Abstract

**Home ketone monitoring in Glycogen storage disorders (GSD 0, III, VI, and IX**

**Introduction**

Optimising metabolic control in GSD 0, III, VI, IX with suppression of ketosis can lead to improved growth outcome. Winstone et al 2014

Traditionally this required an inpatient stay for biochemical monitoring which may poorly reflect home circumstances and is time consuming and costly to families and the NHS.

**Method**

28 patients were randomly selected to partake in a trial of 48 hour home ketone monitoring using a machine previously validated to accurately record low ketones levels (<1.0mmol/L). A food diary and patient satisfaction questionnaire were also completed.

**Results**

20 profiles were successfully completed. 9 had suboptimal profiles according to our clinical practice: 8 with ketosis (>0.4mmol), 1 with poor dietary practise. Dietary changes were discussed by telephone. 9 patients were requested to retest with 4 showing satisfactory control. 2 required further intervention and 3 are yet to return profiles.

All returned questionnaires had a preference for home ketone monitoring.

**Conclusion**

Home ketone monitoring has proved effective in managing this cohort of patients. Patient satisfaction was high with this method of monitoring.