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Deposited on: 1 November 2016
Transformation of the paradigm in intestinal failure; future prognostication and quality of life, not just survival.

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Dr Barclay and Dr Henderson received no additional support in the writing of this article

Dr Barclay and Dr Henderson have no relevant conflicts of interest in the writing of this article

Word Count 718
Figures 1
Dr Barclay reviewed the article on two occasions and primarily wrote the manuscript.

Dr Henderson reviewed the article after acceptance to *the journal* and assisted in the writing of the manuscript.
The use of parenteral nutrition (PN) has been the mainstay of treatment for intestinal failure (IF) for over 40 years. Until relatively recently the major complications of PN, namely sepsis and IF-associated liver disease (IFALD), have been the primary limiters of good long-term outcome (1). The multifactorial developments of the last decade have transformed the paradigm for patients with long term (Type III) IF, often evidenced by the serial publications of the Paris group (2-5). The authors of the article in this issue of JPGN (6) should be congratulated on the outcomes described; 98% 10 year survival, in a cohort with a median duration of 10 months PN, represents a new aspiration for professionals looking after these children. However their stratification of patients by diagnostic category and weaning prognostic factors sheds new light on the possibilities for patients with ‘potentially irreversible IF’ (ultra short segment short bowel syndrome [SBS], neuromuscular disease [NMD] and mucosal disorders [MUD]).

As with previous studies the authors conclude, through multivariate analysis, that SBS characteristics (>40cm small bowel, preserved ileocaecal valve and preserved colon) predict enteral weaning, but non-weaning is no longer predictive for death or IFALD. With only one death in the cohort, the Paris group now describe equitable survival for NMD and MUD despite reduced likelihood of weaning (41% and 14% respectively). This represents a triumph for NMD patients, for whom outcomes and quality of life have lagged behind other IF diagnoses. Indeed the absolute number and proportion of NMD patients on long-term PN has risen considerably in the last two decades (2, 7), but again the authors’ study describes equitable outcomes for the first time. These gains may also be secondary to organisation of specialist services for complex NMD within supra-regional motility units. With the prospect of novel restorative therapies for NMD on the horizon (8), this lends more favour to long term PN as a therapy versus small bowel transplantation in such cases. Whilst small bowel transplantation continues to have an important role for patients with ‘potentially irreversible IF’, the reduction in morbidities and resultant improvement in quality of life demonstrates a shrinking proportion of patients requiring intervention despite year-on-year rises in the incidence of diseases requiring long-term PN (9).

What this paper clearly articulates is an expectation for patients with ‘potentially irreversible IF’ to spend the majority of time in the community, free from major morbidity and particularly IFALD. Their serial publication in this field is also a firm advocate of the role of
the large specialist service dedicated to all aspects of care of ‘potentially irreversible IF’. The evolution of the nutrition support team, led primarily by the paediatric gastroenterology multidisciplinary team (MDT), into larger interdepartmental intestinal rehabilitation programmes (IRP) has been advocated by many large centres and the transformative effect of this is beyond reproach (10). However, despite systematic review, the analyses of the individual elements and organisation of the IRP remain elusive in terms of an evidence-based appraisal of the contributions of specialist PN/lipid prescribing, sepsis prevention, intestinal lengthening surgery, specialist motility expertise and other MDT input (Figure 1). We anticipate potential future publication from the Paris group on individual aspects of their care, to aide professionals to replicate these outcomes.

Whilst these data support the further improvements in long term PN, challenges remain for patients with poorer outcomes, often minor sub-groups of such study, or those with non primary gastrointestinal disorders. Patients with early onset inflammatory and immune-mediated diseases (including autoimmune enteropathy, classified as non-primary gastrointestinal for the purposes of this paper) for example, still report poorer outcomes than the IF group as a whole. Further understanding of the pathogenesis and mechanisms of these diseases will hopefully lead to more targeted therapies, and the ESPGHAN funded GENIUS patient registry is now recruiting to help characterise these diseases(http://www.genius-group.org/). Additionally as all measures for long term PN improve, the boundaries for such therapy will continue to move; mild dysmotility or dysmotility in the context of severe neurodisability are conditions that the NST are being asked to manage more frequently. As it stands the authors’ paper is a welcome addition to the literature and offers us the future agenda for IF, namely the ability to prognosticate for the individual patient and the primary focus to move to quality of life rather than just long term survival.
References


Figure Legend

**Figure 1**: Shifting the paradigm in intestinal failure over the last decade as evidenced by the Paris Group.

IRP; Intestinal Rehabilitation Programme
Shifting the paradigm in IF

- IRP
- Novel PN prescription
- Mortality assessment
- Growth factors
- Oral feeding
- Enteral antibiotics
- Line locks
- Bile analogues
- Intestinal lengthening surgery