# GESTATIONAL DIABETES IN A PATIENT WITH PHENYLKETONURIA: WHEN DIET IS COMPLICATED

**GONZALO MARÍN M, , GONZÁLEZ-MOLERO I, MORILLAS-JIMÉNEZ V, ABUÍN-FERNÁNDEZ J, CONTRERAS-BOLIVAR V, OLVEIRA-FUSTER G.**

**UGC ENDOCRINOLOGÍA Y NUTRICIÓN, HOSPITAL REGIONAL DE MÁLAGA (SPAIN)**

## MEDICAL HISTORY

39-year-old Woman. *Classic Phenylketonuria* diagnosed through neonatal screening in Germany. Good adherence to the diet since childhood, with a good metabolic control. Pregnancy in 2007 with good controls of phenylalanine (Phe).

## EVOLUTION

She planed a new pregnancy, starting off from very good controls (<4mg/dl) and ingestion of 22-24 Phe's portions and supplements without Phe. She did Phe and tyrosine (Tyr) controls weekly, analysis with plasmatic amino acids and dietary survey.

In 24 gestational week, O'Sullivan test was pathological (164mg/dl) and she was diagnosed with *gestational diabetes*. At the beginning, she tried to control carbohydrate intake and improved the glycemic control. In third trimester, low levels of Phe and Tyr did necessarily to increase up to double Phe's rations. Since the patient could not take high biological value proteins, she increased the ingestion of vegetables and changed low-protein cereals, pasta and rice into others with normal protein content. This implied a deterioration of glycemic control that derived in the prescription of two doses of NPH Insulin, and rapid-acting insulin for postprandial control. She needed tyrosine suplements. Delivery was uncomplicated, with a healthy newborn.

Later, she reduced Phe's portions of the diet to her normal. Tyr supplements and insulin terapy was suspended.

| GW | Phenylalanine (µmol/mL) | Tyrosine (µmol/L) | Phe portions | Tirosine (g) | Kcal | Weight (Kg) |
|---|---|---|---|---|---|---|
| 6 | 113 | 43 | 22 | 9,5 | 2500 | 70 |
| 7 | 173 | 88 | 24 | 9,5 | | |
| 10 | 71 | 48* | 26 | 9,5 | | |
| 12 | 200 | 70 | 28,5 | 8 | | |
| 13 | 84 | 43 | 33 | 8 | | 71,3 |
| 16 | 91 | 38 | 35 | 8 | | |
| 19 | 93 | 48 | 37 | 8 | | |
| 20 | 149 | 46 | 47 | 8 | 2880 | 77,7 |
| 24 | 64 | 38** | 42 | 10 | | |
| 25 | 100 | 46 | 47 | 10 | | 82,5 |
| 29 | 67 | 42 | 47 | 10 | | 86 |
| 36 | 116 | 55 | 52 | 10 | 3000 | |
| 38 | 79 | 57 | 52 | 10 | | |
| 41 | 107 | 52 | 52 | 10 | | 94 |

*Changed Vitaflo PKU Cooler 20 to PKU gel 1 per day (1g tyrosine, 70kcal) because of digestive intolerance.

**Added L-tyrosine, 2 per day (1g tyrosine each).

## DISCUSSION

***Maternal phenylketonuria syndrome*** appears in children of mothers affected by seriously or moderated hiperphenylalaninemia with high plasmatic Phe concentrations during the previous months and/or gestation. It is involved in fetus mental retardation, microcephaly, cardiac malformations, esophageal atresia and facial dysmorphia, among others.

Phe crosses the placenta and high plasma levels are difficult to metabolize due to the fetal immaturity. It is indispensable to contribute enough quantity of proteins (low-phenylalanine formula) and energy.

We report a case of a successful increase of Phe's portions based on food and supplements rich in carbohydrates, which complicates the managing of the gestational diabetes and forces us to use the insulin therapy, since it is not possible to reduce the ingestion of carbohydrates because a minor caloric contribution in patients with phenylketonuria would worsen Phe's levels.

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Diabetes (to include epidemiology, pathophysiology)

INMACULADA GONZALEZ

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