An Analysis of Results for the Norwood 1 operation at Evelina London Children's Hospital (ELCH)

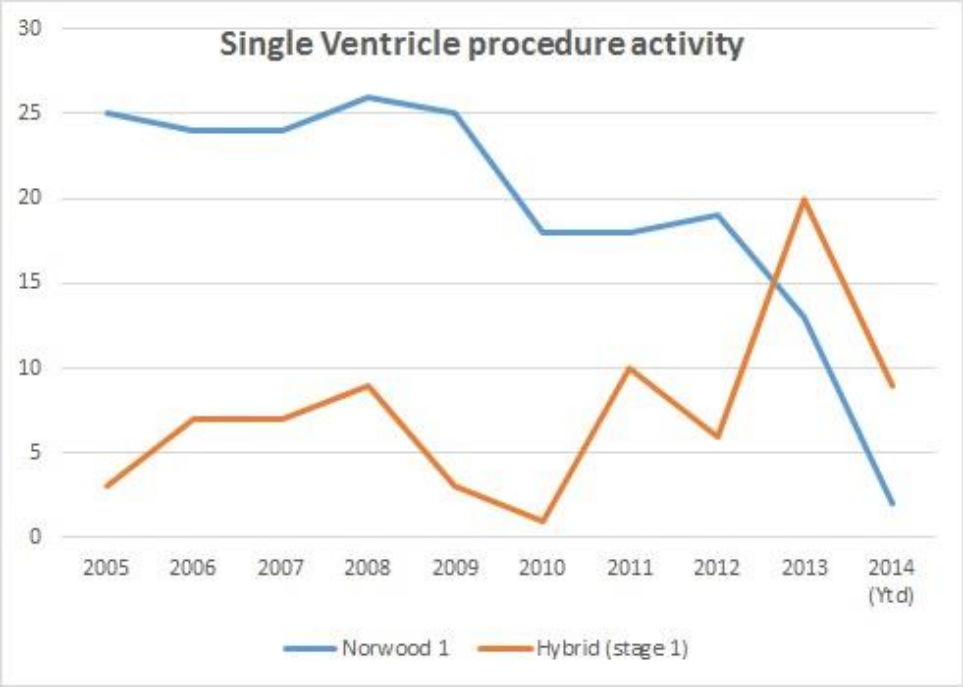
(2011-2014)

Introduction

The 2011-2014 NCHDA data analysis suggests that there is a higher than expected 30-day mortality for the Norwood 1 operation at the ELCH. On the NICOR analysis we fall into a green warning area and this may occur purely by chance (1:40). A review of this procedure group has been carried out.

For the Norwood 1 operation, the initial NICOR analysis showed 13 deaths out of a total of 49 procedures. During validation, we identified 2 cases which had been incorrectly coded and had been inadvertently placed within the Norwood 1 group; these cases had in fact had Combined Stage 1 & 2 operation after an initial Hybrid procedure. These have been recoded with NICOR and we therefore have 47 patients in this cohort with 12 deaths representing a 30-day procedural survival of 74%. The NICOR notification confirmed concerns raised by our own internal monitoring. In fact we had already started comprehensive internal review of our whole single ventricle programme 12 months in advance of the NICOR notification.

Our single ventricle programme has two main arms; those who undergo a primary Norwood 1 repair and those who go down the Hybrid (bilateral PA bands & arterial duct stent) route. As can be seen in the graph below there has been a significant change in our strategy over the last 5 years, with a fall in the number going down the Norwood 1 pathway and an increase in those following a Hybrid pathway.



We therefore feel it is important that both arms of this treatment path are reviewed as it would be potentially misleading to view them as separate surgical programmes. We would ask that NCHDA review how it presents data on single ventricle palliation.

In 2005 we pioneered the “Hybrid” programme in the UK for hypoplastic left heart syndrome (HLHS) due to the high procedural mortality associated with the Norwood 1 operation in very small babies. This was initially aimed at

neonates with a birth weight < 2.5 kg, otherwise fragile neonates who may not tolerate cardiopulmonary bypass or those with other significant co-morbidities.[1]

In the period 2011-14, we have performed 36 Hybrid procedures (mortality 16.6 %) for HLHS or variants and 47 Norwood 1 operations (mortality (25.5%). Despite initially catering to small and fragile infants, we have expanded our Hybrid programme to include larger infants and those with fewer co-morbidities. Despite the encouraging results from the Hybrid procedure, the risk for external referrals is still higher (mortality 20% vs. 14% for local patients).

Consideration of the Hybrid procedure however, introduces further heterogeneity as it is used not only for single ventricle palliation but also as a staging procedure in the borderline left ventricle which may lead to an eventual take down to biventricular repair.

The Hybrid procedure can be followed either with an early subsequent Norwood-type aortic arch reconstruction or a later combined Stage I+ II operation which includes both a Norwood type arch reconstruction and a superior cavo-pulmonary anastomosis as a combined procedure. This complex clinical pathway is not able to be described by the limited EPCC codes used in NICOR's procedural coding. For example, the "Norwood" code includes primary arch reconstruction but also secondary arch reconstruction following the initial Hybrid.

The coding of the Combined Stage I & II would also benefit from using expanded long codes as it may currently fall either into a Norwood category or a Bidirectional Glenn category depending on how the arch reconstruction is coded. As a major UK centre for this procedure, we would welcome an opportunity to work with NICOR to harmonise coding across the UK.

Analysis of the procedural mortality confirms that the outcomes are not surgeon-specific and are equally distributed between two surgeons. In our programme, all single ventricle palliations, both Norwood and Hybrid are performed by all three consultant surgeons.

Despite anomalies in NICOR procedural classification and in conjunction with our concerns about outcomes for high risk single ventricle patients, we embarked in January 2014 on a comprehensive internal review of our whole programme. We believe strongly that outcome is influenced by the whole clinical pathway and is not simply a matter of surgeon performance. For this reason we have broken down the single ventricle programme in to component work streams and looked both at our data but also contemporary practice to devise an institutional approach to this complex patient pathway.

Due to historic subspecialisation, we are aware that extra-regional referrals make up a significant part of our single ventricle workload. We have traditionally not been risk averse and have supported clinicians and families from many other UK centres with single ventricle palliation. These cases may have been referred for many different reasons including having been judged as too complex and/or too high risk for the referring institution or simply due to surgeon availability, bed status or patient/family choice. Amongst the deaths covered in this report, 4/12 were extra-regional cases.

During the analysis period (2011-14) some UK centres did not have an active Norwood programme and we received referrals either ante-natally or post-natally to cover this lack of a local service. Our internal analysis has shown that external referral have a higher risk of mortality compared to patients from within our network. For these reasons we consider it particularly important to look at case selection to ensure that these external referrals, some who have been judged as higher risk in the referring institution are appropriately triaged when referred to us. Although we continue to accept these cases, it should be recognised that accepting a high risk referral

[1] Venugopal PS, Luna KP, Anderson DR, Austin CB, Rosenthal E, Krasemann T, Qureshi SA Hybrid procedure as an alternative to surgical palliation of high-risk infants with hypoplastic left heart syndrome and its variants. J Thorac Cardiovasc Surg. 2010 May;139(5):1211-5

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transfers surgical risk from one institution to another. This may affect 98% control limit (green line) and also not reflect the true national benchmarking (like for like comparison). In any case, we have agreed clear case selection criteria and from January 2015 have commenced a more rigorous case triage process with a separate and focussed MDM for this more complex higher risk group.

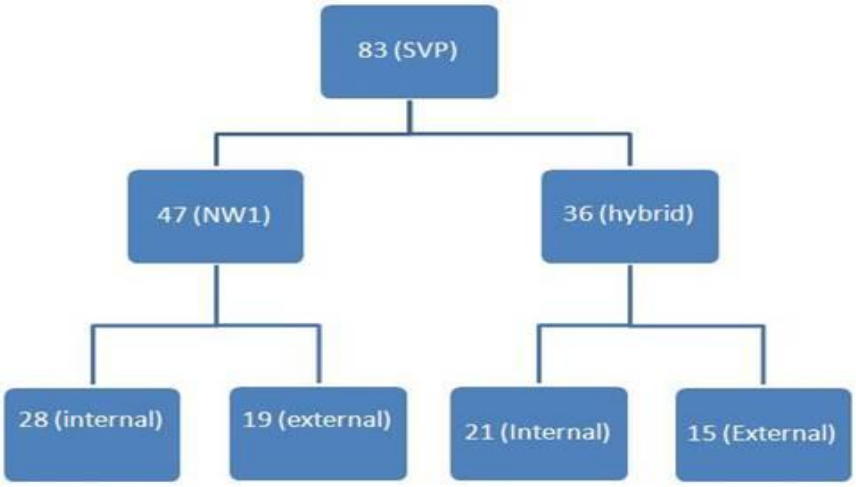

Figure 1: Origin of Single Ventricle Patients (SVP) entering ELCH programme. The centres who have sent us single ventricle patients during the analysis period are : Liverpool/Manchester (7); Bristol/Cardiff (6); Belfast (1); Glasgow (1); Royal Brompton (1); Great Ormond Street (2); Leeds (11), Leicester (3); Oxford (1) and overseas (1).

A second work stream has reviewed and benchmarked our anaesthetic management for the Norwood 1 and Hybrid procedures. We have agreed standardised guidelines for anaesthesia for Norwood 1 and Hybrid procedures. There is a new protocol for transfer and formal handover to intensive care which is now well established. We will audit our compliance with the protocol and will implement any recommendations. We have developed a group which focuses on anaesthesia for Norwood 1 and Hybrid procedures to ensure consistency and to consolidate the implementation of the standardised guidelines.

Post-operative intensive care management has also been scrutinised, but has not seen any identifiable pattern in ICU practice linked to Norwood I mortality. Despite this we have resolved to standardize protocols for the PICU management of single ventricle patients, and involve the PICU team formally in the case selection stage of the clinical management pathway.

Regarding inter-stage mortality, we have instituted the Evelina London Home Monitoring Programme (HMP). This programme identifies all single ventricle palliation patients, institutes a parent/carer education programme before hospital discharge and provides an oxygen saturation (SaO_2) monitor and baby weighing scales to each family at discharge. The parent/carer must monitor and diarise the daily SaO_2, infant weight and feeding pattern. Weekly (often more frequent) telephone contact and fortnightly clinic visits provide very close supervision of these vulnerable infants between Stage 1 and Stage 2 surgery and has seen 0% inter-stage mortality in the first 9 months of the programme.

Our review has highlighted the following aspects of our programme for particular attention:

1. Extra-regional referral. 4/12 procedural deaths following NW1 surgery were in patients referred from external regional cardiac centres. As a methodological observation, the transfer of high risk patients between centres, may have an effect on the funnel plot, particularly when the funnel plot control lines is derived from centres with higher

volumes of low risk cases or who perform large numbers of Sano procedures, where the 30 day mortality is known to be lower although the mortality beyond 1 year is similar. [2] As a unit, we have continued to perform a modified Blalock-Taussig shunt, accepting the higher mortality over 30 days (which is known to equalise later) in order to avoid the incision of the systemic right ventricle required for the Sano shunt. This approach has been supported by a recent randomized trial showing that the right ventricular systolic function is indeed preferentially preserved using this strategy. [3] For this reason, we do not wish to change our surgical approach to improve only the 30-day outcome at the potential detriment of longer term outcomes.

We would welcome an opportunity to work with NICOR/NCHDA to further develop procedural coding categories and increase the number of procedure codes so that data is more robust and more truly represents national activity.

2. **Low birth weight.** 7/12 of the patients in the Norwood 1 30 day mortality group had a birth weight < 3.0 kg. In 2005 we had commenced our Hybrid programme to address this issue of low birth weight single ventricle mortality, but had not sufficiently standardised our approach to only offer the Hybrid to this patient group. Going forward our own internal review has recommended clear guidelines for surgical decision making including birth weight thresholds.

3. The current **NICOR procedure codes** cover a heterogeneous diagnosis population with heterogeneous risk profiles within a limited numbers of procedural codes. We have always been and continue to be staunch supporters of NICOR, and would support a more sophisticated approach to coding particularly as some centres do subspecialise and offer surgery for high risk cases which may have been declined by their local and/or other specialist centres. We look forward to working with NCHDA to agree a way forward with regard to risk stratification particularly for the evolving procedural options now available for patients with a single ventricle.

Our action plan:

1. Our own concerns regarding single ventricle outcomes had already prompted a year-long review of the whole patient pathway from referral to discharge (and beyond). We have produced updated formal guidelines for case selection, surgical decision making and perioperative management. We are intensively examining each case and our cumulative outcome and will perform ongoing clinical reviews at 6 monthly intervals until we see an improvement in our data. Of note, following our internal review and the introduction of new triage and management protocols, we have performed 7 single ventricle palliations (3 Hybrid and 4 Norwood) with 0 % 30 day mortality. (data from 01/12/14 – 13/03/2015)

2. For the fetus diagnosed with a higher risk HLHS (intact interatrial septum, tricuspid regurgitation, aberrant subclavian artery), where we consider post natal intervention to be very high risk, we will consider sending the data for an ante natal second opinion before accepting on to our surgical programme.

[2] Ohye RG, Sleeper LA, Mahony L, Newburger JW, Pearson GD, Lu M, Goldberg CS, Tabbutt S, Frommelt PC, Ghanayem NS et al: Comparison of shunt types in the Norwood procedure for single-ventricle lesions. The New England journal of medicine 2010, 362(21):1980-199

[3] Newburger JW, Sleeper LA, Frommelt PC, Pearson GD, Mahle WT, Chen S, Dunbar-Masterson C, Mital S, Williams IA, Ghanayem NS et al: Transplantation-free survival and interventions at 3 years in the single ventricle reconstruction trial. Circulation 2014, 129(20):2013-2020

3. We have a long history of accepting high risk referrals. Although we will continue to offer this national service, we now insist on a formal MDT review of all clinical data in advance of accepting the transfer of a newborn or a fetus. This process has been further strengthened by a focussed senior oversight panel to review all single ventricle cases to quality assure our agreed internal clinical pathway. We acknowledge that continuing to accept such high risk cases, will transfer risk from the referring centre to the Evelina and that this is not fully compensated by the current NICOR analysis.

4. Should we not see a prompt and positive response to this action plan which commenced in December 2014, we will invite an external review of our programme to further inform a way forward.

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