

# Saturday Newborn Screening

ER = Emergency Regimen

CMG = Clinical Management Guideline

On call metabolic consultant is accessed via Sheffield Children's  
Hospital Switchboard: 0114 271 7000

The information provided here is intended for use by the Sheffield Regional Newborn Screening Network and should be used in conjunction with telephone advice from the Sheffield Metabolic Consultant.

# From 1 April 2017

- Sheffield newborn screening lab will report positive cases with elevated:
  - C8 – MCADD
  - C5 – IVA
  - Leucine – MSUD
- The report will be to the on-call metabolic consultant for Sheffield
- The newborn screening clinical scientist will identify the appropriate community midwife and ask them to visit the family immediately with appropriate literature (signpost BIMDG website)

# Midwife

- To ascertain if baby is already in hospital – if so obtain contact details and pass to on-call metabolic consultant
- Will liaise with the on-call metabolic consultant prior to the visit
- Will access appropriate ‘suspected’ leaflet from the [www.gov.uk](http://www.gov.uk) website (can be accessed with the BIMDG / NBS Guides page)
- Visit the family ASAP
- For MSUD, if not able to visit within two hours, to phone family and tell them to take the baby to hospital immediately. Inform on call consultant

# If not able to contact family

- Midwife to leave note asking family to phone regarding the newborn screening test result asap; Ascertain if the baby is already in hospital
- **For MCADD**
  - Phone at two and four hours later
  - To attempt visit again following day
- **For MSUD**
  - Contact metabolic consultant on call
    - On-call consultant to discuss with SCH duty director regarding alerting police via 999
    - If police contacted, should ensure baby is taken to nearest hospital with 24 hour paediatric cover. Inform metabolic consultant on call if baby sent to hospital
    - On call consult to discuss with paediatric team at hospital
- **For IVA**
  - Discuss with metabolic consultant on call
    - If likely mild, suggest as for MCADD
    - If likely severe, as for MSUD

# MSUD

- Midwife to give quick explanation to parents and MSUD is suspected literature
- Quick assessment of baby's condition, feeding history
- Ask family to take baby to hospital (nearest / where baby was born, all providing 24 paediatric cover provided).
- If baby unwell, to call ambulance via 999
- Inform on-call metabolic consultant
- Metabolic consultant to contact on-call paediatrician at local hospital, give details of baby and MSUD clinical management guideline links (BIMDG NBS page)
- Metabolic consultant to establish availability of PICU bed at SCH. If none available establish availability at QMC or LRI, then BCH, Manchester Children's or GOS and which hospital to transfer to

# MSUD at local hospital

- Follow CMG for MSUD
  - Samples as listed in 4b and plasma amino acids and urine organic acids
  - SCH NBS clinical scientist to liaise with local hospital lab about immediate sample transfer to SCH via taxi of diagnostic samples
  - Start treatment as in 4d
  - Liaise with on-call metabolic consultant to arrange transfer once stable to specialist centre with PICU availability
  - Local hospital to arrange transfer to specialist centre with input from metabolic on-call consultant (specialist centre will be Manchester Children's Hospital if Manchester consultant on call)
  - On-call metabolic consultant to ensure any diagnostic test results from SCH to be transmitted to accepting specialist centre if not at SCH
    - Use BIMDG newborn screening MSUD dietary guidelines
    - Availability of product eg MSUD anamix, valine, isoleucine (both strengths) at SCH, QMC, LRI

# IVA

- Midwife to give quick explanation to parents and IVA is suspected literature
- Quick assessment of baby's condition, feeding history
- Ask family to take baby to hospital (nearest / where baby was born, all providing 24 paediatric cover provided).
- If baby unwell, to call ambulance via 999
- Inform on-call metabolic consultant
- Metabolic consultant to contact on-call paediatrician at local hospital, give details of baby and IVA clinical management guideline links (BIMDG NBS page)

# At local hospital

- Paediatrician to follow IVA CMG
  - If unwell:
    - to inform metabolic consultant immediately. Metabolic consultant to establish availability of PICU bed at SCH, QMC, LRI, MCH, BCH, GOS
    - Metabolic consultant to advise on supply of carnitine, energyvits
    - Follow CMG 4bii
    - Ensure diagnostic samples taken and SCH NBS clinical scientist to discuss transport of samples to SCH if appropriate. Sample to go with baby to SCH (or by courier first thing on Monday morning if baby not going to SCH)
  - Diagnostic samples are: blood spots on Guthrie Card (genetics, acylcarnitines), LiHep plasma (acylcarnitines), plain urine (organic acids)
    - Local hospital paediatrician to facilitate transport to specialist centre once stable. If Manchester consultant on call, to advise if baby to go Manchester
  - If well:
    - Ensure appropriate feeding (**network dietitians guidance**) and normal obs / examination
    - Follow CMG 4biC – metabolic consultant to assist with signposting BIMDG IVA ER. Ensure appropriate history of antibiotics use obtained. To agree with local paediatrician if safe for baby to be managed at home
    - Parents advised if any concerns attend ED with BIMDG IVA ER
    - SCH NBS nurses to arrange for family to see specialist on Monday



# MCADD

- Midwife to:
  - give quick explanation to parents and MCADD is suspected literature
  - Supply family with 'suspected' leaflet
  - Ask family to take baby to local hospital ED and to tell parents that a paediatrician will assess if their baby is well or needs any treatment
  - If baby unwell, to call ambulance via 999
  - Discuss with on-call metabolic consultant
  - Metabolic consultant to contact paediatrician at local hospital to refer and give advice, signpost BIMDG MCADD CMG
  - Otherwise continue to feed normally (**network dietitian guidance**)

# MCADD at local hospital

- Paediatrician to assess if baby is well, has normal exam and obs and feeding satisfactorily (**network dietitian guidance**).
- Follow MCAD CMG step 4 (covers well / unwell)
  - If well, no need for diagnostic samples (will be taken on Monday)
  - If unwell, take diagnostic samples: blood spots on Guthrie Card (genetics, acylcarnitines), LiHep plasma (acylcarnitines), plain urine (organic acids) and follow BIMDG ER
- Inform metabolic on-call consultant of outcome and obtain advice
- Ensure is well at discharge and has BIMDG ER
- Advise continue to feed normally (**network dietitian guidance**)
- Advise if any concerns, go to ED with BIMDG ER
- SCH NBS nurses to arrange specialist contact on Monday (as if usual first contact)

# At SCH

- Babies sent in by midwife
  - Managed by general paediatric on-call team as for local hospital
  - On-call metabolic consultant to support with telephone advice
- Unwell babies / those transferred in
  - Managed by general paediatric on-call team as any sick baby on-call, including transfer to PICU
  - If not requiring intensive care, may be managed by general paediatrics on NSU
  - On-call metabolic consultant to support with telephone advice