Phenylalanine levels in a patient undergoing single anastomosis gastric bypass surgery

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Introduction

- Phenylalanine (Phe) metabolism in patients with phenylketonuria (PKU) undergoing bariatric surgery is not well documented.
- We report the metabolic control in a 36-year-old lady with PKU and morbid obesity who underwent bariatric surgery

Case Presentation

- A 36 year old lady with PKU presented to the Adult IMD clinic
- She was obese with BMI 56kg/m2 (weight 164kg, Height 1.7m), not planning any further pregnancies and wanted to undergo bariatric surgery.
- Concerns were about the effect of rapid weight loss, associated catabolism on phe levels and tolerance to phe free amino acids supplements post operatively due to lack of reported cases of bariatric surgery in the PKU population
- After multidisciplinary discussions, she underwent a single anastomosis gastric bypass surgery.
- Post surgery, there was significant weight loss and phe tolerance appears to have increased with phe levels remaining below target despite increased phe diet and supplements.

Background

- Phenylketonuria (PKU) is an autosomal recessive inborn error of phenylalanine (Phe) metabolism due to mutations in phenylalanine hydroxylase (PAH) gene causing toxic build up of phe in the blood stream.
- It is one of the most common metabolic disorders, frequency (around 1/10,000) among Caucasians.
- It is associated with an abnormal phenotype including growth failure, microcephaly, seizures and intellectual impairment from the neurotoxic effects effect of hyperphenylalaninemia which are linked to tyrosine deficiency.
- Diagnosis in most countries is through newborn screening programs.
- Treatment is low Phe diet with monitoring and control of Phe consumption throughout life and /or use prescribed medications such as Kuvan to avoid neurological complication

Phenylalanine levels in previous pregnancies, pre and post bariatric surgery.

<table>
<thead>
<tr>
<th>Timeline</th>
<th>Weight (Kilograms)</th>
<th>Dietary Phe levels (grams)</th>
<th>PKU supplements (grams)</th>
<th>Phe levels (Range) (µmol/L)</th>
<th>Phe levels (Mean) (µmol/L)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Third trimesters of pregnancies</td>
<td>500</td>
<td>60</td>
<td>374-423</td>
<td>398</td>
<td></td>
</tr>
<tr>
<td>Pre bariatric surgery</td>
<td>172</td>
<td>750</td>
<td>60</td>
<td>229-812</td>
<td>604</td>
</tr>
<tr>
<td>3 - weeks pre-op (on hypo-</td>
<td>164</td>
<td>700</td>
<td>60</td>
<td>373-525</td>
<td>385</td>
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<tr>
<td>caloric liver reducing</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>diet)</td>
<td></td>
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</tr>
<tr>
<td>Post bariatric surgery</td>
<td>140</td>
<td>750</td>
<td>60</td>
<td>296-529</td>
<td>273</td>
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<tr>
<td>5 months</td>
<td>139</td>
<td>1150</td>
<td>60</td>
<td>429-539</td>
<td>528</td>
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<tr>
<td>3 years</td>
<td></td>
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Discussion

- Phe tolerance appears to have increased post-surgery.
- The exact cause is not known.
- Decreased dietary protein absorption is an unlikely explanation, as this would have resulted in higher blood phe due to endogenous protein catabolism.
- Improved compliance, selective decrease in phe absorption due to loss of SLC6A19 in the gut, improved phe oxidation post bariatric surgery could all be possible explanations for apparent increase in Phe tolerance.

Conclusion

- It is possible both, to maintain target phe levels on a significantly hypocaloric diet and to prevent rise in phe levels during weight loss post bariatric surgery.

References