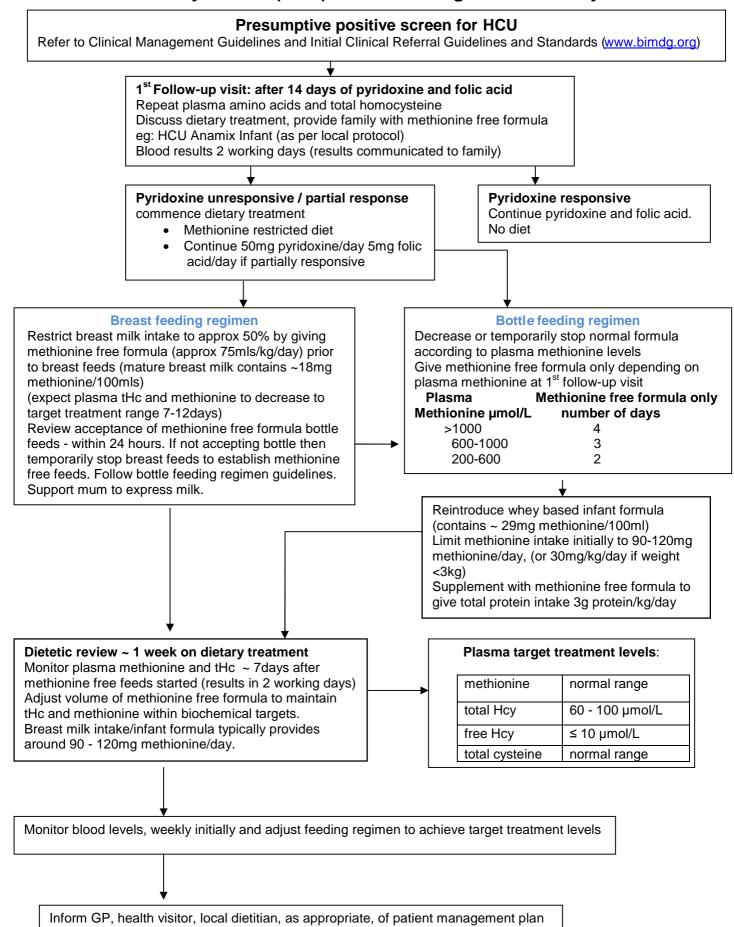
Homocystinuria (HCU) Dietetic Management Pathway



Homocystinuria - Dietetic Management

Dietary management information (as per local centres own resource)

- HCU Dietary information
- Weaning information
 - o low protein 'free' foods
 - o low protein manufactured foods
 - o basic 20mg methionine / 1g protein exchanges
 - o calculating protein exchanges
 - o low protein prescribable products
 - o low protein weaning recipes
 - o second stage L-amino acid supplement
- Prescription letters to GP
 - o methionine free amino acid supplements
 - o low protein manufactured foods & advice regarding prescriptions
- Home delivery services

Follow-up in first year - 'suggested' dietetic management for HCU

| Follow-up visits | Dietetic management pathway |
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| 1 week following introduction of diet | Wt, length (weekly weights) Review feeding – volume of standard infant formula (methionine intake), volume methionine free formula – check total protein intake ~ 3g protein /kg/d Review feeds with results of repeat plasma total homocysteine levels, adjust methionine intake (from infant formula or breast feeds) as necessary to achieve biochemical target levels Monitor plasma cysteine: 1-2 monthly. Supplement if < 170 µmol/L |
| Weekly telephone reviews | Review feeding regimen with results total plasma homocysteine and methionine levels, adjust feeds as necessary to achieve target levels Once achieved target levels consistently (over 4 – 8 weeks) could change to fortnightly monitoring & weights. |
| 4 Months | Wt, length (consider monthly weights) Review feeding – volume standard infant formula (methionine intake), volume methionine free formula – check total protein intake ~ 3g protein /kg/d Weaning stage 1 • Discuss low protein 'free' baby foods |
| 6 Months | Wt, length (monthly or more as necessary) Review feeding – volume standard infant formula (methionine intake), volume methionine free formula – check total protein intake ~ 3g protein /kg/d Weaning stage 2 • expand range of low protein free foods, include prescribable low protein foods • discuss introduction of methionine containing foods (teach methionine / 1g protein exchanges) to gradually replace breast / normal infant formula feeds • gradually introduce second stage methionine free L- amino acid powder (as paste or add to methionine free infant formula). |
| 8 Months | Wt, length (monthly or more as necessary) Weaning stage 3 • expand range of low protein free foods include prescribable low protein foods • continue introduction of methionine containing foods (exchanges) • introduce more texture/finger foods into diet (as per usual weaning practices). • introduce more family low protein meal choices • increase second stage methionine free L- amino acid powder/decrease methionine free formula |
| 12 Months | Wt, length (monthly or more as necessary) Weaning stage 4 • family foods, encourage more variety, • Give advice re: toddler feeding |