Parenteral nutrition in children with severe neurodisability: a panacea?

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A 14-year-old adolescent with complex motor disorder and severe cerebral palsy was referred for further management. Over the last year, he developed poor feed tolerance, increasing dystonia and distress with feeding. There was no improvement with medications or continuous jejunal feed. Symptoms improved with enteral hydration or intravenous fluid. In the absence of identifiable cause for their child symptoms and the apparent feed/symptom association, the family wanted to explore all options, including parenteral nutrition (PN).

Neurodisability is an umbrella term used to describe a heterogeneous group affected by central nervous and/or neuromuscular deficit. Their motor functional output can be objectively assessed by Gross Motor Functional Classification,[1] but the group comprises a diverse and often overlapping range of difficulties. The enteric nervous system, a semiautonomous part of the nervous system responsible for regulating the gastrointestinal (GI) function, is invariably affected with neurodisabilities and can drive some GI complaints.[2] With increasing longevity of children on the extreme end of neurodisability, a new chapter of complex GI symptoms begins to unravel, and novel terminologies, such as GI dystonia, although poorly understood, are fast becoming embedded in medical jargons.

PN is classically used to provide nutrition when the GI tract is unable to support sufficient nourishment. In most children, the requirement of PN is temporary, but minority will need long-term nutritional support, home PN and intestinal transplantation.[3] In the neurologically able child, healthcare professionals can navigate this journey supported with guidelines and a plethora of evidence,[4] but are left to the ambiguity of anecdotal evidence and personal experience to deal with the neurodisabled child who develops declining gut function, leading to variation in practice and divergence of care.

With the right level of multidisciplinary support and nutrition support teams, PN can sustain the nutritional need throughout childhood and transition to adulthood. For particular groups such as short gut, the safe usage of PN allowed vital time for gut growth, adaptation and to research and develop new medications with potential of reversing intestinal failure.[5] In severe GI motility disorders, long-term PN permitted control of symptoms and halting the decline in gut function.[6] It may be reasonable to assume similar effects can be achieved by using PN in children with neurodisabilities.

In most high-income countries, the advanced provision of care to children with severe neurodisabilities allowed 'invasive' support to become acceptable, and complex orthopaedic procedures, neurosurgical operations, intensive care and airway support are considered routine across the spectrum of neurodisabilities. Organs transplantation are increasingly offered to children with significant neurodisabilities.[7 8] An argument will arise why long-term and home PN is not routinely offered to these children.

Feeding is a social event and is an opportunity for many parents to bond and interact with their children from an early age. It is also associated with the relatable pleasurable experience of postprandial satiety. Due to the strong psychosocial rewards from feeding one's child, many parents struggled to move their child from oral to gastrostomy feed.[9] The widely available sterile liquid feeds add another complex layer to parents striving for normal family meal time. Healthcare professional supervision of weight gain and nutrient intake, although beneficial and credited with reducing incidence of malnutrition, may further lessen the act of feeding to another medically dictated task. The boundaries are pushed further with non-physiological feeding (continuous, jejunal and overnight feedings). It is not surprising to see a surge in uptake of blended diet for many children with severe neurodisabilities.

Over the recent years, many paediatricians are faced with cohorts of children who have severe neurodisabilities and escalating GI complaints. Pain, poor feed tolerance and distressing neurological events appear to consolidate around feeding. While it is not possible to prove casual relation in the non-verbal child, cessation of feed can ease some of the distress. The fact that many children have no noticeable improvement from manipulation of feeds or medications will further strengthen the belief of GI origin of symptoms. Reduction of feed below nutritional adequacy or switching to hydration fluids can be associated with starvation in many minds and can trigger an independent emotional stress in many families. If the child is not on an end-of-life pathway and in the absence of psychological support, parents may feel they are unwillingly but actively hastening the demise of their child, which may strain their relationship with healthcare professionals. In this setting, PN may appear as an attractive option, but short-term PN can lead to long-term dependency and home PN, potentially exposing the child to life-threatening complications such as central venous access line sepsis and PN-related liver disease.

The outcome of home PN is generally good with positive outcomes including weight gain, reduction of venous catheter infection and providing an assured and symptom-free route to deliver nutrition and hydration. Particularly appealing in children

whom food is the trigger for unpleasant symptoms such as in paediatric intestinal pseudo-obstruction.[10] In children with irreversible intestinal failure, bowel transplant can be an alternative to long-term PN, and this is usually discussed early on to give the young person and their families sufficient time to consider and deliberate.

Home PN is also a heavy burden medical intervention.[4] The level of training and responsibilities involved may discourage extended family members to come forward, limiting childcare arrangement and further exacerbating parents' feeling of isolation and despair. PN bags and tubing should be protected from light, necessitating the child to be indoors during PN administration. Home PN should ideally be infused overnight to allow children to attend school and to participate in activities during the day. Prolonged PN infusion into the day can complicate parent's daily routines. Unlike enteral feeding, which is easily initiated and completed outside the home, PN connection/disconnection requires strict adherence to hygiene protocols and specialist equipment that are difficult to follow outside the child's home. This may impact parents' ability to work, socialise or even leave the house without meticulous time planning. In children with complex medical need who require multisystem support, initiating home PN may further medicalise the relationship between parents and their children. This is particularly acute in families without comprehensive external package of care where parents are expected to provide most of the child's care.

It is an easily understood concept that most medical interventions are directed towards reversing pathological state and are aimed to return the child to baseline physiological state of symptom freedom. However, for some GI complaints, a different concept arises; it involves moving away from physiological state (eating orally) to a non-physiological state (feeding liquid diet through tubes) not always to control symptoms but mostly to prevent damage (such as aspiration). Feeding can thus become an intervention, 'a treatment strategy' particularly when the child refuses to eat after gastrostomy insertion. Continuous feeding and PN can suppress appetite, and many children stop eating altogether on these modalities, further removing the child from the psychosocial dynamic of eating; this can be challenging to some parents of non-verbal children.

It is important to understand the complexity and the multifactorial consideration that are required throughout the decision-making process when dealing with feeding children with severe neurodisabilities. Clinical decisions about children's care are often made jointly by professionals and parents who commonly work in harmony, relying on parents to make good decisions on behalf of their children. When parents' and clinicians' opinions diverge and disagreement ensue, the child's best interest should be the primary concern of healthcare professionals.[11] All efforts should be used to bridge the difference in opinions, including mediation and independent clinical reviews,[12] but the focus must remain firmly on the child's best interest.[13] In the case of invasive nutrition, this can create an ethical dilemma, and a formal ethical review is increasingly sought and is considered part of home PN assessment pathway is some centres. The conflicting moral considerations in this context arise from at least two of the four pillars of medical ethics, namely, beneficence (do good) and non-maleficence (do no harm), often combined to achieve net benefits over harm and/or benefits with minimum harm, but can also extend to respecting the patient's autonomy (tailored to the individual need of the child as a beneficial treatment for one patient can be harmful to another) and justice (to ensure a fair distribution of resources, respect of the law and people's rights).[14 15] Clinicians should use reflective judgement to ensure there are realistic prospects to reach the desired treatment goal, offer good rationale for the preferred course of action, explore all morally available options, minimise infringement to achieve the desired aim and to impartially treat all concerned parties.[16]

Some of the issues that create the dilemma are the poorly understood mechanisms leading to gut decline in children with severe neurodisabilities, the difficulties in objectively assessing symptom correlation to enteral feeding, whether symptoms are in fact related to the GI tract, and the role of parental anxiety and its impact of the child's distress.[17 18] Although it is acknowledged that raising children with significant neurodisabilities can produce a positive outcome for many families,[19] the highly demanding care burden and multifaceted stains can lead to increased incidence of anxiety and depression among parents of children with neurodisabilities.[20 21] A large European study reported a high level of stress in 26% of parents of children with cerebral palsy compared with 5% in the general population. Parents of children with pain, communication and behavioural difficulties are at higher risk of stress, which may affect their abilities to respond effectively to their children's behaviour or to work productively with healthcare professionals looking after their child.[22] Chronic pain is common in this population of children; parental stress and severe motor impairment are associated with increased parent-reported pain compared with self-reported pain in children with cerebral palsy.[23]

The term 'gut dysmotility' is often used in an attempt to explain the GI origin of symptoms, but such terminology can be misleading and challenging to investigate. It is a non-specific term, has no definitive diagnostic criteria and is often used interchangeably to refer to many motility disorders of the GI systems. Many central nervous system disorders and medications commonly used in comorbidities associated neurodisabilities are known to adversely affect GI motility, leading to gut dysmotility.[24] The currently available motility investigations are designed to investigate the GI motor function and bolus flow across the GI tract.[25] Manometries (oesophageal, antroduodenal, colonic and anorectal) can assess the integrity of neuromuscular function in the respective region but may offer limited data on intraluminal bolus movement and do not provide correlation to symptom.[26] Scintigraphy can objectively quantify the passage of radioactive tracer across different parts of the GI tract with normative values validated for gastric emptying, whole gut and colonic transit times.[27] Unfortunately, GI scintigraphy results have a poor association with clinical symptoms, have low diagnostic yield and do not predict response to therapy.[25] Tests that can assess symptom association are highly specialised and designed to study correlation with specific disorder (such as multichannel intraluminal impedance and pH to study symptom association with gastro-oesophageal reflux disease).[28] Other available motility investigations (transit markers, breath test, wireless motility capsules and MRI) are practically challenging to perform in children with severe neurodisabilities and do not objectively assess symptom association hence cannot explain the patient's symptoms nor can they offer a specific diagnosis. Extrapolating their original indications to our group of patients may not be appropriate.

After many discussions focusing on outcome measures, quality of life associated with different interventions and carefully drawn plan to manage the episodes of heightened symptoms, the parents of the 14 years old child in question have grasped the complexity of intravenous nutrition and the potential negative impact on their child's quality of life. They agreed to continue with enteral feeding and ongoing engagement with the wider multidisciplinary team. A plan to review medications with known adverse effects on the GI tract and a clear guidance to

use gut rest to maintain hydration and comfort was agreed on. A clear communication that is built on trust between families and healthcare professionals was the key to ensure the most appropriate path is followed. A multidisciplinary approach to manage expectation and agreement on outcome measures will allow families to raise their pressing concerns and feel part of joint decision making.

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