## OG003.v1 Hypoplastic Left Heart Syndrome

<table>
<thead>
<tr>
<th></th>
<th>Antenatal Diagnosis</th>
<th>Postnatal Diagnosis</th>
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</table>
| 1 | • All reasonable efforts should be made for an in-utero transfer to London if surgical palliation is requested.  
  • If in utero transfer cannot occur, the neonatal and paediatric ScotSTAR teams must be made aware of the expected due date. | • In the event of a postnatal diagnosis of HLHS, transport to London or Birmingham will be required if surgical palliation is requested. |
| 2 |                                                                                      |                                                                                      |
| 3 | Transport                                                                            |                                                                                      |
|   | • The retrieval of a neonate with HLHS may necessitate the formation of a second paediatric team.  
  • If the retrieval is performed by the Neonatal ScotSTAR team, a second team would not be required.  
  • The transport team identified should have experience in the care or retrieval of patients with HLHS  
  • The Specialist Services Desk of Ambulance Control should be contacted early to enable allocation of air assets. |                                                                                      |
| 4 | Financial & National Service Designation (NSD) Issues for Transport                   |                                                                                      |
|   | • Funding for ongoing treatment outside Scotland needs to be sanctioned by National Services Division (NSD) and sought by the referring paediatric cardiologist.  
  • The cost of the transfer to London or Birmingham and repatriation to Glasgow is paid for by NHS Greater Glasgow & Clyde. |                                                                                      |
# OG003.v1 Hypoplastic Left Heart Syndrome

## 1. Version History

<table>
<thead>
<tr>
<th>Reference Number</th>
<th>CG003</th>
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<tbody>
<tr>
<td>Version</td>
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### Writing group (Chair in bold)

<table>
<thead>
<tr>
<th>Name</th>
<th>Position</th>
<th>Institution</th>
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<tbody>
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<td>Intensivist</td>
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### Associate Medical Director

Andrew Inglis

### Date issued

17th April 2018

## 2. Distribution

### ScotSTAR

<table>
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<th>Services</th>
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<tr>
<td>EMRS</td>
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<td>✓</td>
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<tr>
<td>Neonatal</td>
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### Referring centres via service websites

- ✓

### BASICS Scotland

- X

### Medic 1

- X

### Tayside Trauma Team

- X

### Grampian (GHEMS)

- X

### Rural GPs Association of Scotland

- X

### SAS Air Ambulance Division

for information

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[Logo] ScotSTAR

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3. Scope and purpose

• Overall objectives:
The hypoplastic left heart syndrome (HLHS) program at RHC Glasgow has been paused for further review. During this time, alternative arrangements need to be made to ensure that a child with a diagnosis of HLHS receives appropriate treatment in a centre that is able to offer operative intervention (Birmingham and London) if parents wish to pursue surgical palliation for their child. Ideally, children with an antenatal diagnosis of HLHS are transferred in utero and delivered in or close to an appropriate centre. Alternatively, infants who deliver in Scotland with HLHS need to have a safe and efficient pathway identified for timely transfer.

• Statement of intent:
This guideline is not intended to be construed or to serve as a standard of care. Adherence to guideline recommendations will not ensure a successful outcome in every case, nor should they be construed as including all proper methods of care or excluding other acceptable methods of care aimed at the same results. The ultimate judgement must be made by the appropriate healthcare professional(s) responsible for clinical decisions regarding a particular clinical procedure or treatment plan.

• Feedback:
Comments on this guideline can be sent to: scotamb.CPG@nhs.net

• Equality Impact Assessment:
Applied to the ScotSTAR Clinical Standards group processes.
4.1 Antenatal Diagnosis

• All reasonable efforts should be made for an in utero transfer to London if surgical palliation is requested.

The first operative stage for HLHS is usually a Norwood 1 operation. An antenatal referral is made to the Evelina London Children’s Hospital in the second trimester. The case will be discussed at their multidisciplinary meeting and, if appropriate, accepted for postnatal surgery. A review in London will be undertaken at approximately thirty weeks, which will include foetal cardiology, an obstetric review and foetal cardiac MRI. A date for delivery will be made and the family will return home, only returning to London at the time of delivery. Transport and accommodation will be arranged by foetal medicine and the local health board.

• If in utero transfer cannot occur, the neonatal and paediatric ScotSTAR teams must be made aware of the expected due date.

If in utero transfer is unacceptable to parents or cannot occur, the parents need to be counselled that the infant will require a timely transfer once born to an appropriate centre if they wish for surgical palliation. Delivery will be arranged towards term at the QEUH. The neonatal and paediatric ScotSTAR teams must be made aware of the expected due date of the infant. If delivery of a baby with an antenatal diagnosis of HLHS occurs prior to in utero transfer, the paediatric cardiologist will discuss suitability of transfer with the London team. If clinically appropriate to transfer the infant, the ScotSTAR neonatal and paediatric teams must be notified so that provisions for an appropriate retrieval can be made.

4.2 Postnatal Diagnosis

• In the event of a post-natal diagnosis of HLHS, transport to London or Birmingham will be required if surgical palliation is requested.

If a postnatal diagnosis of HLHS is made, and parents request surgical palliation, neonatal and paediatric ScotSTAR teams need to be notified immediately so a retrieval plan can be generated. This should at the same time as referral to the surgical centre. In the interim, the infant should be transferred to the Paediatric Intensive Care Unit and the agreed HLHS pathway followed.
### 4.3 Transport

- The retrieval of a neonate with HLHS may necessitate the formation of a second paediatric team.
- If the retrieval is performed by the Neonatal ScotSTAR team, a second team would not be required.

Transfer of an infant with HLHS should not compromise the capability of ScotSTAR retrievals for paediatric and neonatal patients within Scotland. The decision on whether to allocate a paediatric or neonatal retrieval team will depend on workload, availability of additional retrieval staff to transport the infant and skill mix of staff available for transport. If an infant with HLHS requires transfer to Birmingham or London, the paediatric or neonatal ScotSTAR team could undertake the transfer. If the paediatric team were involved, they may need to formulate a second team so that the paediatric retrieval capacity in Scotland is not compromised. If the neonatal team (West) in ScotSTAR were to undertake the transfer, the north or south neonatal team could cover any emergency work for the duration.

- The transport team identified should have experience in the care or retrieval of patients with HLHS.
- Transport staff involved in the retrieval should have experience in managing infants with HLHS. A clear and concise plan should be agreed between the treating team (NICU/PICU and cardiology after appropriate discussion with the receiving unit) and the transport team. This should include contingencies such as appropriate escalation of the deteriorating infant during transfer and appropriate pharmacological intervention.

- The Specialist Services Desk of Ambulance Control should be contacted early to enable allocation of air assets
- Availability of air resources is important and it is imperative that SSD is contacted early for planning purposes. These patients are emergency transfers for immediate operative intervention that cannot be provided in Scotland so they should be a priority before SAS air ambulance routine workload. Delaying transfer may compromise the infant and the subsequent transfer would carry more risk. Conflicting priorities regarding air transfer should involve a conference call with the on-call air wing manager, the on-call ScotSTAR manager, the clinical team and SSD.

### 4.4 Financial & National Service Designation (NSD) Issues for Transport

- Funding for ongoing treatment outside Scotland needs to be sanctioned by National Services Division (NSD)

Funding for ongoing treatment needs to be sanctioned by NSD. The local Safehaven team should be contacted by the referring paediatric cardiologist and completed forms sent to Jean Travers: jean.travers@nhs.net to generate an authorisation code. The ScotSTAR team should ensure that this has been completed.

- The cost of the transfer to London or Birmingham and repatriation to Glasgow is paid for by NHS Greater Glasgow & Clyde.

SSD should send the details of the cost for the entire journey to Jamie Redfern: Jamie.Redfern@ggc.scot.nhs.uk for approval.
5. Appendix

Checklist for transfer of infants with HLHS to London or Birmingham

1. Initial referral for transfer of infant with HLHS by ScotSTAR
   Inform ScotSTAR coordinator of referral, who shall decide the following in collaboration with paediatric & neonatal ScotSTAR duty teams:
   • Date of transfer _________
   • Patient accepted in Birmingham Children’s Hospital or Evelina’s London Children’s Hospital
   • ScotSTAR team to complete transfer – paediatrics or neonates
     • If paediatrics – require 2nd transport team
   • NSD sanctioned treatment (paediatric cardiologist in Royal Hospital for Children in Glasgow will do this but must check this has been done)
   • Book air asset for morning of proposed date of transfer

2. Funding
   • ScotSTAR coordinator to request SSD to send transfer costs to Jamie Redfern at Jamie.Redfern@ggc.scot.nhs.uk (NHS Greater & Glasgow & Clyde) for both journey down and repatriation to Glasgow.
   • Ensure that SSD have received the completed form from Jamie Redfern prior to transfer.

3. Prior to transfer
   • On the day prior to the transfer, the retrieval team should get a handover from Royal Hospital for Children, Glasgow. This should include an agreed plan of management if there is deterioration during transfer and bed availability of receiving centre.
   • If bed status at the receiving unit cannot be confirmed until the day of the transfer, the retrieval team should proceed with preparation of transfer to reduce delay if the retrieval goes ahead.