Abstract G474(P) Figure 2 Scatterplot with regression line for the association between mean satisfaction and mean HbA1c at PDU level (2012–2013 NPDA data for England and Wales)

G475 SUPPORTING PARENTS TO MANAGE CHRONIC CHILDHOOD CONDITIONS AT HOME: RESULTS OF A FEASIBILITY RANDOMISED CONTROLLED TRIAL OF A NEW INTERACTIVE HEALTH COMMUNICATION APPLICATION

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Background Families living with chronic childhood conditions face multiple challenges and parents have previously identified the need for an interactive health communication application (IHCA) to improve their condition management ability. We developed, and evaluated in a feasibility randomised controlled trial the OPIS (online parent information and support) IHCA.

Methods Parents of children with chronic kidney diseases were randomly assigned to usual health–professional support for caregiving (control) or usual support plus password-protected access to OPIS for 20 weeks (intervention). We assessed feasibility descriptively in terms of recruitment and retention rates overall; assessed recruitment, retention, and uptake of OPIS and compared family condition management between groups using the Family Management Measure [FaMM] and qualitative interviews. Questionnaire data were analysed using descriptive statistics and qualitative data using Framework Analysis

Results 55 parents of 39 children were recruited. Three-quarters of intervention group parents (19/26, 73%) and control group parents (22/29, 76%) were retained, the overall retention rate was 41/55 (75%). The 41 parents completing the trial were asked to respond to the same 10 questionnaire scales at baseline and 20 weeks later; 10 scores were missing at baseline and nine were missing at 20 weeks. All intervention group parents accessed OPIS and showed a greater improvement in perceived competence to manage their child's condition compared to control group parents (adjusted mean FaMM Condition Management Ability Scale, intervention group 44.5 vs control group 41.9, difference 2.6, 95% CI –1.6 to 6.7). Differences between groups agreed with qualitative findings that OPIS improved parents' management ability.

Conclusions OPIS is being made available as standard practice in the hospital where it was developed and evaluated. A full-

scale national trial of the effectiveness of OPIS is feasible. Our design and methodology can be transferred to the management of other conditions in different contexts.

G476 DIABETES MORTALITY TRENDS 1990 TO 2010 IN THE UK COMPARED WITH THE EU15 AND THE USA

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Aims We investigated whether the UK has higher child and youth diabetes mortality than in comparable European countries and the USA.

Methods We obtained data from the WHO World Mortality Database for the UK, the USA and the EU15+ (the 15 countries of the EU in 2004 plus Australia, Canada and Norway) for 1990 to 2010. Diabetes mortality rates were calculated for 1–14 and 15–24 year olds. Multilevel longitudinal Poisson regression models were constructed including all country-level data from 1990–2010 (378 country years) comparing the UK with the EU15+ and the USA. Graphics show average mortality across the EU15+.

Results Mortality trends are shown in Figure 1 for 1–14 year olds (both sexes) and Figure 2 (males) and Figure 3 (females) for 15–24 year olds. In 1990 the UK had higher mortality than the EU15+ amongst 1–14yo ($p = 0.004$) but not amongst 15–24yo ($p > 0.2$ both sexes). Diabetes mortality did not significantly change in the EU15+ across the study period in any age group. In contrast, amongst 1–14 year olds the UK had a significantly higher rate of decline than the EU15+ (negative slope coefficient, $p = 0.03$). UK mortality rose amongst 15–24 year olds compared with the EU15+ (positive slope coefficients, $p < 0.0001$ both sexes) In 1990, the UK had higher diabetes mortality than the US in 1–14 year olds ($p < 0.0001$) but lower amongst 15–24 year olds ($p < 0.0001$ both sexes). The USA had little change in diabetes mortality amongst 1–14 year olds but a significant rise in diabetes mortality amongst 15–24 year olds ($p < 0.0001$ both sexes). The UK had a greater rate of mortality decline than the USA amongst 1–14 year olds (negative slope

Abstract G476 Figure 1 Total diabetes mortality amongst 1–14 year olds (3 year moving averages)

Abstract G476 Figure 2 Total diabetes mortality amongst 15–24 year males (3 year moving averages)

Abstract G476 Figure 3 Total diabetes mortality amongst 15–24 year females (3 year moving averages)

coefficient $p < 0.0001$) but rising mortality amongst 15–24 year olds (positive slope, $p < 0.0001$ both sexes).

Discussion Diabetes mortality in the UK for children 1–14 years approximates the EU15+ mean and is better than in the USA. However, for 15–24 year olds, the UK has high and rising diabetes mortality compared with the EU15+ from 2000 onwards. Further work is needed to understand the contributions of healthcare factors to the UK's poor diabetes mortality record amongst young people.

G477 IDENTIFICATION OF NOVEL PAR2 MUTATION IN THROMBOTIC MICROANGIOPATHY

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Atypical Haemolytic Uraemic Syndrome (aHUS) is a rare cause of renal failure, occurring as a result of glomerular endothelial cell injury, with a prevalence of 3–5 per million. The clinical

features consist of recurrent episodes of haemolytic anaemia, thrombocytopaenia and renal failure with biopsy evidence of thrombotic microangiopathy.

Mutations in complement regulatory genes account for approximately 70% of cases. More recently, components of the coagulation cascade, such as DGKE, have been implicated in the aetiopathogenesis of aHUS. We have identified a novel mutation in protease-activated receptor 2 (PAR2) in a patient with an aHUS phenotype.

Methods Whole Exome Sequencing (WES) was performed on a child presenting at the age of 3 years with nephrotic range proteinuria and biopsy evidence of thrombotic microangiopathy, thrombocytopaenia and haemolytic anaemia. One year from presentation, his renal function recovered to baseline but he continues to experience episodic haemolytic anaemia and thrombocytopaenia. Of note, there is a familial history of relapsing thrombocytopaenia affecting the patient's father and aunt.

Results WES did not demonstrate any mutations in known aHUS genes. A novel mutations in PAR2 (Y345H) was identified, *in silico* analysis of this classified this as deleterious. This mutation was not detected in databases of healthy controls.

Sequence analysis demonstrated segregation in affected family members (Figure 1)

Western blot analysis suggests reduced PAR2 expression in the patient's serum compared to controls. We also show that PAR2 is expressed in glomerular cells (Figures 2 and 3)

Discussion PAR2 is a G-protein coupled receptor, which is activated by a number of pro-inflammatory and coagulation mediators. Activation of this receptor causes a positive feedback loop, leading to a pro-thrombotic state that may result in pathological platelet activation and endothelial dysfunction. As there are high levels of PAR2 in the kidney, this pro-thrombotic state may lead to the thrombotic microangiopathy seen in aHUS.

PAR2 has additional roles in complement regulation that are less well defined. It is known to cause down-regulation of DAF, which is an important regulator of the complement pathway.

Further analysis of genetically undefined cases of TMA and MPGN are required for PAR2 mutational screening to determine

Abstract G477 Figure 1 Segregation analysis showing PAR2 Y345H in affected