Dietetic led step-wise PKU return to diet care pathway: greater reductions in blood phenylalanine levels and faster return to full diet

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BACKGROUND

In 2017, we developed a step-wise patient-initiated return to PKU diet care pathway (Version 1.0 (V1.0)); and modified it in 2019, to include additional regular fixed dietetic follow-ups (Version 2.0 (V2.0)) (see Figure 1 and 2). The pathway aims to make the multiple dietary and lifestyle changes required less daunting for patients, by introducing one change at a time. Phase 1 is focused on establishing protein substitute and Phase 2, on reducing dietary protein. Progression through the pathway is planned over 6 months; however, patients can progress at their own pace.

We evaluated the impact of additional dietetic follow-ups (V2.0) on return to diet success.

METHODS

Retrospective data was collected from April 2017 to Dec 2020 from patients who had initiated the step-wise approach care pathway in returning to a PKU diet; including demographics, days taken to progress through each phase, and blood phenylalanine results.

To allow for comparison between V1.0 and V2.0, only patients who initiated either V1.0 or V2.0 were included in the analysis. Patients who were involved in both V1.0 and V2.0 were excluded from the analysis.
**Figure 1: Return to Diet Care Pathway V1.0**

**Phase 1**

- **Initiation**
  - Patient requested to return to PKU diet.
  - Baseline nutritional bloods

- **Trial of protein substitutes**
  - Within 1-2 week of initiation

- **GP prescription request**
  - Patient to inform Dietitian of protein substitute preference.
  - Dietitian to write to GP and arrange home delivery

- **Initiate protein substitute**
  - To start protein substitute and to establish routine before reducing dietary protein

- **Kitchen session**
  - Whilst establishing routine with protein substitute, to have low protein diet kitchen session

**Phase 2**

- **Low protein food trial**
  - Within 1 week of the kitchen session, patient to notify of low protein food sample requests. Low protein food samples sent to patient

- **GP prescription request**
  - Patient to inform Dietitian of low protein food needs.
  - Dietitian to write to GP and arrange home delivery

- **Reduce dietary protein**
  - Once patient receives supply of low protein foods, to start reducing protein to exchange allowance.

**Follow-up:** If no contact received from patient within 3 months, book into virtual clinic to assess motivation/barriers/support needed

**Monitoring:** Patients to send monthly bloodspots from start of Phase 2. Dietitian support via email or phone as required

**Figure 2: Return to Diet Care Pathway V2.0**

**Phase 1**

- **Initiation**
  - Patient requested to return to PKU diet.
  - Baseline nutritional bloods

- **Trial of protein substitutes**
  - Within 1-2 week of initiation

- **GP prescription request**
  - Patient to inform Dietitian of protein substitute preference.
  - Dietitian to write to GP and arrange home delivery

- **Initiate protein substitute**
  - To start protein substitute and to establish routine before reducing dietary protein

- **Group session**
  - Whilst establishing routine with protein substitute, to attend a low protein diet group training session

**Phase 2**

- **Low protein food trial**
  - Within 1 week of the group session, patient to notify of low protein food sample requests. Low protein food samples sent to patient

- **GP prescription request**
  - Patient to inform Dietitian of low protein food needs.
  - Dietitian to write to GP and arrange home delivery

- **Reduce dietary protein**
  - Once patient receives supply of low protein foods, to start reducing protein to exchange allowance.

**Follow-up:** If no contact received from patient within 3 months, book into virtual clinic to assess motivation/barriers/support needed

**Monitoring:** Patients to send fortnightly bloodspots from start of Phase 2. Regular Dietitian support via email or phone

**Follow-up:** If no contact received from patient within 3 months, book into virtual clinic to assess motivation/barriers/support needed

**Video clinic review**
- Occurs 3 months after initiation of step-wise return to diet

**Clinic review**
- Six months after initiation of step-wise approach to diet.
  - Nutritional bloods
RESULTS

A total of 37 adults with PKU (22 females and 15 males) were included in the audit. Mean age of the patients at initiation of the pathway was 35.5 years (15-68 years). Six of the patients (16%) were late diagnosed.

Seventeen adults initiated pathway V1.0 a total of 21 times, and 20 adults initiated pathway V2.0 a total of 22 times. Baseline phenylalanine results (on an unrestricted diet) were available for 32 patients*. Mean baseline blood phenylalanine was 1392 µmol/L (846-2051 µmol/L).

The proportion of patients completing Phase 1, Phase 2 and progressing to full PKU diet are outlined in Figure 3. For patients undertaking pathway V1.0, 86% of patients completed Phase 1, 52% complete Phase 2 and 38% progressed to full PKU diet. For patients undertaking pathway V2.0, 86% of patients completed Phase 1, 41% went on to complete Phase 2 and 27% progressed to full PKU diet.

The median number of days to complete full pathway V1.0 and V2.0 was 255 (110-699) and 160 days (66-311), respectively. The median reduction in blood phenylalanine seen in those following pathway V1.0 and V2.0 was 547 µmol/L and 860 µmol/L, respectively.

*\( n = 2 \) were already on protein substitutes when the phenylalanine result was obtained; \( n = 3 \) did not provide a baseline phenylalanine.

Note: For bloodspot phenylalanine results provided by the laboratory as >1100 or >1400, 1100 or 1400 respectively, were used and therefore the mean phenylalanine is likely to be an underestimate of the true baseline value.
CONCLUSION

Additional dietetic follow-ups (V2.0) resulted in a reduction in days taken to return to full PKU diet, and in greater reductions in blood phenylalanine.

The impact of the additional dietetic follow-ups in V2.0 on long-term adherence is yet to be established. Progression to full PKU diet was completed by 11% fewer patients in V2.0 vs V1.0. This is possibly a reflection of other life challenges experienced during the pandemic as the V2.0 care pathway was implemented in 2019.

Our step-wise approach model has received significant positive feedback from patients, specifically in relation to the option of being able to focus on one aspect of the diet at a time. Incorporating behaviour change techniques, alongside dietetic input, will further support patients in making the challenging diet and lifestyle changes needed when returning to PKU diet.