DRIPLINE

PARENTERAL NUTRITION DOWN UNDER

Parenteral Nutrition - Down Under™

ISSUE 4
MAY
2013

Welcome to our May issue of Dripline. We have a variety of topics this issue. We begin a series of articles written by Baxter employees, in both New Zealand and Australia, enlightening us about what is involved in their area of work. We preview a leaflet that has been developed by our Management Committee and designed by Carla, to be available to hospitals for new HPN patients. Find out about our Restaurant Card, and about our latest get-together in Sydney. For those with venous access problems, read about AV fistulas – an article by research doctors in Georgia, USA, and another by one of our own members. There is a story written by American parents, who are also doctors, who credit Omegaven® lipid with saving their young daughter. Read about an exciting new drug, Teduglutide, which has been found to lessen the amount of PN needed in some Short Bowel Syndrome cases. We have an explanation of some of the reasons why our PN is light sensitive. And finally, Karen is a winner in the MedicAlert® competition!

Gillian

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A DAY IN OUR SHOES

WORDS BY *ARTHUR AND JIM*

This is the first of a few articles by various employees of Baxter, both in Australia and New Zealand, written in order to give us a better idea of what is involved in delivering our PN to our doorstep. Every week it faithfully arrives, usually delivered by the same familiar face. But what is involved in getting it to us? The first story is by an Auckland, NZ, driver, and the second by a Sydney, Australia, husband and wife driver team.

STORY ONE, BY *ARTHUR*

My day starts at 4am in our Auckland depot. The planning of the route and loading takes around 1hour. Home PN patients are a priority, so when planning my day I make sure that the delivery will be done as soon as possible. Occasionally that means driving an extra 100+kms, which might end up as being an extra hour or two in a working day, but this is a very common thing for us to do.

We cover a large portion of North Island, from nearly the top (Cape Reinga) to the East Coast (Waihau Bay). Some days, by the time I get back to Auckland, I would have covered a distance which easily exceeds 1000kms.
Our patients are very important to us, and we try to provide the best service in the world. We deliver on time, and we deliver to our customers' requested area of the house. It does not matter if it is a room on the second floor of a house, or a basement; sometimes it's a fridge in a shed. We unpack the package and place it on a dedicated shelf as requested by the customers.
This is a very busy and very important job which I truly enjoy, so being friendly comes naturally.

STORY TWO, BY *JIM*

Hi, my name is Jim and my wife's name is Dimi. We have been with Baxter for 9 years.

Some of you might know us as the Baxter drivers who deliver your PN in Sydney, along with David who has been with us for 2 years and Ashleigh almost a year.

We love our job as it gives us the opportunity to meet people.

Our day starts at Baxter at 6am for Jim and 7am for Dimi. [NB Jim and Dimi both drive a van] It consists of getting our run sheet from our Transport co-ordinator Andree, who sets out our daily run.

We do not just deliver PN; we also deliver drugs to hospitals, day surgeries & vet clinics.

We are also available to Baxter for emergency call-outs for deliveries at night or weekends, wherever and whenever they are needed. After so many years, the people we deliver to are not just patients; in our eyes we would like to consider them our friends. We enjoy catching up once a week for a friendly chat, as well as supplying their weekly PN.

Baxter is a great place to work, as there are many people who care about what they do, from our Management to our Team Leaders and Technicians who prepare the products for us to deliver.

In short, this is our daily routine; we look forward to seeing you all. ✳

Dimi & Jim

PLEASE SIR, CAN I HAVE LESS?

WORDS BY *KAREN*

I'm not sure about the rest of you who can eat, but I'm always rather reticent to make a fuss when I'm ordering food at a cafe/ restaurant. I would prefer to order something smaller on the menu or simply ask for a take-away container at the end for the inevitable large amount of left-overs.

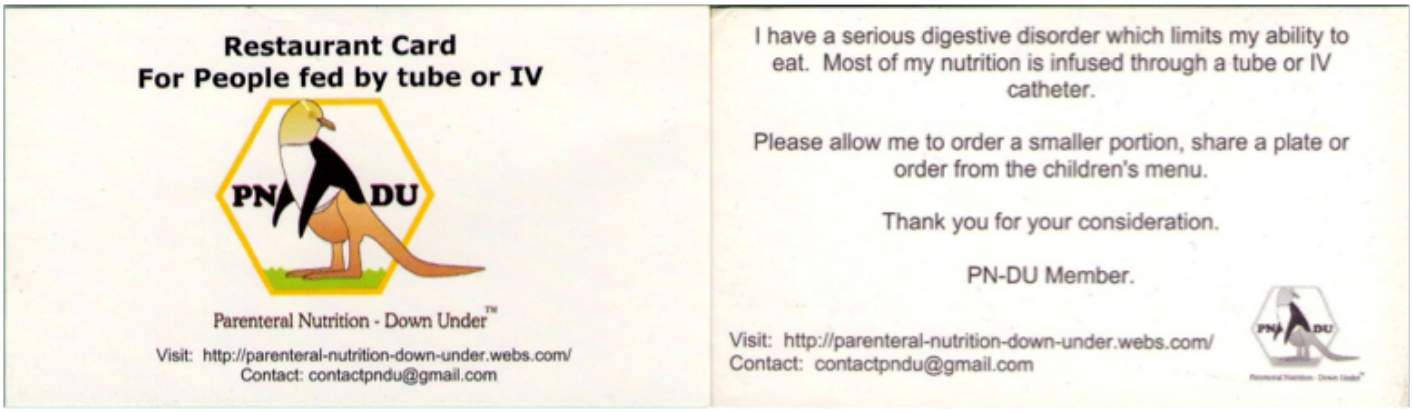
Recently, my sisters, Mum and I went out for brunch for my Mum's Birthday to the Old Bell Tower in Tamworth, NSW, a very nice cafe. I really did feel like the full breakfast but knew it wouldn't be such a great meal for take-away left-overs, so my two sisters ordered, taking with them the PN-DU Restaurant Card to show to the staff.

Well the staff couldn't have been more accommodating! I had a fantastic half-size breakfast with a little bit of everything PLUS it was half the price!

So a big thumbs up to the Old Bell Tower (www.theoldbelltower.net.au) in Tamworth NSW for accommodating an HPNer.

If anyone knows of other cafes/restaurants who are more than happy to accommodate HPNers, please let us know. And if you haven't got one of our PN-DU Restaurant Cards, email me at contactpndu@gmail.com with your postal address and I'll mail one out to you. We're very grateful to Oley in the US for the idea.

Our card reads (on the front):
Restaurant Card
For People fed by tube or IV
Parenteral Nutrition - Down Under (plus contact details and logo)
On the back:
I have a serious digestive disorder which limits my ability to eat. Most of my nutrition is infused through a tube or IV catheter.
Please allow me to order a smaller portion, share a plate or order from the children's menu.
Thank you for your consideration. PN-DU Member ✳



The birthday lunch... Happy birthday, Wendy! Note the half-size 'full' breakfast!

PICNIC AT THE PARK

WORDS BY *GILLIAN*

PN-DU's first Sydney picnic last November was such a success that we wanted to have another one. We decided to use the same venue, Blaxland Riverside Park, Silverwater, as it is fairly central to most areas. We met on Saturday, 9th March, but this time, only five HPNers could meet, as two others had conflicting engagements. Karen's mum, Wendy, was visiting from Tamworth, so it was lovely to meet her, and she felt the same, being able to put faces to names that she has heard or read about. Apart from Karen and her mum, there were Jane; Sal, with husband Matthew and two of her four children – Callum and Bradden; Miranda with Ariel; and Gillian with husband Ray.

We'd like to thank Baxter for putting the flyer, with details about the day, into the PN packs of Sydney HPNers. This gave other HPNers, who might not otherwise know of our group's existence, the chance to come if they wished. Unfortunately, no-one new came along…but maybe next time.

Gil, our treasurer, was planning to fly in from Auckland just for the picnic in order to meet some of us, but sadly, last minute work demands prevented this. Again, maybe next time!

The week leading up to our picnic was quite rainy, but we had a fine, if hot and cloudy day. Karen chose a spot near one of the car parks, under some shady trees, so we were able to be quite comfortable for the few hours we met.

The highlight of the picnic was seeing little Ariel, two years old, now walking, if a bit unsteady at times. As Ariel has 24/7 enteral nutrition, as well as overnight PN, she has had to learn to walk with a backpack weighted with pump and feed, which would change her centre of balance. She was also quite happy and smiled and laughed at us all – although her favourites were Callum and Bradden, who captivated her by pulling silly faces. It was wonderful that Miranda and Ariel were able to attend, as Ariel had been in hospital for 4 days, and had just returned home the day before the picnic! She had been vomiting continuously, and had a temperature, but as the doctors couldn't find any infection in her cultures, it was deemed to be a nasty virus, probably picked up in the playground from other littlies, but hitting her harder than other children.

One of the great benefits of meeting in person is being able to easily talk, rather than type, or have sometimes broken conversations over Skype. Being able to share our good times, and our problems, with people who understand how difficult, or simply frustrating, life can be living with a chronic condition, seems to hearten us all and bring us closer together.

We all look forward to the next picnic! ✳

The 5 HPNers – Sal, Gillian, Jane, Karen, Ariel (front)

Left: Ariel, and her mum, Miranda
Above: Wendy, Jane, Sal and Matt, Karen, Miranda and Ariel, Ray

HPN AND ARTERIOVENOUS FISTULA (AVF)

WORDS BY *A PN-DU MEMBER*

Caveat

I am writing this from my experience, that of being both someone who has assisted in inserting/forming and repairing an AV, and as a patient who has 'worn' and used one for 12 yrs. So what I write is from my personal aspect and can't be considered wrong or right.

Just what does it have to do for someone on long term PN, or those needing a central line access? It's another access route to be considered!

There is plenty of information out there, but:-

Most data and research come via the dialysis side of things which is where AVF are most commonly used. The other specialities have been very slow in utilising them, but this is now slowly changing and little satellites of usage are emerging around the world. Cystic Fibrosis disease is one that is starting to use AVF as an ongoing vascular access site. The Netherlands has the biggest centre successfully employing them for HPN.

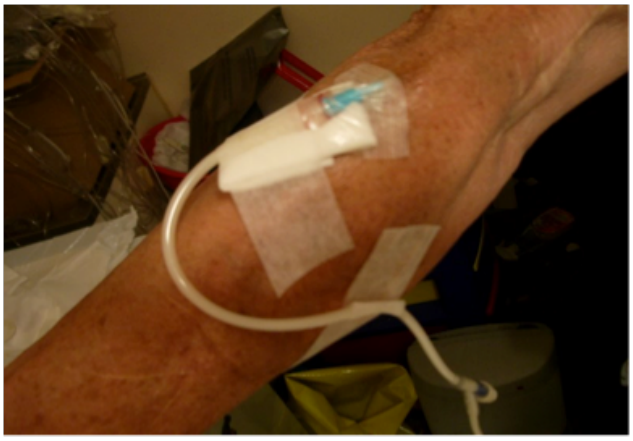
I feel that the hesitation by other groups to advocate AVF has come from the often dramatic and sensational way that the dialysis teams care and look after them. Which of course has meant that the stricter the policy the better for the patient and longer life of maintaining them. But, to the outsider it's all a bit too much, Over the Top, so they are dissuaded.

In the 1980's several HPNers did use AVF-G (grafts) with varying results. This set the idea in motion as to their usefulness, and some papers written at that time concluded that they were not ideal. In response to those –yes, they are high maintenance, and yes, a team effort is required, but they provide a good access, and a good Quality of Life for the patient. Surgical techniques, interventional radiology (IR), diagnostic support, and bio-engineering principles all have greatly improved since then.

I have gathered a series of links at the end of this article for those interested in further information.

What is an A-V

This is jargon for arterio-venous meaning the joining of an artery to a vein, and can occur from trauma, be congenital or surgically performed. It is done to bring the larger flow blood vessel to the surface so that it can be accessed easily.



A-V
- - - -

What is the 'F'?

This is the direct connection, either an end to end, side to end or side to side join so forming a **fistula.**

This is the best situation as there is no dead flow area, everything is normal tissue, there is no foreign material, but it can take up to 2 yrs to mature, although mostly about 6 months, but sometimes it can be used after 3 months. That is, the join from the artery settles to be of suitable diameter that the vein tolerates the higher flow rate, and toughens up for frequent needle insertions! So it should be humming nicely along. Known as "a thrill to feel and a bruit to hear"!

What is the 'G'?

This is the **graft** which is a tubular pipe connecting the artery to the vein.

This is an another option to the fistula if, as often is the case, the patient's arm or thigh is not ideal for a direct connection, due to smaller blood vessels or a mismatch in position. A deeper vein may be needed, to attach the artery to for the best flow rates. A plastic tube, usually of PTFE, approximately 6-8mm in diameter, and looking like thick surgical stockings, is used. This has properties that allow it to be stabbed by a needle, but will close over afterwards and heal, as the body grows tissue around and within. Some grafts may be made by a vein stripped from another area, such as the leg, although this is not commonly done.

How is it decided if it's a suitable access

An A-V needs to be formed from 'good' vasculature, so those veins/arteries ruined by peripherally inserted central catheters (PICC), or peripheral intravenous lines (PIVs), poor health status of the patient, both physically and environmentally, are all taken into consideration. The hospital needs to have a vascular department, and a team able to follow and treat complications of the AV, including Surgical, I.R and Sonography. The patient needs to be able to self access, and understand the care needed.

Why don't many HPNers have an AV offered as an access choice?

Hospitals often work under the "silo system", that is each speciality keeps to itself, so crossing boundaries for care can be difficult. Many PN-IV specialists are still quoting research from the mid 1980's when discussing central vascular access, and this has been an obstacle.

In the young child, the team are so busy caring for the very sick child and often not expecting a full life span, that looking down the line in 10-20yrs' time is not thought of. Therefore the good easy veins are used for central venous access devices (CVADs) e.g. PICCs, and damage is then possibly done to these important upper limb veins with stenosis possibly occurring and possible scarring of the blood vessel.

In the adult, the damage might have been done prior to the permanent or long term need for HPN. Sometimes the hospital is not prepared to set up a team to care for it.

Explain the 'flow ability'

Think of an upgrade of a road system by your local council. It is no use having a big sport stadium that can take 50,000 people if the only way to get there is via a narrow winding road. PN needs a good blood flow rate of at least 400 ml/min to dilute the concentrate or hypertonic solution.

So can a 2 lane be turned into a 4 lane highway? Is the land structure and contour able to have a freeway that is suitable for fast transportation, with no hairpin bends, which cause cars to flick up stones as they go round. In the same way, blood gets bruised and flicks onto the side walls if the graft/fistula has too sharp a curve graduation, or if narrowing occurs. Just as leaving the sport complex with only a few access gates open makes for bad tempered drivers, so blood cells get grumpy, leading to thrombus formation.

Also the vein, having a thinner wall than the artery, needs to be able to adjust to the extra traffic flow!

What's the difference between having a CVAD and an AVF

The AVF creates no foreign body reaction, reducing chances of infection; and has no outside contact except when being accessed, thus giving freedom during the day, for those who are on night feeds. The alternative A-V graft, if technically placed properly, is also superior to the central catheter. It can be dilated and 'fixed up', surgically or with IR, several times before needing replacement or a new site. Also this does not interfere with the large central veins where the catheter can cause mechanical phlebitis from rubbing on the inner wall, or cause obstruction if the diameter of catheter is greater than 50% of the vessel it is in, impeding flow of the vein. SVC (superior vena cava) syndrome is an example of this.

How is the AVF put in?

It can be done as a day-stay procedure or in an overnight observation ward. For the fistula, a small incision done under local anaesthetic is usually sufficient, or maybe an arm block. For the graft, there are three or four incisions. One at the artery site, one at the venous connection, and two at the bend of the graft being put in. This can be done under local or arm block or light anaesthetic, depending on the surgeon and the patient's wishes.

The two end sites are identified and exposed (artery and vein) then the graft tube is tunnelled under the skin and subdermal layer, pulled through and down and then back around, flushed and joined up.

I get told that PN has to be infused into the heart

No, the heart does not need to be the first organ to 'see' PN solution; it needs a high flow vessel for diluting the concentration so that chemical phlebitis does not occur. For some who are on a fluid restriction regimen and on high strength glucose and/or other added medications, then a blood flow rate which the heart produces of >600 ml/min might be needed, but most on HPN would cope with the >400 ml/min blood flow rate of the AVF.

What are the main complications?

Common complications are bleeding and infection. It is also possible that it might not mature, that is, the vein might not adjust to the increased pressure.

Because the lymph system sits beside the blood vessels, often lymphoedema of the arm can occur, which is usually controlled with elevation and massage. Nerves also run alongside and nerve damage is possible. Steal syndrome is where the hand can go cold as blood is 'stolen' for going to the V of the AV and insufficient is going down to the hand! Reducing the opening at the A end and controlling the flow to the V, can often fix it, but in some cases the AV will have to be closed off. Tests to check for the flow of the other artery to the hand should be done before surgery! For mild cases, often wearing a fingerless mitten or woollen tubular sleeve to keep the limb warm helps.

For the ongoing problems of thrombosis and stenosis see the nih.gov link below..

Can the AV graft be removed?

No; if it stops working, it is just left there. The only time it is removed is in cases of infection and then that means a large incision and careful removal of the infected area or whole graft.

How can I find out if I am eligible for an AVF?

Firstly, the hospital has to be willing. They then need to look at the patient's history of vascular assault, (how many IVs and central lines the patient has had) then have a duplex scan done to see what vessels are available, and their blood flow ability.
What if I want to ask more?

Either ask your team to put you in contact with the dialysis team in your area, or send questions to the editor.

Sites to visit

http://kidney.niddk.nih.gov/kudiseases/pubs/vascularaccess/vascularaccess_508.pdf
Explains the needs, types, formation and maintenance of an AVF for dialysis patients

http://www.surgeryencyclopedia.com/A-Ce/Arteriovenous-Fistula.html
Explains management for both medical team and the patient.

http://www.network13.org/FRM/Section_05/FF_Tools/C03-Early_Referral_to_Surgeon/08-Surg_Anatomy_Upper_Arm.pdf
How the surgeon will decide on the best AVF type and position

http://samples.jbpub.com/9781449652609/99069_ch05_6101.pdf
Good anatomy and explanation of physiology of arteries and veins.

http://www.kidney.org/professionals/kdoqi/guideline_uphd_pd_va/va_guide4.htm
Discusses all the nitty gritty pressures and problems relating to dialysis.

Another article about AV Fistulas, citing a research project.

ARTERIOVENOUS FISTULAS FOUND SAFE FOR TPN IN SMALL STUDY

19-Apr-12 Elsevier Global Medical News BY M. ALEXANDER OTTO

LAS VEGAS (EGMN)–Long-term total parenteral nutrition doesn't necessarily require a long-term, infection-prone central venous catheter through which to deliver it.

Georgia researchers have found that arteriovenous fistulas (AVFs) work well and are safe for TPN (total parenteral nutrition) administration; patients can even learn to deliver TPN through the fistula themselves at home.

For the handful of patients in the pilot study, the switch to AVFs improved their quality of life dramatically, said lead investigator Dr. Jonathan Woody, because they no longer had to manage the complications of catheter-based access, which include infection, thrombosis, catheter fracture, and other problems.

"AVF for TPN is a life changer" that has the potential to become the "new gold standard" for long-term TPN access, he said at the annual meeting of the Society for Clinical Vascular Surgery.

Four women and one man (mean age, 56 years) were chosen for the project because of recurring catheter infections. The patients had been on TPN for months or years, four of them because of short gut syndrome following bowel resection, and one because of severe diabetic gastroparesis.

They were given standard, dialysis-type AVFs, but the segments used were generally shorter because TPN requires one needle puncture, not two, said Dr. Woody, a vascular surgeon with Athens (Georgia) Vascular Specialists.

Three patients received brachiocephalic fistulas, two of which were used for TPN. The third, despite initial plans, ultimately was used only for dialysis.

Another patient received a basilic vein transposition, which was used for TPN. The cephalic vein transposition in the fifth patient failed, probably because of the necessary use of a vein smaller than 3 mm in diameter.

The three patients who used their AVFs for TPN were first taught how to care for their fistulas, to access them nightly through 16- or 18-gauge IV cannulas, and to cycle TPN overnight with a standard IV pump. Patients and family members were comfortable doing it themselves once they were trained, Dr. Woody said.

One of the three TPN patients died because of underlying illness, but one has been using the fistula for more than a year, and one for more than 5 years, both with standard TPN concentrations. There have been no cases of phlebitis.

In short, AVFs are safe and effective for TPN, and they eliminate recurrent catheter infections and associated medical costs. "Patients requiring long-term TPN should be referred for AVF," Dr. Woody said.

After hearing his presentation, several vascular surgeons in the audience said the approach seemed like a good idea and that they planned to try it.

But while creating the fistulas is something "we all know how to do," Dr. Woody said, "the more challenging and more important aspect of this endeavour is engaging others."

Gastroenterologists, general surgeons, patients, payers, and medical organizations have little awareness that AVFs are an option for long-term TPN, and that patients need to be referred early while they still have suitable veins. "We must educate [them] about the benefits," Dr. Woody said.

Until that happens, pharmacists may be reluctant to release a TPN bag for fistula administration because they've never heard of it before, and without endorsement from medical organizations, insurance companies may refuse to cover it.

Dr. Woody said he has no relevant financial disclosures. ✳

OUR NEW HOSPITAL LEAFLET

The PN-DU Management Committee has produced a flyer, to be available in hospitals, to promote awareness of PN-DU and our role as a support group for people with Intestinal Failure who need HPN. We'd like to thank Carla for the bright, eye-catching design. The Management Committee is now considering how best to introduce it to hospitals, so that it is used and doesn't languish, forgotten, on shelves. ✳

PARENTERAL NUTRITION DOWN UNDER: PN-DU

PN-DU is a support group that has been set up specifically by and for Australian and New Zealand Home Parenteral Nutrition (HPN) consumers and their families/carers

"Our aim is to Support, Research and Inform Consumers, Carers, and Providers of Parenteral Nutrition for Intestinal Failure"

Together we can provide comfort, strength and reassurance to each other and together we can have a louder and more influential voice on the future development of HPN products, HPN therapy and Home Care Services.

If you would like to find out more about PN-DU and receive a new member pack (including information, newsletter, invitation to receive a cuddly Pendoo for kids or mug/pen for adults), please contact us at contactpndu@gmail.com

Parenteral Nutrition - Down Under™

WHAT IS PARENTERAL NUTRITION OR PN?

PN means being fed intravenously, when it is not possible to eat normal food because of intestinal failure.

WHAT IS HOME PARENTERAL NUTRITION OR HPN?

HPN is receiving your PN in your own home, usually after you have been trained by your hospital team.

WHAT IS INTESTINAL FAILURE OR IF?

IF is reduction of gut function to below the minimum necessary for absorption of nutrients, water, vitamins and electrolytes. This means it becomes impossible to eat or absorb goodness from food of any kind – whether it is normal food or special formulas, modified diets or tube feeds – so you need PN to live.

WHERE CAN I GET MORE INFORMATION ON HPN?

You should refer all medical questions to your hospital team but if you want to discuss the experiences of others like you who are on HPN you can join PN-DU.

We want to encourage all HPN consumers in Australia and New Zealand to join us and exchange your experiences, challenges and achievements.

Check out our website to join us and post questions or comments:
www.parenteralnutritiondownunder.com

or you can email us at:
contactpndu@gmail.com

Parenteral Nutrition - Down Under™

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HI! I'M DOWN-UNDER-DAN AND I'M ON PARENTERAL NUTRITION

Useful Information for Adults and Carers of Kids with Intestinal Failure going Home on Parenteral Nutrition

OUR JOURNEY AS PARENTS

WORDS BY *SHIRLEY HUANG, MD, AND JAMES YOO, MD*

L: Janie on her first birthday
R: Janie with her big brtoher, Ryan

I am a pediatrician working in an academic children's hospital where I see predominantly children with special health care needs. My husband, Jim, is a colorectal surgeon. Together, we have a daughter, Janie, with short bowel syndrome who is dependent on home parenteral nutrition (HPN). We would like to share our journey as physician parents with you.

Imaginings

Life changed dramatically for us when we became parents. Two years after our son, Ryan, was born, we had a second child, our daughter, Janie. During both pregnancies, we enjoyed imagining what our child would be like. All parents have hopes and dreams for their children.

When you have a child with special needs, those hopes and dreams change. We used to wonder: Would Janie be smart or funny? Would she be good at sports? Will she get along with her big brother? As she got sicker and sicker, the questions we asked about Janie changed. We wondered: How big will her scar be? Does she really need a stoma? Will she ever be able to eat? Will she live? Does she know how much we love her?

Realities

Janie was born premature at thirty-four weeks gestation, shortly after being diagnosed as small with no amniotic fluid. She weighed just three pounds at birth. Initially, besides being small, she appeared normal. Within a day, however, she was diagnosed with a distal ileal atresia.

Janie underwent her first of six surgeries at four days of life. We were relieved that they were able to resect the area and reconnect the bowel, "solving" her problem with the initial surgery. She remained in the neonatal intensive care unit (NICU) for two months as a "feeder and grower," off and on PN.

When we were discharged, she was quite jaundiced and we were scheduled to come back daily for studies and weight checks. Despite this, we were thrilled to have her home with us, and to finally have her big brother meet her. Ryan was less than two years old at the time and had not been allowed in the NICU to visit her.

Complications

Over the next two months, we were in and out of the hospital for failure to thrive and malabsorption. Sometimes, we would be home for less than a day, sometimes we would make it for a week. Finally, when Janie was almost four months old and still weighing about six pounds, the decision was made to take her back to the operating room (OR) for a resection of her initial anastomotic site, which was thought to be the cause of her difficulties.

We hoped this would solve her problems and would allow us to go back to a normal life. We anticipated a simple resection and re-anastomosis like her original surgery, but instead she came back from this surgery with a central venous catheter, a G-tube, and an ileostomy.

Unfortunately, things became more complicated when, within a day, she was taken back to the OR. They found the majority of her bowel was necrotic, requiring a massive resection and jejunostomy. We thought Janie would die that night. She was made DNR ("do not resuscitate") and after her return to the NICU, we were left to spend time with her behind drawn curtains.

Janie managed to pull through, but in addition to severe liver disease, jaundice, and failure to thrive, she now had short bowel syndrome and would be PN-dependent for the remainder of her predicted short life. Our medical team gave us two options: to consider a multivisceral transplant or take her home knowing she would probably not make it to her first birthday. For us as parents, we had decided transplant was not the route we wanted to take.

Alternatives

This is when we began looking into Omegaven®. We had heard about it in passing from one of the neonatologists, but it was not available in our city at the time. When we realized it had originally been used at Boston Children's Hospital, I thought perhaps I would know the investigators as I had completed my pediatric residency there. As luck would have it, the primary investigator was a pediatric surgeon, Dr. Mark Puder, who had been Jim's senior resident when Jim was an intern.

We contacted Dr. Puder ourselves, looked into the published research, and spoke with medical colleagues at other institutions with clinical experience in Omegaven. Some of our own doctors discouraged us and told us we were searching for miracles. We were. We took a chance and had Janie transferred to Boston Children's Hospital, a difficult decision that involved moving our family across the country.
We spent the next six months in Boston, where Janie was treated with Omegaven. Slowly, her liver failure improved and Janie got better. We stayed there until she was well enough to come back home with us. As parents and as physicians, we have no doubt that Omegaven saved Janie's life, and we look forward to the day when it will be available, anywhere, for any child who needs it. [See editor's note below]

Toddlerhood

At fifteen months of age, we returned to Boston to have Janie's intestines reconnected. When we returned home, we were finally able to cycle her HPN to run over twelve hours instead of twenty-four. This came just in time, as chasing after a newly walking toddler connected to a backpack on your back all day has its challenges! Luckily, once Janie got going, she didn't mind balancing and wearing her own backpack. With this time came a lot of hope for us that things were on the upswing, that we had gone through the worst of it, and that she would continue to get better.

Over the next year, there were a lot of bumps. She still wasn't able to tolerate enteral feeds. We thought she would need another surgery to "fix" a persistently dilated loop of bowel that was believed to be the problem, but ultimately the surgery was canceled. She was changed over to a GJ-tube and tried on another prokinetic medication. We made little progress in advancing her enteral feeds and she was still on full HPN.

Life Today

Though it was a difficult year, we also saw Janie grow and develop. While we continue to hope that someday she may come off HPN, we don't wait for this day. We look at the big picture and marvel at all the wonderful things that are happening in the midst of our struggles.

When Janie was two and a half years old and our son had his fourth birthday, his birthday wish was for Janie to start talking so they could talk together. Over the past year and a half, she not only began talking, but now talks nonstop with never-ending energy! Ryan may at times regret that birthday wish, but he will never tire of having a sister in his life.

At the end of the day, we have a happy four-year-old daughter. And we have been able to answer many of the questions we used to have about Janie before she was even born. Would Janie be smart or funny? Well, Janie is smart (gifted, actually!), and she is funny. She makes us laugh every day. Will she be good at sports? Not really. Will she get along with her big brother? Usually, and when they do it is a beautiful thing. How big will her scar be? Her scars are big, but that just means she is growing. Will she ever be able to eat? She is eating better and is growing and thriving. Will she live? Life is precious; we take every day as it comes. Does she know how much we love her? Of course she does, because we tell her every single day.

LifelineLetter Editor's Note: Omegaven is an omega-3 based lipid solution made from fish oil. Progress is being made in identifying dose requirements, primarily for babies. It must be emphasized that use of this lipid in older children and adults with PN-related liver disease has not been fully explored. The mechanism for development of PN-related liver disease is uncertain; it appears that the type of lipid used, the amount of lipid used, or the frequency of lipid infusion may be related to the pathophysiology of liver disease. Whether the improvement that has been seen in babies and very young children on Omegaven is related to the ability to decrease the Intralipid infusion or to a specific characteristic of the fish oil, Omegaven does seem to play a role in some specific consumers. HPN consumers need to be following the lipid story as it develops over time.

We at Oley are very pleased that this beautiful family (and especially Janie) is doing so well after a difficult course. We were privileged to meet them at the Oley meeting in California this summer, and are happy to say Dr. Huang and Dr. Yoo's presentation is available on the conference DVD (http://www.oley.org/video_dvd.html).

LifelineLetter, September/October 2012

PN-DU Note: Omegaven® is registered in New Zealand (Reg. No: TT50-6977) and is marketed by Fresenius Kabi Ltd, but is currently not yet registered in Australia.

Smoflipid®, another lipid emulsion containing fish oil, is registered in Australia and New Zealand and is also marketed by Fresenius Kabi Ltd (more information on Smoflipid® in the next issue).

PN-DU would like to thank Oley for permission to use this article, published in their journal, LifelineLetter. ✳

THE ROLE OF TEDUGLUTIDE IN THE TREATMENT OF SBS

WORDS BY *PALLE BEKKER JEPPESEN, MD, PHD*

[Adapted with permission from the author and Oley by Gil Hardy PhD from original article in Oley's Lifeline Letter, Jan/Feb, 2013]

Recently, the U.S. Food and Drug Administration (FDA) and the European Medicines Agency (EMA) approved the glucagon-like peptide 2 (GLP 2) **analogue** teduglutide for the treatment of adults with short bowel syndrome (SBS). SBS results from surgical resection, congenital defects, or disease-associated loss of intestinal absorption.

The symptoms of SBS vary based on the amount and the health of the remaining bowel, and the specific part of the small bowel that has been removed. Many SBS patients with intestinal insufficiency are able to compensate for their malabsorption by changing their diet and increasing oral intake. However, SBS patients with intestinal failure (SBS-IF), need parenteral nutrition (PN) and/or IV fluids (IV) to maintain nutrient, fluid, electrolyte, trace element and/or vitamin balances.

Although typically life-saving in SBS-IF patients, PN has been associated with catheter-related bloodstream infection, central venous thrombosis, and embolism. In addition, PN and chronic dehydration may contribute to progressive IF-associated liver and renal disease. These burdens, combined with the symptoms of malabsorption (e.g., diarrhoea, large stomal output, stomal problems, fear of faecal incontinence, flatulence, and abdominal pain), may cause restrictions in the lifestyle of SBS-IF patients and may lead to significant impairment of their quality of life. Therefore, treatments of SBS-IF aim to maximize the absorptive capacity of the remaining intestine; minimize symptoms of malabsorption; and avoid, eliminate, or minimize the need for PN.

Current Approaches to Managing SBS

The fundamental principle of SBS management is to decrease fluid secretion in the upper bowel and to maximize the contact time between the digested nutrients and the intestinal mucosa, thereby increasing absorption of the nutrients. In general, SBS patients are encouraged to compensate for malabsorption by adjusting their diet and by increasing oral intake. Frequent meals and snacks are encouraged. Historically, treatment strategies have included changes in dietary composition, for example high-carbohydrate, low-fat diets; the use of ***preferred luminal substrates*** (e.g., glutamine or medium-chain triglycerides). However, the effects of these interventions have not been evaluated in long-term, ***placebo-controlled*** studies. Continuous tube feeding has been suggested to improve the absorption of macronutrients, but it may be accompanied by increased faecal fluid and electrolyte losses, which can aggravate abdominal discomfort and diarrhoea, and further increase the need for IV fluids and electrolytes.

The extremely salty taste of oral rehydration solutions with a high sodium concentration may keep people from using them over the long term. Agents such as codeine, loperamide, and tincture of opium slow intestinal motility and antisecretory drugs such as H2-receptor antagonists, proton pump inhibitors, or somatostatin analogues have been shown to reduce gastric acid secretions, jejunostomy fluid output, and diarrhoea, but no effect on absorption has been established. Therefore, there is a high unmet need for medical treatments in SBS-IF patients.

Studies

A ***double-blind placebo-controlled study*** was recently performed by Jeppesen PB et al, (Gastroenterology 2012;143:1473–81). Forty three SBS patients were ***randomized*** to a 0.05 mg/kg/day dose of teduglutide and forty-three patients received placebo for up to 24 weeks. Findings were significant. Twenty-seven of the patients who received teduglutide achieved a 20 percent to 100 percent reduction of PN/IV at weeks 20 and 24, compared to thirteen of the patients who received a placebo (62.8% versus 30.2%). In patients completing the study, twenty-one patients treated with teduglutide (54%) were able to reduce their PN/IV by at least one day, compared to nine of those given a placebo (23%).

Risks

The studies have revealed that teduglutide treatment may be associated with adverse events, mainly of gastrointestinal origin (abdominal distension, abdominal pain, nausea, and stoma enlargement). In general, teduglutide should not be prescribed to patients who have had malignancies within the last five years. Cases of cholecystitis, cholangitis, cholelithiasis, and pancreatitis have been reported in the clinical studies; in such cases continued teduglutide treatment should be reassessed.

Which SBS patients will benefit from teduglutide and how?

The only SBS patients who will be able to discontinue PN completely are probably those who are on the borderline between intestinal insufficiency and intestinal failure. It is estimated that these patients may account for 10 to 15 percent of the SBS-IF patients. In patients with intermediary degrees of IF, the consequences of improved intestinal absorption resulting from teduglutide treatment may vary according to the individual patient. Some may prefer days off PN, even at the risk of becoming slightly dehydrated, whereas others will prefer a larger day-to-day stability, requiring smaller PN volumes and possibly shorter daily infusion times.

Conclusion

Teduglutide treatment has the potential to optimize intestinal absorption, decrease malabsorption and accompanying symptoms, reduce the need, burdens and complications related to PN and IV fluids, and ultimately improve the health-related quality of life in SBS-IF patients.

Glossary
Words that are italicized in the article are defined below, as they are used in the context of this article.

analogue—a manmade compound that is chemically related to the normally synthesized product

double-blind study—neither patient nor researcher know whether the drug or a placebo is being given

intestinal barrier function—the ability of the intestine to control absorption of certain nutrients

intestinal mucosa— lining of the intestine that contains, among others, specialized cells for absorption also known as enterocytes

placebo—an inactive substance that looks similar to another active substance

placebo-controlled—results of subjects receiving the drug are compared to the results of subjects who receive a placebo instead

preferred luminal substrates—nutrients that are preferred for fuel (energy) by the intestinal cells

randomized—chosen at random, usually with the researcher being unaware of which subjects are allocated to specific groups

Editor's notes:
* The full article is also available, with a full reference list, at www.oley.org/lifeline/The_Role_of_Teduglutide_in_the_Treatment_of_SBS.html

* As far as we are aware Teduglutide is not yet licensed in Australia or New Zealand.

PN-DU would like to thank Oley for the use of this article, published in their journal, LifelineLetter. ✳

SPLAT! - A DEHYDRATION WARNING

WORDS BY *GILLIAN*

Since I was the one who put together last issue's article about dehydration, from comments made on our HPNers' forum, and requested the article about Oral Rehydration Solutions by Suzie Daniells, it is ironic that last month I blacked out due to severe dehydration, split my chin to the bone, and chipped the enamel off a tooth!

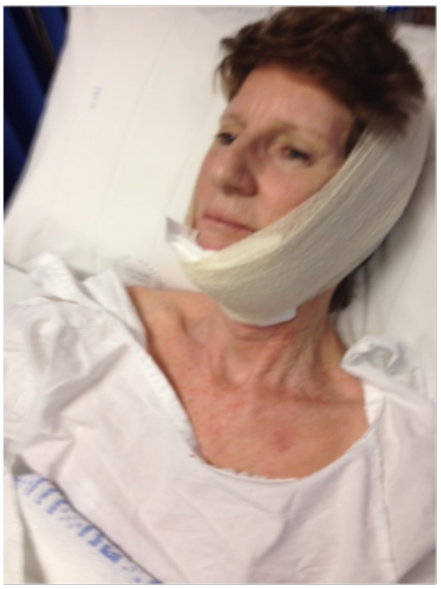
I had been experiencing light-headedness when standing, increasingly often, to the point where it was happening most times I stood. It only lasted for a matter of seconds, then passed, so had no real impact on me, I thought. I realised it was probably due to either dehydration or low blood pressure, and had tried to make an appointment to see my GP as a first port of call, but she was on holidays, and I wasn't due to see my hospital team for a while, so I let it go. Big mistake!

I didn't realise how serious it was, because I was still getting my usual PN every night, plus drinking throughout the day – you rarely see me without a water bottle clutched in my hand – and I didn't actually feel thirsty. One night, I stood up without experiencing dizziness, walked through the house, talking to my daughter, then began to feel quite dizzy, grabbed hold of furniture to steady myself, and woke on the floor half a minute later with blood everywhere!

My daughter said I'd begun to speak gibberish, and she rounded the corner of the room in time to see me fall. My chin was split for a few centimetres, to the bone, but luckily some first aid with a bandaid held it together and the bleeding stopped while we drove to the hospital. To my amazement, I was called through immediately after registering and given a blood test, which showed severe dehydration. As it turned out, the split chin was the least of their concerns, eventually being given five stitches at midnight, five hours after being admitted. Their more urgent concerns involved treating me with extra saline, as well as doing heart and neurological tests, which proved to be clear. My husband, Ray, brought my PN into the emergency department for me to set myself up – it must have looked strange to other patients, seeing me doing my own scrub and dressing pack etc routine!

After being kept overnight, I saw my PN team, who decided to up the volume of PN for a month, and gave me a few extra saline packs to have at home. A couple of weeks later, I saw my dentist to have my tooth fixed, which luckily was a simple, relatively cheap, bonding job.

I've learnt not to ignore warning signs that something is not as it should be. How much more simple life would have been if I'd acted a few weeks earlier, when the light-headedness first began to happen, rather than ignore it and let it escalate. ✳



WHY WE SHOULD PROTECT OUR PN FROM LIGHT

WORDS BY *GIL HARDY PHD*

Intravenous lipid emulsions (IVLE) are an essential component of parenteral nutrition (PN) mixtures, but several new IVLE are now marketed in Australasia. Our recent paper (1) outlines the rationale for the different IVLE formulations available for use in PN admixtures, and reviews some of the factors influencing stability and efficacy of lipid-based PN regimens. We will summarise these data in a future issue of DripLine. In this article we evaluate some of the technologies for minimizing peroxidation of IVLE and for maximizing stability of PN admixtures. And finally, an explanation of some of the reasons our PN is light sensitive.

Ideally all lipid-PN should be assessed regularly for stability, but this is not practicable. Research has therefore centred on detecting or measuring stability "markers" such as precipitates, vitamin oxidation, changes in amino acid content and emulsion particle size distribution. The chemical constituents of IVLE and their relative stability during or after compounding, storage and administration have not been studied in any depth, but oxidised lipids have been identified in a variety of foods, especially those stir fried with sunflower oil(2) and the oxidised products have been implicated in the aetiology of coronary heart disease.

Polyunsaturated fatty acids (PUFA) form up to 50% of the total lipids in some IVLE and can peroxidise to harmful hydroperoxides under the right conditions. A PUFA can be attacked by free radicals, to have a destabilising effect on the emulsion, as well as being potentially toxic in their own right. A major neutraliser of free radicals is vitamin E or d-α-tocopherol. Tocopherols inhibit lipid peroxidation by scavenging lipid peroxyl radicals much faster than the radicals can react with PUFA, thus breaking the chemical chain reaction. During this process α-tocopherol is itself converted into a free radical which in turn is recycled by reaction with vitamin C (ascorbic acid). In theory then, if there is sufficient α-tocopherol and ascorbic acid in PN mixtures then lipid peroxidation can be kept to a minimum. However, at the moment we do not know enough about these complex systems to establish the optimum concentrations for these antioxidants. But by studying different IVLE under a variety of storage conditions we can better understand the dramatic influence that light, oxygen and temperature can have on peroxide formation. Nevertheless, light protection and oxygen exclusion by using multi-layer containers and cover bags can decrease peroxide formation.

The dangers of not protecting PN products from light during storage, and especially during administration, are now well demonstrated. While the opacity of the IVLE in a PN mixture may protect the light-sensitive vitamins in a bag from degradation, under the influence of light, lipid peroxides can still form and further interact with the same vitamins to eventually destabilise the admixture. In theory, as more peroxides are formed the pH of admixtures will drop, and we have previously predicted the adverse effect of low pH on mixture stability(4). In practice, in such complex nutrient mixtures, interactions between the free radicals, α-tocopherol, ascorbic acid, other vitamins and amino acids should limit the extent of any chain reactions and hence the level of peroxides formed. Moreover, the newer IVLE, based on olive or coconut oil are lower in PUFA and consequently less susceptible to oxidative degradation. It will be interesting however, to investigate the interactive effect(s) of these alternative lipids when PN mixtures are compounded, stored and administered under optimum conditions. Until we understand more about these systems it is advisable to light protect PN mixtures at all times.

The pharmaceutical industry, universities and hospital pharmacists have a mutual interest and responsibility for the quality of PN admixtures and should be encouraged to develop new analytical techniques for assessing, and in time predicting, the long term stability of HPN products.

1. Hardy G and Puzovic M. *Nutr Clin Pract.* 2009;24:616-625
2. Wilson R, Payne J A, Smith R, Shephard M J and Riemersma.. *Anal. Biochem.* 1996 .

WE HAVE A WINNER!

WORDS BY *KAREN*

We all love winning something and I was pretty excited to find out I was a winner in the MedicAlert® Foundation's recent "Free to be me" photo competition. I can't remember winning anything since I entered a bunch of my Mum's spinach (I think I planted it, but she watered it!!) in the Tamworth Show and won grand champion in the vegie section! Well that was several decades ago now, so I was thrilled to receive the phone call from MedicAlert® just the other day!!

Entrants were required to send in a photo of themselves wearing their MedicAlert® bracelet or pendant and write in 25 words or less how it enabled them to be 'free to be me'. There were 3 winners, each receiving a $100 gift voucher. You can check out the winning entries here - https://www.facebook.com/AustraliaMedicAlertFoundation?fref=ts (just scroll down till you find them - you can still see it without a Facebook account). It looks like from the other winning entries that there was some very cute competition. Obviously I didn't take the photo of me – Mum did. Thanks Mum… again!!

You'll remember there was an article in PN-DU's last newsletter about MedicAlert® and the service they provide for those of us with medical conditions, treatments, etc, needing to be flagged in the event of a medical emergency. Thanks MedicAlert® for the great service and for the great prize!

One Happy Winner – Karen ✳

Karen's winning photo

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If you feel able to contribute to our support group, you may wish to make a donation. Donations are currently only tax deductible in New Zealand. We are grateful to our sister charity IPANEMA (Charities Commission Registration CC21178) which receives donations on our behalf.

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