Severe Chronic Neutropenia International Registry	**Patient ID Number**: ___/___ ___/___ ___/___ ___ ___ ___ ___ **Patient Initials**: ___ ___ ___

REGISTRATION
PATIENT DETAILS

FOR REGIONAL REFERRAL CENTER USE ONLY
Registration: ☐ Approved Date:____/_____/____ (DD/MON/YY)
☐ Not approved
RRC telephone contact with primary MD: Date:____/_____/____ (DD/MON/YY)
Reviewers Signature:_____
Status: _____

Person completing form: _____ (please print)

Date of Signed Consent: _____/_____/_____ (DD/MON/YY)

Patient: _____ Parent /Legal Guardian (*if applicable*): _____

Address: _____ Address: _____

City/Village: _____ City/Village: _____

State/Province: _____ State/Province: _____

Zip/Postal Code: _____ Country:_____ Zip/Postal Code: _____ Country:_____

Telephone Number:( )( ) _____ Telephone Number:( )( ) _____

Birth Date: ____/_____/____ (DD/MON/YY) Sex: ☐ M ☐ F Race: ☐ Caucasian ☐ Black ☐ Asian ☐ Other: (Specify)_____

Neutropenia Code: ☐ Date of Onset: _____/_____/_____ (DD/MON/YY) Date of Diagnosis: _____/_____/____ (DD/MON/YY)

Congenital: 01 - If known:
- ☐ Severe Congenital Neutropenia / Kostmann Type
- ☐ Severe Congenital Neutropenia (Autosomal Dominant)
- ☐ Severe Congenital Neutropenia with Immunodeficiency
- ☐ Congenital White Cell Aplasia
- ☐ Shwachman - Diamond
- ☐ Glycogen Storage Disease
- ☐ Myelokathexis
- ☐ Other _____

Cyclic: 02 - Must provide documentation of regular cycling with 3 X/week counts for 6 weeks and recurring infections.

Idiopathic: 03 -

Other: 04 - Please specify _____

Anti-neutrophil Antibodies detected: ☐ Yes ☐ No ☐ Not Tested

REFERRING PHYSICIAN

Name: _____ Institution Name: _____

Institution Address: _____

City/Village: _____ State/Province: _____

Country: _____ Zip/Postal Code: _____

Telephone Number:( )( )_____ Fax Number: ( )( )_____

Severe Chronic Neutropenia International Registry	**Patient ID Number**: ___/___ ___/___ ___/___ ___ ___ ___ **Patient Initials**: ___ ___ ___

REGISTRATION
SIGNIFICANT CLINICAL HISTORY OF INFECTIONS

BASELINE (PRE-GROWTH FACTOR/CYTOKINE)					
(An episode is a discrete occurrence with a beginning and an end)	FREQUENCY OF EPISODES				
	None √	1-3 per Year √	4-12 per Year √	>12 per Year, repeated or continuous √	Unknown √
Mouth Ulcers/Gingivitis/ Periodontitis	☐	☐	☐	☐	☐
Skin Infections/ Abscess	☐	☐	☐	☐	☐
Sinusitis	☐	☐	☐	☐	☐
Pharyngitis/Bronchitis	☐	☐	☐	☐	☐
Otitis	☐	☐	☐	☐	☐
Pneumonia	☐	☐	☐	☐	☐
Bacteremia	☐	☐	☐	☐	☐
Liver Abscess	☐	☐	☐	☐	☐
Colitis/Enteritis	☐	☐	☐	☐	☐
Peritonitis	☐	☐	☐	☐	☐
Urinary Tract Infections	☐	☐	☐	☐	☐
Other *(specify)*	☐	☐	☐	☐	☐
Other *(specify)*	☐	☐	☐	☐	☐
Other *(specify)*	☐	☐	☐	☐	☐

Severe Chronic Neutropenia International Registry	**Patient ID Number**: ___/___ ___/___ ___/___ ___ ___ ___ **Patient Initials**: ___ ___ ___

REGISTRATION
SIGNIFICANT CLINICAL HISTORY OF NON-INFECTIOUS EVENTS

Clinical Problems	Problem prior to any growth factor (cytokine)? No √	Problem prior to any growth factor (cytokine)? Yes √	Problem while taking any growth factor (cytokine)? No √	Problem while taking any growth factor (cytokine)? Yes √	Is this a current problem? No √	Is this a current problem? Yes √
Splenomegaly	☐	☐	☐	☐	☐	☐
Hepatomegaly	☐	☐	☐	☐	☐	☐
Hematuria/ Proteinuria	☐	☐	☐	☐	☐	☐
Glomerulonephritis	☐	☐	☐	☐	☐	☐
Arthritis	☐	☐	☐	☐	☐	☐
Vasculitis	☐	☐	☐	☐	☐	☐
Malignancy *(specify)*	☐	☐	☐	☐	☐	☐
Other clinical problem *(specify)*	☐	☐	☐	☐	☐	☐
Other clinical problem *(specify)*	☐	☐	☐	☐	☐	☐

Severe Chronic Neutropenia International Registry	**Patient ID Number**: ___/___ ___/___ ___/___ ___ ___ ___ **Patient Initials**: ___ ___ ___

REGISTRATION
GROWTH AND DEVELOPMENT/PHYSICAL ASSESSMENT

Date of Assessment: _____/_____/_____ (DD/MON/YY)

Height: _____ cm or _____ ft _____ in

Weight: _____ kg or _____ lb _____ oz

Spleen: ☐ Palpable _____cm bcm
☐ Not Palpable
☐ Not Assessed

Liver: ☐ Palpable _____cm bcm
☐ Not Palpable
☐ Not Assessed

REPRODUCTIVE ASSESSMENT

Number of live births: _____ Number of still births: _____ Number of miscarriages/terminations: _____

Has the Patient, or his/her partner, experienced any fertility problem? ☐ No ☐ Yes ☐ Unknown

If yes, please describe nature of problem:_____

_____

Is patient or partner of male patient pregnant? ☐ No ☐ Yes, estimated delivery date: ___/_____/____ (DD/MON/YY)

FOR PATIENTS <18YEARS, PLEASE INDICATE GROWTH AND DEVELOPMENT

Please enter patient's Tanner Score: ☐ (1,2,3,4,5)

Patient's growth and development
☐ Normal
☐ Abnormal *

* Patients with abnormal growth and/or development should undergo appropriate endocrine evaluation.

TANNER SCORE

	Stage	Pubic Hair	Penis	Testes
BOY	1	None	Preadolescent	Preadolescent
	2	Scanty, long, slightly pigmented	Slight enlargement	Enlarged scrotum, pink texture altered
	3	Darker, starts to curl, small amount	Longer	Larger
	4	Resembles adult type, less in quantity; coarse, curly	Larger; glans and breadth increase in size	Larger, scrotum dark
	5	Adult distribution, spread to medial surface of thighs	Adult	Adult

	Stage	Pubic Hair	Breasts
GIRL	1	Preadolescent	Preadolescent
	2	Sparse, lightly pigmented, straight medial border of labia	Breast and papilla elevated as small mound, areolar diameter increased
	3	Darker, beginning to curl, increased amount	Breast and areola enlarged, no contour separation
	4	Coarse, curly abundant but less amount than in adult	Areola and papilla form secondary mound
	5	Adult feminine triangle, spread to medial surface of thighs	Mature; nipple projects, areola part of general breast contour

Severe Chronic Neutropenia International Registry	**Patient ID Number**: ___/___ ___/___ ___/___ ___ ___ ___ **Patient Initials**: ___ ___ ___

REGISTRATION
TREATMENT HISTORY
PREVIOUS GROWTH FACTOR (CYTOKINE)

☐ Treatment History Unknown ☐ No Previous Growth Factor ☐ Yes, Previous Growth Factor

If Yes, please list growth factor and specify brand name:	Initial Start Date (DD/MON/YY)	Discontinued[1] (list all that apply)	Comments List any problems or doses > 50 mcg/kg/day
G-CSF:	____/_____/____		
Other Cytokine:	____/_____/____		
Other Cytokine:	____/_____/____		

CURRENT GROWTH FACTOR (CYTOKINE)

Current Growth Factor ? ☐ No ☐ Yes If Yes, please list growth factor specifying brand name:	Initial Start Date (DD/MON/YY)	Current Dose		
		Quantity	Units [2]	Frequency [3]
G-CSF:	____/_____/____			
Other Cytokine:	____/_____/____			
Other Cytokine:	____/_____/____			

OTHER MEDICATIONS/TREATMENTS FOR NEUTROPENIA

Medication / Treatment	None √	Past √	Current √
Steroids	☐	☐	☐
Gamma Globulin	☐	☐	☐
Other, specify: _____	☐	☐	☐
Other, specify: _____	☐	☐	☐
Bone Marrow Transplant Date: ____/_____/_____ (DD/MON/YY)	☐		

[1] Discontinuation Codes
1 = Ineffective
2 = Pt chose to withdraw
3 = Lost to follow-up
4 = Toxicity--specify in comments field
5 = Neutrophil recovery
6 = Death
9 = Other, specify in comments field

[2] Units Code List
MCG = Microgram
MCG/KG = Microgram / kilogram
M L = Millilitre (cc)
U = Units
U /M^2 = Units / meter square

[3] Frequency Code List

qd =	Once a day	qtd =	Every third day
bid =	Twice a day	qwk =	Once a week
tid =	3 times a day	biw =	Twice a week
qid =	4 times a day	tiw =	3 times a week
qod =	Every other day	prn =	As needed

Severe Chronic Neutropenia International Registry	**Patient ID Number**: ___/___ ___/___ ___/___ ___ ___ ___ **Patient Initals**: ___ ___ ___

REGISTRATION
BONE MARROW AND CYTOGENETIC RESULTS
AND
BONE DENSITY ASSESSMENT

⇒

BONE MARROW

Bone Marrow Evaluation: ☐ No ☐ Yes ⇒ Please attach report.

Date of evaluation: _____/_____/_____ (DD/MON/YY)

Please send a stained and unstained slide from bone marrow aspirate, if available.

If slides are not presently available, have these been requested? ☐ No ☐ Yes

Expected date Registry will receive the slides: _____/_____/_____ (DD/MON/YY)

CYTOGENETICS

Cytogenetics Evaluation: ☐ No ☐ Yes ⇒ Please attach report.

Date of evaluation: ____/____/____ (DD/MON/YY)

Have any cytogenetic abnormalities been noted in previous evaluations?

- ☐ No previous evaluation
- ☐ Unknown if previous evaluations done
- ☐ No: date of previous normal evaluation: ____/_____/____ (DD/MON/YY)
- ☐ Yes: date of previous abnormal evaluation: _____/_____/_____ (DD/MON/YY)

If Yes, specify abnormality: _____

_____

BONE DENSITY ASSESSMENT

Bone Evaluation Done: ☐ No ☐ Yes ⇒ Please attach report.

Date of evaluation: _____/_____/_____ (DD/MON/YY) ☐ Normal ☐ Abnormal

Method: ☐ X-ray ☐ QCT ☐ DEXA ☐ Other, specify _____

Fractures: ☐ No ☐ Yes, specify: ☐ Spontaneous OR ☐ Accidental

Severe Chronic Neutropenia International Registry	**Patient ID Number**: ___/___ ___/___ ___/___ ___ ___ ___ **Patient Initials**: ___ ___ ___

REGISTRATION

BASELINE HEMATOLOGY - ATTACH REPORTS (ANC<500)

If it is not possible to attach reports, please complete this form. CBC's should be reported no more frequently than once per week within the 3 months prior to initiation of growth factor treatment or registration if patient is untreated.

		DATE DRAWN →	____/_____/____ (DD/MON/YY)		____/_____/____ (DD/MON/YY)		____/_____/____ (DD/MON/YY)	
		TYPE/UNITS	Measurements	Units[1]	Measurements	Units[1]	Measurements	Units[1]
		RBC x10^6/mm^3						
		Hemoglobin g/dl						
		Hematocrit %						
		MCV fL						
		Platelets x10^3/mm^3						
		WBC x10^3/mm^3						
		Absolute or Percentage	☐ A or ☐ P		☐ A or ☐ P		☐ A or ☐ P	
Differential		Bands/Stabs						
		Seg. Neutrophils						
		Neutrophils [2]						
		Lymphocytes						
		Monocytes						
		Eosinophils						
		Basophils						
		Metamyelocytes						
		Myelocytes						
		Promyelocytes						
		Myeloblasts						
		Atypical Lymphocytes						
		Large Unstained Cells						
		Other (specify) _____						

[1] Complete if units are different from those stated.

[2] Includes segs + bands

Severe Chronic Neutropenia International Registry	**Patient ID Number**: ___/___ ___/___ ___/___ ___ ___ ___ **Patient Initals**: ___ ___ ___

REGISTRATION
ADDITIONAL PATIENT INFORMATION

(Please complete and review with your physician before returning to the Data Coordinating Center.)

( ) Family History Unknown Relationship to Patient	Enrolled in Registry √	Neutropenia √	Living √	Deceased √	Leukemia √	Other Blood Disorder (specify) √
Mother						
Father						
Brothers (fill in initals of all brothers) 1.						
2.						
3.						
4.						
5.						
Sisters (fill in initals of all sisters) 1.						
2.						
3.						
4.						
5.						
Other Affected Family Members: (indicate relationship, eg. maternal grandmother, etc.)_____ _____						
Other Affected Family Members: (indicate relationship, eg. maternal grandmother, etc.)_____ _____						

Are the parents of SCN patient related by blood to each other (e.g., 2nd cousins)?

☐ No ☐ Yes, specify: _____

Severe Chronic Neutropenia International Registry	**Patient Id Number** ____/_____/_____/_____ **Patient Initials** _____ _____ _____

REGISTRATION
GLYCOGENOSIS TYPE IB

BIRTH TYPE

☐ Single ☐ Identical twin ☐ Fraternal twin, gender: ☐ same ☐ different

☐ Other multiple: _____

GENOTYPE

Genotype ☐ Not tested ☐ Tested

☐ Mutation of G6P-Transporter Gene: _____

GSD-IB RELATED SYMPTOMS

Unknown	☐ No ☐ Yes	
Asymptomatic	☐ No ☐ Yes	
Low birth weight	☐ No ☐ Yes	
Skeletal abnormalities	☐ No ☐ Yes:	_____
Muscular hypotonia	☐ No ☐ Yes	
Xanthomas or lipomas	☐ No ☐ Yes	
Malignant adenoma	☐ No ☐ Yes	
Epilepsy	☐ No ☐ Yes	
Dental problems	☐ No ☐ Yes:	_____
Growth retardation	☐ No ☐ Yes	
Failure to thrive	☐ No ☐ Yes	
Hematological abnormalities	☐ No ☐ Yes:	_____
Malignancy	☐ No ☐ Yes:	_____
Other dysfunction	☐ No ☐ Yes:	_____
		_____
Supplementary feeding	☐ No ☐ Yes:	☐ tube feeding ☐ percutaneous endoscopic gastrostomy (PEG)

SERUM PARAMETERS

C-Gluc/GD/e	_____ mmol/L	Date [DD/MON/YY]: _____
Lactate	_____ mmol/L	Date [DD/MON/YY]: _____
Alanine	_____ µmol/L	Date [DD/MON/YY]: _____
Creatinine	_____ µmol/L	Date [DD/MON/YY]: _____
Urea	_____ mmol/L	Date [DD/MON/YY]: _____
Cholesterol	_____ mmol/L	Date [DD/MON/YY]: _____
Triglycerides	_____ mmol/L	Date [DD/MON/YY]: _____

RADIOLOGY RESULTS (please attach reports)

Pancreas	☐ CT ☐ U/S	☐ Normal	☐ Abnormal:	_____
Liver	☐ CT ☐ U/S	☐ Normal	☐ Abnormal:	_____
Ribs		☐ Normal	☐ Abnormal:	_____
Long bones		☐ Normal	☐ Abnormal:	_____
Dental radiology		☐ Normal	☐ Abnormal:	_____

PSYCHOLOGY

Overall functioning	☐ Normal	☐ Concerns:	_____
Concentration power	☐ Normal	☐ Concerns:	_____
Mental development	☐ Normal	☐ Concerns:	_____
General behaviour	☐ Normal	☐ Concerns:	_____
Social competence	☐ Normal	☐ Concerns:	_____
Other issues	_____		