ○conclusive ○not conclusive ○not done

liver ○Yes ○no amyloid detected ○no biopsy performed

date of biopsy dd.mm.yyyy

congo red ○positive ○negative ○not done

immunohistochemistry ○kappa ○lambda ○aa ○attr ○other ○not conclusive ○not done

please specify

electron microscopy ○conclusive ○not conclusive ○not done

ms-based proteomics ○conclusive ○not conclusive ○not done

skin ○Yes ○no amyloid detected ○no biopsy performed

date of biopsy dd.mm.yyyy

congo red ○positive ○negative ○not done

immunohistochemistry ○kappa ○lambda ○aa ○attr ○not done ○other ○not conclusive

please specify

electron microscopy ○conclusive ○not conclusive ○not done

ms-based proteomics ○conclusive ○not conclusive ○not done

nerve ○Yes ○no amyloid detected ○no biopsy performed

date of biopsy dd.mm.yyyy

congo red ○positive ○negative ○not done

immunohistochemistry ○kappa ○lambda ○aa ○attr ○other ○not conclusive ○not done

please specify

electron microscopy ○conclusive ○not conclusive ○not done

ms-based proteomics ○conclusive ○not conclusive ○not done

lung ○Yes ○no amyloid detected ○no biopsy performed

date of biopsy dd.mm.yyyy

congo red ○positive ○negative ○not done

immunohistochemistry ○kappa ○lambda ○aa ○attr ○other ○not conclusive ○not done

please specify

electron microscopy ○conclusive ○not conclusive ○not 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 ○positive ○negative ○not done

immunohistochemistry ○kappa ○lambda ○aa ○attr ○other ○not conclusive ○not done

please specify

electron microscopy ○conclusive ○not conclusive ○not done

ms-based proteomics ○conclusive ○not conclusive ○not done

other ○Yes ○no amyloid detected ○no biopsy performed

specify other biopsy 1

affected tissue

date of biopsy dd.mm.yyyy

congo red ○positive ○negative ○not done

immunohistochemistry ○kappa ○lambda ○aa ○attr ○other ○not conclusive ○not done

please specify

electron microscopy ○conclusive ○not conclusive ○not done

ms-based proteomics ○conclusive ○not conclusive ○not done

More

symptoms leading to diagnosis

heart failure ○Yes ○No ○not known

syncope/-s ○Yes ○No ○not known

stroke ○Yes ○No ○not known

atrial fibrillation ○Yes ○No ○not known

edema ○Yes ○No ○not known

chronic kidney failure ○Yes ○No ○not known
(creatinine clearance, nephrotic syndrome)

weight loss	○ Yes ○ No ○ not known
weight gain	○ Yes ○ No ○ not known
fatigue	○ Yes ○ No ○ not known
bleeding diathesis	○ Yes ○ No ○ not known
anemia	○ Yes ○ No ○ not known
carpal tunnel syndrome	○ Yes ○ No ○ not known
peripheral neuropathy	○ Yes ○ No ○ not known
autonomic neuropathy	○ Yes ○ No ○ not known
gastrointestinal disease ***(diarrhea, hepatomegaly, splenomegaly, etc.)***	○ Yes ○ No ○ not known
pathologic fracture	○ Yes ○ No ○ not known
osteolytic lesions	○ Yes ○ No ○ not known
macroglossia	○ Yes ○ No ○ not known
periorbital bleeding (racoon-eyes)	○ Yes ○ No ○ not known
other	○ Yes ○ No ○ not known

fever	○ Yes ○ No ○ not known
fatigue	○ Yes ○ No ○ not known
neuropathy	○ Yes ○ No ○ not known
cardiac failure	○ Yes ○ No ○ not known
chronic kidney disease	○ Yes ○ No ○ not known
other	○ Yes ○ No ○ not known

other symptom leading to diagnosis 1

please specify

More

symptoms of inflammation

abdominal pain / peritonitis	○ Yes ○ No ○ not known
inflammatory joint pain	○ Yes ○ No ○ not known
pleuritis and/or pericarditis	○ Yes ○ No ○ not known
myalgia	○ Yes ○ No ○ not known
rash / skin	○ Yes ○ No ○ not known
fever without any of the listed symptoms above	○ Yes ○ No ○ not known

chronic infection

presence of chronic infection
○ Yes ○ No ○ not known

please specify

date of diagnosis
dd.mm.yyyy

chronic inflammatory disease

presence of chronic inflammatory disease	○ Yes ○ No ○ not known
rheumatological disease	○ Yes ○ No ○ not known
please specify	
date of diagnosis	dd.mm.yyyy
chronic inflammatory bowel disease	○ Yes ○ No ○ not known
please specify	
date of diagnosis	dd.mm.yyyy
other	○ Yes ○ No
please specify	
date of diagnosis	dd.mm.yyyy

hereditary periodic fever syndromes (HPFS)

presence of hereditary periodic fever syndrome
○ Yes ○ No ○ not known

3)

please specify

date of diagnosis
dd.mm.yyyy

number of attacks
/per year

mutation analyse done	please 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 for the corresponding items marked above.

1)

AL (immunoglobulin light chain amyloidosis)
AA (serum amyloid a amyloidosis)
other (ATTR, AApo etc.)

2)

not known (congo red positive, no further 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 variant)
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)
AFib (fibrinogen α, variants)
ACys (cystatin c, variants)
ABri (ABriPP, variants)
other

3)

FMF (familial mediterranean fever)
HIDS (hyperimmunoglobulinemia D with periodic fever syndrome)
TRAPS (TNF receptor-associated periodic syndrome)
MWS (muckle-wells syndrome)
FCAS (familial cold autoinflammatory syndrome)
CINCA (chronic infantile neurologic cataneous articular syndrome)
other

laboratory values (V1.01)

baseline period: (do not fill in)

data entry range from: **until:**

paraprotein

paraprotein examination performed?
○ Yes ○ No

affected paraprotein

affected paraprotein 1)
affected light chain 2)

immunofixation

serum immunofixation ○ positive ○ negative ○ not done
date of immunofixation dd.mm.yyyy
urine immunofixation ○ positive ○ negative ○ not done
date of immunofixation dd.mm.yyyy

immunoglobulins

serum m-gradient ○ Yes ○ No ○ not known
please specify g/l not done ☐
urine bence jones protein ○ positive ○ negative ○ not known
please specify g/l not done ☐
total IgG g/l not done ☐
total IgM g/l not done ☐
total IgA g/l not done ☐
total IgD g/l not done ☐
total IgE kU/l not done ☐
date of data collection dd.mm.yyyy

total free light chain

total free light chain lambda mg/l not done ☐
total free light chain kappa mg/l 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 mg/dl not done ☐
SAA (serum amyloid a) mg/l not done ☐
date of data collection dd.mm.yyyy

immunology

β-2 microglobulin	mg/l	not done ☐
date of data collection	dd.mm.yyyy	

hematology

Hb	g/l	not done ☐
leucocytes	G/l	not done ☐
platelets	G/l	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 entries for the corresponding items marked above.

1)

IgG
IgM
IgA
IgD
IgE
light chain only
none
not known

2)

lambda
kappa
none
not known

medical history (V1.01)

family history

amyloidosis in family history?
○ Yes ○ No ○ not known

please specify type of amyloidosis 1)

please specify

affected relative 1

please specify 2)

More

past / actual medical history

cardiovascular system

coronary heart disease ○ Yes ○ No ○ not known
congestive heart failure ○ Yes ○ No ○ not known
cerebrovascular disease ○ Yes ○ No ○ not known
peripheral arterial disease ○ Yes ○ No ○ not known
pulmonary embolism ○ Yes ○ No ○ not known
deep venous thrombosis ○ Yes ○ No ○ not known

other organ disorders

chronic pulmonary disease ○ Yes ○ No ○ not known
chronic kidney disease ○ Yes ○ No ○ not known
hepatopathy ○ Yes ○ No ○ not known
gastrointestinal disease ○ Yes ○ No ○ not known
neurologic disorder ○ Yes ○ No ○ not known
solid tumour ○ Yes ○ No ○ not known
please specify

endocrinology

diabetes mellitus ○ Yes ○ No ○ not known

infectious disorders

viral hepatitis ○ Yes ○ No ○ not known
human immunodeficiency virus ○ Yes ○ No ○ not known
infectious disease ○ Yes ○ No ○ not known

hematologic disorders

bleeding diathesis ○ Yes ○ No ○ not known

MGUS (monocl. gammopathy of unknown significance) ○ Yes ○ No ○ not known

date of diagnosis [ ] dd.mm.yyyy

multiple myeloma ○ Yes ○ No ○ not known

date of diagnosis [ ] dd.mm.yyyy

lymphoplasmocytic lymphoma ○ Yes ○ No ○ not known

date of diagnosis [ ] dd.mm.yyyy

other lymphoma ○ Yes ○ No ○ not known Please specify [ ]

date of diagnosis [ ] dd.mm.yyyy

other

○ Yes ○ No ○ not known

other past / actual medical history 1

please specify

[ ]

More

Signature

Place, date [ ] Signature [ ]

Possible entries

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

1)

AL (immunoglobulin light chain amyloidosis)
AA (serum amyloid a amyloidosis)
ATTR (transthyretin amyloidosis)
SSA (senile systemic amyloidosis)
AFib (fibrinogen amyloidosis)
AApoAI (apolipoprotein a1 amyloidosis)
other

2)

mother
father
sister
brother
daughter
son
maternal aunt
paternal aunt
maternal uncle
paternal uncle
maternal grandmother
paternal grandmother
maternal grandfather
paternal grandfather

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)

- biopsy of affected organ demonstrates amyloid

or

- biopsy at an alternate site demonstrates amyloid 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
○ Yes ○ No ○ not known

NYHA
1)

date of examination
dd.mm.yyyy

atrial fibrillation
○ Yes ○ No ○ not known

involvement: mean septum thickness >12mm (echocardiography), no other cardiac cause; consensus criteria: Gertz et al. AJoH 79:319-328 (2005) (link above: definition of organ involvement)

○ Yes ○ No ○ not known

NYHA
1)

date of examination
dd.mm.yyyy

atrial fibrillation
○ Yes ○ No ○ not known

vital parameters **not done** ☐

heart rate bpm

blood pressure 3)

standing position: systolic /

standing position: diastolic mmHg

sitting / lying position: systolic /

sitting / lying position: diastolic mmHg

date of measurement dd.mm.yyyy

blood parameters **not done** ☐

2) ng/l (=pg/ml) ☐

troponin T µg/l ☐

date of data collection dd.mm.yyyy

transthoracic echocardiogram

echocardiography (TTE) performed?
○ Yes ○ No ○ not known

date of examination
mm.yyyy

left ventricle / left atrium **not done** ☐

LVEF biplan % ☐

diastolic function **not done** ☐

E/e'

right ventricle **not done** ☐

right ventricular hypertrophy ○ Yes ○ No ○ not known

EDVI [ ] ml/m² ☐

LVMMI [ ] g/m² ☐

rTh [ ] ☐

septum thickness [ ] mm ☐

left atrial dilatation ○ Yes ○ No ○ not known

LA ESDI [ ] cm/m² ☐

LAVI [ ] ml/m² ☐

diastolic dysfunction [ ] 4)

speckle tracking

apical sparing? ○ Yes ○ No ○ not known

global peak longitudinal strain [ ] % not done ☐

ΔPsyst RV/RA [ ] mmHg ☐

pericardial effusion ○ Yes ○ No ○ not known

further investigations of the heart (MRI / right heart catheter)

MRI performed?
○ Yes ○ No ○ not known

pathologic late gadolinium enhancement
○ Yes ○ No ○ not known

date of performance
[ ] dd.mm.yyyy

right-heart catheter performed?	**mean wedge pressure**	**not done**	**mean pulmonal pressure**	**not done**	**CVP**	**not done**	**date of performance**
○ Yes ○ No ○ not known	[ ] mmHg	☐	[ ] mmHg	☐	[ ] mmHg	☐	[ ] dd.mm.yyyy

kidney

involvement: 24h urine protein > 0.5g/day, predominantly albumin. Gertz et al. AJoH 79:319-328 (2005). (link above: definition of organ involvement)

○ Yes ○ No ○ not known

renal involvement
○ Yes ○ No ○ not known

renal replacement therapy ○ Yes ○ No ○ not known

please specify [ ] 5)

start of dialysis [ ] dd.mm.yyyy

serum parameters	**not done**	**morning urine**	**not done**	**24h urine**	**not done**
serum creatinine [ ] µmol/l	☐	urine creatinine [ ] mmol/l	☐	24h urine creatinine [ ] mmol/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	☐
[ ] 6)					
date of data collection [ ] dd.mm.yyyy		date of data collection [ ] dd.mm.yyyy		date of data collection [ ] dd.mm.yyyy	

renal sonography performed? ○Yes ○No ○not known

date of performance ☐ dd.mm.yyyy

length diameter right kidney ☐ cm not done ☐

length diameter left kidney ☐ cm not done ☐

digestive system

gastrointestinal involvement

○Yes ○No ○not known

involvement: total liver span >15cm in the absence of heart failure or alkaline phosphatase >1.5 times institutional upper limit of normal. Gertz et al. AJoH 79:319-328 (2005). (link above: definition of organ involvement)

○Yes ○No ○not known

symptoms

constipation ○Yes ○No ○not known

diarrhea ○Yes ○No ○not known

malabsorption ○Yes ○No ○not known

sonography

			not known	
liver size	○ enlarged ○ normal ○ not known	max. diameter ☐ cm ☐		ascites ○Yes ○No ○not known
spleen size	○ enlarged ○ normal ○ not known	max. diameter ☐ cm ☐		

date of sonography performance ☐ dd.mm.yyyy

liver parameters

alkaline phosphatase ☐ U/l not done ☐

upper limit of normal ☐ U/l not known ☐

lower limit of normal ☐ U/l not known ☐

date of data collection ☐ dd.mm.yyyy

nerve system

nerve system involvement

○Yes ○No ○not known

involvement:

peripheral: clinical; symmetric lower extremity sensorimotor peripheral neuropathy

autonomic: gastric-emptying disorder, pseudo-obstruction, voiding dysfunction not related to direct organ infiltration. Gertz et al. AJoH 79:319-328 (2005). (link above: definition of organ involvement)

peripheral sensoric

○Yes ○No ○not known

peripheral motoric

○Yes ○No ○not known

autonomic

○Yes ○No ○not known

ENMG

performed? ○Yes ○No ○not known

NIS-LL

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

FAP-scale (TTR-amyloidosis)

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

result [ ] 7)
date of performance [ ] mm.yyyy

muscle power grading [ ] total points ☐
sensory/reflex activity grading [ ] total points ☐
date of performance [ ] mm.yyyy

FAP-scale [ ] 8)
date of performance [ ] mm.yyyy

lung

lung involvement
○ Yes ○ No ○ not known

involvement: interstitial radiographic pattern. Gertz et al. AJoH 79:319-328 (2005). (link above: definition of organ involvement)
○ Yes ○ No ○ not known

pulmonary radiography

interstitial radiographic pattern ○ Yes ○ No ○ not known

function test		**not done**
FVCex	[ ] l	☐
FEV_1	[ ] l	☐
SO_2 (pulsoxymetry)	[ ] %	☐

date of pulmonary function test [ ] dd.mm.yyyy

soft tissue

soft tissue involvement
○ Yes ○ No ○ not known

involvement: if any of the following listed symptoms are positive. Gertz et al. AJoH 79:319-328 (2005). (link above: definition of organ involvement)
○ Yes ○ No ○ not known

physical findings
tongue enlargement ○ Yes ○ No ○ not known
periorbital bleeding (racoon eyes) ○ Yes ○ No ○ not known
arthropathy ○ Yes ○ No ○ not known
claudicatio ○ Yes ○ No ○ not known
skin ○ Yes ○ No ○ not known
myopathy / pseudohypertrophy ○ Yes ○ No ○ not known
lymph node ○ Yes ○ No ○ not known
carpal tunnel syndrome ○ Yes ○ No ○ not known

coagulation disorders

coagulation disorders
○ Yes ○ No ○ not known

coagulation disorders factor X deficiency ○ Yes ○ No ○ not known

factor VII deficiency ○ Yes ○ No ○ not known

dysfibrinogenemia ○ Yes ○ No ○ not known

hyperfibrinolysis ○ Yes ○ No ○ not known

acquired platelet function disorder ○ Yes ○ No ○ not known

other ○ Yes ○ No ○ not known

please specify

Signature

Place, date

Signature

Possible entries

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

1)

not known
no dyspnea
I
II
III
IV

2)

NT-pro BNP
BNP

3)

sitting / lying position
standing position
both performed
not done

4)

normal
grade I - relaxation dysfunction
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 + demyelination
conduction block
normal
not known

8)

stage I
stage II
stage III
not known

organ response (V1.01)

data entry range
any time since last follow up, nearest to actual date of follow up.
always indicate date of data collection!

Follow up period: (do not fill in)

data entry range from: dd.mm.yyyy **until:** dd.mm.yyyy

response criteria: Gertz MA et al. AJoH, 79:319-328 (2005)

heart

vital parameters **not done**

heart rate bpm ☐

blood pressure 2)

standing position: systolic /

standing position: diastolic mmHg

sitting / lying position: systolic /

sitting / lying position: diastolic mmHg

date of data collection dd.mm.yyyy

blood parameters **not done**

1) ng/l (=pg/ml) ☐

troponin T µg/l ☐

date of data collection dd.mm.yyyy

NYHA

3)

date of examination dd.mm.yyyy

transthoracic echocardiogram

echocardiography (TTE)

performed? ○ Yes ○ No ○ not known

date of examination

left ventricle / left atrium **not done**

LVEF biplan % ☐

EDVI ml/m² ☐

LVMMI g/m² ☐

rTh ☐

septum thickness mm ☐

left atrial dilatation ○ Yes ○ No ○ not known

LA ESDI cm/m² ☐

diastolic function **not done**

E/e' ☐

diastolic dysfunction 4)

speckle tracking

apical sparing? ○ Yes ○ No ○ not known **not done**

global peak longitudinal strain % ☐

right ventricle

right ventricular hypertrophy ○ Yes ○ No ○ not known **not done**

ΔPsyst RV/RA mmHg ☐

pericardial effusion ○ Yes ○ No ○ not known

LAVI [ ] ml/m² ☐

kidney

renal replacement therapy ○ Yes ○ No ○ not known

please specify [ ] 5)

start of dialysis [ ] dd.mm.yyyy

serum parameters	**not done**	**morning urine**	**not done**	**24h urine**	**not done**
serum creatinine [ ] µmol/l	☐	urine creatinine [ ] mmol/l	☐	24h urine creatinine [ ] mmol/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	☐
[ ] 6)					
date of data collection [ ] dd.mm.yyyy		date of data collection [ ] dd.mm.yyyy		date of data collection [ ] dd.mm.yyyy	

sonography of the kidneys

performed? ○ Yes ○ No ○ not known

date of performance [ ] dd.mm.yyyy

length diameter right kidney [ ] cm not done ☐

length diameter left kidney [ ] cm not done ☐

liver

liver parameters

alkaline phosphatase [ ] U/l not done ☐

upper limit of normal [ ] U/l not known ☐

lower limit of normal [ ] U/l not known ☐

date of data collection [ ] dd.mm.yyyy

nervous system response and progression

ENMG

performed? ○ Yes ○ No ○ not known

result [ ] 7)

changes in result since last follow up [ ] 9)

date of performance [ ] mm.yyyy

NIS-LL

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

muscle power grading [ ] total points ☐

sensory/reflex activity grading [ ] total points ☐

date of performance [ ] mm.yyyy

FAP-scale (TTR-amyloidosis)

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

FAP-scale [ ] 8)

date of performance [ ] mm.yyyy

hematologic response and progression

paraprotein examination performed

○ Yes ○ No ○ not known

not done

immunofixation

serum immunofixation ○ positive ○ negative ☐

urine immunofixation ○ positive ○ negative ☐

date of immunofixation [ ] dd.mm.yyyy

total free light chain

total free light chain kappa [ ] g / l ☐

total free light chain lambda [ ] g / l ☐

date of FLC data collection [ ] dd.mm.yyyy

m-gradient / bone marrow

serum M-gradient [ ] g / l ☐

bone marrow examination performed ☐ yes

bone marrow plasma cells [ ] %

date of examination [ ] dd.mm.yyyy

hematologic data have to be entered in the next form "treatment since last follow up"

laboratory values

not done

Hb [ ] g/l ☐

leucocytes [ ] G/l ☐

platelets [ ] G/l ☐

plasma cells [ ] % in peripheral blood ☐

neutrophiles [ ] % in peripheral blood ☐

lymphocytes [ ] % in peripheral blood ☐

date of data collection [ ] dd.mm.yyyy

CRP [ ] mg/l ☐

SAA (serum amyloid a) [ ] mg/l ☐

date of data collection [ ] dd.mm.yyyy

physical examination

not done

weight [ ] kg ☐

date of weight measurement [ ] dd.mm.yyyy

fever attacks

fever attacks

since last follow up ○ Yes ○ No ○ not known

number of attacks per year [ ] /year

symptoms of inflammation

symptoms of inflammation

abdominal pain / peritonitis ○Yes ○No ○not known

joint pain ○Yes ○No ○not known

pleuritis / pericarditis ○Yes ○No ○not known

myalgia ○Yes ○No ○not known

rash / skin ○Yes ○No ○not known

other ○Yes ○No ○not known

please specify [ ]

Signature

Place, date [ ] Signature [ ]

Possible entries

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

1)

BNP
NT-pro BNP

2)

sitting / lying position
standing position
both performed
not done

3)

not known
no dyspnea
I
II
III
IV

4)

normal
grade I - relaxation dysfunction
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 + demyelination
conduction block
normal
not known

8)

stage I
stage II
stage III
not known

9)

improved
worsened
no change
not known

Participants and roles (V1.01)

Please note: This form is only filled in for the **PRODUCTIVE** area. Data entry will be done by the CTC DM.

Participants at center

No.	Given name	Surname	Role	First Access to productive	Withdrawn?	Access until
1				dd.mm.yyyy	☐	dd.mm.yyyy

More

Signature

Place, date

Signature

Participants and roles (V1.01)

Please note: This form is only filled in for the **SETUP** area. Data entry will be done by the CTC DM.

Center

Registered (by CTC) dd.mm.yyyy

Hospital / Organisation

Department

City

Language menus in secuTrial (standard for all participants) ○ Deutsch ○ Englisch ○ Französisch ○ Spanisch

Release for productive = enter study data? ○ Automatisch durch CTC DM ○ Initial durch Sponsor

Principal Investigator

Current PI of center

Salutation ○ Mr ○ Ms

Given name

Surname

E-Mail

PI has changed? ☐

Previous PI

More

No.	Salutation	Given name	Surname	PI until
1	○ Mr ○ Ms			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

No.	Given name	Surname	User-ID	Role	Required qualification	Training / RUS-Log ok?	Pseudonym test patient

1 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

Premature Study End (V1.01)

Please fill in the form 'Study End' first!

Course

Premature Study End

Date of premature study end dd.mm.yyyy

Withdrawal of informed consent

Please note:
If the patient has withdrawn consent the forms should be locked. For this use the items below.

Has the patient withdrawn the informed consent?
- ○ Yes
- ○ No

Date of withdrawal dd.mm.yyyy

Reason for discontinuation

Origin

Cause for discontinuation lies with
- ○ Patient
- ○ Study team
- ○ Authorities
- ○ other

If OTHER: Please explain

Further explanation for premature termination.

Please explain the reasons for discontinuation (tick all that apply)

	Applies
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	☐

Comment (optional)	**Please specify reason for WITHDRAWAL**	**Please specify OTHER REASON**

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 checkox and saving the form. This will be documented in the audit trail.

Lock patient, no further data entry ☐

Signature and Confirmation

Confirmation of the investigator (Prüfperson)

With my electronic signature (username and password) I confirm the **completeness** and **correctness** of the data entered in this CRF.

Date dd.mm.yyyy

Name of investigator (Prüfperson)

Signature

Date (handwritten after printout) **Signature (handwritten after printout)**

Please **print out** this form, **sign** it and put it in the **patient folder**.

Signature

Place, date Signature

Study End (V1.01)

Study End

Please give the date of the last study visit conducted.
With premature study end, please give also the date of discontinuation in the appropriate form.

Has the patient completed the study according to protocol?

- ○ Yes
- ○ No

Date of last study visit conducted

dd.mm.yyyy

Signature and Confirmation

Confirmation of the investigator (Prüfperson)

With my electronic signature (username and password) I confirm the **completeness** and **correctness** of the data entered in this CRF.

Date dd.mm.yyyy

Name of investigator (Prüfperson)

Signature

Date (handwritten after printout)

Signature (handwritten after printout)

Please **print out** this form, **sign** it and put it in the **patient folder**.

Signature

Place, date

Signature

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.