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A case study outlining the dietary management of an unplanned Maternal PKU (mPKU) pregnancy.

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Patient Details

Age:
24 years

Gender:

Diagnosis:
Phenylketonuria (PKU) diagnosed on newborn screening.

Relevant history:
No previous pregnancies.
Works part time in retail

The patient called the metabolic unit to inform them that she was pregnant (estimated gestation 5/40).

Relevant Medical History

The patient had good metabolic control during childhood, discontinued a protein restricted diet aged 18 years and had since been lost to metabolic follow up.

Her diet included standard portions of high protein foods such as meat, eggs and dairy on a daily basis. Already taking a pregnancy specific multivitamin which provided 400µg folic acid.

Plasma Phenylalanine (Phe):
No information at time of initial presentation.

Target Phe for preconception and pregnancy at managing centre:
120-250µmol/L

Initial dietary advice provided (over the phone). Two days of dietary education arranged and the patient was able to attend the unit within 24hours.

Action	Rationale
Advised to avoid all high protein foods and consume as many Phe exchange free foods as possible	The patient did not have any protein substitute or low protein foods at home. She was encouraged to base her meals around low Phe fruits and vegetables to minimise dietary Phe intake.
Advised to fortify her diet with fats, oils and sugars.	To encourage adequate energy intake to meet requirements and prevent catabolism.

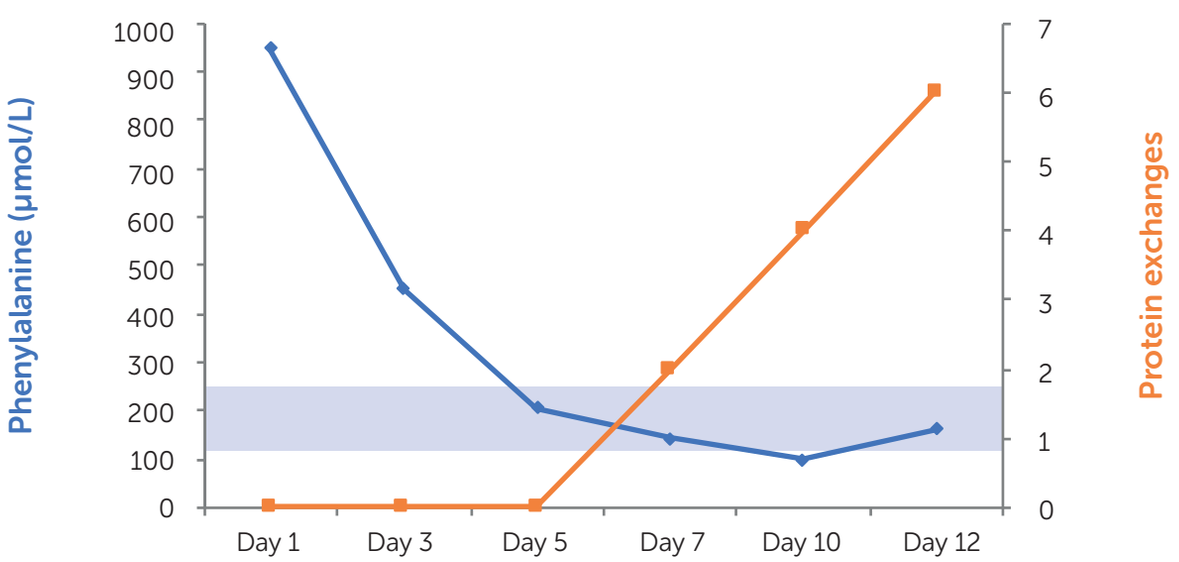
Measurements taken at the metabolic clinic:

Anthropometry:
Weight: 71kg Height:1.70m BMI 24.6 kg/m^2

Protein requirements:
71 x 0.83 x 1.2 +1 = 72g/day (WHO, 2007)
51 x 1.15 = 59g/day (Maritz, 2010)

Action	Rationale
Bloodspot Phe taken when the patient attended the metabolic clinic.	Same day blood sample results reported if sample received before 1pm.
Bloodspot technique training provided by a metabolic clinical nurse specialist.	The patient had not performed a home bloodspot test for many years. Good bloodspot technique is important to ensure results are as accurate as possible.
Commenced on 0g dietary protein (0 x 50mg Phe exchanges).	Once the patient was provided with sufficient supply of low protein prescription foods she could then comply with a very low protein diet whilst maintaining a sufficient energy intake. The patient had not had any previous pregnancies and her plasma Phe was unknown, it was decided to commence 0 exchanges as a temporary measure to bring the plasma Phe level down within target range as quickly as possible.
Commenced on 80g PE from protein substitutes (4 x PKU air20)	The initial dose of protein substitute is higher to help reduce the plasma Phe level quickly. The patient chose to take ready to drink PKU air20 red and white as she found it more convenient and preferred the flavours.
Advised to stop pregnancy multivitamin.	4 x PKU air20 provides sufficient micronutrients and DHA to meet requirements for pregnancy and so additional supplementation was not needed at this time.
Advised to start 400µg folic acid only supplement.	Recommended for the first 12 weeks of pregnancy in addition to folic acid provided in protein substitutes.

A baseline bloodspot Phe level was 948µmol/L. Within 5 days, the Phe level had decreased below 300µmol/L. Phe exchanges were then gradually introduced, titrated to blood results to maintain levels between 120-250µmol/L.

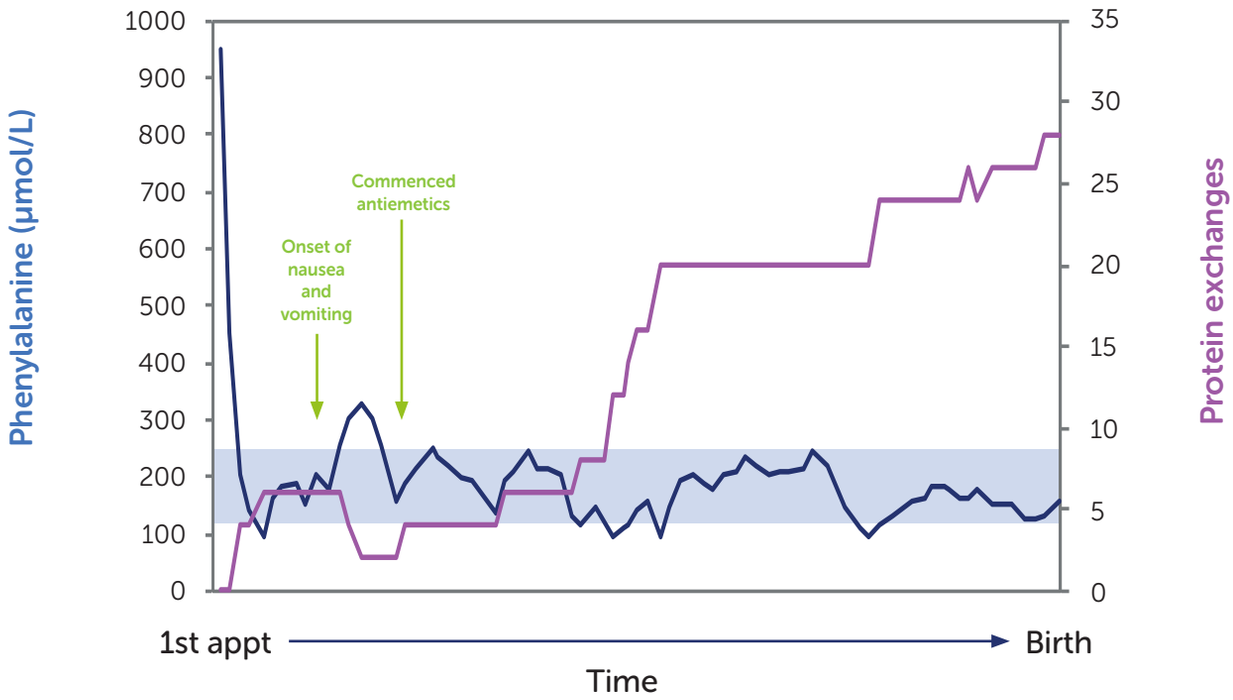


At 8 weeks gestation, the patient developed pregnancy-associated nausea and vomiting. This led to difficulties in consuming all the prescribed PKU air20 and Phe levels rose above 300µmol/l. For more tips on managing nausea, refer to the VIA resource 'A practical guide to maternal PKU'.

Action	Rationale
Reduce dietary protein (Phe exchanges)	To reduce Phe level as quickly as possible to protect the developing foetus from exposure to Phe
Ensure adequate energy intake	When dietary Phe is reduced some women struggle to consume enough calories. The patient was encouraged to include additional oil/butter in cooking and to consume calorie-containing drinks (including fruit juice, permitted sugar containing drinks and low protein milk)
Offered to change presentation of protein substitute.	The patient recalled previously tasting powder and tablet presentations of protein substitute but decided not to change her protein substitute as she did not feel this exacerbated her nausea or vomiting.
Metabolic Consultant contacted the patient's physician to request antiemetics to be initiated	Suitable antiemetics for pregnancy-associated nausea and vomiting are proactively encouraged if metabolic control is compromised by nausea and/or vomiting in pregnancy.

Phe levels quickly decreased back into target range and were maintained in range for the duration of pregnancy.

In the second trimester (from 17/40) Phe tolerance increased, necessitating an increase in dietary Phe (Phe exchanges). An increase of 100mg phe (2 x 50mg Phe exchanges) was advised when bloodspot Phe measured <150µmol/L. As the pregnancy progressed and Phe tolerance continued to increase, the patient struggled to keep track of individual 1g protein (50mg Phe exchanges). To make counting easier she included 300mg Phe portions (6 x 50mg Phe exchanges) and replaced manufactured low protein staple foods such as bread, rice, and pasta with protein containing versions. At the end of pregnancy, total protein intake was 88g (60g protein equivalent from Phe substitute, plus 1400mg Phe (28 x 50mg Phe exchanges). Maternal weight gain throughout pregnancy was 13.8kg.



A healthy male infant was born at 39/40 + 5 days, weighing 3.1kg (9th – 25th centile) with a head circumference of 33cm (2nd – 9th centile). He was initially exclusively breast fed, and moved onto a combination of mixed feeding (breast and infant formula) after 3 weeks. The patient chose to discontinue her Phe-restricted diet and protein substitutes post-partum and return to her pre-pregnancy unrestricted diet and was seen in the metabolic clinic 6 months after delivery. She has continued to remain engaged with follow up and is seen on an annual basis. Contraception and planning for any future pregnancies, if wished, is discussed as part of each clinic appointment.

This case study demonstrates that with the dedication of the patient and support from their metabolic unit, good metabolic control can be achieved within the first 10 weeks of pregnancy even if Phe levels were not in range at the time of conception. As part of antenatal care the patient had a detailed foetal cardiology scan at 24/40 gestation which showed no abnormalities so further cardiology follow up was not arranged after the infant was born. The infant continued to have routine neonatal and paediatric checks, and will be offered developmental follow up at the managing centre at age 2, 4, 8 and 14 years which is routine care for all infants born to women with PKU at this centre.

Reference:

Maritz C, Ellerton C. 2010. Dietary Management of Maternal Phenylketonuria: A Practical Guide.

World Health Organization (WHO), Food and Agriculture Organization of the United Nations (FAO), United Nations University (UNU). (2007). Protein and amino acid requirements in human nutrition. WHO Technical Report . Series 935, 1-265

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All information correct at the time of print

November 2018