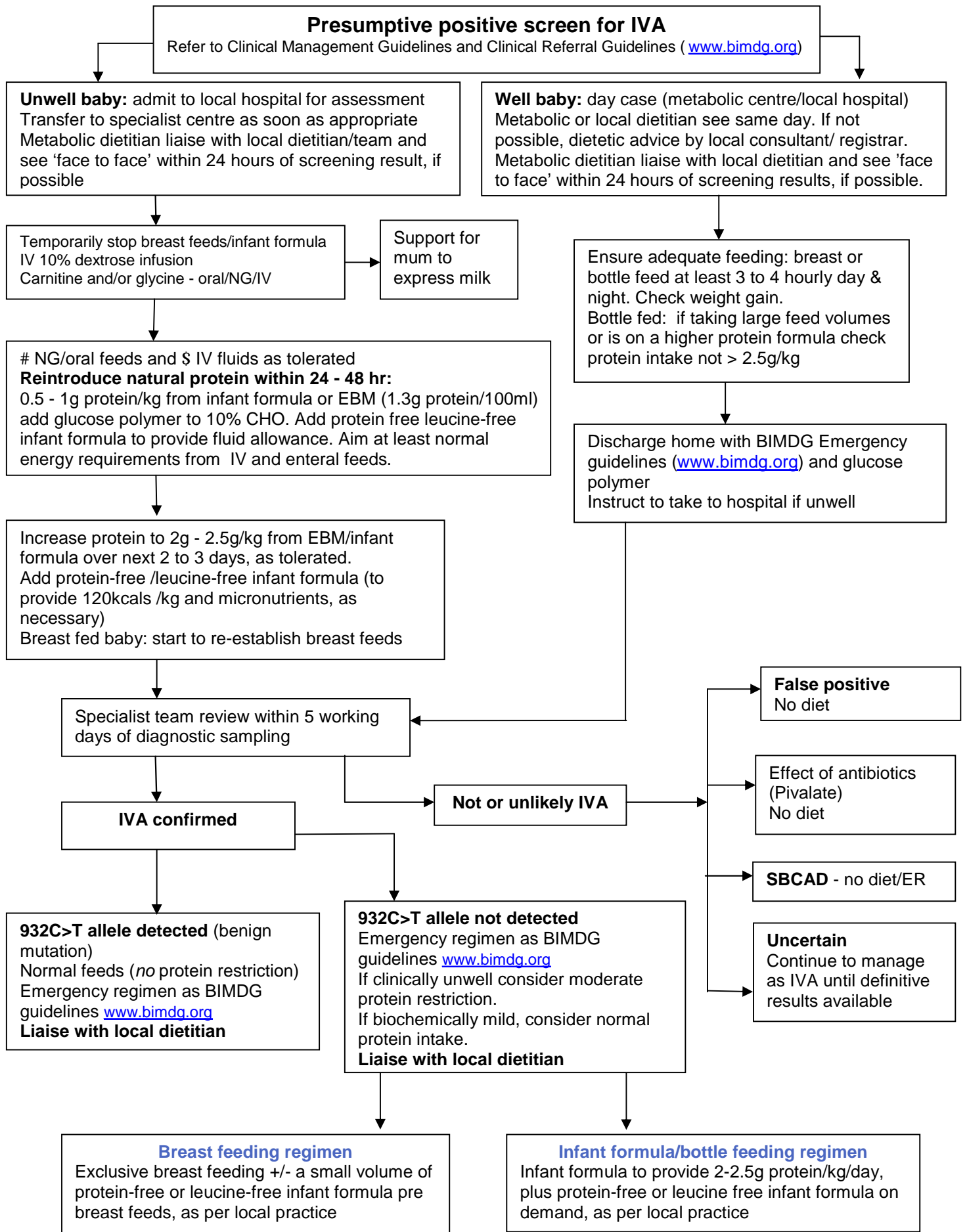


Isovaleric Acidaemia (IVA) Dietetic Management Pathway



Isovaleric acidaemia - Dietetic management

Dietetic products used in IVA

Protein free infant formula – Energivit, Nutricia www.nutricia.co.uk

15% dilution (eg: 90ml cooled boiled water, add 3 level unpacked scoops)
(use scoop in tin = 5g powder)

| | Per 100g | Per 100ml |
|---------------|-----------|-----------|
| Energy (Kcal) | 492 | 74 |
| Protein (g) | nil added | nil added |
| CHO (g) | 66.7 | 10 |
| Fat (g) | 25 | 3.75 |

Leucine Free infant formula – IVA Anamix Infant, Nutricia www.nutricia.co.uk

15% dilution (90ml cooled boiled water, add 3 level unpacked scoops)
(use scoop in tin = 5g powder)

| | Per 100g | Per 100ml |
|------------------------|----------|-----------|
| Energy (Kcal) | 457 | 69 |
| Protein equivalent (g) | 13.1 | 2.0 |
| CHO (g) | 49.5 | 7.4 |
| Fat (g) | 23.0 | 3.5 |

Emergency regimen products

Glucose polymer powder eg: Maxijul, SOS 10 sachets

Dietary management information (as per local centres own resource)

Emergency regimen information

- metabolic team and local hospital contact details
- when to use, preparation

Dietary leaflets

- feed regimen
- low protein 'free' foods (fruits, veg and others),
- low protein prescribable foods
- calculating protein exchanges
- 1g protein exchange list

Prescription letters to GP

- protein free formula
- low protein manufactured foods
- glucose polymer

Home delivery services