

Appendix 1

Case Ascertainment Process

To identify the HLHS patients for whom we had HSCIC life status tracking within NCHDA we deployed the following process for categorising records and patients:

Step A: inclusion of possible HLHS cases

We initially included for consideration all patients from England and Wales born after 1 January 2000 where either:

- The patient record contained at least one of the following cardiac diagnosis codes relating to HLHS or hypoplastic left ventricle (HLV):
 - HLHS (diagnosis code: 01.01.09);
 - aortic atresia (09.15.03);
 - mitral atresia (06.02.01)21;
 - ventricular imbalance: dominant right ventricle and HLV (07.08.42); or
 - left ventricular hypoplasia (07.07.00)
- The patient record included procedure codes consistent with a Stage 1 (Norwood type) procedure

Step B, exclusion of patients with other diagnoses

In line with the International Society for Nomenclature of Paediatric and Congenital Heart Disease (ISNPCHD) definition of HLHS¹, we then removed patients with diagnostic codes indicative of hypoplastic left heart related malformations¹ such as atrioventricular septal defect, double inlet ventricle and double outlet right ventricle rather than HLHS (see Table A1 of the appendix for the list of such codes).

Step C, exclusion of patients with procedures inconsistent with a diagnosis of HLHS

Procedure records were reviewed by two surgeons (MM and VTT) and two other clinicians (KB and RF) with considerable experience of the IPCCC system and patients were excluded if

- any procedure code was inconsistent with a diagnosis of HLHS (for instance various types of biventricular repair);
- there was no procedure in the first 3 months of life consistent with a HLHS treatment pathway essential to survival for HLHS patients;
- the sequence of procedures was inconsistent or implausible (for instance a patient having a Stage 2 procedure without having first a Stage 1 procedure);

Step D, exclusion of patients with anomalous data

If the dates of birth implied by different procedure records in the dataset were inconsistent, the patient was excluded from the analysis.

We reiterate that, since the NCHDA is a procedure-based dataset, patients with HLHS who received only comfort care following birth and did not undergo any interventional treatment are not included.

Ascertainment of survival

Procedure records were linked using the pseudonymised NHS number (a unique identifier for patients in England and Wales) and/or pseudonymised hospital ID number to give patient specific histories of intervention and survival.

The survival of patients in NCHDA is inferred from recorded life status at discharge from the tertiary cardiac centre following each procedure and, for residents of England and Wales,

from death certification data requested at least every six months from the UK Health and Social Care Information Centre (HSCIC) using each patient's NHS number, their name and other identifiers.

Each patient history contained information on age of operation and age and life status at discharge for all procedures for that patient, augmented for residents of England and Wales by life-status tracking up to 2015. These were checked for consistency, with a submitted status of dead at discharge trusted over a later HSCIC status of alive because of known delays in the updating of HSCIC life status for deaths referred to a coroner. For a small number of patients, patient history indicated that the patient was lost to follow up (e.g. moved country) with no recorded procedures for a period infeasible in the context of HLHS, then their survival was censored at the age of their most recent discharge from hospital.