University Hospitals Bristol NHS
NHS Foundation Trust

Arterial shunt operations for congenital heart disease

An analysis of results at the Bristol Royal Hospital for Children (BRHC) from 1999-2013

Introduction

The 2010-2013 NCHDA data analysis suggests that at the Bristol Royal Hospital for Children (BRCH) there is a higher than expected 30-day mortality for the arterial shunt operation. The CCAD analysis shows there are 6 deaths out of a total of 27 patients, representing a 30-day survival of 77.8%.

For the first time, the NCHDA analysis produced in April 2014 risk-adjusted the mortality data. This data is shown in a funnel plot which compares UK institutions by volume using the average national mortality for this procedure as the comparison target. However, when comparing institution's performance, it is essential to understand that this risk-adjustment, although a welcome step, is rudimentary and does not include important factors such as the severity of primary diagnosis or other co-morbidities such as prematurity, low weight, genetic syndromes etc. Further analysis, using these factors, is required to fully understand an organisation's performance. It is also important to state that the funnel plot methodology is prone to false positives through repeated testing. This is acknowledged in the notification letter from CCAD.

This report summarizes the results of arterial shunt operations done at BRCH since 2000. It is generated from several sources of information:

1) NCHDA -based, centrally validated data;

2) BRCH internal data, and

3) audit of BT shunts performed between 2008-2012.

It is important to note that BRHC's internal data collection is contemporary whereas NCHDA centralisation and validation is approximately one year behind individual units' data.

Context

One significant factor in this more detailed consideration is the effect that small variations in the results for one year can have on the statistics over a number of years when overall totals are low. Significantly, the BRHC survival for arterial shunts in 2012-2013 was 81.8%, improving to 2013-2014, 92.3%.

Another significant factor in understanding shunts is that NCHDA requires every operation to be allocated to one category. As a consequence, far more complex operations can be allocated along with much simpler operations. A significant example relevant to the BRHC data is that a complex procedure called a unifocalisation may be allocated as a shunt if at the end of the procedure it has

not been appropriate to perform a complete repair but the patient's blood supply to their lungs comes from an arterial shunt.

In addition there are factors that are recognised to significantly increase the risk of an arterial shunt procedure. First is low birth weight. Shunts are created from fixed sizes of tube and, if the tube is too big, too much blood flows down it. Therefore, the smaller the child, the higher the risk that the procedure carries. Two children who died in the BRHC patient group fell into this category.

Second are high-risk abnormalities of the heart. Because of the high pressure in the right ventricle, abnormal arterial connections are formed with the coronary arteries. These are not normal coronary arteries and the filling of them depends on the pressure in the right ventricle rather than the systemic circulation. It is well recognised that patients with this abnormality are at much higher risk undergoing an arterial shunt than other patients. One child who died in the BRHC patient group fell into this category.

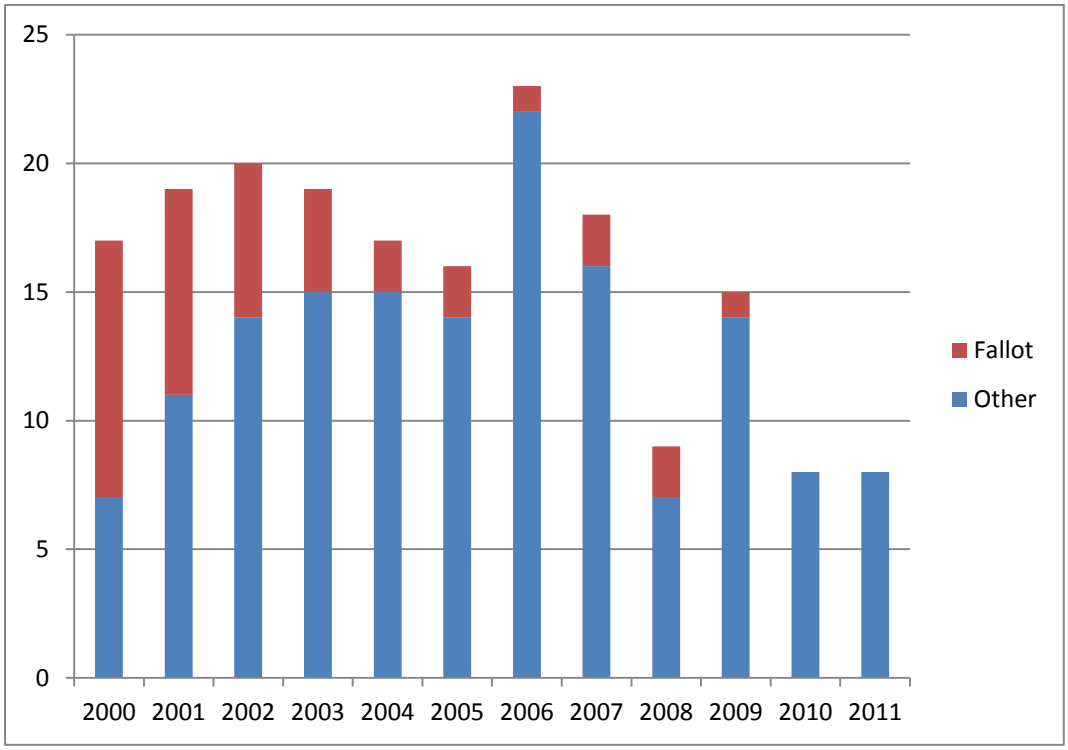
Finally, there are other abnormalities that the child has been born with, co-morbidities, that significantly impact on patient outcome.

1. **NCHDA -submitted data**

One hundred and ninety-eight shunts and shunt-related procedures were performed in England from 2000 - 2011. The number of arterial shunts performed at BRCH has significantly decreased over time, both in absolute number and as a percentage of total procedures done at the hospital. This is because of changes in practice and the management of some patients.

With recent improvements in outcomes after primary repair operations in younger children, we have moved away from undertaking arterial shunt operations in some conditions. This has particularly been our experience with infants with pulmonary atresia and tetralogy of Fallot where we now offer a full repair operation in preference to a shunt operation (Fig 1). This is due to a change in our philosophy of treatment for tetralogy of Fallot, whereby primary complete repair is now favoured over initial shunting. Of particular importance in this is the fact that patients with tetralogy of Fallot and pulmonary atresia have 2 ventricles and therefore are far more robust if they undergo a shunt operation. As a consequence, fewer of our patients overall undergo an arterial shunt procedure and those that do have complex, high risk abnormalities that only have one ventricle. Despite our change in management of this condition, the NCHDA data shows there has been no change in our mortality for tetralogy of Fallot and the BRHC currently has 100% survival for this operation (https://nicor5.nicor.org.uk/CHD/an_paeds.nsf/0/DFE20B65CF749CF180257C77000BFDE5?OpenDocument?Benchmark).

Figure 1. Change in indication for BT shunts



P<0.001 for the proportion change over time

2. BRCH HeartSuite data

A search of the BRHC database using the key terms 'shunt' and 'arterial' showed we had performed 182 operations over the period 1998-2013. The discrepancy between NCHDA data and internally collected patient records is mainly as a result of the NCHDA -specific algorithms that allocate diagnoses and procedures.

These data show that 42 shunts and shunt-related procedures were performed in 36 patients at the BRHC from 2010 – 2013. Of these, 7 patients did not survive to 30 days, which translates in a survival of 80.6% (or 83.8% if the denominator is the entire number of procedures rather than patients). This figure is below the national average.

3. Audit of BT shunts

Through an internal audit system, BRHC clinicians have previously reviewed their approach to, and results from, the arterial shunt procedure.

The first audit was performed in 2008, at which the specific problem identified was acute shunt blockage. Through the audit, clinicians identified modifiable factors, such as prehydration for children with high haematocrits. After implementing these changes, results improved.

More recent analysis identified that overcirculation, too much blood going to the lungs was now affecting outcomes. The problem of overcirculation is one that every unit in Britain has experienced. Therefore, representatives of surgeons, anaesthetists, and intensivists at BRHC telephoned

colleagues in other centres to determine if there is anything that could be done differently. The centres that were chosen were ones that have always had good results from this procedure, and one that had previously had poor results but subsequently showed improved outcomes.

What we found has led us to write new guidelines, emphasising;

- avoidance of catecholamines
- maximising vasodilatation
- maintaining a high haematocrit
- low threshold for using cardiopulmonary bypass
- possibly clipping shunt for first night
- median sternotomy for surgical approach.

Another important aspect associated with the arterial shunt is the recognition that, of those children who died following the procedure, a high proportion did well during their hospital stay but died following discharge from hospital within 30 days of surgery. The reasons for this are multifactorial but all may be addressed by using a home monitoring system.

Back in summer 2009, clinicians at BRHC started a home monitoring programme for patients with hypoplastic left heart syndrome when we started our Norwood programme. As these two groups of patients have a similar physiology, we have adopted the Norwood protocol for patients with shunts. Appropriate information leaflets have been prepared to inform parents what to do if their child falls outwith the expected parameters. Since adopting this protocol no child has died between discharge from hospital and undergoing the next stage of their surgical pathway.

Clinicians at BRHC have been aware for a period of time that the arterial shunt operation is a high risk procedure. In recent years the NCHDA national data has shown that it is the second most risky operation congenital cardiac surgeons perform. We have therefore moved away from doing this operation, instead preferring to use stents implanted in the cardiac catheterisation laboratory or performing a complete repair in suitable children even though they are very small. One consequence of this is that we now only perform the shunt operation in very high risk patients.

Using this approach we have had excellent results for primary complete repairs (see data above for tetralogy of Fallot). However, this overall improved outcome is not reflected in the NCHDA data as the data reflects an operation rather than the heart abnormality the operation was performed for. It should also be observed that following shunts there is a significant mortality beyond the 30 days quoted by NCHDA for shunts which makes the modified approach at BRHC even more beneficial.

Every death that occurs in the BRHC paediatric cardiac services unit is carefully and completely considered and discussed in our Child Death Reviews, and any lessons learnt are implemented.

4. Summary Points

We recognise that the 2010-2013 NCHDA data~~set~~ suggests that at BRCH there is a higher than expected 30-day mortality for the arterial shunt operation. Our intent has been to fully understand the reasons for this and seek to further improve our practice, whilst at all times ensuring that the patients' welfare is of paramount importance.

Our preliminary review has highlighted the following key findings:

- The number of arterial shunts done at BRCH has decreased over time, both in absolute number and as a percentage of total procedures.
- There has been a trend towards primary complete repair, *rather than palliative shunt*, for Tetralogy of Fallot and other 2-ventricle abnormalities such as pulmonary atresia/VSD, resulting in shunts only being performed in patients with high risk, single ventricle abnormalities.
- Of the 6 patients who died (2010-2013) following an arterial shunt:
    - 2 were of low birth weight
    - 2 died of blocked shunts at home, remote from the operation
    - 1 had MAPCAs (multiple abnormal vessels to the lungs) but ended up having a shunt to provide blood flow to the lungs
    - 1 had high risk anatomy (RV-PA sinusoids, Coronary artery stenosis)

The significance of these factors is explained in the report.

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