





British Inherited Metabolic Disease Group

## RECOMMENDED MANAGEMENT

### CLINICAL ASSESSMENT

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- r
- r
- r
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### INITIAL INVESTIGATIONS

• **Blood Tests:**

- 
- 
- r r r l

• **Urine Tests:**

- L
- r r r r
- r r

• **Other tests as indicated by clinical findings**

### TREATMENT

- **Analgesia**
- r r

l r

- **Maintain fluid and calorie intake**

r r r

- **Treat symptoms as indicated**

r r r r r



- **Consider treatment with Haem arginate (Normosang®)**

- **Indications**
- **Contraindications**
- **Warnings**
- **Promptly**
- **Adverse effects**

**Haem arginate carries a risk of significant extravasation injury with thrombophlebitis at the site of administration being a common issue.**

- **Indications**
- **Contraindications**
- **Warnings**
- **Adverse effects**

**stop the infusion immediately**