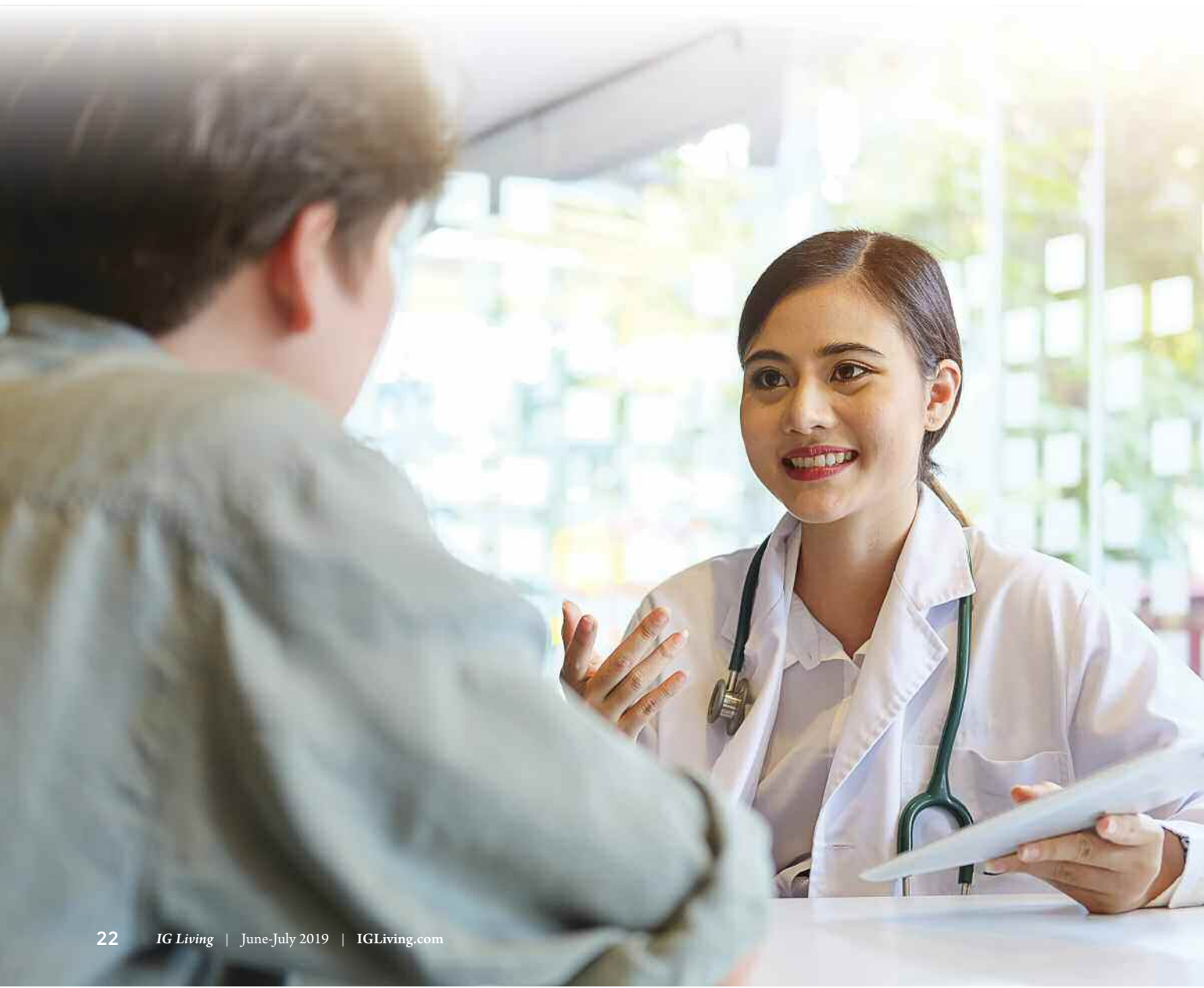


Organizing the Chronically Ill Chaos

This system of organizing medical data will help doctors and specialists understand a patient's current state of care.

By Denna McGrew



ORGANIZED CHAOS is honestly the best I can hope for most days. By the numbers, my family is as complex as it gets since three of the four of us are chronically ill. Together, we have:

- 6 rare diseases
- 15 other major diagnoses
- 2 primary care physicians
- 14 specialists
- 8 to 12 monthly treatment days
- 4 to 6 doctor visits per month
- 10,000 miles travelled annually to see specialists

That’s a lot to manage for anyone! As such, I’ve learned a few things along the 17-year journey of our chronic life, including two main strategies for getting the best possible outcomes.

The first strategy, and most important, is to find the right specialist for your own disease. For our family, that means travel. I cannot overstate the importance of finding the doctor for whom your rare disease is of research interest. This “unicorn” of a specialist is one who has dedicated his or her career to learning about and treating your disease (even though this disease is one that most doctors need to Google).

Once you have that unicorn specialist on board, you must implement the second strategy: Arm him or her with your medical information in a way that makes it easy to understand and utilize. I’ve developed a system that works for my family and one that our doctors report finding useful. In fact, a couple of them have commented that my son wouldn’t be alive today without this system.

In the rare disease community, patients see many specialists who may be located all over the country. Busy doctors do not have the time to keep each other in the loop, so that arduous chore falls to the patient and/or caregiver (such as the patient’s parent). I think of it as being a little like football: The specialist is the coach and calls the plays, whereas the patient or caregiver is the quarterback, implementing and carrying out the plays determined by the coach. Rare diseases further complicate matters since there are multiple coaches, so it’s our job to make sure they have all the information so they can call the right plays.

My system involves keeping a single document in Microsoft Word that I update before every appointment and hand a hard copy to the doctor (Figure). Here are the basic components:

1) *Header*: In the header section of the document, include the patient’s name, date of birth and the date on which the document was created or most recently updated/ revised, as well as the page number and number of pages. This header will automatically be present on every page, which helps if the pages get separated. If the person preparing the document is someone other than the patient, consider adding that information to the header as well. For example: Prepared by Denna McGrew, Jenna’s mother.

Figure. Medical Information Sheet

Patient Name DOB 0/0/0000 Created/Revised 0/0/0000; Page 1 of 1	<u>1</u>
DIAGNOSES Diagnosis (date) Diagnosis (date)	<u>2</u>
MEDICATIONS <u>Prescription:</u> Drug, dosage, frequency <u>PRN (as needed):</u> Drug, dosage, frequency <u>OTC (over the counter):</u> Drug, dosage, frequency <u>Recently tried medications:</u> Drug, dosage, frequency and reason stopped	<u>3</u>
ALLERGIES Drug and reaction (anaphylaxis, hives, etc.) Environmental allergies and reaction Food allergies and reaction	<u>4</u>
DOCTORS Name, specialty, location, phone/fax/email	<u>5</u>
MAJOR SYMPTOMS Symptom Symptom	<u>6</u>
NARRATIVE (reverse chronological order) 0/0/0000 Summarize medical event in short form such as doctor visit, lab performed, medications prescribed and reason 0/0/0000 Summarize medical event in short form such as doctor visit, lab performed, medications prescribed and reason	<u>7</u>
Developed by Denna McGrew; denna@dennamcgreww.com	

Hizentra[®]

Immune Globulin Subcutaneous
(Human) 20% Liquid

1st and only Self-administered Ig for CIPD Maintenance

#1 Ig Prescribed for PI

**My Life,
My Way
With
Hizentra**

Hizentra is an Ig* therapy that provides proven protection for PI and CIPD with the convenience of self-administration, so you can focus on everyday living

*Ig=immunoglobulin

Important Safety Information

WARNING: Thrombosis (blood clots) can occur with immune globulin products, including Hizentra. Risk factors can include: advanced age, prolonged immobilization, a history of blood clotting or hyperviscosity (blood thickness), use of estrogens, installed vascular catheters, and cardiovascular risk factors.

If you are at high risk of blood clots, your doctor will prescribe Hizentra at the minimum dose and infusion rate practicable and will monitor for signs of clotting events and hyperviscosity. Always drink sufficient fluids before infusing Hizentra.

See your doctor for a full explanation, and the full prescribing information for complete boxed warning.

Hizentra is a prescription medicine used to treat:

- Primary immune deficiency (PI) in patients 2 years and older
- Chronic inflammatory demyelinating polyneuropathy (CIPD) in adults

Treatment with Hizentra might not be possible if your doctor determines you have hyperprolinemia (too much proline in the blood), or are

IgA-deficient with antibodies to IgA and a history of hypersensitivity. Tell your doctor if you have previously had a severe allergic reaction (including anaphylaxis) to the administration of human immune globulin. Tell your doctor right away or go to the emergency room if you have hives, trouble breathing, wheezing, dizziness, or fainting. These could be signs of a bad allergic reaction.

Inform your doctor of any medications you are taking, as well as any medical conditions you may have had, especially if you have a history of diseases related to the heart or blood vessels, or have been immobile for some time. Inform your physician if you are pregnant or nursing, or plan to become pregnant.

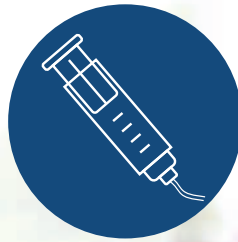
Infuse Hizentra under your skin *only*; do not inject into a blood vessel. Self-administer Hizentra only after having been taught to do so by your doctor or other healthcare professional, and having received dosing instructions for treating your condition.

Why Choose Hizentra?



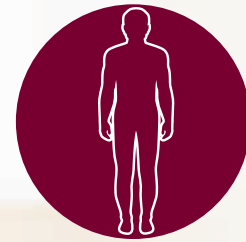
Choose where you infuse

Self-administration with Hizentra means you and your doctor can decide where you can infuse. Convenient dosing routines mean you won't have to adjust or cancel your plans due to IV infusion appointments.



No IV infusions

IV infusions can be challenging for people who have hard-to-find or damaged veins. Hizentra allows you to infuse just under the skin, not into a vein, after training from your doctor.



Proven safety

Hizentra has an established safety profile and demonstrated tolerability. In clinical trials, the most common side effects were redness, swelling, itching, and/or bruising at the infusion site; headache, chest, joint or back pain; diarrhea; tiredness; cough; rash; itching; fever, nausea, and vomiting. These are not the only side effects possible.



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Immediately report to your physician any of the following symptoms, which could be signs of serious adverse reactions to Hizentra:

- Reduced urination, sudden weight gain, or swelling in your legs (possible signs of a kidney problem).
- Pain and/or swelling or discoloration of an arm or leg, unexplained shortness of breath, chest pain or discomfort that worsens on deep breathing, unexplained rapid pulse, or numbness/weakness on one side of the body (possible signs of a blood clot).
- Bad headache with nausea; vomiting; stiff neck; fever; and sensitivity to light (possible signs of meningitis).
- Brown or red urine; rapid heart rate; yellowing of the skin or eyes; chest pains or breathing trouble; fever over 100°F (possible symptoms of other conditions that require prompt treatment).

Hizentra is made from human blood. The risk of transmission of infectious agents, including viruses and, theoretically, the Creutzfeldt-Jakob disease (CJD) agent and its variant (vCJD), cannot be completely eliminated.

The most common side effects in the clinical trials for Hizentra include redness, swelling, itching, and/or bruising at the infusion site; headache; chest, joint or back pain; diarrhea; tiredness; cough; rash; itching; fever, nausea, and vomiting. These are not the only side effects possible. Tell your doctor about any side effect that bothers you or does not go away.

Before receiving any vaccine, tell immunizing physician if you have had recent therapy with Hizentra, as effectiveness of the vaccine could be compromised.

Please see brief summary of full prescribing information for Hizentra on adjacent page. For full prescribing information, including boxed warning and patient product information, please visit Hizentra.com.

You are encouraged to report negative side effects of prescription drugs to the FDA. Visit www.fda.gov/medwatch, or call 1-800-FDA-1088.

IgIQ: Support Programs to Help You Get Started

Call IgIQ to find out about programs such as:



Free 1-month supply of Hizentra and free at-home nurse visits



Most patients with commercial insurance pay \$0 out-of-pocket*



Protection if you lose insurance*



Your single source for Ig solutions

1-877-355-IGIQ (4447)

*Other terms and conditions apply. Call IgIQ to learn more.

HIZENTRA®, Immune Globulin Subcutaneous (Human), 20% Liquid
Initial U.S. Approval: 2010

BRIEF SUMMARY OF PRESCRIBING INFORMATION

These highlights do not include all the information needed to use HIZENTRA safely and effectively. See full prescribing information for HIZENTRA.

WARNING: THROMBOSIS

See full prescribing information for complete boxed warning.

- **Thrombosis may occur with immune globulin products, including HIZENTRA. Risk factors may include: advanced age, prolonged immobilization, hypercoagulable conditions, history of venous or arterial thrombosis, use of estrogens, indwelling vascular catheters, hyperviscosity, and cardiovascular risk factors.**
- **For patients at risk of thrombosis, administer HIZENTRA at the minimum dose and infusion rate practicable. Ensure adequate hydration in patients before administration. Monitor for signs and symptoms of thrombosis and assess blood viscosity in patients at risk for hyperviscosity.**

INDICATIONS AND USAGE

HIZENTRA is indicated for:

*Treatment of primary immunodeficiency (PI) in adults and pediatric patients 2 years and older.

* Maintenance therapy in adults with chronic inflammatory demyelinating polyneuropathy (CIDP) to prevent relapse of neuromuscular disability and impairment.

-Limitation of Use: Maintenance therapy in CIDP has been systematically studied for 6 months and for a further 12 months in a follow-up study. Continued maintenance beyond these periods should be individualized based on patient response and need for continued therapy.

For subcutaneous infusion only.

DOSAGE FORMS AND STRENGTHS

0.2 g per mL (20%) protein solution for subcutaneous injection

CONTRAINDICATIONS

- Anaphylactic or severe systemic reaction to human immune globulin or components of HIZENTRA, such as polysorbate 80
- Hyperprolinemia (type I or II) (HIZENTRA contains the stabilizer L-proline)
- IgA-deficient patients with antibodies against IgA and a history of hypersensitivity

WARNINGS AND PRECAUTIONS

- IgA-deficient patients with anti-IgA antibodies are at greater risk of severe hypersensitivity and anaphylactic reactions.
- Thrombosis may occur following treatment with immune globulin products, including HIZENTRA.
- Aseptic meningitis syndrome has been reported with IGIV or IGSC, including HIZENTRA treatment.
- Monitor renal function, including blood urea nitrogen, serum creatinine, and urine output in patients at risk of acute renal failure.
- Monitor for clinical signs and symptoms of hemolysis.
- Monitor for pulmonary adverse reactions (transfusion-related acute lung injury [TRALI])
- HIZENTRA is made from human plasma and may contain infectious agents, e.g., viruses, the variant Creutzfeldt-Jakob disease (vCJD) agent and, theoretically, the Creutzfeldt-Jakob disease (CJD) agent.

ADVERSE REACTIONS

The most common adverse reactions observed in ≥5% of study subjects were local infusion site reactions, headache, diarrhea, fatigue, back pain, nausea, pain in extremity, cough, upper respiratory tract infection, rash, pruritus, vomiting, abdominal pain (upper), migraine, arthralgia, pain, fall and nasopharyngitis.

To report SUSPECTED ADVERSE REACTIONS, contact CSL Behring Pharmacovigilance at 1-866-915-6958 or FDA at 1-800-FDA-1088 or www.fda.gov/medwatch.

DRUG INTERACTIONS

The passive transfer of antibodies may interfere with the response to live virus vaccines, and lead to misinterpretation of the results of serological testing.

Based on March 2018 revision

2) *Diagnoses*: In this section, all the patient's diagnoses should be listed followed by the year in which the diagnosis was made. This section can be in reverse chronological order or in order of the severity of the diagnoses (my preference).

3) *Medications*: This section potentially contains four separate components. The first is prescriptions and should include all medications taken on a routine or scheduled basis. Next, list medications taken on an as-needed basis. Then, list all over-the-counter medications, vitamins and supplements. Finally, list any medications that have been tried and stopped, and indicate the reasons they were stopped and the dates. Each of these sections should include the same information about the medication: drug name, dosage, frequency and route of administration. For example: Armour Thyroid, 120 mg, one per day in the morning; Gammagard IVIG, 20 grams every two weeks via IV.

4) *Allergies*: For those with known allergies, this section is quite important. List the allergen and the reaction. Include drugs/medications, environmental allergens, foods, medical supplies such as latex or adhesives. Note the specific reaction the patient has when exposed to each individual allergen.

5) *Doctors*: List each doctor or specialist. Include their specialty, location and contact information such as address, phone and fax number, as well as their email if available. This facilitates specialists being able to contact one another should they choose to do so.

6) *Major symptoms*: Indicate the major presenting symptoms in this section. Include any problems that cause the patient difficulties in daily life such as severe fatigue, recurrent infections, syncope episodes and tachycardia.

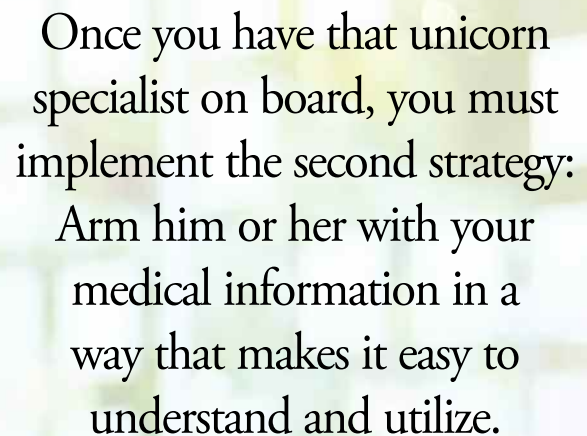
7) *Narrative*: This is the historical section that will be developed over time. I started my children's documents when they were born since they were both sick from birth and their narratives are quite complete. For myself, I began getting very ill in 2011, and was diagnosed with common variable immunodeficiency in 2013. To compile my narrative when I started it in 2012, I had to do some digging. I pulled out my insurance explanation of benefits statements and used them to reconstruct the journey over the previous couple of years. Thankfully, today that information is more easily accessible via the insurance company's website.

This narrative should be a short summary of any major event that occurs, including doctor visits, labs, imaging and other tests, and episodes of illness or flares. Include the date and then a relevant and concise summary. Keep it

short and with the most recent events at the top (reverse chronological order).

This document will become your "go-to" for every doctor visit and can guide the appointment, thus saving time and confusion on the part of the provider. Our specialists literally sit knee to knee with us and go through the narrative items that have transpired since the last visit. And, the medical assistants use the medication section to update that information in their systems. It's a simple way to make sure everyone is on the same page.

To ensure I have these documents with me at all times, I save the latest version as a PDF file and keep it in the free online document sharing tool "Drop Box," which I can access from my computer, phone or iPad. That way, if I'm away from home and need to have a conversation with a provider, I have my cheat sheet. I can quickly email the PDF document to a specialist upon request.



Once you have that unicorn specialist on board, you must implement the second strategy: Arm him or her with your medical information in a way that makes it easy to understand and utilize.

This chronic life is not easy, but developing a system to wrangle the chaos can take away some of the stress. As caregivers and patients, the more we put on paper, the less we try to keep in our heads, and that equals reduced stress. Spend some time on the front end developing your systems, and enjoy the fruits of that labor for years to come. ■

DENNA MCGREW is a rare disease warrior for herself and her two children and often writes while sitting on one of Florida's beautiful Treasure Coast beaches.