



Early management and preparation for transport of infants with hypoplastic left heart syndrome born outside Leeds Children's Hospital.

“Hypoplastic left heart syndrome management”

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Approved: June 2022
Review Due: June 2025

Purpose

To guide the early management of transferring infants with hypoplastic left heart syndrome.

Intended Audience

Embrace team call handlers, nurses, ANPs, doctors and regional paediatric teams.

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ABBREVIATIONS

HLHS	Hypoplastic left heart syndrome
LCH	Leeds Children's Hospital
TPN	Total parenteral nutrition
NEC	Necrotizing enterocolitis

1. OBJECTIVES

These guidelines are designed to establish a safe streamlined approach to the early management, referral and transport of infants with hypoplastic left heart with the aim of ensuring successful transfer to Leeds Children's Hospital for definitive surgical care.

2. BACKGROUND INFORMATION

Infants with hypoplastic left heart syndrome (HLHS) are amongst the most challenging subgroup of cardiac patients to care for, with the period of acute risk lasting from birth until the start of stage II palliation¹. Because these infants often respond poorly to minor external stress or subtle changes in their management the process of transport is a particular risk. It must be planned carefully with clear communication between consultants in the referring, receiving and transport teams. Most babies born with HLHS in Yorkshire and the Humber will be both born and cared for at Leeds Children's Hospital (LCH), however, occasionally some babies will need to be transferred from LCH to Evelina or Birmingham Children's Hospital.

3. GUIDELINE

3.1 Pre-natal management

- When the diagnosis of HLHS is made antenatally, delivery is planned at Leeds Children's Hospital. Surgery is usually planned for day 3 or 4 of life but may be required earlier if circulatory instability particularly when this is related to excessive pulmonary blood flow.

3.2 Immediate management for an expected HLHS delivery outside of LCH

- Ensure both Embrace and local Consultant aware of impending delivery
- Delivery room management to be coordinated by local Consultant or senior Specialist Trainee
- Target oxygen saturations 75-80%, avoid additional oxygen unless saturations less than 70%
- Intravenous access to be obtained on arrival to the NICU
- Double lumen umbilical venous catheter (UVC) is the preferred route of venous access. This should be placed by the most experienced person available
- Siting two peripheral venous lines is an alternative if UVC access not successful
- Obtain blood for baseline venous gas, lactate, full blood count, electrolytes, glucose, coagulation, group and save
- Dinoprostone 10 nanograms/kg/min should be commenced immediately after the first route of intravenous access is obtained as per the regional guideline https://www.networks.nhs.uk/nhs-networks/yorkshire-and-humber-congenital-cardiac-network/guidelines-and-protocols/paediatrics/dinoprostone-for-maintenance-of-duct-patency-in-chd-guideline/file_popview
- Ventilation should not be routine in the stable, non-acidotic patient
- Contact paediatric cardiology via Embrace
- Request a maternal blood sample early to avoid delays later on

3.3 Continued care at referring centre prior to transfer to Leeds Children's Hospital (LCH)

- All infants initially admitted to NICU
- Continue to target saturations of 75-80%, avoid additional oxygen unless saturations less than 70%
- Check blood gases, glucose and lactate
- If capillary gases are abnormal then check a venous (via UVC) or arterial sample
- No enteral feeds in first 24 hours because of risk of gut ischaemia and NEC
- Insertion of peripheral lines and other procedures can be a significant stress leading to fluctuations in systemic vascular resistance secondary to agitation and pain
- Senior clinicians should obtain venous access and multiple attempts should be avoided
- Use of sucrose is recommended for peripheral lines and long line insertion
- Correctly placed umbilical lines should be left in place for transport
- **Management of low systemic cardiac output:** Four distinct causes should be sought and systematically excluded. Senior Paediatric Cardiologist input is essential to guide management.
 - Excessive pulmonary blood flow (high saturations) - strongly consider ventilation prior to transport, ensure adequate filling, consider inodilator (Milrinone)
 - Impaired systemic vascular function +/- significant tricuspid regurgitation (normal or low saturations) - consider Dobutamine and or Milrinone
 - Restrictive ASD (usually low saturations) - needs emergency intervention.
 - Restrictive duct (any saturations) - increase Dinoprostone
 - Consider sepsis and other causes of low cardiac output
- **Indications for intubation** (liaise with cardiology/Embrace)
 - Apnoeas
 - Poor systemic circulation with rising lactate
 - Pulmonary over circulation (saturations >90% with systemic hypoperfusion (lactic acidosis)
- Intubation can cause considerable instability to infants with a duct-dependent systemic circulation
- Ideally intubation should be performed by a senior paediatrician
- 20ml/kg fluid should be available and consider low dose Dobutamine infusion (5micrograms/kg/min) prior to induction
- Insert an arterial line (umbilical or peripheral) in all ventilated patients
- Target blood gases:
 - pH – normal
 - PaO₂ 4.5 – 6.0
 - PaCO₂ 4.5 – 6.0

3.4 Making the referral

- Consultant to consultant conference call via Embrace to include cardiology, neonatology, Embrace and receiving unit PIC consultants
- **THINK**
 - Is intubation required for transfer?
 - Is there adequate venous access for transfer?
 - Are further vasoactive drugs required for transfer?

3.5 Management for transfer

- Check blood gas, glucose and lactate
- Clinical re-assessment
- Inform Embrace and receiving team of any significant change
- Check photocopy of notes and drug card, Badger summary, cardiology referral letter and PACS transfer of radiology images
- Maternal blood sample hand labelled with NHS number
- Continue clear maintenance fluids and prepare some additional fluids in labelled 50ml syringes
- Consultant delivered transfer by Embrace

4. MONITORING ARRANGEMENTS

Audit and critical incident review

Joint case reviews with Leeds Teaching Hospitals Trust

5. REFERENCES

¹Early management of infants with hypoplastic left heart syndrome 2008 L Shekerdeman. <https://www.rch.org.au/uploadedFiles/Main/Content/picu/HLHS.pdf>

²The intensive care of infants with hypoplastic left heart syndrome. Arch Dis Child Fetal Neonatal Ed 2005;90:F97-F102

6. VERSION CONTROL STATEMENT

Version	Date	Author	Status	Comment
1	1/7/2013	C. Harness	Original	
2	28/11/2016	C Harrison	Amended	Approved by Embrace Guideline Group
3	03/07/2017	C Harrison	Amended	Approved by Embrace Guideline Group
4	15/09/2019	J Oldfield	Amended	
5	26/05/21	S Hancock	Amended	Approved by Embrace Guideline Group

7. APPENDICES

None