



SCID
Compass
An educational program of the
Immune Deficiency Foundation

B-cell Reconstitution and IgG Infusion for SCID

August 31st, 2022



IDF MISSION

Improving the diagnosis, treatment, and quality of life of people affected by primary immunodeficiency through fostering a community empowered by advocacy, education and research.



HOUSEKEEPING

- Attendees will not have access to their microphone or webcam throughout the event.
- To see the full slides, you can adjust the settings on the speaker view panel on the top of the Zoom screen and select "side-by-side" in the dropdown option.
- Please submit all questions for the presenter via the Q&A box

DISCLAIMER

Immune Deficiency (IDF) education events offer a wide array of educational presentations, including presentations developed by healthcare and life management professionals invited to serve as presenters. The views and opinions expressed by guest speakers do not necessarily reflect the views and opinions of IDF.

The information presented during this event is not medical advice, nor is it intended to be a substitute for medical advice, diagnosis or treatment. Always seek the advice of a physician or other qualified health provider with questions concerning a medical condition. Never disregard professional medical advice, or delay seeking it based on information presented during the event.



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Where are you in your SCID journey?

Wherever you are on your journey with Severe Combined Immune Deficiency (SCID), use the links below to find the information and support you need.

[SCID Compass Home / SCID Compass Home](#)



UNDERSTAND SCID

Go here if you're just getting started.

[Learn More](#)



EXPLORE TREATMENT OPTIONS

Go here to learn more about treatment options.

[Learn More](#)



NAVIGATE HOSPITAL STAY

Find out what to expect as your child undergoes treatment.

[Learn More](#)

¿En qué parte del trayecto de la IDCG está?

Dondequiera que esté en el trayecto de la inmunodeficiencia combinada grave (IDCG), use los siguientes enlaces para buscar la información y el apoyo que necesita.

[Scid Compass / SCID Compass Home](#)



ENTENDER LA IDCG

Vaya aquí si está comenzando.

[Más Información](#)



EXPLORAR LAS OPCIONES DE TRATAMIENTO

Vaya aquí para aprender más sobre las opciones de tratamiento.

[Más Información](#)



ORIENTARSE SOBRE LA ESTADÍA EN EL HOSPITAL

Descubra qué esperar cuando su hijo comience el tratamiento.

[Más Información](#)

B cell reconstitution and IgG replacement

SCID Compass Lunch & Learn

Manish Butte, MD/PhD, UCLA Health

Victoria Dimitriades, MD, UC Davis Health

SCID

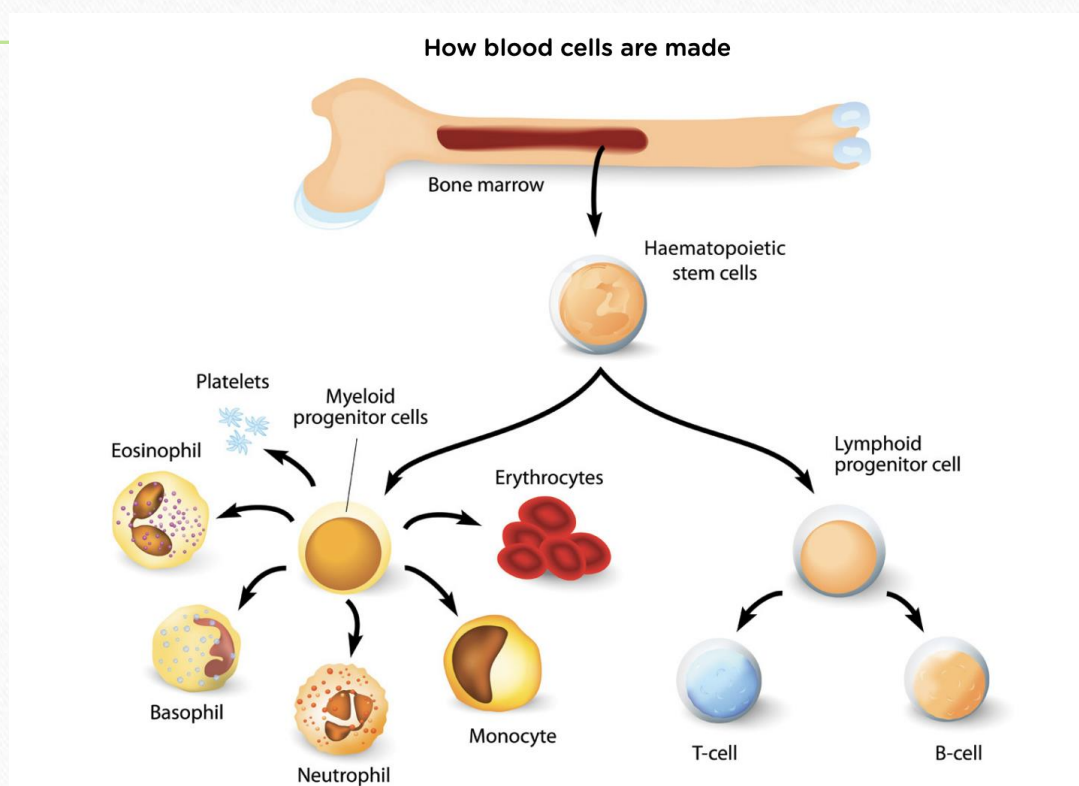
How we started



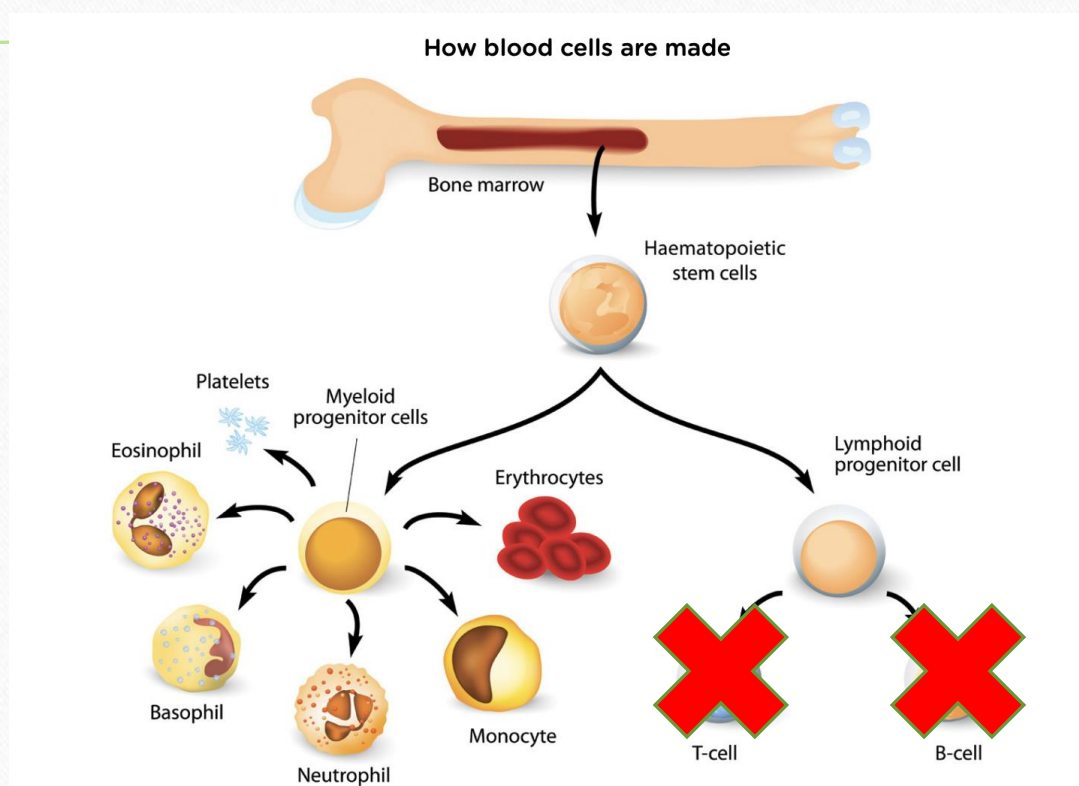
How it's going



Immune system development from bone marrow stem cells

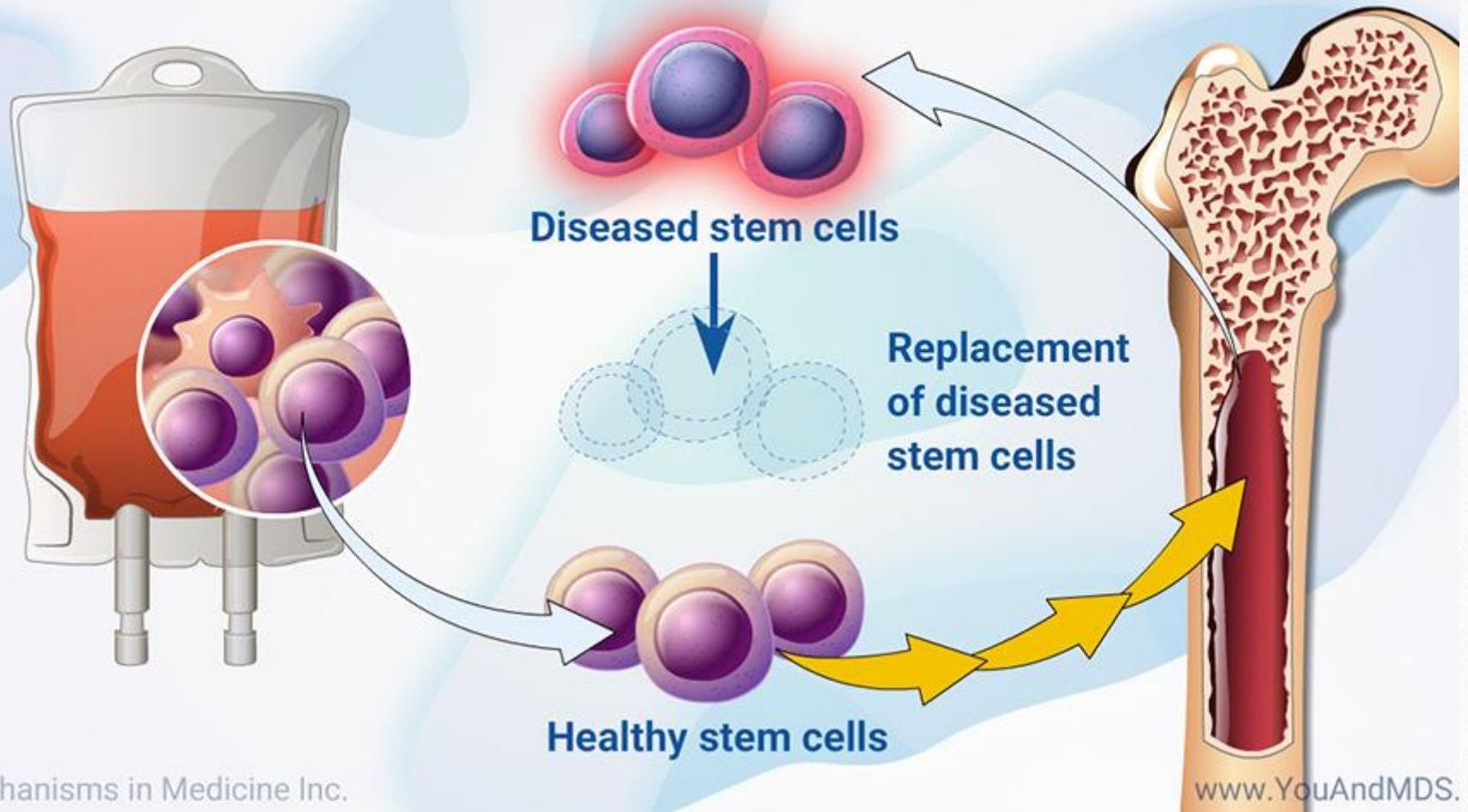


Immune system **with SCID**



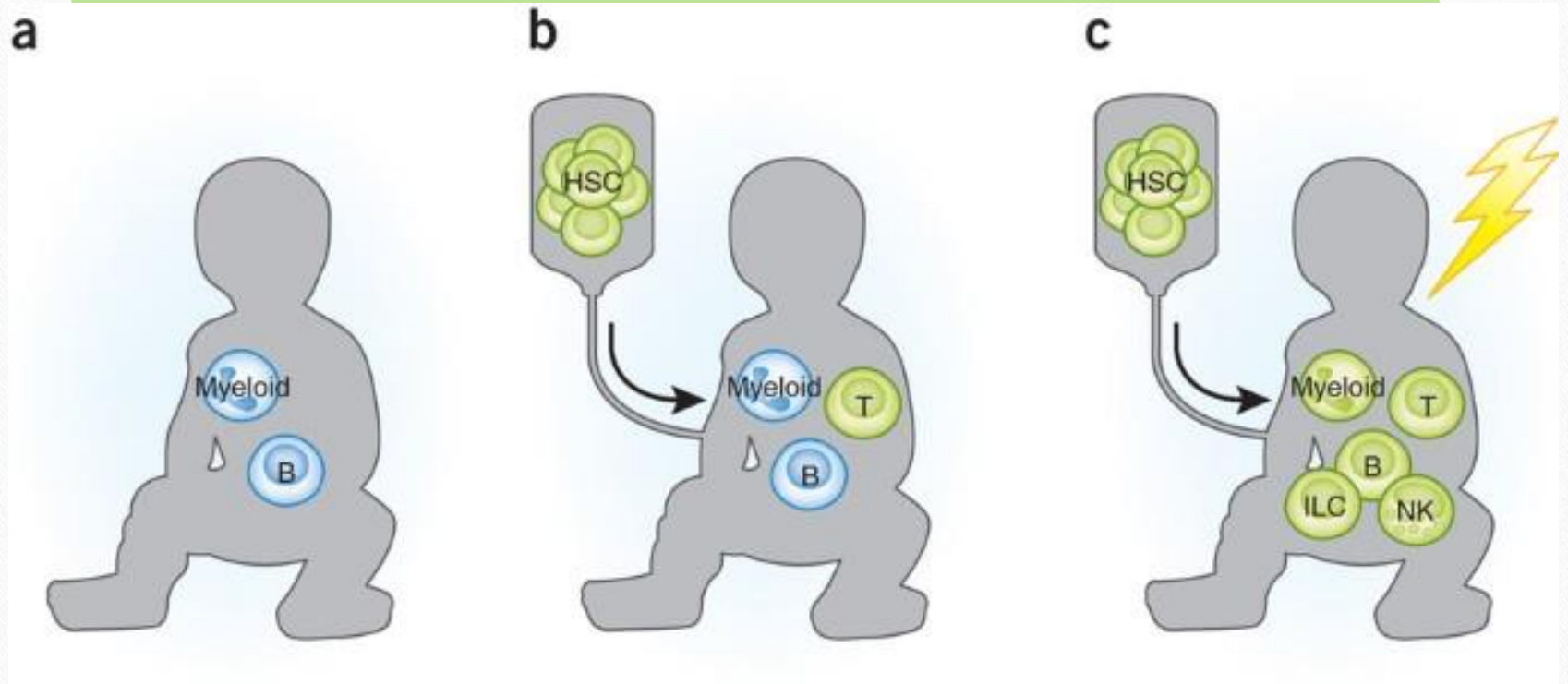


What is blood and marrow transplant (BMT)?

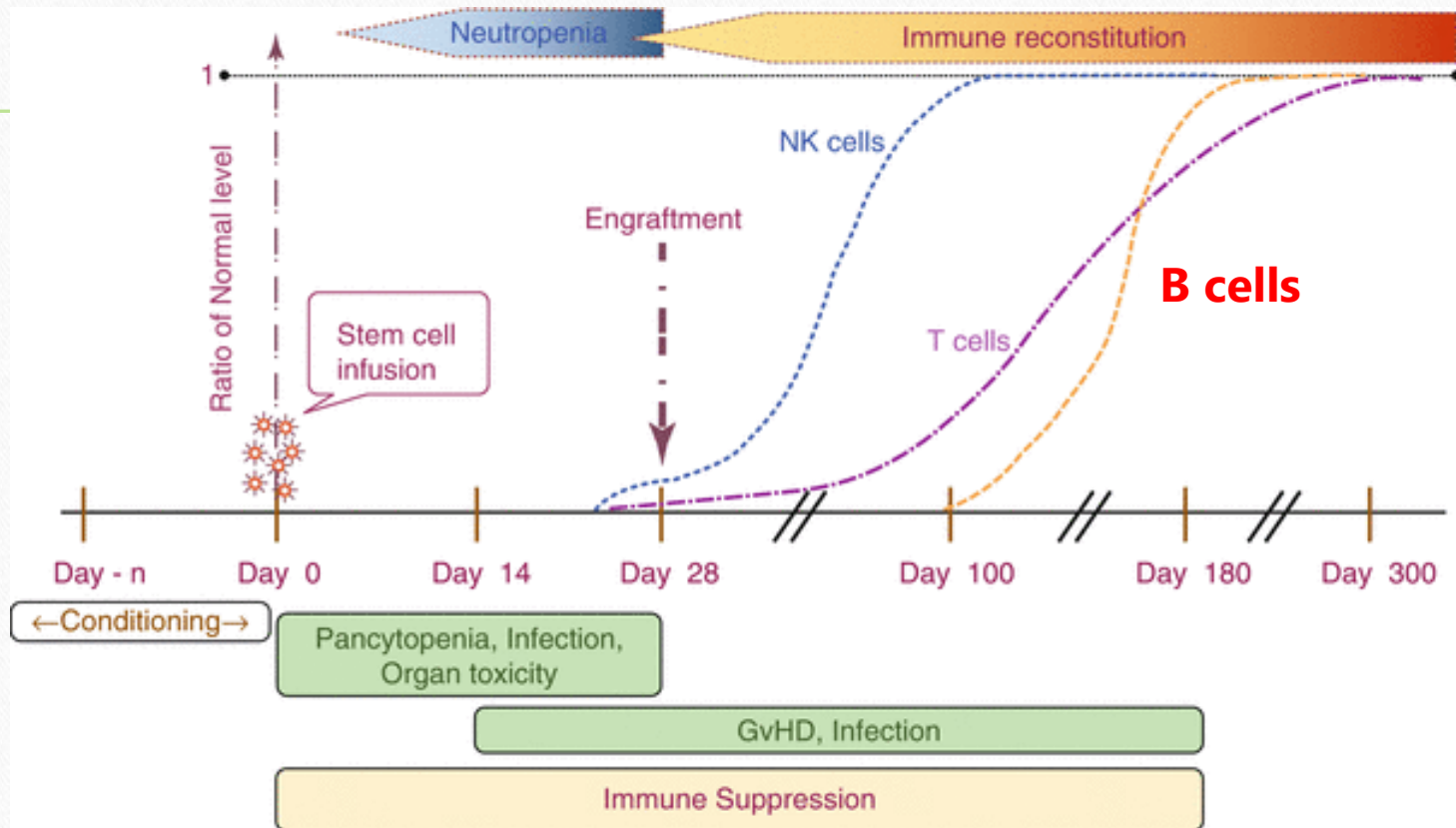


Immune expectations after transplant

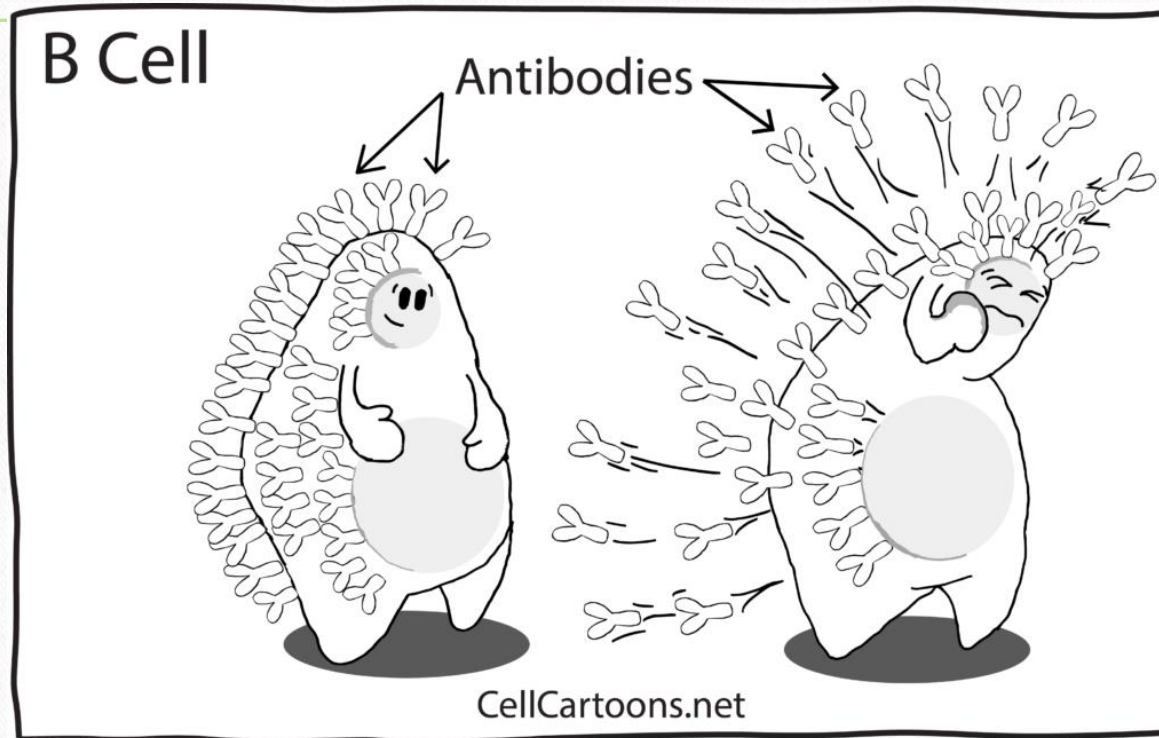
SCID: no T cells



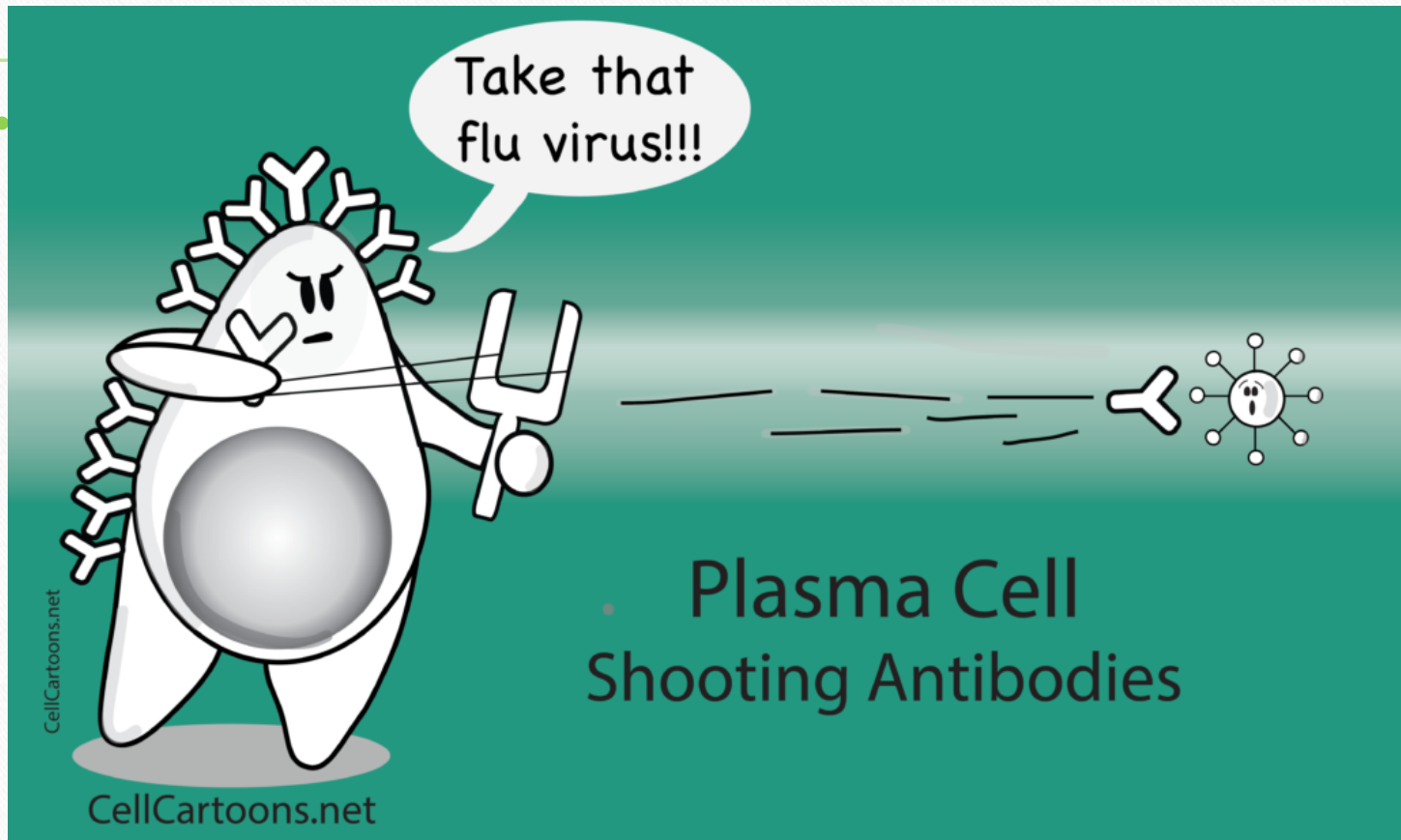
After transplant reconstitution timeline



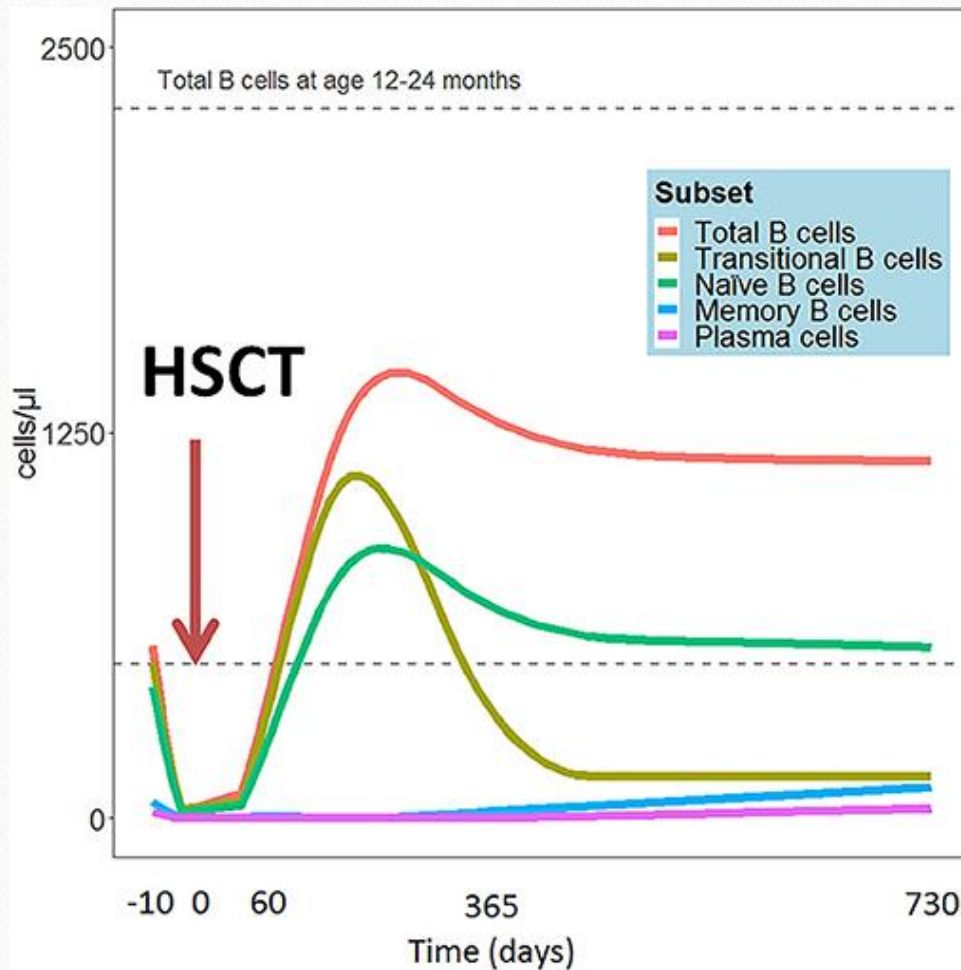
B cells make antibodies



B cells mature to plasma cells



B cell reconstitution after transplant

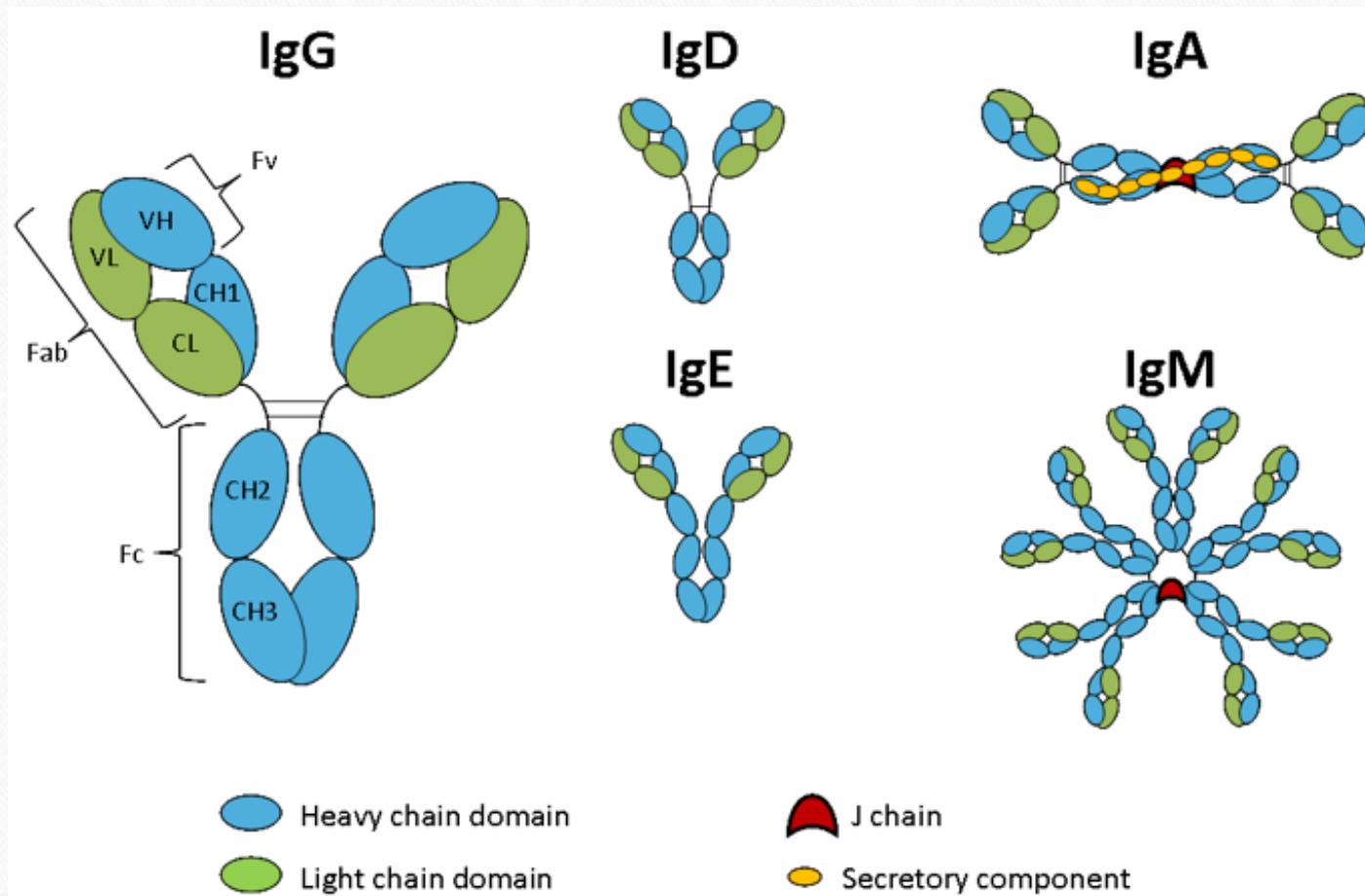


- Overshoot of B cells is common (partially from graft itself)
- Memory B cells are low for 2 years
- Vaccine responses are often very low for 1-2 years

What are the factors that influence B cell reconstitution after SCID BMT?

- Genetic cause of SCID? Yes
- Conditioning regimen? Yes (71% with, 15% without)
- Can you have functioning B cells without conditioning? Yes but unlikely
- Is busulfan needed to get B cell reconstitution? Jury is still out, likely yes busulfan helps a lot

The different types of immunoglobulins



	IgA	IgD	IgE	IgG	IgM
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% total Ig in serum	10-20	<1	<1	70-85	10
Where found in body	Found in bodily secretions.	Found on B-cell surface	Attach to basophils and mast cells.	Blood & extracellular fluid	Blood & extracellular fluid
Functions	Protect external openings	Unknown; maybe antigen detection	Allergic response and defend infection by large parasite	Long term Ab that protect the body	Appear earlier in the infection and offer valuable defense during critical stage of the infection

IgG Replacement Facts

- Products are meant to replace only the IgG component of the immune system protection (not IgA or IgM)
- Generally, IgG antibodies protect you from recurrent infections of the ears, sinuses, and lungs
 - IgG supplementation contains many kinds of antibodies to viruses as well, but this tends not to be the main *impact* of replacement



How it's made

- IgG is a blood product; it utilizes up to 1000 different donors to make a pool of antibodies
 - Donors must be screened and their first donation is held; they return for a second screening 3-6 months later, and then donations can be released.
- Ultrafiltration process is different between companies, but the products are generally very similar
 - Some patients need one specific type of product, but most patients do well on any of the products

Dosing

- Based initially on the weight of the patient (usually around 500 mg for every kg of weight)
- Adjusted based on how well the patient is protected from infections
 - Are they having issues with ear, sinus, lung infections?
 - Are they having symptoms which are towards the end of their window, just before their next treatment?

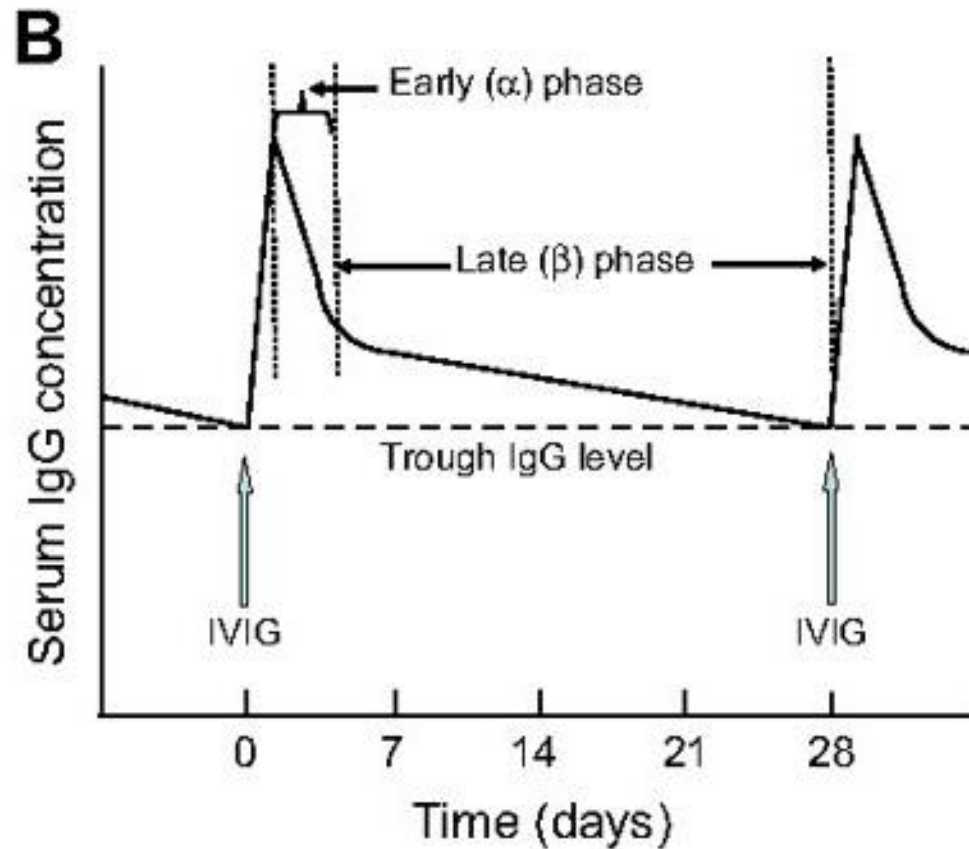
Intravenous Immunoglobulin (IVIg)

- In-the-vein therapy

- Needs IV placement each month (may use indwelling catheters in the beginning)
- Sometimes premedication is given, though not always necessary; topical anesthesia can be used
- Hydration prior to infusion is important to reduce side effects
- Usually given every 3-4 weeks, depending on needs
 - Can take 3-4 hours for infusion appointment- bring distractions!



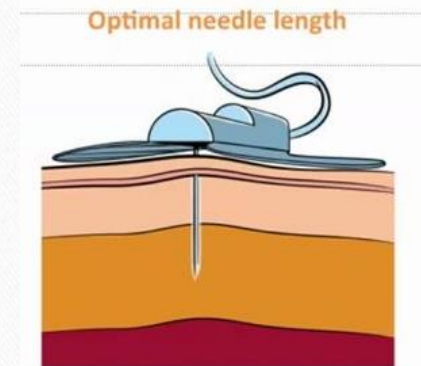
Intravenous Immunoglobulin (IVIg)



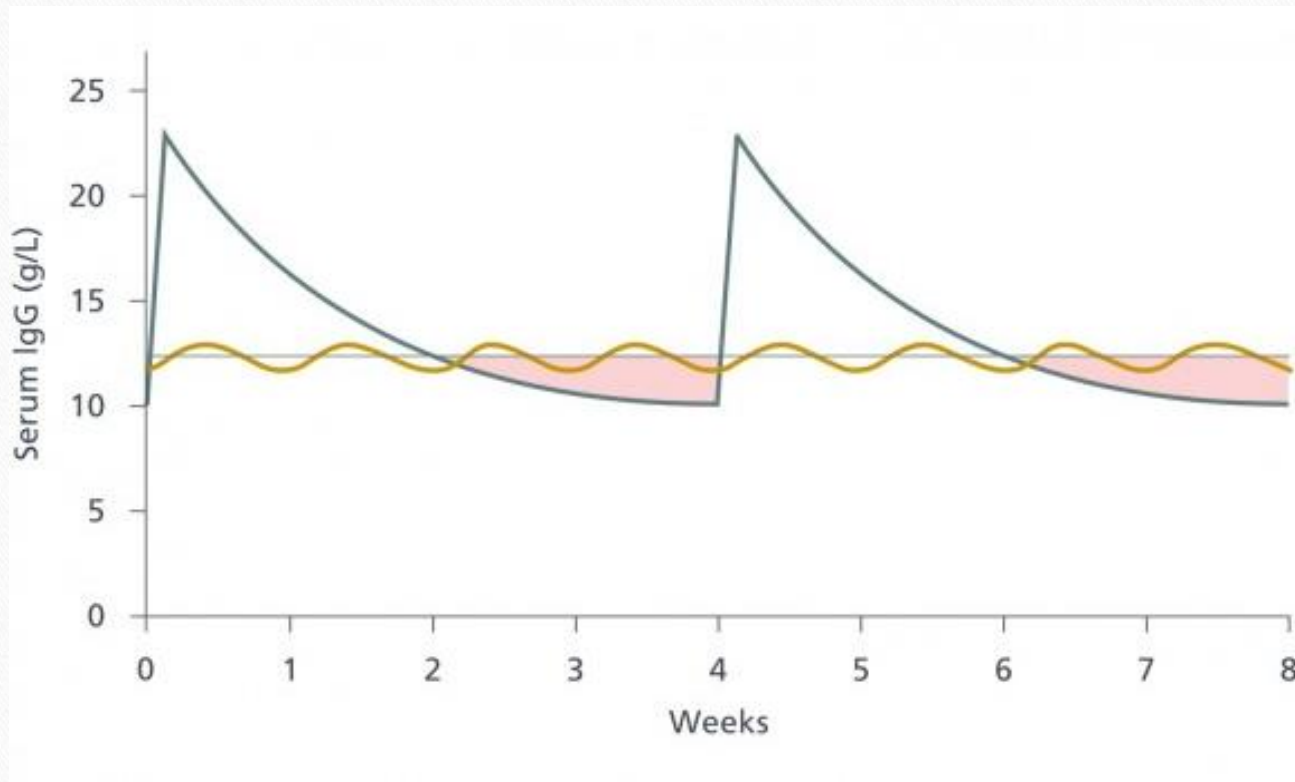
Subcutaneous Immunoglobulin (SCIg)

- Under-the-skin therapy

- Can be done at home with needle placement by a caregiver (can keep medicine refrigerated at home)
- Sometimes premedication is given, though not always necessary; topical anesthesia can be used
- Usually given every 1-2 weeks, depending on needs
 - Lasts 30 minutes to 2 hours, depending on dosing and product
- Can adjust locations, number of sites
- Need to be able to schedule yourself consistently



Subcutaneous Immunoglobulin (SCIg)



Following Up



- Labs: Blood counts, chemistry, IgG level every 6-12 months (may be more frequent with your team)
- Review any infections
 - Adjust dosing if needed
- Review IgG side-effects
 - headaches, fatigue, local reactivity, time to resolve skin swelling, etc.
- Remember:
 - You cannot test for antibodies to vaccines or for infections while on IgG replacement, as this will reflect what is in the therapy
 - Sometimes you can have false-positive antibody testing to thyroid or anti-nuclear antibodies (ANA), which are common in the population

Side Effects

- IVIg:
 - headache, which occurs at a frequency of 8%–15%
 - Infrequent: fatigue, aches, fever/chills, and nausea/diarrhea
- SCIg
 - Headache at rates of 1.4%–4%
 - Fatigue, etc, is at a much lower rate
 - Local adverse events such as discomfort and pain, erythema, swelling/edema, and pruritis
 - Often can be modified by needle and location adjustments



Vaccines while on IgG replacement

- ****Check with your team first****
- In general, once cleared to restart non-live vaccines, yearly influenza is also recommended for all patients on IgG replacement
- Live vaccines should be specifically recommended by your care team
 - Caregivers should be vaccinated, in general, to protect our patients; In cases where caregivers are receiving live vaccines, they should check with the team first

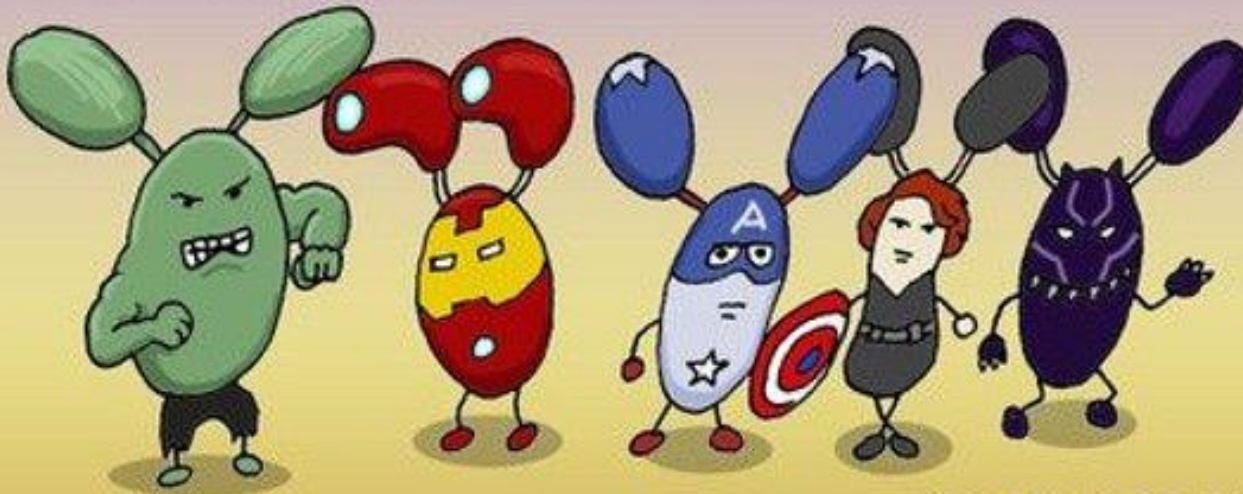
To Sum Up...

- Supplementation is important until the B cells can return to making antibodies again
- There is flexibility in how you receive IgG replacement which can keep you safe and still be compatible with your daily activities
- The goal is to eventually be able to stop needing IgG infusions once the immune system is replenished 😊

Dzu-Doodles

ANTIBODIES

AFFINITY WAR



THE
BULK

IgG Man

CAPTAIN
IMMUNOGLOBULIN

V(D)J WIDOW

BLACKANTIGEN

THANK YOU!

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**Q&A SESSION:
YOUR QUESTIONS ANSWERED**

**Have more
Questions?**



primaryimmune.org/ask-idf

800-296-4433



NEXT PROGRAM

**SCID Compass Lunch & Learn:
ADA SCID Gene Therapy
Update with Donald Kohn, MD**

**September 14th, 2022
2:00-3:00 PM ET**
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www.scidcompass.org/events