donating a methyl group, thus converting homocysteine to methionine. Limited literature exists outlining the risk of metabolic encephalopathy associated with hypermethioninaemia presenting in children with classical Homocystinuria. Here we present one such case, made more remarkable by the absence of treatment with betaine.

**Case description** Our patient was diagnosed on newborn screening on day 4 of life with classical Homocystinuria and had been promptly commenced on a treatment regime. At four weeks of age he presented to his local hospital with poor feeding, vomiting, and lethargy. Following this acute presentation, he was started on IV antibiotics but a source of infection could not be established. Hyponatraemia, hypertension and proteinuria were noted. His renal ultrasound revealed increased echogenicity bilaterally with no evidence of renal vein thrombosis. His brain magnetic resonance imaging (MRI) showed areas of diffusion restriction bilaterally with normal MR spectroscopy and MR angiogram. Intensive care treatment was required, including intubation and ventilation, and antihypertensive treatment. No underlying thrombo-embolic cause could be established. His blood tests which were taken at the time of his acute presentation to his local hospital revealed a methionine of 1329 μmol/L with a normal cystine (23 μmol/L) and a raised total homocysteine (118 μmol/L) and free homocysteine (11 μmol/L), it was therefore concluded that his presentation was metabolic in origin. These levels improved promptly with further natural protein restriction, increased calorie intake, including intravenous dextrose and electrolytes. The patient made a full recovery; his brain MRI findings normalised.

**Discussion** Hypermethioninaemic encephalopathy is a rare complication of classical Homocystinuria. It has previously been described in association with betaine treatment. We are unaware of any previous publications of this complication in patients not on this medication. In addition to monitoring for long-term complications, we suggest that methionine should be monitored regularly in patients with classical Homocystinuria, in particular during any episodes of acute deterioration.