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

FOR REGIONAL REFERRAL CENTER USE ONLY

Registration: ☐ Approved ☐ Not approved Date:____/______/____ (DD/MON/YY)

RRC telephone contact with primary MD: Date:____/______/____ (DD/MON/YY)

Reviewers Signature:________________________________

Status: ______

REGISTRATION
PATIENT DETAILS

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)?		Problem while taking any growth factor (cytokine)?		Is this a current problem?	
	No √	Yes √	No √	Yes √	No √	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

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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 /M2 = Units / meter square

[3] Frequency Code List
qd = Once a day
bid = Twice a day
tid = 3 times a day
qid = 4 times a day
qod = Every other day
qtd = Every third day
qwk = Once a week
biw = Twice a week
tiw = 3 times a week
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: ______/______/______ ☐ Normal ☐ Abnormal
(DD/MON/YY)

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

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

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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: ______________________________

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REGISTRATION
SHWACHMAN-DIAMOND-SYNDROME

BIRTH TYPE

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

☐ Other multiple: ____

SBDS GENOTYPE

Genotype ☐ Not tested ☐ Tested :

Allele 1: ☐ 258+2T>C ☐ 183TA>CT ☐ [258+2T>C +183TA>CT]
☐ Other (please specify) : ______________________________

Allele 2: ☐ 258+2T>C ☐ 183TA>CT ☐ [258+2T>C +183TA>CT]
☐ Other (please specify) : ______________________________

Method : ☐ Panel (number of mutations in panel): _____ ☐ Sequencing

SDS RELATED SYMPTOMS

Unknown	☐ No	☐ Yes
Asymptomatic	☐ No	☐ Yes
Low birth weight	☐ No	☐ Yes
Skeletal abnormalities	☐ No	☐ Yes: ______________________
Abnormal liver function	☐ No	☐ Yes
Pancreatic insufficiency	☐ 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: ______________________

PANCREATIC FUNCTION

Taking pancreatic enzymes	☐ No	☐ Yes
Pancreatic stimulation	☐ Normal	☐ Abnormal
Diabetes mellitus	☐ No	☐ Yes

Faecal fat balance	_____ % of intake	Date: __________ (DD/MON/YY)
Faecal elastase	_____ µg/g	Date: __________ (DD/MON/YY)
Serum trypsinogen	_____ µg/L	Date: __________ (DD/MON/YY)
Serum pancreatic isoamylase	_____ µg/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	________________		