Svenska barncancerregistret – VPH

Follow up - formulär

Version 210921

Report date: | 2 | 0 |___|___|___|___|___|___|
Registered by:
Reporting clinical:
Forename:
Surname:
Date of Birth: | 2 | 0 |___|___|___|___|___|___| - |___|___|___|___|
Gender: ☐ **Female** ☐ **Male**
City:
Country:

Patient status

Date follow up

| 2 | 0 |___|___|___|___|___|___|

Patient status (One option)

- ☐ Alive
- ☐ Dead
- ☐ Lost in follow up
- ☐ Opted out

If patient is dead only answer below

Date of death

| 2 | 0 |___|___|___|___|___|___|

Main cause of death:

_____

Patients care pathway

HCP List:

_____

Physician ID:

_____

Bone marrow aspiration

Patient status (One option)

- ☐ Never performed
- ☐ Normal bone marrow
- ☐ Normal marrow cellularity with a paucity of erythroid precursos (DBA)
- ☐ Other bone marrow findings - specify

If answer above is other bone marrow findings please specify:

RBC Parameters

Date of the hematological parameters:

| 2 | 0 |___|___|___|___|___|___|

RBC number (10^6/mm3, 10^12/L:

_____

Hb (g/dL, g/L):

_____

Reticulocytes (One option)

- ☐ Reticulocytes absolute count
- ☐ Reteiculocytes %
- ☐ Data missing

If Reticulocytes absolute count, please fill in below:

Reticulocytes absolute (10^3/mm3, 10^9/L):

_____

If Reteiculocytes % , please fill in below:

Reticulocytes (/1000 RBC, %):

_____

WBC number (10^3/mm3, 10^9/L):

_____

Platelet number (10^3/mm3, 10^9/L):

_____

Blood transfusion

Transfusion status in the present (One option)

- ☐ Yes (Regular)
- ☐ Yes (Occasional)
- ☐ No
- ☐ Unknown

If Transfusion status in the present is Yes (Regular), please fill in below:

Date of first (regular) transfusion

| 2 | 0 |___|___|___|___|___|___|

Indication for regular transfusion (One option)

- ☐ Cerebral Vasculopathy
- ☐ Chronic Anemia
- ☐ Extramedullary hematopoiesis
- ☐ Fatigue
- ☐ Heart failure
- ☐ Hepatobiliary problems
- ☐ Leg ulcers
- ☐ Recurrent acute chest syndrome
- ☐ Priapism
- ☐ Pulmonary Hypertension
- ☐ Quality of life
- ☐ Recurrent Vaso-occlusive Crisis
- ☐ Shortness of breath
- ☐ Other, specify free text

Year

| 2 | 0 |___|___|

Length at registration (cm):

_____

Weight at registration (kg):

_____

Goal Hb pre-Tx (g/L):

_____

Tx numbers/year:

_____

Amount of ml per each transfusion event (mL):

_____

Blood consumption (mL/Kg/year):

_____

Adverse event

☐ No ☐ Yes

If Yes, please specify:

_____

Alloimmunization

☐ No ☐ Yes

If Transfusion status in the present is Yes (Occasional), please fill in below:

Tx numbers/year:

_____

Amount of ml per each transfusion event (mL):

_____

Adverse event

☐ No ☐ Yes

If Yes, please specify:

_____

Alloimmunization

☐ No ☐ Yes

Delayed haemolytic transfusion reactions (DHTR)

☐ No ☐ Yes

Main causes (triggers):

- ☐ Acute Aplastic Crisis
- ☐ Acute Chest Syndrome
- ☐ Acute Haemolysis
- ☐ Acute painful Vaso-occlusive Crisis
- ☐ Mental stress (school exams, other)
- ☐ Holiday
- ☐ Priapism
- ☐ Splenic Sequestration
- ☐ Stroke
- ☐ Sun-exposure (PKD)
- ☐ Travel outside europe
- ☐ Other, specify free text

If Other, please specify:

_____

If Transfusion status in the present is No, please fill in below:

Main causes (triggers):

- ☐ Acute Aplastic Crisis
- ☐ Acute Chest Syndrome
- ☐ Acute Haemolysis
- ☐ Acute painful Vaso-occlusive Crisis
- ☐ Mental stress (school exams, other)
- ☐ Holiday
- ☐ Priapism
- ☐ Splenic Sequestration
- ☐ Stroke
- ☐ Sun-exposure (PKD)
- ☐ Travel outside europe
- ☐ Other, specify free text

If Other, please specify:

_____

Delayed haemolytic transfusion reactions (DHTR) ☐ No ☐ Yes		

Iron overload

Date
| 2 | 0 |___|___|___|___|___|___|

Average serum ferritin (12 months):
_____

Transferrin saturation %:
_____

ALT (U/L):
_____

Total bilirubin (mg/dL, µmol/L):
_____

Creatinine (mg/dL):
_____

Liver MRT2*

☐ Not performed ☐ Performed

If Liver MRT2* is performed please fill in below

Date
| 2 | 0 |___|___|___|___|___|___|

Liver MRT2* mg iron/g liver dry weight

☐ No value recieved ☐ mg iron / g liver dry weight

If above option is mg iron / g liver dry weight please specify below:
_____

Liver iron MRT2*

☐ No value recieved ☐ ms (milliseconds)

If above option is ms (milliseconds) please specify below: _____

Cardiac iron MRT2*

☐ Not performed ☐ Performed

If Cardiac iron MRT2*is performed please fill in below

Date
| 2 | 0 |___|___|___|___|___|___|

Evidence of heart iron overload

☐ Yes ☐ No

Ejection fraction

☐ Normal ☐ Decreased

Cardiac iron T2* (ms milliseconds):

_____

Heart SQUID

☐ Not performed ☐ Performed

Date

| 2 | 0 |___|___|___|___|___|___|

Heart SQUID result (One option)

- ☐ No excess iron
- ☐ Mild iron overload
- ☐ Moderate iron overload
- ☐ Heavy iron overload
- ☐ Very heavy iron overload
- ☐ Iron content in mg - specify

If Heart SQUID result is Iron content in mg, please specify below:

_____

Liver SQUID

☐ Not performed ☐ Performed

Date

| 2 | 0 |___|___|___|___|___|___|

Liver SQUID mg/g dry weight:

_____

Fibroscan

☐ Not performed ☐ Performed

Date

| 2 | 0 |___|___|___|___|___|___|

Fibroscan result (One option)

- ☐ No fibrosis
- ☐ Mild fibrosis
- ☐ Moderate fibrosis
- ☐ Severe fibrosis
- ☐ Very severe fibrosis
- ☐ Elastography/fibroscan median value – specify

If Fibroscan result is Elastography/fibroscan median value, please specify kPA below:

_____

Liver biopsy

☐ Not performed ☐ Performed

Date
| 2 | 0 |___|___|___|___|___|___|

Liver biopsy result (One option)

- ☐ No excess iron
- ☐ Mild iron overload
- ☐ Moderate iron overload
- ☐ Heavy iron overload
- ☐ Very heavy iron overload
- ☐ Iron content in mg – specify

If Liver biopsy result is Iron content in mg, please specify below:

_____

Chelation

Chelation

☐ Yes ☐ No ☐ Data missing

If yes, please fill in below

Drug (Several options)

- ☐ Deferroxamine
- ☐ Deferasirox
- ☐ Deferiprone
- ☐ Other, specify free text

If Other, please specify:

_____

Year
| 2 | 0 |___|___|

If there is one drug, please fill in below:	If there are a second drug, please fill in below:
Dose 1 (mgs daily): _____	**Dose 2 (mgs daily): _____**
Dose 1 (mgs/Kg): _____	**Dose 2 (mgs/Kg): _____**

Previously given treatments

☐ No ☐ Yes

If Yes, please specify products:

_____

Organ damage

Complications of bones and extremities

☐ Yes ☐ No ☐ Data missing

If Yes, please specify below (Several options)

- ☐ Bone fractures
- ☐ Bone abnormalities
- ☐ Osteoporosis
- ☐ Leg ulcers
- ☐ Osteonecrosis
- ☐ Extramedullary hematopoiesis
- ☐ Other, specify free text

If Other please specify:

_____

Cardiac and pulmonary disease

☐ Yes ☐ No ☐ Data missing

If Yes, please specify below (Several options)

- ☐ Acute chest syndrome (SCD only)
- ☐ Arrhythmia
- ☐ Heart failure
- ☐ Increased TRV
- ☐ Ischaemic heart disease
- ☐ Pulmonary Hypertension
- ☐ Other, specify free text

If Other please specify:

_____

Neurologic disease

☐ Yes ☐ No ☐ Data missing

If Yes, please specify below (Several options)

- ☐ Haemorrhagic stroke
- ☐ Ischemic stroke
- ☐ Seizures polyneuropathy
- ☐ Other, specify free text

If Other please specify:

_____

Endocrinologic disease

☐ Yes ☐ No ☐ Data missing

If Yes, please specify below (Several options)

- ☐ Delayed puberty
- ☐ Diabetes
- ☐ Growth hormone disturbance
- ☐ Hypocortisolism
- ☐ Hypogonadism
- ☐ Hypoparathyroidism
- ☐ Hypothyroidism
- ☐ Infertility
- ☐ Tyroid disease
- ☐ Other, specify free text

If Other please specify:

_____

Liver and Kidney disease

☐ Yes ☐ No ☐ Data missing

If Yes, please specify below (Several options)

- ☐ Hypertension
- ☐ Liver cirrhosis
- ☐ Liver disease
- ☐ Liver fibrosis
- ☐ Microalbuminuria
- ☐ Renal failure
- ☐ Other, specify free text

If Other please specify:

_____

Visual and hearing diseases specified

☐ Yes ☐ No ☐ Data missing

If Yes, please specify below (Several options)

- ☐ Blindness
- ☐ Hearing loss
- ☐ Retinopathy
- ☐ Visual disturbances
- ☐ Other, specify free text

If Other please specify:

_____

Acute complications

☐ Yes ☐ No ☐ Data missing

If Yes, please specify below (Several options)

- ☐ Diagnosis or symptoms of sepsis
- ☐ Septic shock
- ☐ Meningitis
- ☐ Hospital admission because of pneumonia
- ☐ Any ICU admission
- ☐ Any emergency hospital admission
- ☐ Other, specify free text

If Other please specify:

_____

Spleen

Splenomegaly

☐ Yes ☐ No ☐ Data missing

Splenectomy

☐ Yes ☐ No ☐ Data missing

If yes, please fill in date

Date of splenectomy (Year)
| 2 | 0 |__|__|

Serological data

HIV antibodies

☐ Positive ☐ Negative ☐ Not done ☐ I don't know

HCV antibodies

☐ Positive ☐ Negative ☐ Not done ☐ I don't know

HBV antibodies

☐ Positive ☐ Negative ☐ Not done ☐ I don't know

Cholelithiasis

Cholelithiasis

☐ Yes ☐ No ☐ Data missing

Cholecystectomy

☐ Yes ☐ No ☐ Data missing

If yes, please fill in date

Cholecystectomy Date (Year)
| 2 | 0 |__|__|

Osteoporosis

Skeletal survey for osteopenia/osteoporosis done

☐ Yes ☐ No ☐ Data missing

Osteoporosis diagnoses

☐ Yes ☐ No ☐ Data missing

If yes, please fill in date

Year of diagnosis of osteoporosis (Year)
| 2 | 0 |__|__|

Extramedullary hematopoiesis

Investigation done

☐ Yes ☐ No ☐ Data missing

If yes, please specify method (One option)

☐ CT Scan ☐ MRI

Symptomatic requiring treatment?

☐ Yes ☐ No ☐ Data missing

Treated for extram erythropoiesis?

☐ Yes ☐ No ☐ Data missing

If yes, please fill in treatment

☐ Radiotherapy ☐ Increase transfusions ☐ Other

Pulmonary hypertension

Pulmonary hypertension

☐ Yes ☐ No ☐ Data missing

If yes, please specify method of diagnosis (One option)

☐ Echo ☐ Angiografi

Treatment given

☐ Yes ☐ No

If yes, please specify treatment:

_____

BMT/HSCT transplant & Gene therapy

HSCT transplant

☐ No ☐ Yes

If HSCT transplant is yes, please fill in below

Date (Year)

| 2 | 0 |__|__|

HSCT outcome

☐ Cured ☐ Not cured

If HSCT transplant is No, please fill in below

HSCT transplant

- ☐ Family refusal
- ☐ Lack of availability of facilities
- ☐ Lack of guidelines for this disease
- ☐ No clear indication
- ☐ No indication
- ☐ Other, free text

If above answer is Other, please specify:

_____

Gene therapy given?

☐ Yes ☐ No ☐ Data missing

If Gene therapy given is yes, please fill in date

Date

| 2 | 0 |___|___|___|___|___|___|

Gene therapy outcome

☐ Cured ☐ Not cured

Specific treatment

Specific treatment

☐ Yes ☐ No ☐ Data missing

If specific treatment is Yes, please specify Drug:

_____

Regular blood exchange (erythrocyte apheresis) given?

☐ Yes ☐ No ☐ Data missing

Number of treatments given last 12 months:

_____

Drugs given?

☐ Yes ☐ No ☐ Data missing

If drugs given is yes, please fill in drugs to the medication form.

Patient involved in a Clinical Trial protocol

☐ Yes ☐ No ☐ Data missing

If above answer is Yes, please specify clinical trial below:

_____