



Hirschsprung Disease Patient on Home Parenteral Nutrition during COVID-19 Pandemic

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Abstract

Hirschsprung Disease (HD) is a rare neonatal congenital disorder with an absence of parasympathetic ganglion cells in the distal colon. Adequate nutrition support is important in HD management. Parenteral Nutrition (PN) and Home Parenteral Nutrition (HPN) is an alternative nutrition support for the HD patient to grow in psycho-familial environment. A female patient weight 3 kg was born with no bowel opening, abdominal distension and bilious aspirate in 2014. Biopsies and laparoscopic biopsies had confirmed the presence of ganglion cells up to 20 cm distal to the stoma. She was diagnosed with Hirschsprung Disease-Total Colonic Aganglionosis. She underwent laparotomy, adhesiolysis and serial transverse enteroplasty STEP procedures for bowel lengthening initiative. She started with 2-in-1 parenteral nutrition *via* internal jugular vein in 2015 and discharge with home parenteral nutrition in 2017. Prior to discharge, patient mother has demonstrated competence in HPN management. Her family made the necessary adjustment and renovation of their house to provide a dedicated room for PN administration. Discussion and arrangement have been made with related authorities for PN supply, routine home visit, emergency health management, PN transportation and routine medical follow-up. She was 13 kg at the beginning of HPN program, and currently her weight is 15.8 kg, active and no other medical issues. PN solution and lipid solution were packed in separate bags and infused using infusion pump for 12 h each starting at 6.00 pm daily. The HPN treatment continue during COVID-19 pandemic. She had never been infected by COVID-19 and not yet vaccinated. HPN is a strategy to support patient live a normal life and should remain uninterrupted yet flexible, even in the era of pandemic.

Introduction

Hirschsprung Disease (HD) is a rare neonatal congenital disorder. The common manifestation of HD is an absence of parasympathetic ganglion cells in the distal colon. Patients with HD will have bowel obstruction due to lack of peristalsis and loss of relaxation of the anal sphincter. This may lead to severe constipation and life-threatening enterocolitis [1]. Patients with HD are usually diagnosed and undergo surgery during the neonatal period [2]. Surgical intervention with resection of aganglionic bowel is the principal treatment for Hirschsprung disease, and has evolved since 1950 [1]. Transanal Endorectal Pull-Through (TERPT) has become one of the most common surgical procedures for HD in recent decades [2].

Another important aspect in HD management is an adequate nutrition support through oral, enteral or intravenous route [3,4]. Intravenous Nutrition (IVN) or Parenteral Nutrition (PN) is an artificial nutrition alternative, for nutrition support when gastrointestinal fails to functions as it should [5]. It contains amino acids, dextrose, and lipid emulsions as the main nutrients, and electrolytes, vitamins, and trace elements [6]. As HD has been classified as chronic intestinal failure and require long term PN, Home Parenteral Nutrition (HPN) is the best option for the patient to grow in psycho-familial environment [7]. HPN enables the patients to have a high quality of life in their own environment [8]. HPN involves providing IVN through central intravenous route in a homecare setting, is a life-saving therapy for patients with chronic intestinal failure who cannot meet their nutritional requirements *via* an oral or enteral route [9].

Case Presentation

A female patient was born at 36 weeks and 5 days of gestational pregnancy age at Hospital Sultan Haji Ahmad Shah Pahang (HoSHAS) in December 2014. Her body weight was 3.04 kg during the Spontaneous Vaginal Delivery (SVD), had no bowel opening, abdominal distension and

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Table 1: Summary of parenteral nutrition regimen delivered to patient from 2017 until 2022.

PN contents	May 2017	Jun-17	Jun-18	Jun-19	Jun-20	Jun-21	Jan-22
Macronutrient							
Amino acids (g/kg/day)	3	3	3.5	3.5	3.1	3	3
Glucose (g/kg/day)	11.6	12.4	15.1	13.4	11.76	6.6	6.6
Fat (g/kg/day)	2	2	2	2	2.8	2.9	2.9
Electrolytes							
Sodium (mmol/kg/day)	5	5	5	5.1	2.97	3.1	3
Potassium (mmol/kg/day)	5	5	6.5	5.5	2.03	2.1	2
Calcium (mmol/kg/day)	1	1	1	1	1.01	1.1	1
Magnesium (mmol/kg/day)	0.5	0.5	0.5	0.51	1	1.03	1.01
Phosphate (mmol/kg/day)	0.05	0.05	0.3	0.26	0.1	0.10	0.1
Acetate (mmol/kg/day)	1.75	0	0	0	0	0	0
Micronutrient							
Water soluble vitamins (ml/day)	10	10	10	10	10	10	10
Fat soluble vitamins (ml/day)							
Trace elements (ml/day)	13	10	10	10	10	10	10
Total volume (ml/kg/day)	88	81.1	100.8	92.3	47.3	67.1	67.1
Total calorie (kcal/kg/day)	78.6	80.7	94.5	87.5	87.7	90.8	90.8
Duration of PN infusion (hour)	18	20	20	20	12	12	12
Duration of lipid infusion (hour)	18	20	20	20	12	12	12

bilious aspirates. A lower GI contrast study was done and showed a tapering and no contrast beyond ascending colon. She was referred to University Malaya Medical Centre (UMMC) for suspicion of bowel atresia and further medical management on day three of life.

A laparotomy, appendectomy, multiple bowel biopsies and a jejunostomy were performed on January 01st, 2015 that is day five of her life. During the operation, there was no evidence of bowel atresia but had microcolon and a flexure at the transitional zone, 45 cm from the Duodenojejunal (DJ)-flexure). Results of the biopsies revealed the presence of ganglion cells 20 cm from the distal to the stoma, but absence of ganglion cells 40 cm distal to the stoma. She was diagnosed with Hirschsprung Disease-Total Colonic Aganglionosis. The repeated laparoscopic biopsies on March 2015 had confirmed that ganglion cells present up to 20 cm distal to the stoma. The stoma was re-sited distally; 78 cm distal to DJ-flexure, due to the exteriorized ileostomy was ganglionic as confirmed by the frozen section biopsy of the stoma edge intraoperatively.

In February 2016, at the age of 14 months, she underwent laparotomy, adhesiolysis and serial transverse enteroplasty STEP procedures for bowel lengthening initiative. Prior to the lengthening procedure, the total length of the proximal small bowel to the ileostomy was 130 cm and the enteroplasty process managed to secure another 31 cm length. The proximal bowel was generally dilated but peristalsis was seen. She was started with 2-in-1 total Parenteral Nutrition (PN) in mid-January 2015 through right internal jugular vein. The central venous catheter was re-inserted in June 2015 to the left internal jugular vein. She has been under close monitoring for her nutritional requirements and intake, fluid and electrolyte status, and catheter care during her hospitalization in UMMC.

Finally, in June 2017, the UMMC team has come to agreement that the nutritional requirements and the PN regimen had been stabilized and optimized. She was discharge from UMMC for home

parenteral nutrition. Prior to discharge, her mother has demonstrated competence in performing HPN management; line care and management, setting up enteral and parenteral nutrition under strict aseptic technique and also capable of performing ostomy care. Her family also has made all the necessary adjustment and renovation of their house in Kemaman, Terengganu to provide a dedicated room that is safe and clean for PN administration.

Further discussion and arrangement have been made by UMMC with Hospital Sultanah Nur Zahirah, Kuala Terengganu (HSNZ) for PN supply, with Kemasik Health Clinic, Terengganu (KKK) for home visit and emergency health management, and Hospital Kemaman, Terengganu (HKM) for PN transportation from HSNZ to HKM and routine medical follow-up. All of this arrangement is crucial as the patient will stay 250 km away from UMMC, while HKM, the nearest hospital to patient house, didn't equipped with the facility deemed suitable for compounding PN. HSNZ on the other hand, is about 103 km from HKM.

Discussion

Pre-discharge preparation

Patient's mother underwent home PN training, coordinate by UMMC Surgery Department in 2017. Prior to discharge, an assessment was conducted and she was able to perform and competence in hand washing and gloving techniques, setting up the dressing table, handling PN administration and discontinuation, handling the PN infusion pump and central line care.

A series of discussion were conducted with patient family on the technical aspect of the HPN therapy. Although PN was supplied by the hospital, but some of the consumables were self-purchase by the family. There were also equipment and consumable items, sponsored by Department of Social Welfare Malaysia, Terengganu and Islamic religious and Malay Customs Council, Terengganu such as infusion

Table 2: Summary of nutrient required and delivered to patient via parenteral route.

Macro & Micronutrients	Start (2017) 2 years old			Current (2022) 7 years old		
	Requirement		Delivered	Requirement		Delivered
	per day	by weight		per day	by weight	
Energy (kcal/kg/day)	65-75	845-975	1049 (107%)	65-75	962-1110	1011 (91%)
Amino acids (g/kg/day)	2.5	32.5	39 (120%)	2.0	29.6	43 (143%)
Glucose (g/kg/day)	4.3-8.6	55.9-111.8	151 (135%)	4.3-8.6	63.6-127.3	95 (75%)
Lipids (g/kg/day)	3 (max)	39	36 (92%)	3	44.4	44.4 (100%)
#Vitamins and trace elements (daily dose)	Complete	Complete	Complete	Complete	Complete	Complete
*Fluid (ml)	*	1150 ml	1144 (99%)	*	1240 ml	960 ml (77%)

#Vitamin and trace elements: Soluvit® and Vitalipid®

*Fluid requirement = (100 ml × 10 kg) + (5 ml × 10 kg) + (25 ml × [Weight – 20 kg])

Peptamen Junior; 4 scoops + 120 ml × 3-5/day 464 kcal, 13.7 g Protein, 62.8 g CHO and 17.5 g Fat in 500 ml water.

Table 3: Summary of patient body weight from 2017 until 2022.

		May 2017	Jun-17	Jun-18	Jun-19	Jun-20	Jun-21	Jan-22
Age	Year	2	2	3	4	5	6	7
	Month	5	6	0	0	0	0	0
Weight-for-age (kg)*		16.3	15.8	15.3	15.3	15.3	15.5	15.8
Actual body weight (kg)		13	13	11.5	13	14.8	14.3	15

*The standard growth chart for Malaysia girl age 2-20 years old adapted from National Center for Health Statistics, USA

pump and also syringe pump.

As home PN therapy require a lot of preparation, UMMC had organize teams to help patient family on technical aspects especially for the house preparation. The team visited the house to ensure the room or space for home PN administration, electricity supply, refrigerator, sink, water supply and telephone were in place. All of the aspect is crucial in reducing the risk for contamination and infection to the patient. Pre-discharge home visit was done on Jun 2017 by Kemaman Hospital team, while the post-discharge home visit was conducted by Kemasik Health Clinic team every 3 to 6 months.

The patient was 2 years old and the body weight was 13 kg at the time she was enrolled into the HPN program. Apart from PN, she also taking oral food and received enteral nutrition support. Macronutrient and micronutrient regimen for nutrition support were developed by the UMMC Surgery team. The team reviewed and make the required adjustment according to patient clinical development, relevant policy and also references.

Post-discharge

Monitoring of the treatment progress were scheduled and followed by the medical team and the patient family strictly over the years. Patient current body weight was 15.5 kg to 15.8 kg. Her body weight is on borderline as compared to the Malaysian standard growth chart for girl age 2 to 20 years old [7]. After almost 6 years on PN, not much improvement seen on her body weight. She was physically small compared to other child with similar age, but she is active and had no other medical issue.

As the patient growing up, her PN regimen was re-calculated, re-developed and delivered to suit her new nutritional requirement based on age and weight. Patient was required to be ward for every change made to the PN regimen, to monitor the responds. Discussions have been made by health professional regarding her parenteral nutrition treatment including the use of All-In-One (AIO) PN. Even though changing from TIO to AIO is safe and feasible [8], TIO PN still the most preferred PN among the medical team.

PN solution and lipid solution were packed in separate bags and infused using infusion pump daily. The duration of infusion is 12 h each and normally started around 6.00 pm to 8.00 pm. When first started, the duration of infusion was 18 h and the lipid was in syringes. As the lipid volume increase, there were more syringes to be infuse. The family were required change the syringe four to five time during the infusion time. There is also a need to reduce the duration of infusion as the patient already going to school at age of 6 years old.

The routine of HPN treatment for this patient continue during COVID-19 pandemic. The frequency of attending clinic and blood monitoring were rescheduled and reduced, due to traveling restriction policy in Malaysia. Supply of the PN, lipid and related consumable from HSNZ continue as usual. The family facing challenges in purchasing and supply of other consumable item such as glove, mask and disinfectant solution, as what the whole world facing at that time.

Physical home visit and aseptic technique monitoring was withheld during COVID-19 pandemic 2019 till 2021. The activities continued *via* phone and video calls [9]. Monitoring aseptic technique is crucial and should be monitored regularly to reduce the contamination risk during PN infusion handling. The physical visit and monitoring activities resume on July 2022 by HKM and HSNZ pharmacists. The patient had never been infected by COVID-19 since 2019 and not yet vaccinated. COVID-19 vaccination is still voluntarily in Malaysia since 2021.

There was no standard guidelines for HPN service in Malaysia at the time this patient started HPN. Guidelines developed by ASPEN, ESPGHAN/ESPEN and NICE were the source of information available during those days. It is quite a challenge for medical team to manage HPN patients with very little expertise, experience and information. In 2021, Ministry of Health Malaysia has published the Home Parenteral Nutrition: A Guide for Pharmacist 2021, to help pharmacist and medical team in managing HPN programs.

Conclusion

HPN is an alternative nutrition strategy delivered to support

patient live a normal life outside the health institution. All activities related to HPN program should remain uninterrupted and flexible enough to secure health and medical safety even in the era of pandemic.

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