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Original article

Long-term outcome of patients with systemic sclerosis requiring home parenteral nutrition☆

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SUMMARY

Background & aims: Patients with systemic sclerosis may develop intestinal failure requiring home parenteral nutrition. However, few outcome data have been reported. This study aimed to review the outcome of patients with systemic sclerosis receiving home parenteral nutrition.
Methods: Records of all patients with systemic sclerosis who commenced home parenteral nutrition, at a national intestinal failure unit were retrospectively reviewed. Disease characteristics, survival and outcome data were evaluated.
Results: Twenty five patients (20% male; median age: 55 years) were included over a 22-year period (37,200 central venous catheter days). All patients had small intestinal involvement. Prior to home parenteral nutrition, 16 failed enteral feeding. Nine patients were trained to self-administer their home parenteral nutrition; carers/relatives were trained for the remainder. Cumulative survivals on home parenteral nutrition at 2, 5 and 10 years were 75%, 37%, and 23%. Sixteen patients died from causes unrelated to home parenteral nutrition. Two patients were weaned off home parenteral nutrition. Seven patients survive on home parenteral nutrition (median: 41 months; range 9–178). Central venous catheter-related complications were low; these included occlusion (0.70 episodes per 1000 central venous catheter days), sepsis (0.19 episodes per 1000 central venous catheter days) and central venous thrombosis (0.11 episodes per 1000 central venous catheter days).
Conclusions: This is the longest, largest reported series of patients with systemic sclerosis receiving home parenteral nutrition. It shows that home parenteral nutrition can be used safely and effectively in patients with very severe systemic sclerosis-related gastrointestinal involvement.

1. Introduction

Any part of the gastrointestinal (GI) tract may be involved in patients with systemic sclerosis (SSc) [1]. Over 90% of patients have some GI involvement, with manifestations including reflux, dysphagia, gastroparesis, small bowel dysmotility with or without bacterial overgrowth, pseudo-obstruction, constipation and faecal incontinence [1,2].

Eighteen percent of patients with SSc are reported to be at high risk of malnutrition, often – but not solely – as a result of their GI involvement [1,3]. Indeed, 3–4% of patients with SSc die from GI-related causes, and malnutrition leading to intestinal failure (IF) is a principal contributor to this mortality [4,5]. Thus, malnutrition should be addressed through the provision of tailored nutritional support. Few reports describe the successful use of percutaneous enteral feeding in patients with SSc, and most success is reported in patients without small intestinal disease [6–9]. For patients with malabsorption as a result of small bowel disease, home parenteral nutrition (HPN) may be a viable alternative. However, HPN is a

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complex intervention with significant associated risks and data evaluating the long-term outcome of HPN in the SSc population are sparse. Indeed, to-date, only 3 papers describe the short term outcomes of series of patients with SSc requiring HPN (8, 12 and 15 patients followed for a median of 40, 16 and 30 months respectively) [10–12]. A fourth paper describes nutritional support in 5 patients with SSc (4 on HPN) [8]. Other evidence for HPN use derives from case studies of 1–2 patients [13–15]. Thus, little is actually known about the long-term outcome of patients with SSc on HPN. Furthermore, there are minimal data describing the complications associated with HPN provision in patients with SSc versus other groups of patients with IF.

This study, the longest and largest series of patients with SSc requiring HPN, aimed to review the disease characteristics and survival and outcome data of all patients with SSc, who commenced HPN, at a national U.K. IF referral centre, over a 22 year period.

2. Materials and methods

This study was conducted at a centre which houses both a tertiary rheumatology referral centre for patients with SSc and a National IF Unit. The details of all patients requiring HPN are stored on a prospectively-maintained database. This database was used to identify all those patients with SSc who received HPN between May1990 and October 2012. All available records were reviewed for each patient. Any information pertaining to demographics, SSc characteristics, GI manifestations and HPN characteristics (including survival, complications and management) were recorded. The functional ability of patients with SSc was assessed using a validated 11 item self-reporting questionnaire [16]. Results of this questionnaire from within 2 years of HPN initiation were also noted. All surviving patients gave their consent for this review of their records (NRES Committee North West, U.K.).

Survival data for all other patients on HPN for over 3 months, at the same national IF unit, were available for comparison to the SSc cohort.

2.1. Statistical analysis

The difference between the mean SSc functional statuses, recorded within 2 years of HPN initiation, of patients who were trained to care for their own catheter and those who were not trained, was assessed using an unpaired Student's t test, with a significant difference accepted as a p value of ≤ 0.05. The difference between the mean times from SSc diagnosis to HPN initiation of patients with different disease sub-types was assessed using an unpaired Student's t test, with a significant difference accepted as a p value of ≤ 0.05.

The cumulative probability of survival for all patients with SSc was calculated using the Kaplan–Meier method (SPSS version 20), censoring patients upon discontinuation of HPN (weaning) or HPN continuance at the end of follow up.

Survival data were available for all other patients, at the same national IF unit, who had survived on HPN for more than 3 months. Data were unavailable for those patients who had received HPN for less than 3 months. Therefore, for the comparison of survival on HPN between the patients with and without SSc, any patients with SSc, surviving on HPN for less than 3 months, were excluded. All patients, who had received HPN for more than 3 months, were censored upon discontinuation of HPN (adaptation, surgical intervention or transplantation), loss to follow-up or HPN continuance at the end of follow up. A log rank test was performed in order to identify whether survival differed significantly between the groups (patients with and without SSc). In addition, three Cox regression analyses were performed to compare survival between the groups after adjusting for differences in age and HPN start date. Fitting the models in a sequence is informative as it shows how the hazard ratio (HR) for the patient group changes after introducing each potential confounder, thereby highlighting possible explanations for any apparent differences in survival other than the underlying disease. The first Cox model included only the patient group (with or without SSc), giving an estimate of the hazard of death for patients with SSc compared to patients without SSc, unadjusted for other potential confounders. The second model extended this by introducing age, giving a HR for patients with SSc compared to those without, after taking age differences into account. The final model extended this further with the addition of HPN start date; HPN start date was coded as an integer, with the first year in the dataset [1978] set to zero and unit increases for each subsequent year. This adjustment for HPN start date was made in order to adjust for any underlying trend resulting from changes to the treatment over time.

Using the same data sets, a log rank test sub-analysis was conducted to determine whether survival significantly differed between the patients with SSc-related dysmotility and the patients with non-SSc related dysmotility. In addition, two Cox regression analyses (unadjusted and age-adjusted) were performed to estimate the hazard of death for patients with SSc compared to patients with non-SSc related dysmotility.

3. Results

3.1. Demographics of patients with SSc

Twenty-five patients (5 (20%) male) with SSc commenced HPN during the 22 year period (1990–2012) and were managed on HPN over 37,200 intravenous central venous catheter (CVC) days. Early data from 7 of these patients were included in an earlier series publication [11]. The median age at HPN commencement was 55 years (range 24–76). HPN use has increased over time. During the periods 1990–1994, 1995–1999, 2000–2004, 2005–2009 and 2010–2012, the number of SSc patients commencing HPN was 2, 3, 6, 9 and 5 respectively.

3.2. Systemic sclerosis characteristics

Nineteen of the 25 patients (76%) had limited cutaneous SSc (lcSSc) and 6 (24%) had diffuse cutaneous SSc (dcSSc)[17]. Ten (40%) patients were anti-centromere positive and 4 (16%) were anti-topoisomerase positive, while 10 (40%) were anti-centromere and anti-topoisomerase negative. The autoantibody status was unavailable for 1 patient.

Thirteen (52%) patients had evidence of digital pitting, reflecting the severity of digital vasculopathy. Six patients were recorded as having had a digital amputation either before or after starting HPN. Ten patients had cardiac disease.

3.3. Time to HPN commencement

The median interval from the onset of SSc, as defined by the patient's recall of the date of their first non-Raynaud's clinical feature, to HPN commencement was 102 months (range 14–389; $n = 23$). There was no evidence of any difference between disease sub-types for mean interval to HPN commencement (dcSSc 101 months (standard deviation 77); lcSSc 116 months (standard deviation 100), $p = 0.74$).

3.4. GI disease characteristics

All patients underwent multiple GI investigations, prior to, and following, commencement of HPN. All patients had evidence of

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