

**An Analysis of Results for the Norwood
Operation at Evelina London Children's Hospital
(ELCH)
2012-2015**

Introduction

This report addresses the concerns raised by the 2012-2015 National Congenital Heart Disease Audit (NCHDA), which suggests a higher than expected 30 day mortality for patients undergoing the primary Norwood procedure at the Evelina London Children's Hospital (ELCH). During this time frame a Norwood was undertaken as a primary procedure in 23 patients, 16 remained alive at 30 days with 7 dead resulting in a 30 day survival of 70%.

These concerns were first raised in the 2011-2014 data analysis. For that time period 47 patients had a primary Norwood procedure with a 30 day survival of 74%. A detailed report was produced to address this and an action plan devised and is available on the NICOR NCHDA website.

Given that this is three year rolling data it was anticipated that the outlier status would persist until previously made changes have an opportunity to have a clinical impact. In the 2014-2015 calendar year only three neonatal Norwood procedures were performed with 100% 30 day survival. Although positive this is an insufficient number to have an impact on figures over 3 years.

This report follows on from the previous report and will:

- Summarise the status of treatment of hypoplastic left heart syndrome at ELCH
- Update on progress from the action points outlined in the previous report
- Describe new and ongoing actions

The report represents the collective opinion of the congenital cardiac unit at the Evelina, comprising paediatric cardiology, cardiac surgery, anaesthesia and intensive care

Treatment of Hypoplastic Left Heart Syndrome at ELCH

As previously described our treatment pathway for patients with hypoplastic left heart syndrome has 2 main arms. There are those who undergo palliation with a classical Norwood Procedure (Aortic arch reconstruction, Damus connection and modified BT shunt) or a Hybrid procedure (Arterial duct stent and bilateral pulmonary artery bands). Additionally patients undergoing the Hybrid procedure may undergo supplementary procedures as clinically relevant, for example; Balloon atrial septostomy, open septectomy, PA band adjustment or stent dilation.

Indications for inclusion into the Hybrid program include: low birth weight (<2.75kg), post natal collapse, unfavourable vascular anatomy for a Norwood procedure, potential for biventricular repair.

Patients that undergo a Hybrid procedure will then proceed down a pathway with a further 2 or 3 stages depending on underlying anatomical factors and clinical condition (underlying ventricular function, degree of tricuspid regurgitation, vascular anatomy, additional comorbidities). The 2 Stage pathway following Hybrid consists of a Norwood procedure combined with a superior cavo-pulmonary connection, followed by a total cavo-pulmonary connection later in childhood. The three stage pathway following Hybrid consists of a late Norwood procedure, followed by a superior cavo-pulmonary connection, then the total cavopulmonary connection later in childhood.

Activity

As detailed in the previous report there has been a marked change in our total single ventricle activity and Norwood procedures in the last decade. This is illustrated in figure 1.

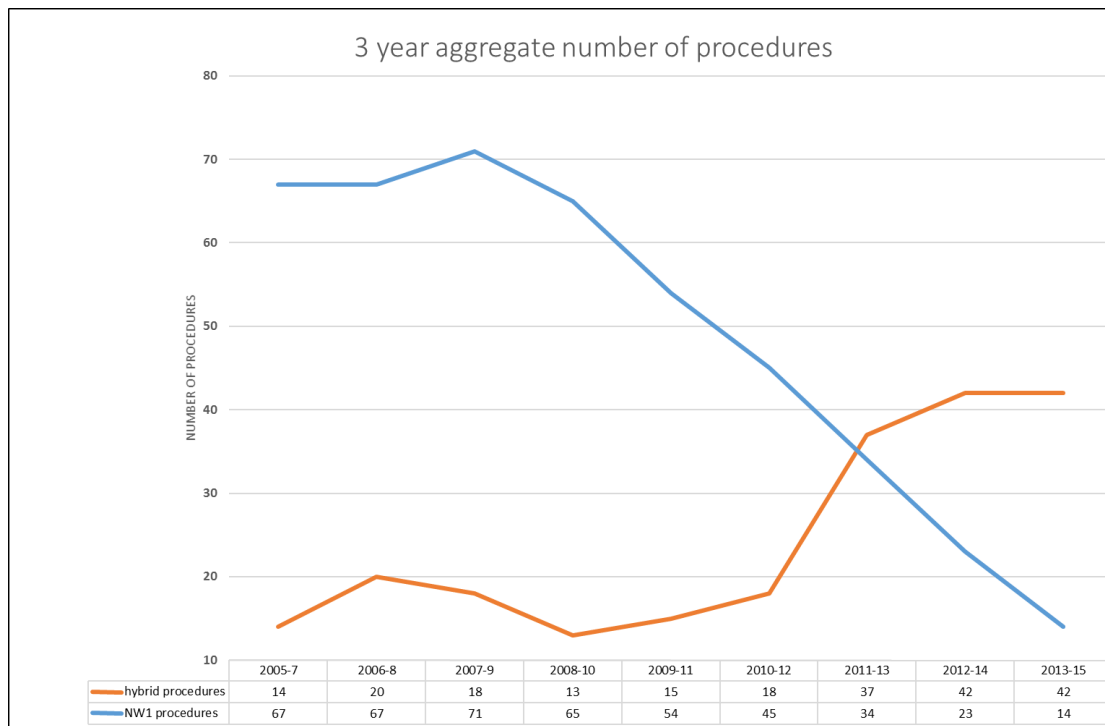


Figure 1: Activity at ELCH for both the Hybrid (orange) and Primary Norwood (blue) in each 3 year cohort as per NICOR funnel plots

This figure illustrates two reasons for the change in activity.

1. A reduction in total number of patients, felt to be due to a fall in external referrals with more centres taking on their own management of hypoplastic left heart syndrome.
2. The increasing uses of the hybrid as a primary palliation for HLHS instead of the Norwood

Consequently the number of primary Norwood procedures within this cohort (23) is significantly less when compared to the previous 2011-2014 cohort (47).

However it should be noted that because of the Hybrid procedure and the subsequent stages there are a group of patients undergoing a classical Norwood that do not feature in this data because they have had a prior procedure. In the time 2012-2015 time frame there were an additional 10 patients who underwent a Stage 1 Norwood Procedure following the Hybrid Of these 10 patients, 9 remained alive at 30 days resulting in a 30 day survival of 90%, with cumulative survival for all Stage 1 Norwood procedures (without censoring) of 76%. This does not include those patients who following the Hybrid procedure then went on to have comprehensive Norwood combined with superior-cavo-pulmonary connection who have been coded separately.

Patients that entered the Norwood and Hybrid program are illustrated in table 1.

Table 1: Patients entering Norwood or Hybrid Program

Year	Norwood Procedure		Hybrid Procedure				
	Cases	Deaths (%)	Cases	Death Post Hybrid* (%)	Delayed Norwood (30d death)	Combined Norwood§ (30d death)	Other procedure¶ (30d death)
2012-13	14	5 (35%)	6	1 (16%)	4 (1)	1 (0)	-
2013-14	6	2 (33%)	19	3 (15%)	3 (0)	8 (2)	5 (0)
2014-15	3	0 (0%)	16	4 (25%)	5 (0)	4 (1)	3 (0)
Totals	23	7 (30%)	41	8 (20%)	12 (1)‡	13 (3)	8 (0)

* Death post hybrid prior to any further intervention

§ Norwood procedure combined with superior cavo-pulmonary connection (±pulmonary artery repair)

¶ Other procedure includes full intra-cardiac repair or further single ventricle palliation not involving Norwood

‡ Two of the Norwood procedures performed post hybrid were in the 2015-2016 year

Surgeon Specific Data

The primary Norwood procedure is performed by all three of our surgeons. The majority in this cohort by the two more senior surgeons. Statistical analysis of the individual operator 30 day mortality has shown no significant difference in mortality rates between surgeons

Update on 2014 Internal Review

As mentioned in the previous response we had conducted a comprehensive internal review of our hypoplastic left heart programme starting in January of 2014, prior to the notification of the previous potential outlier status. This had been undertaken in response to our own concerns about the management and results of this patient group.

This was a multi-disciplinary review conducted by paediatric cardiology, cardiac surgery, intensive care and paediatric cardiac anaesthesia. It examined all elements of the patient pathway. These were:

- Case selection
- Pre-operative care
- Anaesthesia (at the time of Norwood or Hybrid surgery)
- Post operative management
- Inter-stage monitoring

Guideline Development

As a consequence of this review we developed guidelines on:

- Hypoplastic left heart case selection
- Pre-operative management
- Post operative management

These were put into place in December 2014 and formalised through Trust governance processes in 2015.

Additionally there were specific anaesthetic guidelines developed

- Anaesthetic management for the Norwood procedure
- Anaesthetic management for the Hybrid procedure

Process Management

The following processes were put into place

1. Dedicated MDT

All patients who have been referred (internally or externally) for consideration of inclusion onto the Norwood or Hybrid program are discussed at a dedicated, patient specific MDT. This is attended by key representatives ('a core group') from cardiology, cardiac surgery and intensive care. However all clinicians within the department are welcome to attend. In all cases a quorum was achieved

2. Scheduling

Agreement was reached to enable Hybrid or Norwood cases to be the first case of the day, even if this involves late re-arranging of the cardiac surgical operating list.

3. Core anaesthetic team

A small group of paediatric cardiac anaesthetists have been identified who have a specific interest in the anaesthesia of patients with hypoplastic left heart syndrome. The purpose of this group is to provide consistency and advice to the wider paediatric cardiac anaesthetic team. This group were responsible for producing the anaesthetic guidelines.

4. Mandatory intra-operative epicardial echo

A system of mandatory intra-operative imaging has been introduced, with a dedicated group of specialist imaging cardiologists on an independent rota providing support. For the Norwood patients this means an epicardial echocardiogram is performed at the end of the procedure. In addition to this from September 2015 we have introduced the Technical Performance Score (TPS) based on studies at Boston Children's Hospital.

5. Facility for exit angiography.

If concerns are raised about possible structural abnormalities on completion of surgery then emergency exit angiography would be considered. This is facilitated by having the case first on the operating list. This has not yet been required.

6. Structured PICU handover

A system of structured handover from the theatre to the PICU team has been fully embedded. This handover process undergoes regular audit and refinement.

7. ECMO

Agreement was obtained amongst the team that ECMO would be considered in the post Norwood patient, with the view to undertaking cardiac catheterisation and surgical revision of residual lesions if indicated.

Inter-stage monitoring

With regard to interstage monitoring the Evelina London Home Monitoring programme was launched in June 2014 and has been active since the last 18 months. This is an intensive programme where following 1:1 education sessions from cardiac specialist nurses, parents and/or carers are provided with training and equipment for daily heart rate and oxygen saturation measurement, weight and feed volumes. Acceptable criteria are determined and any breach merits urgent clinical review. This is combined with fortnightly visits to a dedicated home monitoring clinic and interval weekly telephone calls. There is also a dedicated telephone line for queries and engagement with local paediatric teams.

These guidelines and actions were all put into place during the course of 2014 to 2015, therefore only covering the final months of this 3 year patient cohort.

Areas for attention from last report

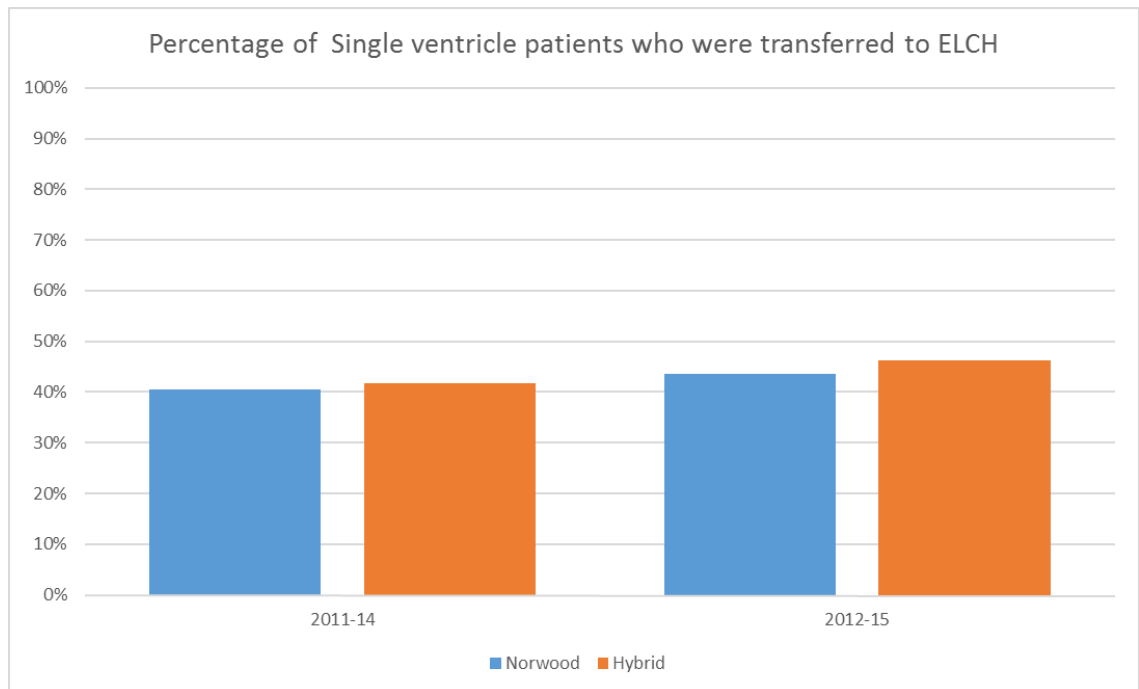
Our 2011-14 report highlighted particular areas for attention

1. Extra regional referrals
2. Low birth weight
3. Current NICOR (NCHDA) procedure codes

1. Extra-regional Referrals

In the 2011-14 cohort, of the 47 patients who had a Norwood procedure 19 (40%) were extra-regional referrals i.e. referred from other tertiary cardiac centres. For the same time frame 15 of the 36 (41%) patients who had a Hybrid as the primary procedure were extra-regional.

In this 2012-15 cohort of the 23 patients undergoing the Norwood procedure 45% had been referred from other tertiary cardiac centres. There is no difference in the mortality between patients from our own area and those referred from elsewhere.



2. Low birth weight

In the 2011-14 cohort 7/12 patients in the mortality group were of a low birth weight (<3.0kg). We recognised that we had not sufficiently standardised our approach to account for birthweight.

When the Hybrid procedure was initially instituted, one of the selection criteria was birth weight <2.5kg. There remains debate in the literature about the ideal selection criteria for a hybrid procedure and when this is indicated with weight as the primary indication. Following the program review we elected to modify the weight criteria for the hybrid procedure to 2.75kg or lower, and apply this consistently. This commenced in January of 2015 so applies to the final 3 months of this cohort

3. NICOR Procedure codes

At present the NICOR codes remain procedure rather than diagnosis based and are not stratified for risk. Additionally exclusion criteria apply for previous procedures. This generates two important points.

1. The code for Norwood is not diagnosis specific, so although a Norwood has been performed this may have been for a variety of complex conditions (hypoplastic left heart syndrome, tricuspid atresia with VSD and VA discordance, double inlet left ventricle variants).
2. In addition to our group of 23 patients who had a Stage 1 Norwood procedure there were and additional 10 patients who underwent a Stage 1 Norwood at a later part in their clinical course following a previous hybrid procedure.

Therefore the way the data is currently presented does not answer the question 'What is the outcome for patients with hypoplastic left heart syndrome in this institution'

We would be very happy to work with the NHCD A to agree a more sophisticated data collection and analysis to move towards more diagnosis based reporting.

Update on previous actions

The last report identified four specific actions. An update on these is provided.

1. Ongoing clinical reviews at 6 monthly intervals to examine data

We have instituted quarterly Paediatric Combined Cardiac Services Meetings attended by paediatric cardiologists, cardiac surgeons, paediatric cardiac anaesthetists and intensivists. The date and time is set to accommodate as many personnel as possible. All patients that enter the Hybrid and Norwood program are presented for review of their clinical course and outcome. Areas of concern are noted and acted on. Mortality is discussed at a separate specific meeting.

2. Antenatal second opinion for high risk cases

Since the publication of the last report there has been no fetus identified with the higher risk HLHS (eg intact atrial septum) requiring data to be sent for a second opinion.

3. Formal MDT review of all clinical data before acceptance of an outside referral

All external reviews are subject to an MDT prior to acceptance. This MDT must include cardiologists (including interventional cardiologists), cardiac surgeons and intensivists. The MDT discussion and outcome is documented formally and uploaded to the electronic patient record in the event of transfer, or sent back to the referrer in the event the patient is not accepted.

This MDT is open to all members of the team however is strengthened by ensuring the representatives of a 'core group' are present at each meeting.

4. External review

Although patient numbers have fallen, there has been a positive response to the action plan. The subject of an external review has been considered, however the collective feeling within the group and at senior managerial level outside of the department is that this is not necessary at this stage. However as a team we welcome scrutiny of the workings of the department and have invited an independent review from the Associate Medical Director of the Guys and St Thomas' NHS Foundation Trust. This is ongoing at the time of the report.

Our Future Action Plan

1. Review of surgical activity

As previously detailed we have invited our Associate Medical Director, a senior transplant surgeon (non cardiac) with experience in surgical unit review, to examine the workings and processes of the department.

2. Continued structured patient specific MDT

For every patient potentially entering the Norwood or Hybrid programme, an additional MDT is convened outside of the normal weekly combined cardiac surgical meeting. This is to ensure that the team is focussed on an individual case, and discussion will not commence until all relevant members of the group are present. This MDT structure will continue for at least the next 12 months.

3. Collate data on optimal treatment for HLHS

At present there is insufficient clinical evidence to guide the optimal approach to patients with hypoplastic left heart syndrome. Although we have developed criteria for the choice of one method of treatment vs. the other, it is yet to be determined whether either is superior, and if so in what clinical context. We will be examining all available data (our own, national and international) to ensure that our surgical treatment is as evidence based as possible, taking long term outcomes into account in addition to short term survival. This will include the choice of primary option and where a hybrid has been undertaken the subsequent clinical pathway.

4. Establish a mortality and morbidity review group

Review of all cardiac cases, what went well, what could be done better, collation and dissemination of learning points to whole team. This is in addition to the existing M&M and incident reporting processes.

5. Peer Support

As a broad team we strive to provide excellence in all aspects of the care we provide. It is important therefore in this situation when there is extra scrutiny of results above and beyond what we normally face, that we maintain morale and ensure that all team members are, and feel, appropriately supported. We will strive to do this through active and supportive leadership and ensuring that all members of the team are able to contribute to the ongoing development of the department.

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