

# Severe Chronic Neutropenia International Registry (SCNIR / Registry)

July 14, 2023



# Severe Chronic Neutropenia Registry

- Neutrophils
  - Fights infection.
  - Bone marrow responds to infection speeding up production of neutrophils fighting infection.
- Neutropenia
  - Condition when bone marrow does not produce enough neutrophils.
- Severe Chronic Neutropenia
  - Neutrophil level is consistently below  $0.5 \times 10^9/\text{L}$  on a continuing basis or in cycles.
  - Bone marrow production of new neutrophils is suppressed or slowed down.
    - Infection overwhelms neutrophils
    - Develop serious bacterial infection

# Severe Chronic Neutropenia

- Types of Severe Chronic Neutropenia (SCN)
  - Congenital Neutropenia
    - Generally low neutrophils; bone marrow morphology evaluation reveals left shifted or arrested marrow
  - Cyclic Neutropenia
    - Neutrophils cycle in a 21 day pattern; marrow is similar to congenital neutropenia to normal depending on cycle
  - Idiopathic Neutropenia
    - Generally low neutrophils; bone marrow morphology evaluation reveals near normal marrow
  - Autoimmune Neutropenia
    - Generally low neutrophils; bone marrow morphology evaluation reveals near normal marrow

How to Enroll

**Frequently Asked Questions** ▸

Glossary of Terms

Helpful Links

KnippeRx

Neutropenia Health Tips

Sports Kit

School Kit

Best Choice of Syringes for You ▸

G-CSF Dosing for Specific Neutropenia ▸

Working With Health Professionals

How Neupogen Helps Maintain Your Neutrophil Count

Calculating an Absolute Neutrophil Count

mail or telephone.

and bolyard@uw.edu.

685-8055.

ents please obtain the tracking

gistry) is a global organization

## About

The Registry

To ensure a timely response please send

Our telephone number is 206-543-7218.

The phone number to KnippeRx Pharma

If you send us a package (US Post Office, number and email the team the tracking

Our Address has changed, the correct address is:  
SCN Registry/Dr. David C. Dale  
1959 NE Pacific Street, Box 356422  
Seattle, WA 98195

The Severe Chronic Neutropenia International Registry (SCNIR) is a global organization dedicated to finding the causes, consequences and best treatments for severe chronic neutropenia (SCN).

## SITE SEARCH



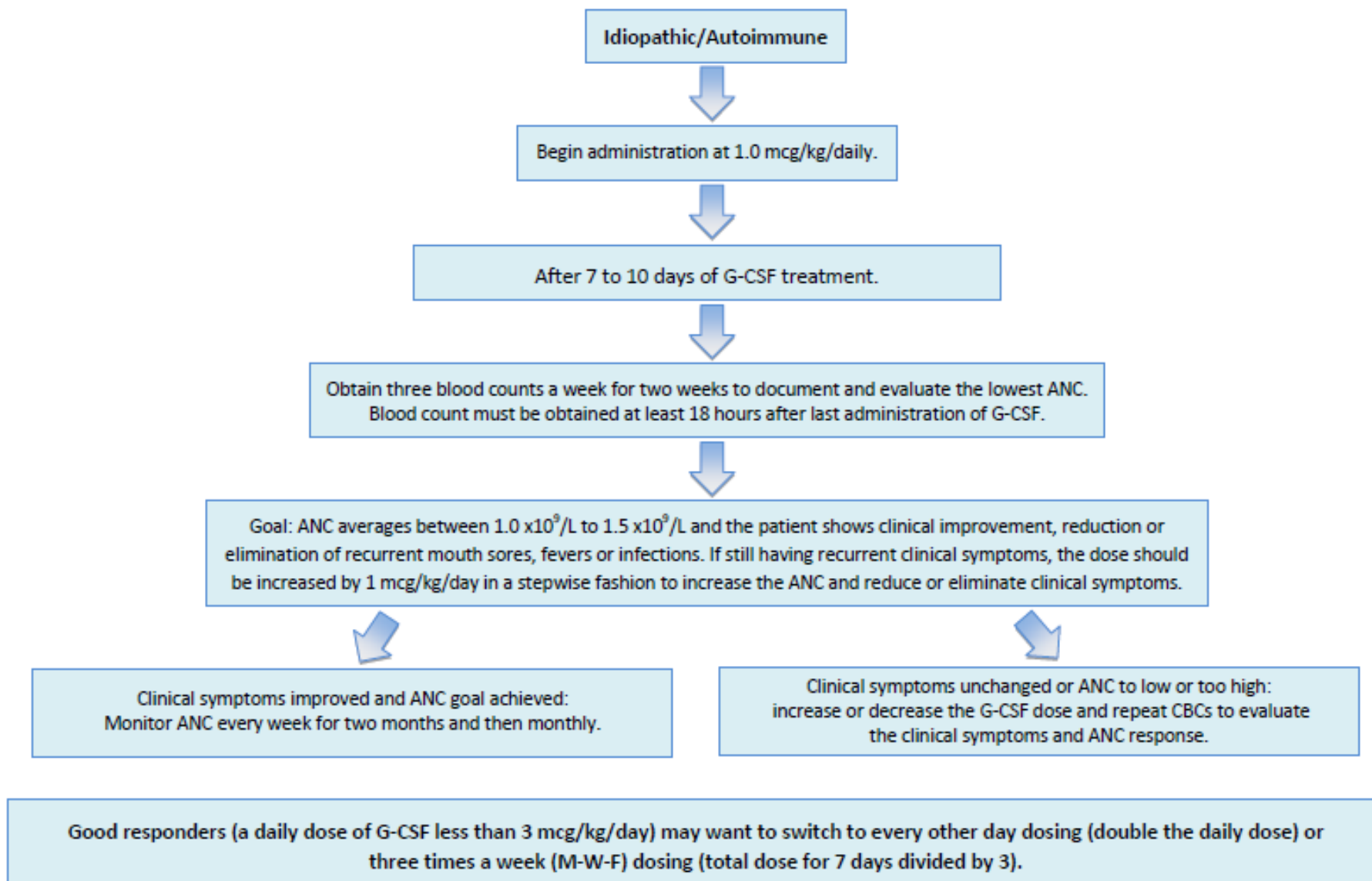
## CORONAVIRUS

For questions concerning the coronavirus disease (COVID-19) please ask your doctor or visit the [CDC website](#)

## PARTNERS

- [National Neutropenia Network \(NNN\)](#)
- [Ella Jewell Foundation](#)

# Flow Sheet for Autoimmune/Idiopathic



# Flow Sheet for Cyclic Dosing

Cyclic

Begin administration at 2.0 mcg/kg/daily.

After 7 to 10 days of G-CSF treatment.

Obtain three blood counts a week for two weeks to document and evaluate the lowest ANC.  
Blood count must be obtained at least 18 hours after last administration of G-CSF.

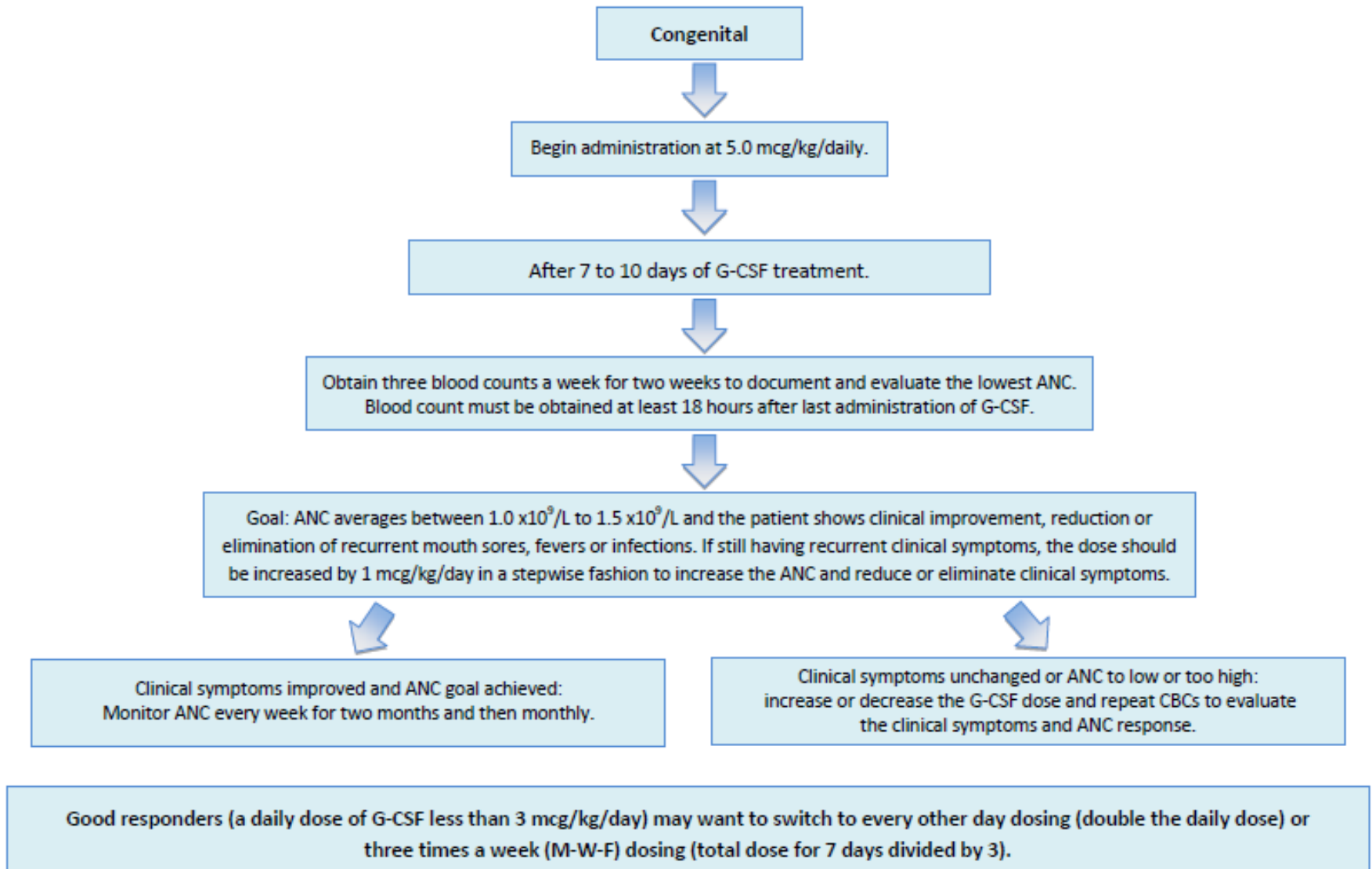
Goal: ANC averages between  $1.0 \times 10^9/L$  to  $1.5 \times 10^9/L$  and the patient shows clinical improvement, reduction or elimination of recurrent mouth sores, fevers or infections. If still having recurrent clinical symptoms, the dose should be increased by 1 mcg/kg/day in a stepwise fashion to increase the ANC and reduce or eliminate clinical symptoms.

Clinical symptoms improved and ANC goal achieved:  
Monitor ANC every week for two months and then monthly.

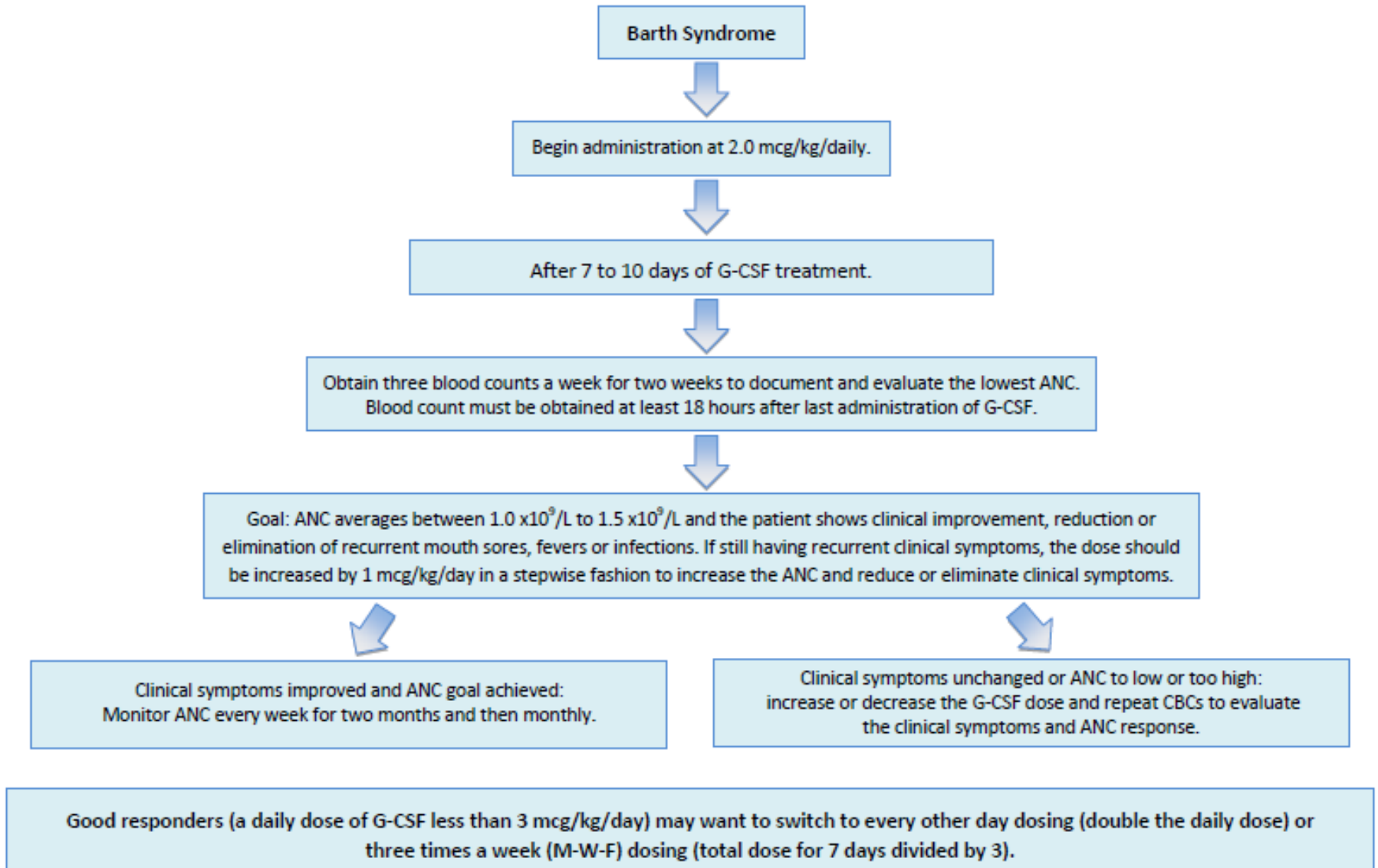
Clinical symptoms unchanged or ANC too low or too high:  
increase or decrease the G-CSF dose and repeat CBCs to evaluate the clinical symptoms and ANC response.

Good responders (a daily dose of G-CSF less than 3 mcg/kg/day) may want to switch to every other day dosing (double the daily dose) or three times a week (M-W-F) dosing (total dose for 7 days divided by 3).

# Flow Sheet for Congenital Dosing



# Flow Sheet for Barth Dosing





# Flow Sheet for Cohen Dosing

Cohen Syndrome



Begin administration at 2.0 mcg/kg/daily.



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## Complete Blood Counts (CBCs or FBCs)

CBCs should be drawn just prior to the next shot or at least 16 hours from the last injection of Neupogen®. Using this method the CBC will show the lowest absolute neutrophil count the patient is experiencing.

Daily Injections	Every Other Day Injections	Three Times a Week Injections	Twice a Week Injections
Just prior to next injection or 16 hours after last injection	Just prior to next injection or more than 36 hours after last injection	Just prior to next injection or more than 2 days since last injection	Just prior to next injection or more than three days since last injection

Sports Kit	
Item	Purpose
Bottled water	Clean cut or scrape
Betadine or anti-bacterial hand cleaner	Clean cut or scrape
Band-Aids (different sizes)	Cover cut or scrape
Cold pack (that you can activate)	Reduce swelling
Emergency numbers	To contact parents in case of serious injury
Sun screen	Prevent sunburn
Blister pads	Protect skin
Neosporin	Antibacterial ointment for cut or scrape
Consent for emergency treatment	Allows for immediate emergency treatment
Insurance information	May be required for treatment
School Kit	
School	
Emergency numbers to contact parents	
Allergy list (if any)	
Physician and telephone number	
Current medication list	
Consent for emergency treatment	
Insurance information	
Classroom	
Item	Purpose
Tooth brush / floss	Remove food from under gums (popcorn)
Betadine or anti-bacterial hand cleaner	Clean cut or scrape
Band-Aids (different sizes)	Cover cut or scrape
Cold pack (that you can activate)	Reduce swelling
Sun screen	Prevent sunburn
Blister pads	Protect skin
Neosporin	Antibacterial ointment for cut or scrape

## Effective Communication Between Patient / Parents and Professionals Managing the Medical Maze

Name		Birthdate	
Existing Medical Conditions			
Previous Physicians			
Name		Telephone #	
Allergies or Sensitivities			
Medication, Food, Other		Specific Reaction	
Current Medications			
Name		Start Date	Dose
History			
Date	Major Infections – Where – What type	Antibiotics taken - Specify	Hospitalized?
			Yes    No
			Yes    No

## Doctor Visit

Suggestions for questions

- Labs – obtain results and ask about implications
- Medications – should there be any changes
- Precautions to take
- Medical status – any changes
- Treatments – anything new

Questions for the Health Care Professional	Doctors Response / Parents Notes
1.	
2.	

# Health Care Kit

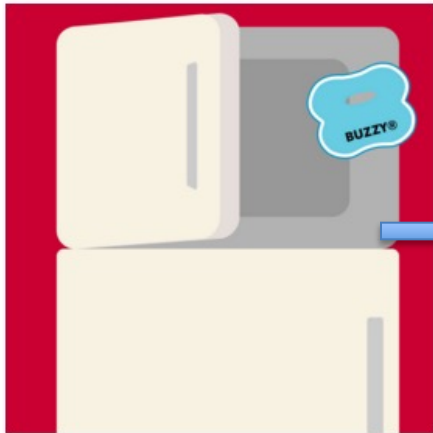


N95 Rated Mask

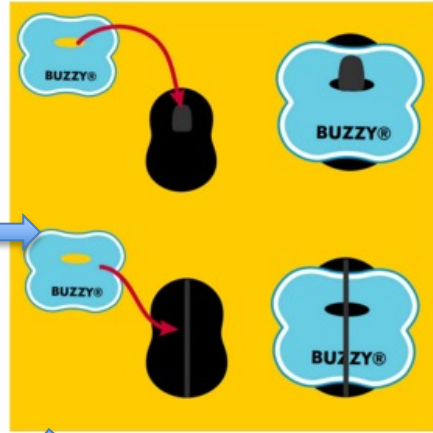
# Oral Care



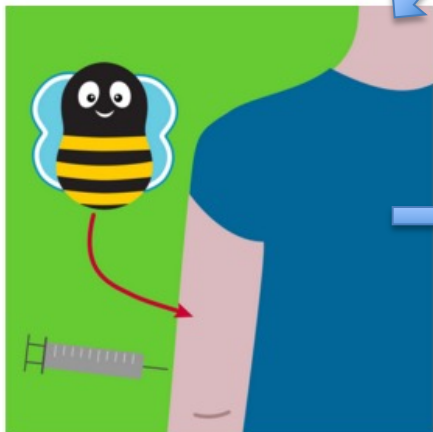
# Buzzy



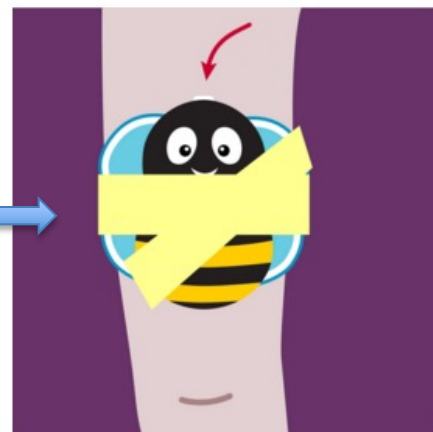
Freeze Ice Wings



Attach Wings to Buzzy®

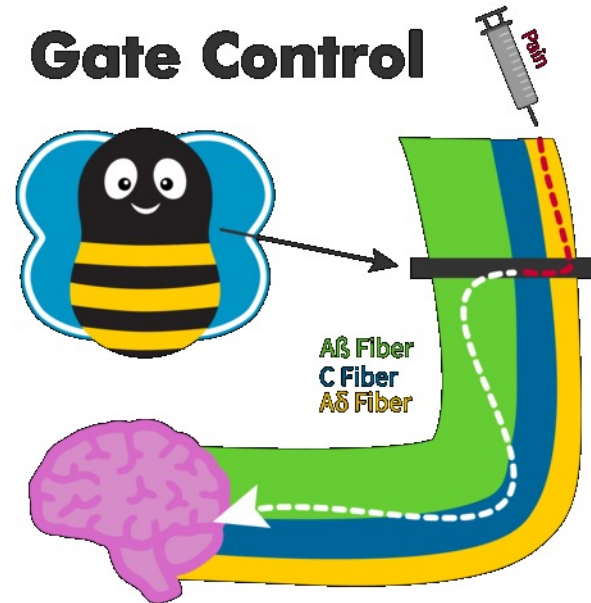


Place Buzzy®



Turn Buzzy® On

## Gate Control





# SCNIR Recommendation

- For Persons Who Do Not have Neutropenia
  - CBCs every year to evaluate for neutropenia
- For Persons Who have Neutropenia
  - CBCs twice a week for 2 weeks
    - Evaluate the severity of neutropenia-is it severe?
    - Is neutropenia intermittent or consistently severe?
    - Evidence of fevers and/or infections?
    - Work with your doctor to determine if G-CSF treatment is needed

# SCNIR Recommendation

- Initiate G-CSF Treatment
  - CBCs once or twice a week, until dose response is achieved
  - The goal for the Absolute Neutrophil Count is about  $1.5$  to  $2.5 \times 10^9/\text{L}$  (1500 to 2500)
- G-CSF Dose Check (until ANC goal is reached)
  - CBCs every 3 months, to evaluate dose response
- G-CSF Reconfirmation
  - CBCs twice a week for 2 weeks, once a year to reconfirm dose is adequate, to maintain ANC about  $1.5$  to  $2.5 \times 10^9/\text{L}$

# G-CSF Products

Product	How is it Available	Dose
Neupogen	Single-dose vial Single-dose prefilled syringe	300 or 480 micrograms
Granix	Single-dose vial Single-dose prefilled syringe	300 or 480 micrograms
Zarzio	Single-dose prefilled syringe	300 or 480 micrograms
Nivestim	Single-dose vial Single-dose prefilled syringe	300 or 480 micrograms
Accofil	Single-dose prefilled syringe	300 or 480 micrograms

# Amgen Foundation

The Amgen SupportPlus (previously Amgen360) assistances with out-of-pocket expenses

SCN Patients with:

Insurance

No insurance or limited insurance

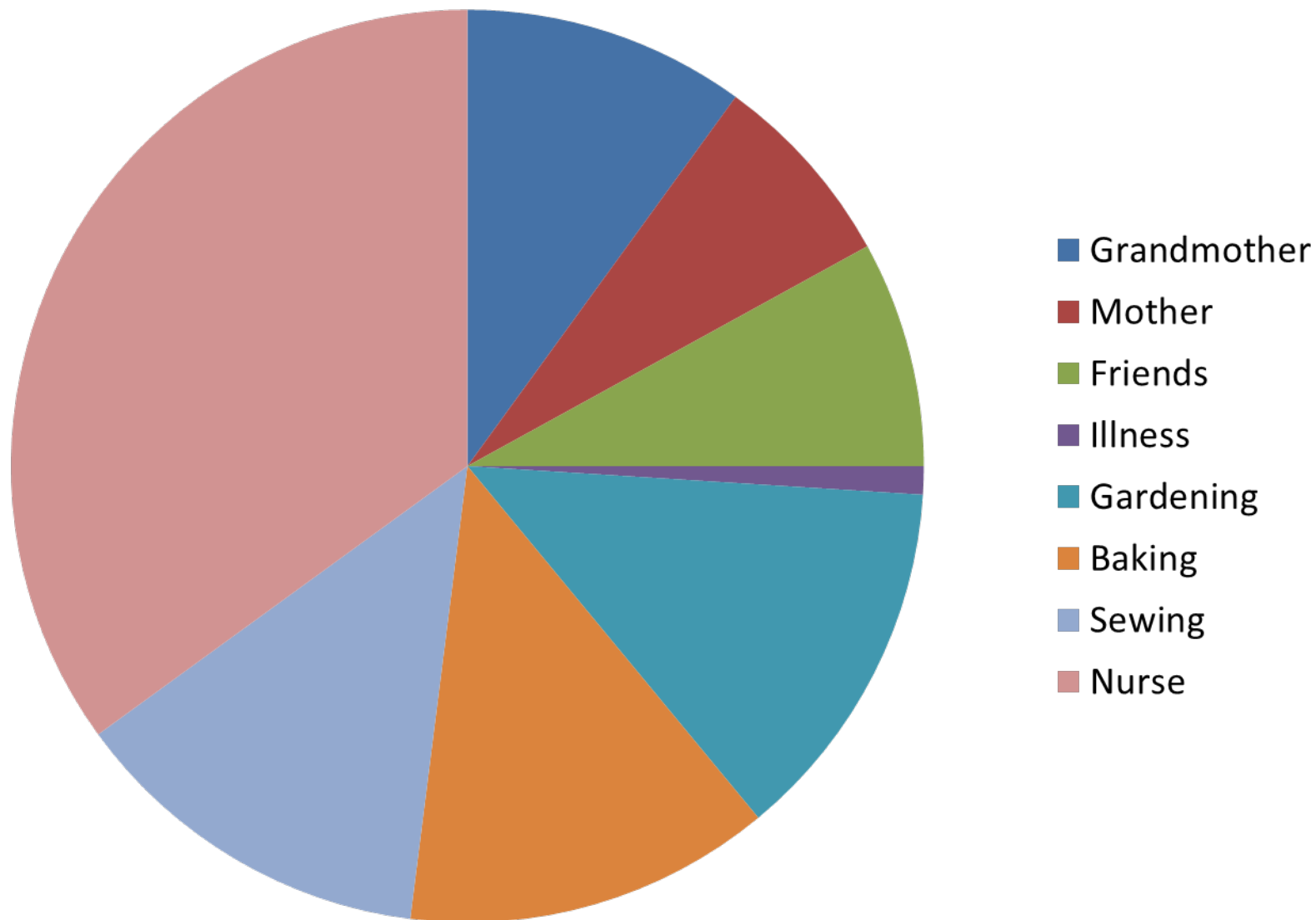
Government insurance (Medicaid, Medicare).

Call Amgen Support:

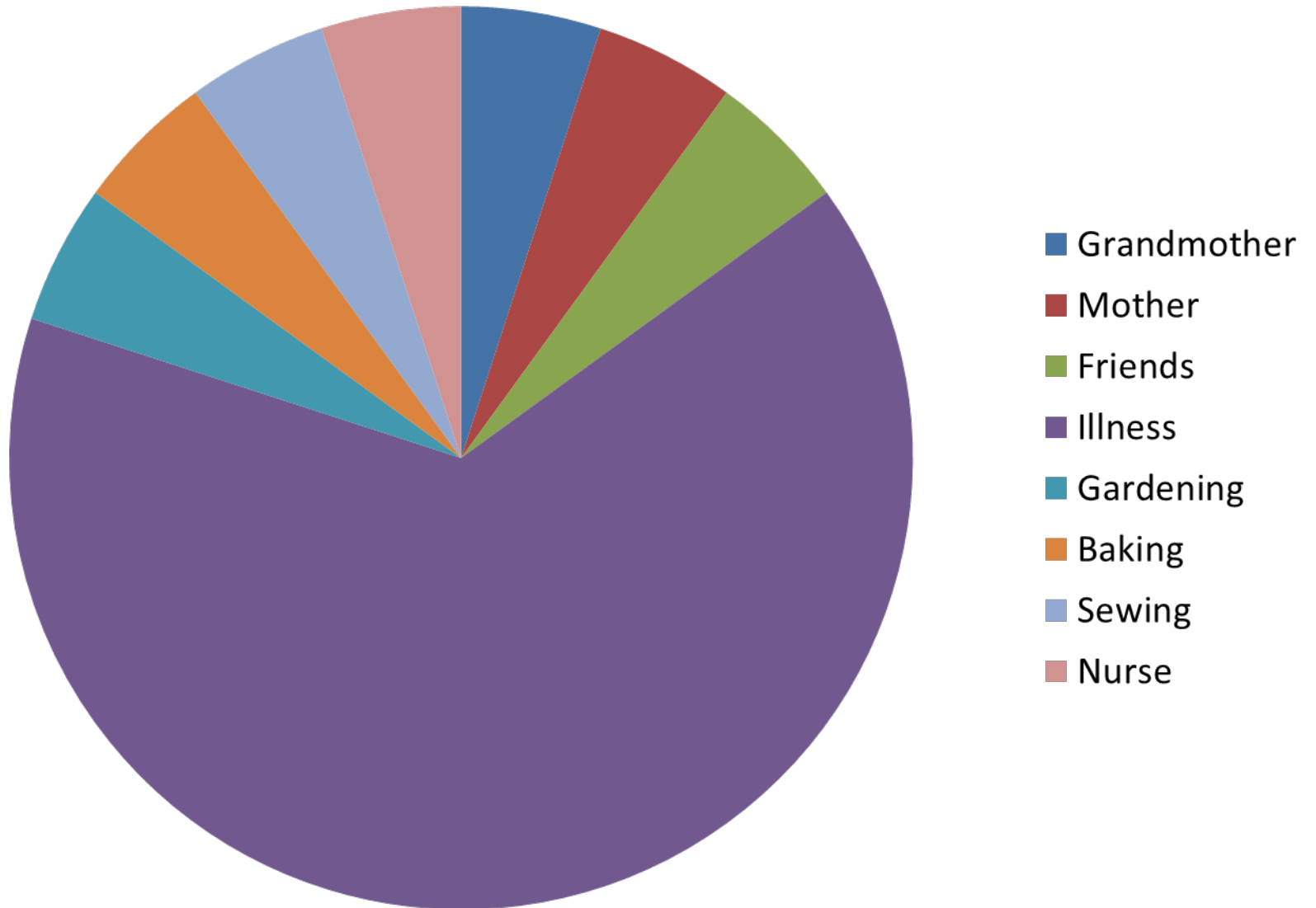
866-264-2778 and talk to the Nurse Partners

Website <https://www.amgensupportplus.com/patient/neupogen>

## My Energy Utilization

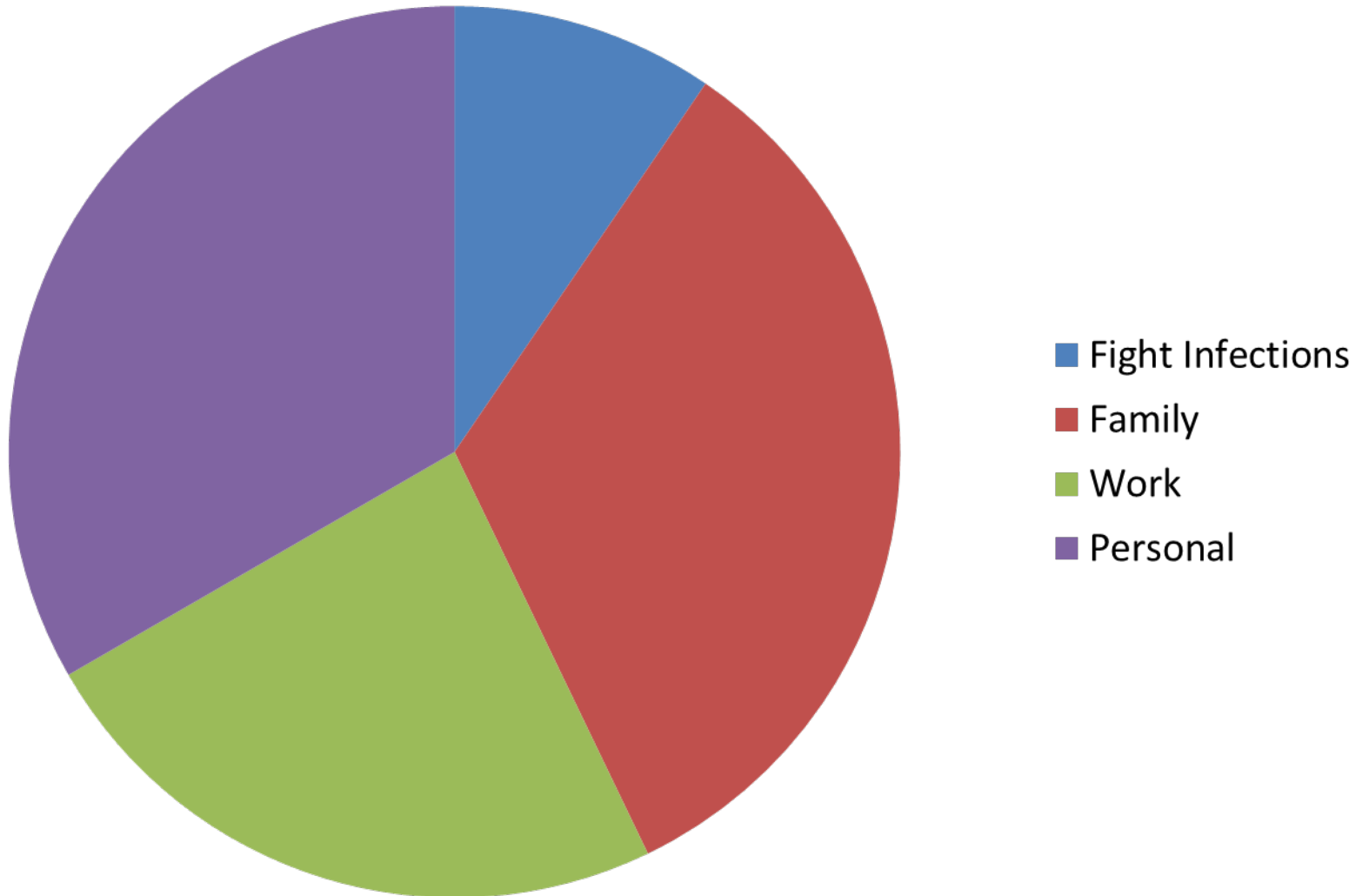


## My Energy Utilization With Covid

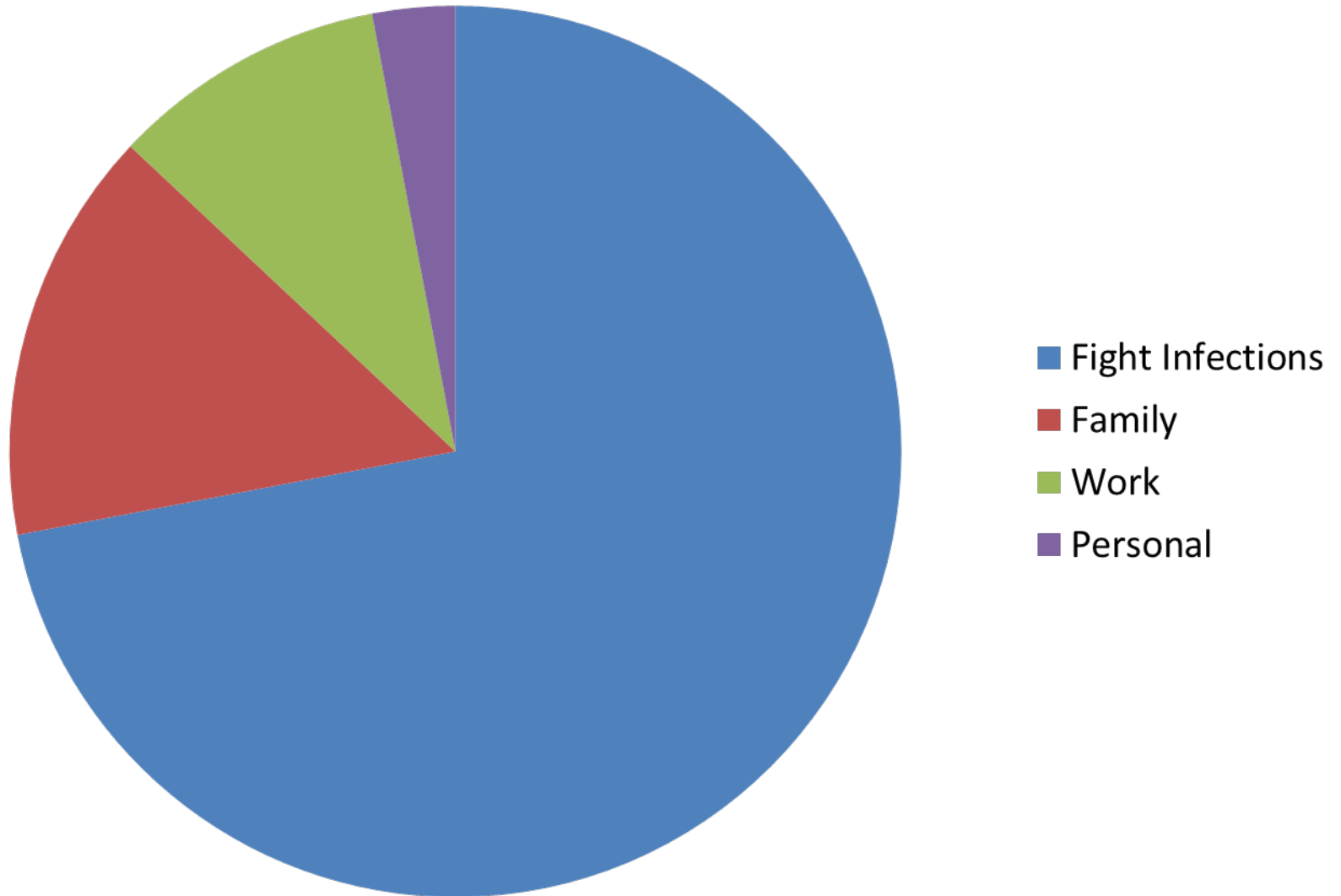


# Energy Utilization

## Normal Neutrophil Counts



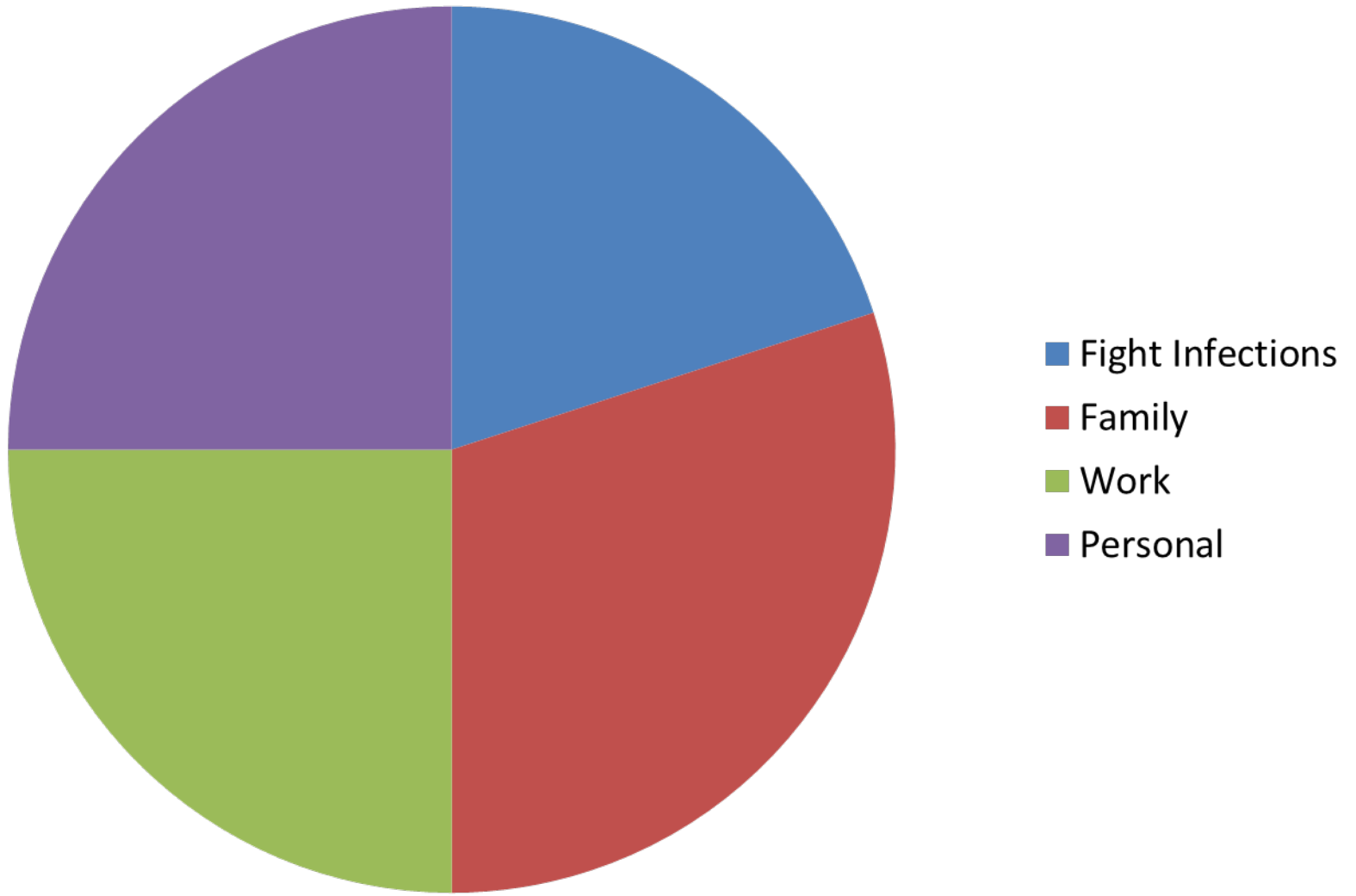
## Energy Utilization Low Neutrophil Counts



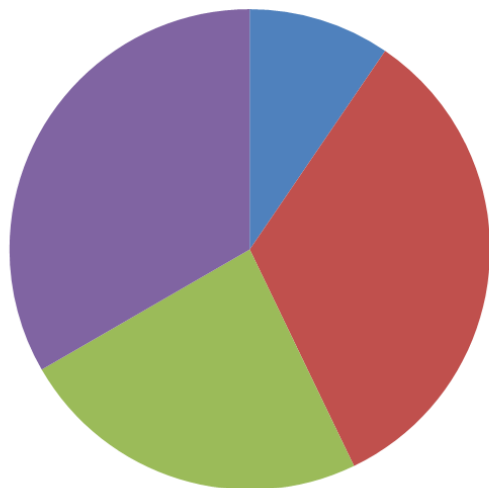


# Energy Utilization

## Normalized Neutrophil Counts

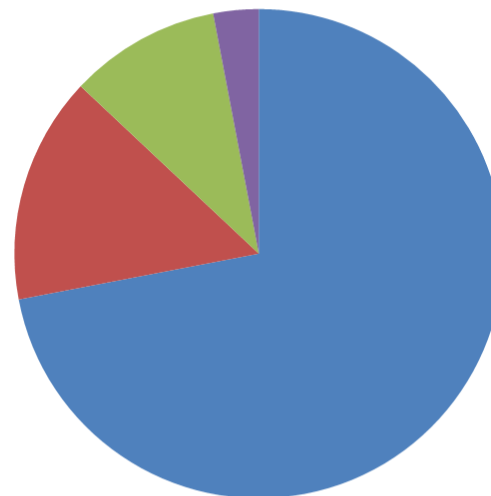


**Energy Utilization**  
**Normal Neutrophil Counts**



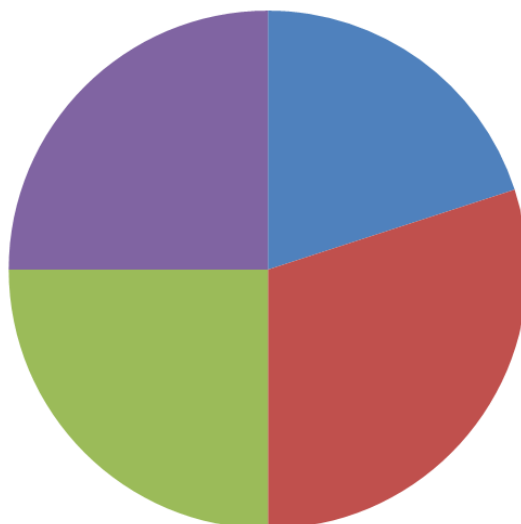
■ Fight Infections  
■ Family  
■ Work  
■ Personal

**Energy Utilization**  
**Low Neutrophil Counts**



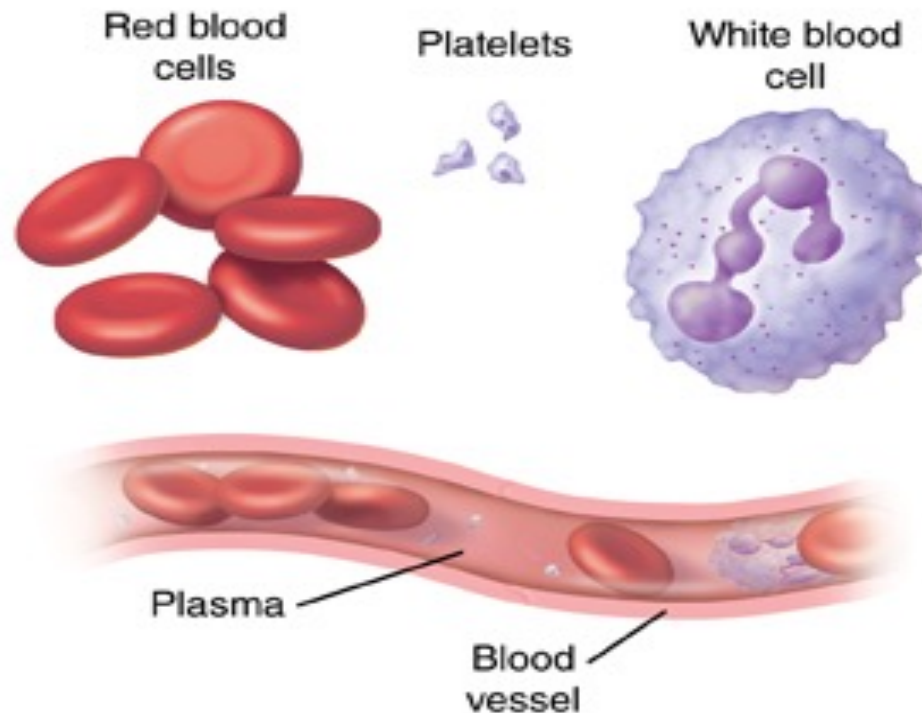
■ Fight Infections  
■ Family  
■ Work  
■ Personal

**Energy Utilization**  
**Normalized Neutrophil Counts**



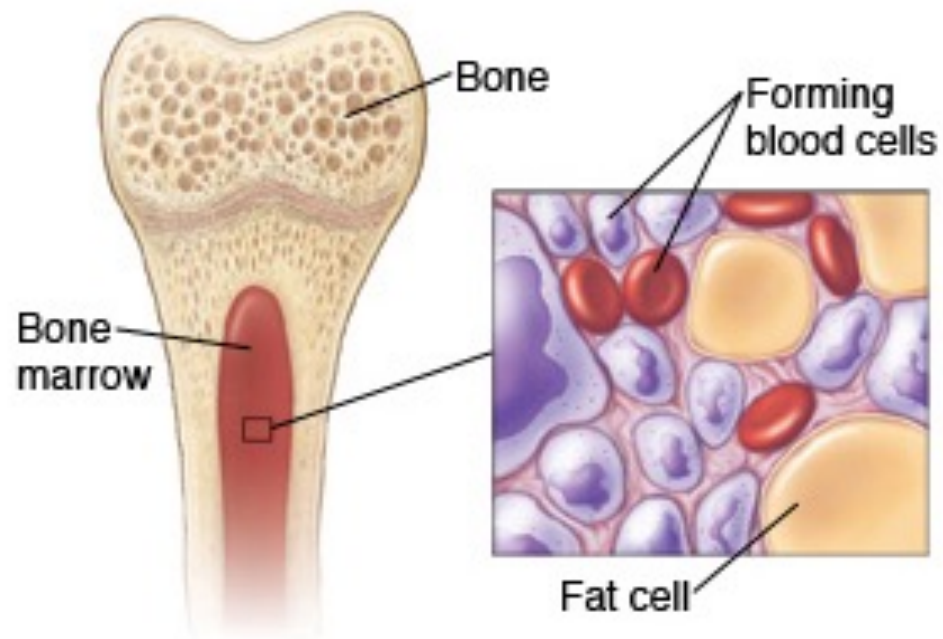
■ Fight Infections  
■ Family  
■ Work  
■ Personal

# Understanding Blood Components

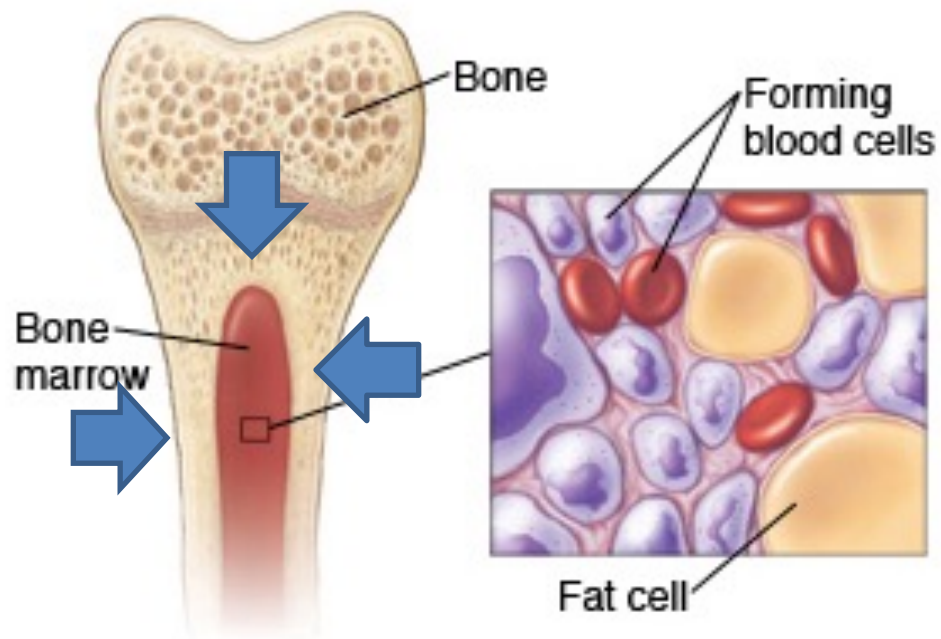


# Neutrophils

## Made in the Bone Marrow



# Bone Pain



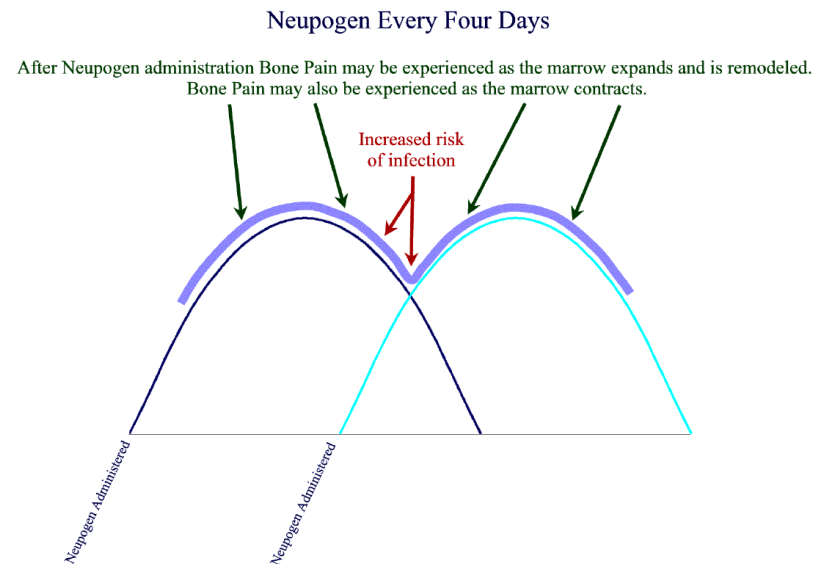
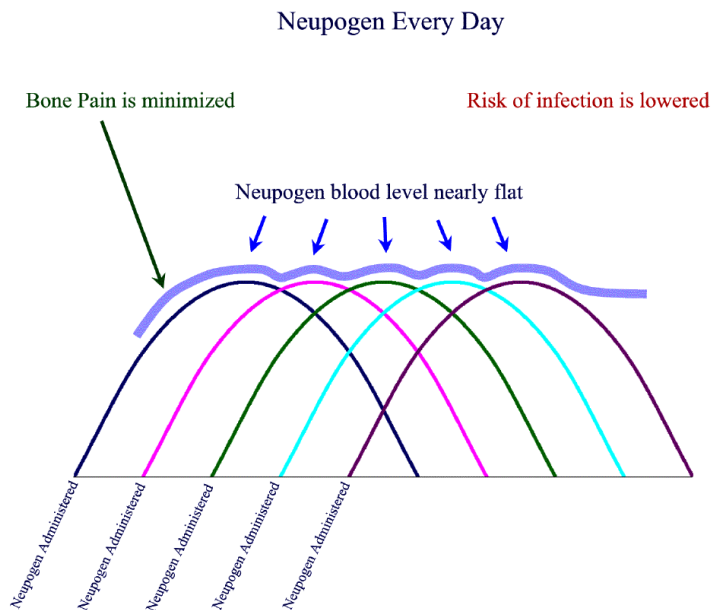
## Bone pain in rhGCSF Recipients (1995)

SG Romanick-Schmiedel, AA Bolyard, DC Dale, VJ Gauthier

- 20 patients
- G-CSF administration for 1-8 years
- Typical Pain
  - Generally lower back, sternal area or femur
  - Described as throbbing or pressure
  - Varies in intensity
  - May last hours or days
  - Coincides with dramatic increase in neutrophil count
  - Resolving as the neutrophil count falls

# Strategies to Reduce Bone Pain

- More frequent G-CSF dosing



- Lowest G-CSF dose possible, does a smaller dose help?
  - Keep records
  - Monitor ANC and Bone Pain

# Fatigue is Real

- Common in people with neutropenia, treated or not with Neupogen
- Strategies
  - Ask for help
    - Family - negotiate family obligations
    - Friends
    - Physician - SSI (disability) or FMLA paperwork
    - Work - do you need a flexible work schedule
  - Define when most tired
    - Morning
    - Evening
    - Is there a time you can rest?
  - Review goals, Reduce stress
    - What has to be done now
    - What can wait to be done
    - What can removed from the list



# Depression is Real

- Common with chronic illness
- Strategies
  - Seek support
    - Friends
    - Family
    - Counseling
  - Find your Joy in Life
    - Books
    - Cooking
    - Crafts
    - Exercise
    - Gardening
    - Music

# Know your G-CSF dose in micrograms

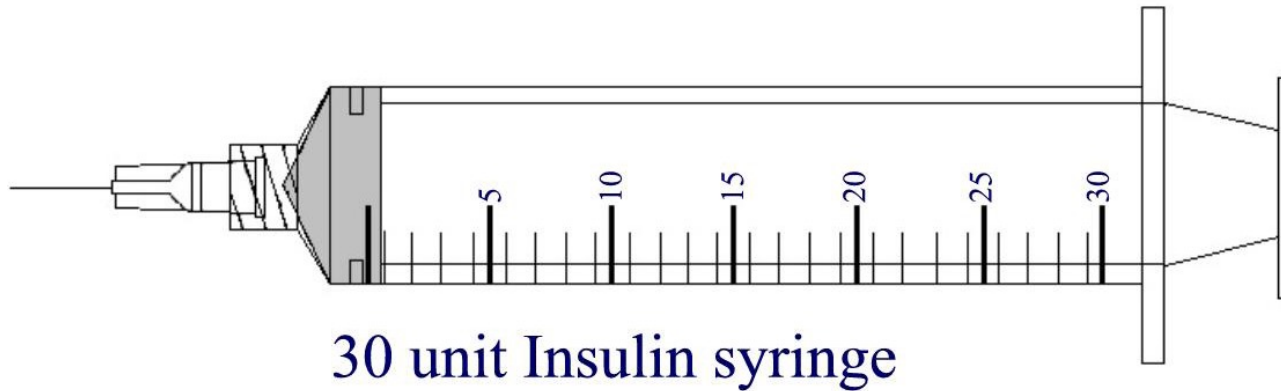
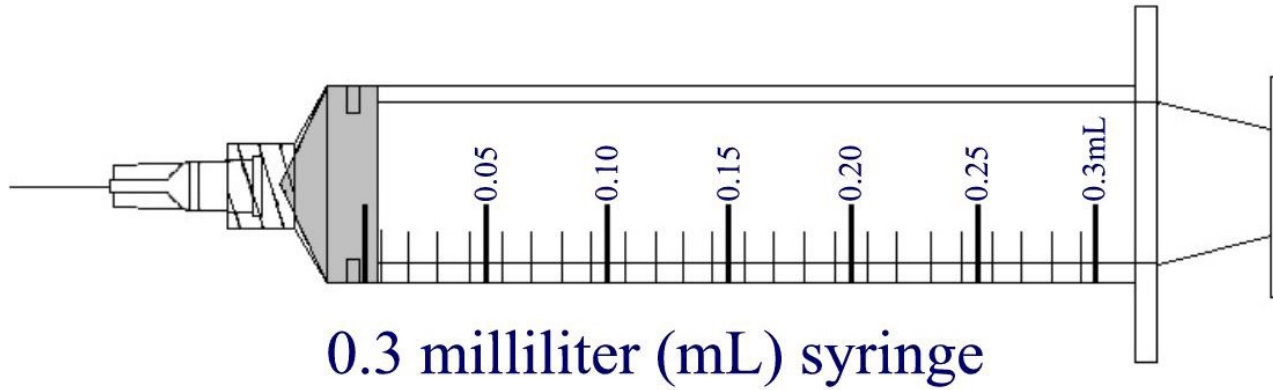
Communicate the dose of G-CSF to health care practitioners

- How often is G-CSF administered
  - Daily
  - Every Other Day
  - Every Third Day
  - Other
  
- Dose of G-CSF in micrograms per injection
  - 0.1 milliliter (ml) = 30 micrograms
  - 10 units = 30 micrograms

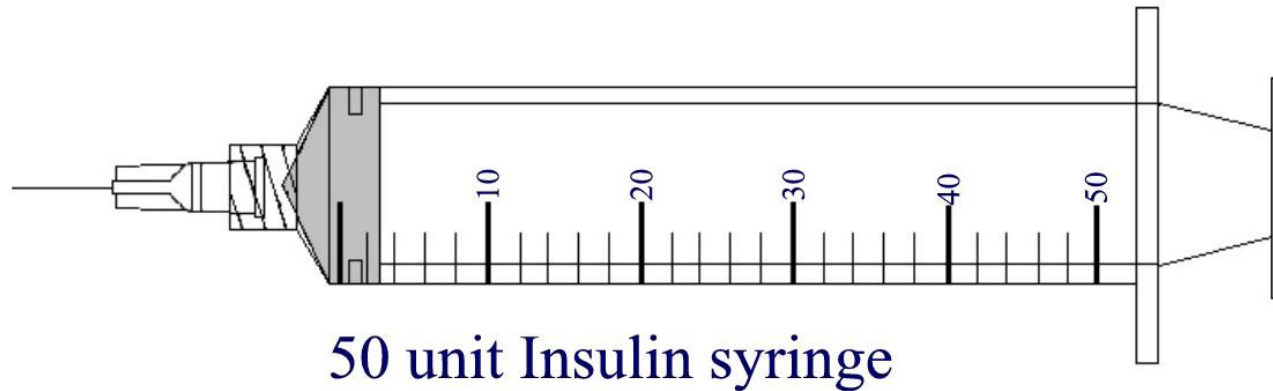
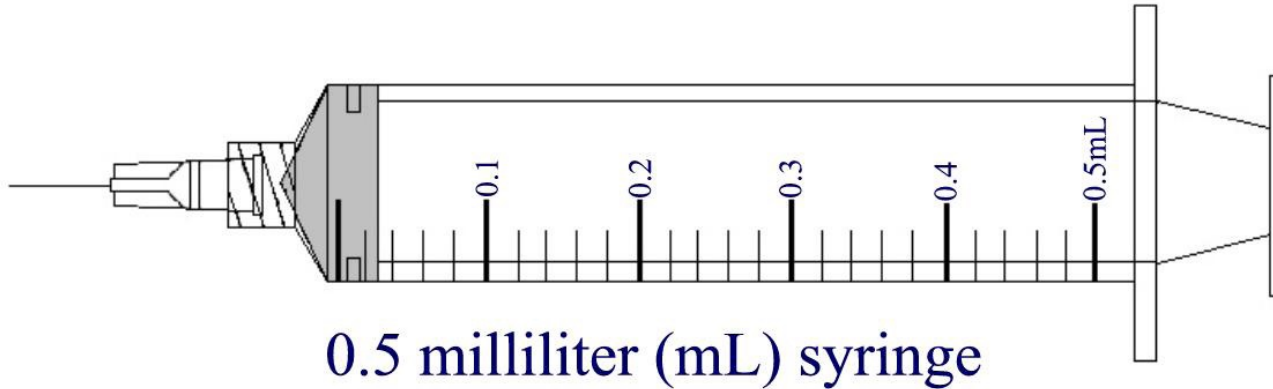
## Neupogen (G-CSF) @ 300 micrograms per milliliter concentration

Micrograms (mcg)	Milliliter (ml) syringe	Insulin syringe
<b>3 micrograms</b>	0.01 milliliters	1 unit
<b>6 micrograms</b>	0.02 milliliters	2 units
<b>9 micrograms</b>	0.03 milliliters	3 units
<b>12 micrograms</b>	0.04 milliliters	4 units
<b>15 micrograms</b>	0.05 milliliters	5 units
<b>18 micrograms</b>	0.06 milliliters	6 units
<b>21 micrograms</b>	0.07 milliliters	7 units
<b>24 micrograms</b>	0.08 milliliters	8 units
<b>27 micrograms</b>	0.09 milliliters	9 units
<b>30 micrograms</b>	0.10 milliliters	10 units
<b>45 micrograms</b>	0.15 milliliters	15 units
<b>60 micrograms</b>	0.20 milliliters	20 units
<b>90 micrograms</b>	0.30 milliliters	30 units
<b>120 micrograms</b>	0.40 milliliters	40 units
<b>150 micrograms</b>	0.50 milliliters	50 units
<b>180 micrograms</b>	0.60 milliliters	60 units
<b>240 micrograms</b>	0.80 milliliters	80 units
<b>300 micrograms</b>	1.00 milliliters	100 units

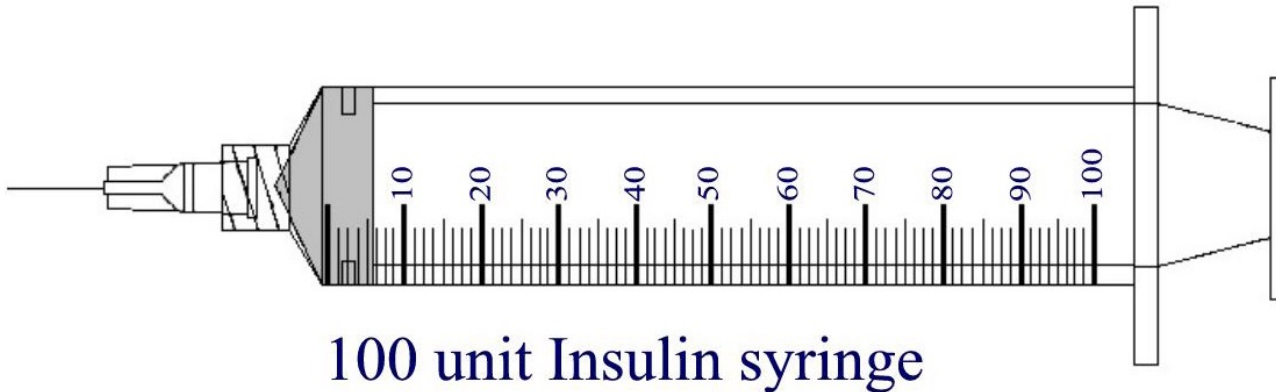
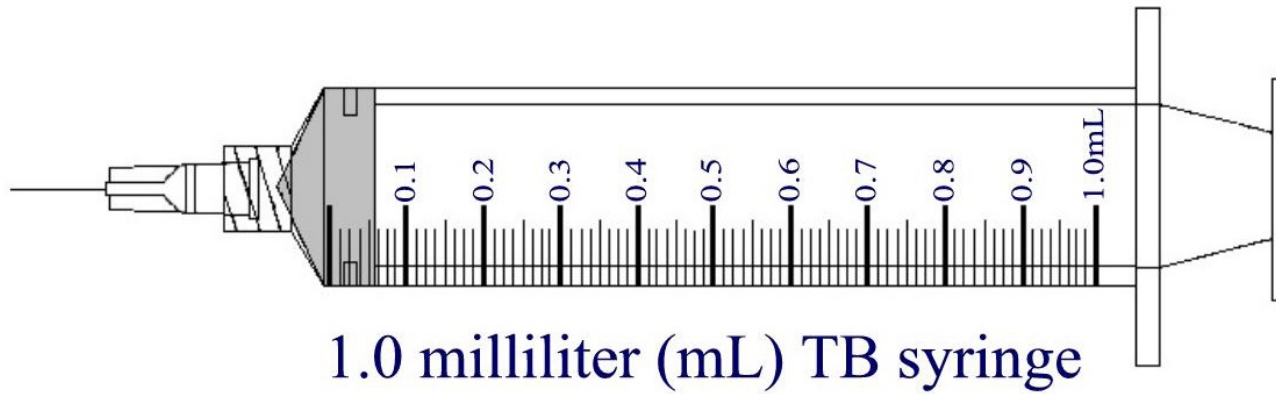
# Syringes



# Syringes



# Syringes



# How to determine your dose of G-CSF in Micrograms

## Which syringe do you use

Insulin syringe	Micrograms (mcg)
1 unit	3 micrograms
2 units	6 micrograms
3 units	9 micrograms
4 units	12 micrograms
5 units	15 micrograms
6 units	18 micrograms
7 units	21 micrograms
8 units	24 micrograms
9 units	27 micrograms
10 units	30 micrograms

Milliliter (ml) syringe	Micrograms (mcg)
0.01 milliliters	3 micrograms
0.02 milliliters	6 micrograms
0.03 milliliters	9 micrograms
0.04 milliliters	12 micrograms
0.05 milliliters	15 micrograms
0.06 milliliters	18 micrograms
0.07 milliliters	21 micrograms
0.08 milliliters	24 micrograms
0.09 milliliters	27 micrograms
0.10 milliliters	30 micrograms

# Take Aways

- Review Information located on the Website
- Know your G-CSF dose in micrograms
- Keep information in a binder for doctor appointments or when you travel
- Contact Amgen SupportPlus