Case report

Intestinal failure in children and young people with neurodisabling conditions

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ABSTRACT

Gastrointestinal dysmotility is common in children and young people with neurodisabling conditions. In this article we seek to highlight the increasing difficulties faced by paediatricians in managing intestinal failure in this patient group. It is becoming clear that, as the median age for survival increases, intestinal failure is a significant problem, and can in some cases become lifelimiting. The ethical issues around starting children with life-limiting conditions on parenteral nutrition (PN) are extremely complicated, not least because we are ignorant of the mechanism of intestinal failure in these children, and indeed, which of these children might be able to return to enteral feeding after a period of PN. Our article highlights these issues, drawing on our experience of a particularly difficult case, which we hope will stimulate further discussion among paediatricians providing care for children with neurodisabling conditions.

INTRODUCTION

Gastrointestinal (GI) problems are common in children and young people with neurodisabling conditions and GI dysmotility is present in the vast majority. This commonly manifests as foregut and colonic motility dysfunction, and can be complicated by such factors as kyphoscoliosis and unsafe swallow.1 GI involvement in neurodisability is not surprising given the density of enteric nerve complexes interacting with the central nervous system.

Developments in managing neurodisabling conditions such as cerebral palsy (CP) have led to improved outcomes. Brooks et al have shown an increase in the median age of survival of 4-year-olds with severe CP from 11 years to 17 years over the past three decades. Although the condition itself is non-progressive, as children with severe CP are surviving longer it has become clear that GI dysmotility may evolve with time, potentially becoming a lifelimiting factor as enteral feeding becomes very difficult or even impossible. This takes paediatricians into unchartered territory when offering feeding advice for these patients, with little published guidance.

The traditional approach to managing GI complications has been multifaceted, taking into account such aspects as seating position and laxative therapy. The demographic of children with neurodisabling conditions is evolving however, and so too their GI problems. At the Noah's Ark Children's Hospital for Wales, we have managed several cases over the past few years that highlight some of these new challenges and we present one.

What is already known on this topic?

- Young people with severe neurodisabling conditions are living longer.
- Progressive intestinal failure is becoming apparent in this patient group.
- There is little published guidance on management strategies.

What this study adds?

- A description of the clinical and ethical problems faced when managing intestinal failure in young people with neurodisabling conditions.
- Stimulus for discussion among professionals facing similar scenarios in clinical practice.
- An ethical discussion on the use of parenteral nutrition as a life-sustaining treatment.

Such case as an example to prompt discussion among professionals.

CASE HISTORY

ND was born at term without complication and was bottle-fed from birth. Developmental delay was apparent from 8 months and extensive investigation revealed no specific cause. He was diagnosed with CP and went on to develop epilepsy and severe learning difficulties with autistic features.

ND initially suffered with gastro-oesophageal reflux, and then vomiting and constipation through his early childhood, sometimes opening his bowels at 3 weekly intervals. Aged 13 years, he presented to a Cardiff hospital with an acute abdomen. X-ray revealed grossly dilated bowel loops in keeping with obstruction and in view of his ongoing sudden deterioration, he was taken to theatre for life-saving surgery. At laparotomy, necrotic patches of colon were identified and a subtotal colectomy was performed. Histology later confirmed complete infarction from the transverse colon to the resection margin distally.

The postoperative period was complicated by respiratory failure and gross generalised muscle weakness. Nasogastric (NG) feeds were not tolerated and nasojejunal (NJ) feeding was also unsuccessful. After multidisciplinary discussion and with the agreement of the parents, a central venous catheter (CVC) was inserted and he was commenced on parenteral nutrition (PN). This was anticipated to be a temporary measure during his postoperative recovery.

Attempts at reintroducing enteral feeds failed and he spent a total of 3 weeks on paediatric intensive care. Shortly after he was discharged to a general paediatric ward, he had treatment for suspected line sepsis and this set the trend for a prolonged, difficult admission. He remained an inpatient for over a year, during which time he continued to have intestinal failure (IF) with recurrent episodes of sepsis, including disseminated fungal sepsis.

At one stage he briefly tolerated NJ feeding, but soon developed abdominal distension with dilated bowel loops and so his feed was stopped. Mechanical obstruction was excluded and investigations suggested severe dysmotility through the remaining gut.

After further discussion among professionals and with the family, PN was recommenced. Regular multidisciplinary meetings were held over the following 5 months with the family, reviewing the clinical and ethical issues of PN use for ND, before a decision was made to instigate palliative care. By then it was felt unlikely that his gut would recover and his parents were in agreement to remove the CVC.

ND was given NG tube feeds and allowed some oral intake for comfort. He very slowly started to tolerate increasing volumes and over the coming 3 months, was able to reach full enteral feeds. He gradually gained weight, had fewer infections, and with physiotherapy input, started to develop a degree of mobility.

He went home over a year after his initial presentation with a package of care, on full enteral feeds, and was for full resuscitation should he become acutely unwell. He is now 19 years old, doing remarkably well and enjoying a good quality of life with his family.

DISCUSSION

This case demonstrates the uncertainty faced when managing GI problems in young people with neurodisability. In the presence of foregut and colonic dysmotility, ND had a life-threatening GI insult, leaving him with a dysfunctional small bowel. One could speculate that this was caused by a sigmoid volvulus secondary to chronic constipation, an impacted distal loop twisting on its mesenteric pedicle causing a closed-loop obstruction.

While on PN, evidence of infection was monitored vigilantly and treated aggressively. Postoperative complications no doubt contributed to the protracted recovery time, particularly septic ileus, and the removal of the CVC could potentially have removed the source of a persistent low grade sepsis.

Six months post operation, ND depended on PN as a lifesustaining treatment (LST), and therein lay the ethical dilemma.

The Royal College of Paediatrics and Child Health suggest careful consideration is given to the role of PN as an LST for children with neurodisabling conditions, and to discuss decisions on its provision fully with the family.4 They also highlight the importance of maintaining the provision of high quality care, and basing decisions to limit LST on the same basis as one would for non-disabled children.4 The parents of ND were closely involved in discussions around commencing PN and through regular meetings, their views were taken into consideration up to and beyond the decision to withdraw treatment. The professionals involved detailed an advanced care pathway with the family, agreeing on a management plan for each of the medical possibilities.

This case is similar to many others and raises the difficult question of when it is justified to commence PN in a child with a life-limiting condition who develops IF? If commenced, when is it appropriate to withdraw PN? The added complexity in answering these questions comes from the uncertainty around the natural progression of IF, the possibility of resolution and the difficulties in accurate prognosis.

ND now enjoys a reasonable quality of life; however we have seen several patients who continue to rely on long-term PN as they approach adolescence. One could argue that it is the responsibility of the healthcare team to introduce PN in the child's best interest in the acute situation, and to continue until enough information is available to reach a high degree of confidence regarding outcome. The alternative view is that PN should not be commenced in such situations, as this is prolonging the dying process without any overall benefit to a child who might warrant palliative care.

In this relatively novel scenario it is not clear what the mechanism driving the developing IF is, and therefore difficult to predict whether gut function will recover. How do we assess which children will recover, which will have a stepwise deterioration, and which will never recover? These questions are yet to be answered and outcomes will vary from child to child.

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