Outcome after extended arch repair for aortic coarctation.

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Abstract

Objectives: To assess survival and long term arch patency rates in a consecutive group of children after extended arch repair for coarctation of the aorta.

Background: Extended arch repair were developed with the aim of minimising recurrent arch obstruction. Existing follow up data is still lacking.

Methods: Review of 191 consecutive children (154 (81%) under 1 year of age) operated on between 1990 and 2002 by a single surgeon using extended arch reconstructive techniques. For assessment of survival patients were divided into 3 groups (1=coarctation alone n=104, 2=coarctation+VSD n=38, 3=coarctation in association with complex intracardiac anomalies n=49). A prospective and systematic clinical and echocardiographic evaluation of the aortic arch was undertaken.

Results: Median time to follow up was 4.2 years (1-10.6). Overall actuarial survival was 92, 88 and 88% at 2, 5 and 10 years. Mortality was significantly higher in those patients with complex intracardiac anatomy. Recurrent arch obstruction occurred in 7/165 (4.2%) of all patients, 4/139 (2.9%) of term and 3/10 (30%) premature infants (p<0.001).

Conclusions: Survival after extended arch reconstruction for coarctation is excellent. At long follow up recurrent arch obstruction is rare, with prematurity the only risk factor.

Introduction
Corrective surgery for coarctation of the aorta was first described 60 years ago. Classical surgical techniques do not address arch hypoplasia or encroachment of ductal tissue beyond the anastomosis and consequently recurrent arch obstruction is relatively common. Recurrent arch obstruction assumes greater importance as a measure of successful outcome after coarctation repair given the excellent survival rates in the modern era. Of all current strategies, extended arch reconstruction (both extended end to end and end to side anastomoses) best addresses both the problem of ductal extension through the aorta and associated arch hypoplasia. Reports of extended arch repair thus far have been in selected groups of patients with relatively short follow up times.

We report a systematic clinical review of over a decade of single surgeon experience using extended arch repairs in unselected children of all anatomical subgroups.

Patients and methods
The Yorkshire heart centre is a regional cardiothoracic centre serving a local population of 5.5 million people with stable referral patterns. Between January 1990 and January 2002, 191 children underwent surgical repair of coarctation of the aorta by a single cardiac surgeon (KGW) utilising an extended arch repair (EAR) (which can be subdivided into either extended end to end or extended end to side-see below). 112 (59%) were male and the group heavily biased towards infants with 154 (81%) under one year of age. The median age at operation was 1 month (1d-15.2 years), median weight 3.7 kg (1-58kg).

During the same period an additional 10 patients underwent subclavian flap repair (all before 1994) and 4 patch augmentation of coarctation (all older patients). There was no difference in anatomy or demographics between those patients undergoing subclavian flap and EAR repair, the mixture of techniques early in the series representing transition from one surgical strategy to another.

Additional cardiac lesions
144 (75%) patients had intracardiac lesions.

For the purposes of survival analysis patients were divided into the following three groups:

Group 1- 104 patients (52%), “Isolated” coarctation without associated major intracardiac lesions –but including “minor” abnormalities (e.g. bicuspid aortic valves and atrial septal...
defects). Median age and weight at surgery 2 months (2 days-15 years) and 4.2 kg (1-58kg) respectively.

**Group 2** - 38 patients (20%). Coarctation and a ventricular septal defect. Median age and weight at surgery 2 weeks (1 day-10.8 years) and 3.2 kg (2.1-25kg) respectively.

**Group 3** - 49 patients (28%). Coarctation with associated complex intracardiac anomalies (table 1). Median age and weight at surgery 10 days (1 day-4 years) and 3.4 kg (2-22kg) respectively.

<table>
<thead>
<tr>
<th>Table 1</th>
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<tbody>
<tr>
<td><strong>Significant associated cardiac anomalies</strong></td>
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<tr>
<td>-------------------------------------------------------------------</td>
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<tr>
<td>Significant mitral valve abnormality</td>
</tr>
<tr>
<td>Shones complex</td>
</tr>
<tr>
<td>TGA (+/- VSD)</td>
</tr>
<tr>
<td>TGA + Dextrocardia</td>
</tr>
<tr>
<td>Single ventricle (DORV, DILV etc)</td>
</tr>
<tr>
<td>AVSD</td>
</tr>
<tr>
<td>Anomalous PV drainage</td>
</tr>
<tr>
<td>Atrial isomerism + VSD</td>
</tr>
<tr>
<td>Severe sub aortic stenosis</td>
</tr>
<tr>
<td>Cor triatriatum</td>
</tr>
<tr>
<td>Sinus venosus ASD</td>
</tr>
<tr>
<td>Ebsteins anomaly</td>
</tr>
<tr>
<td>ccTGA</td>
</tr>
<tr>
<td>AP window</td>
</tr>
<tr>
<td>Dextrocardia</td>
</tr>
<tr>
<td>Complex intracardiac anomalies (total)</td>
</tr>
</tbody>
</table>

Key:

- VSD = Ventricular septal defect
- TGA = Transposition of the great arteries
- DORV = Double outlet right ventricle
- DILV = Double inlet left ventricle
- PV = Pulmonary vein
- AVSD = Atrioventricular septal defect
- ccTGA = Congenitally corrected transposition of the great arteries
- AP = Aorto-pulmonary

Transverse arch hypoplasia (defined as a diameter of the aorta between the common carotid and left subclavian artery of less than 50% of the ascending aorta) was identified retrospectively from either the operation note or the pre operative echocardiogram. Transverse arch hypoplasia was present in 79 (41%) of patients (52% of infants under 1 year) and was more common in those patients undergoing EETS repair through a median sternotomy (30/50 patients vs 49/141, p<0.002).

**Operative techniques**

Extended arch reconstruction (EAR) includes two similar techniques (figures 1 and 2), both based on the principles of wide anastomosis with maximal ductal excision.

1.) Extended end to end arch repair (EETE) (figure 1) was used in 141 patients (73%) through a left lateral thoracotomy. Dissection and mobilization of the subclavian artery, descending aorta,
ductus arteriosus and the aortic arch up to the innominate artery was performed. The aortic arch, subclavian artery and the descending aorta were clamped and the narrowed segment of the aorta excised with particular attention to complete excision of all ductal tissue. The anastomosis was constructed with continuous 7/0 polypropylene.

2.) Extended end to side repair (figure 2) was used in 50 patients. The procedure was performed via median sternotomy using cardiopulmonary bypass and deep hypothermic circulatory arrest. The innominate, left carotid and subclavian arteries and descending aorta were dissected out and extensively mobilized. The ascending and descending aorta were clamped and the circulation arrested. Head vessels were snared and the ductus ligated and excised. An incision slightly larger than the diameter of the descending aorta was made on the convexity of the proximal aortic arch opposite the innominate artery. The descending aorta was moved upward and the anastomosis created with continuous 7/0 polypropylene.

Non invasive assessment and patient follow up

In addition to review of routine follow up clinic notes and echocardiograms, patients were recalled for evaluation in a specific study clinic held between 1999 and 2003. 96% of patients attended the clinic. 6 (6/71, 3.5%) patients were lost to follow up. In the remaining 165 patients a protocol of clinical and echocardiographic assessment was carried out by a single paediatric cardiologist (JT). All patients were assessed at least 12 months after surgery as follows:

a.) Femoral pulses were assessed as follows:
   Grade 3-Normal
   Grade 2-Slightly reduced compared with brachial pulse
   Grade 1-Clearly reduced femoral pulsation
   Grade 0-Absent

b.) A mean of three upper and lower limb blood pressure measurements were made using an automated oscillometric blood pressure machine (Critikon, General electric corp., USA) with a blood pressure cuff appropriate to each patient. Age and sex appropriate centiles for were derived from published normal data.16

c.) Echocardiography: A full 2D echocardiographic examination was performed using a Sonos ultrasound machine (HP, Minnesota, USA). Ascending and descending aortic velocities were taken from the suprasternal notch using a dedicated 1.9 MHz continuous wave Doppler transducer. Peak systolic, and time to half peak systolic and time to half peak diastolic velocities were calculated from the descending aortic traces.17

Residual or recurrent arch obstruction

Recoarctation was defined as either 1) 20 mmHg difference between mean arm/leg blood pressure or 2) Grade 0 or 1 femoral pulses or 3) A descending aortic Doppler gradient of >25 mmHg with a deceleration ½ time >100ms or 4) Previous reintervention to the repair site based on clinical judgement at that time.

Statistics

Statistical analysis was performed using GB Stat version 6.5 (Dynamic microsystems, USA). Outcome variables were survival and recurrent arch obstruction. The Fisher exact test was used to compare univariate categorical variables. Actuarial estimates for survival and re/residual coarctation were made using the Kaplan-Meier method using the Cox proportional hazards model to establish variables independently associated with outcome. A p value of <0.05 was considered significant.
Results

Follow up
Median time to follow up was 4.2 years (1-10.6).

Mortality (table 2)
9 deaths occurred within 30 days of the original operation. Early mortality was 2/104 (1.9%), 2/38 (5.3%) and 5/49 (10.2%) for groups 1, 2 and 3 respectively with a statistically significant difference between groups 1 and 3 (p<0.04).
There were 11 late deaths (> 30 days) in the 182 survivors at a median of 6.9 months (1.3-50.8) after surgery. Late mortality was 2.9%, 5.6% and 13.6% for groups 1, 2 and 3 respectively (significant difference between groups 1 and 3, p<0.03). Actuarial survival for the whole cohort at 2, 5 and 10 years was 92%, 88% and 88% (figure 3).
The majority of deaths occurred in patients with complex congenital heart disease (73%) who had undergone procedures in addition to coarctation repair.
Of the 3 late deaths occurring in the group with coarctation alone (group 1) 2 patients had chromosomal abnormalities and died of non cardiac causes (hydrocephalus and chronic lung disease) and the third patient died 3.6 years after surgery from unexplained pulmonary hypertension having earlier undergone reintervention for recoarctation.

<table>
<thead>
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<th>Table 2</th>
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<tr>
<td>Cause of death</td>
</tr>
<tr>
<td>N</td>
</tr>
<tr>
<td><strong>Hospital mortality (&lt;30 days)</strong></td>
</tr>
<tr>
<td>Failure to recover from operation</td>
</tr>
<tr>
<td>Sepsis</td>
</tr>
<tr>
<td>Ventricular failure</td>
</tr>
<tr>
<td>Blocked ET tube and arrest</td>
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<tr>
<td>Total</td>
</tr>
<tr>
<td><strong>Late mortality (&gt;30 days)</strong></td>
</tr>
<tr>
<td>Pulmonary hypertension</td>
</tr>
<tr>
<td>Parental decision (refused further surgery)</td>
</tr>
<tr>
<td>Sepsis</td>
</tr>
<tr>
<td>Sudden unexplained death at home</td>
</tr>
<tr>
<td>Ventricular failure</td>
</tr>
<tr>
<td>Chronic lung disease</td>
</tr>
<tr>
<td>Hydrocephalus (Trisomy 9 mosaic)</td>
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<tr>
<td>Total</td>
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</table>

Neurological injury
Paraplegia occurred in 1 patient following a significant intraoperative bleed from the anastomotic site. A second patient had transient lower limb weakness but recovered fully and now walks normally. Both of these patients underwent EETE repair through a lateral thoracotomy. A third patient had a small segmental cerebral infarct on CT scan following coarctation repair, arterial switch and VSD closure but has no long term sequelae.
Reintervention to the coarctation site
Reintervention was required in 7/165 (4.2%, 95% CI 1.9-12.1) surviving patients (median age and weight at surgery 2 weeks (1 week-2.2 months) and 2.5kg (1-4.3kg) respectively) for residual or recurrent arch obstruction a median of 5 (2-9) months following surgery. All of these patients had diminished (grade 0 or 1) femoral pulsation, and systemic hypertension in addition to echocardiographic abnormalities (see below). 3/7 were premature infants and 3/7 had transverse arch hypoplasia. 6/7 patients with recurrent arch obstruction had discrete stenosis suitable for balloon angioplasty and 1 (premature) infant a long segment stenosis in which angioplasty was not attempted. In this case a re-operation was performed. Freedom from reintervention for recurrent arch obstruction (all patients) was 96% at 1, 5 and 10 years respectively, with no significant difference between the three anatomical groups.

Prematurity was the only significant risk factor (p<0.001) for reintervention (figure 5). All 7 patients with residual or recurrent arch obstruction had undergone EETE repair thorough a lateral thoracotomy.

Clinical and echocardiographic data (table 3)

Blood pressure
Lower limb systolic blood pressure was significantly higher than upper limb (p<0.001). Mean indexed centile for blood pressure based on mean upper limb systolic readings was 75 (SD 20). 3 patients were on antihypertensive treatment (all with no angiographic evidence of recoarctation) 35/166 (21%) of patients were above the 90th centile for blood pressure. In 34% of these patients (all under 5 years of age) acquiring calm blood pressure readings was noted as difficult at the time of assessment.

Doppler echocardiography
Echocardiographic data are shown in table 2. 10 patients had descending systolic aortic Doppler velocities marginally in excess of 3 m/s (maximum 3.1 m/s) with no other clinical signs of recurrent arch obstruction (normotensive, grade 2 or 3 femoral pulses, no diastolic prolongation of the descending aortic Doppler trace). In 4/10 of these patients cardiac catheter or magnetic resonance imaging showed complete luminal patency. No MRI or catheter data is available in the other 6 patients.

Retrospective review of echocardiographic data in those patients who had required reintervention for recurrent or residual arch obstruction showed time to half peak diastolic velocity to be in excess of 100 ms in all 7 cases (median 130 ms, range 115-185ms). The mean time to half peak diastolic velocity for the rest of the group was 70 ms (SD14).

| Table 3
| Follow up data |
|---|---|
| **Echocardiography** | |
| Ascending aortic velocity (m/s) (SD) | 1.9 (0.57) |
| Descending aortic velocity (m/s) (SD) | 2.2 (0.51) |
| Echocardiographic gradient (mmHg) (SD) | 6.1 (7.8) |
| Time to half peak diastolic velocity (m/s) (SD) | 70 (14) |

| Femoral pulses (grade) (Median, range) | 4 (3-4) |

<table>
<thead>
<tr>
<th><strong>Blood pressure</strong></th>
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<tbody>
<tr>
<td>Upper limb systolic (Mean, SD)</td>
<td>109 (14)</td>
</tr>
<tr>
<td>Upper limb diastolic (Mean, SD)</td>
<td>58 (10)</td>
</tr>
<tr>
<td>Lower limb systolic (Mean, SD)</td>
<td>123 (17.5)</td>
</tr>
<tr>
<td>Lower limb diastolic (Mean, SD)</td>
<td>68 (12.5)</td>
</tr>
<tr>
<td>Upper limb systolic centile (Mean, SD)</td>
<td>75 (20)</td>
</tr>
<tr>
<td>Upper to lower limb gradient (mmHg) (Mean, SD)</td>
<td>-15 (17.6)</td>
</tr>
</tbody>
</table>


Discussion

Surgery for coarctation of the aorta has changed considerably since the original description in 1945.1 Although there is no consensus on optimal management strategy, extended arch repairs are increasingly utilised.7-14 This approach allows the surgeon to fully resect ductal tissue and to address tubular hypoplasia using a large anastomosis incorporating only autologous tissue. There are a number of reports following extended arch repair techniques,7-14 the majority in small numbers of patients generally focusing on those patients with isolated coarctation7-13. Deaths are now rare after arch repair alone—in this regard our study is comparable with other recent data (95% survival at 3 years).7-14 In this study survival rates in patients with associated complex congenital heart disease are similar to those described by Wood et al 13 but superior to those reported by Conte et al 12 both of whom used palliative pulmonary artery banding instead of a one stage repair.

Survival after isolated aortic arch repair is rarely a problem in the modern era and the focus has shifted to minimising the risk of recurrent arch obstruction. Reports of surgical repair using techniques that do not fully address duct encroachment and tubular hypoplasia (subclavian flap augmentation and end to end anastomosis) describe recurrent arch obstruction rates of up to 41%.2-5 In our study actuarial freedom from recurrent or residual arch obstruction was 96% for the whole patient cohort at up to over 10 years of follow up. This medium term data adds to that of others showing excellent early arch patency rates after extended arch repairs.7-14 With recurrent or residual arch obstruction a relatively rare event after extended arch repairs this study probably lacks the statistical power to identify multiple risk factors for arch obstruction. There are conflicting reports regarding the relative risk of low birth weight and prematurity in the genesis of recurrent or residual arch obstruction.18,19 In this study weight at operation was not a risk factor for recurrent or residual arch obstruction but premature infants were significantly at risk with only 70% remaining free from residual or recurrent arch obstruction at a year following surgery. An EETE repair through a lateral thoractomy was utilised in all of these infants and recoarctation was presumably related to both the size of the aorta and inadequate ductal tissue resection (ductal tissue is difficult to differentiate from normal aortic endothelium in these patients).

No residual or recurrent arch obstruction was found in the 50 patients undergoing extended end to side repair (EETS) through a median sternotomy under cardiopulmonary bypass. This approach was generally selected because of the need to address other intracardiac defects through a median sternotomy on cardiopulmonary bypass, although there was an increased prevalence of transverse arch hypoplasia in these patients. An EETS repair allows greater mobilisation and exposure of the aortic arch and therefore a more aggressive resection and hence a wider anastomosis. Although the recoarctation rate was not significantly different between this group and those patients undergoing repair using the extended end to end (EETE) approach (performed through a lateral thoracotomy), the complete absence of recoarctation after the more radical EETS arch repair supports the theory that the extent of resection and the size of the anastomosis are crucial in dealing with anatomical issues that may lead to recurrent or residual arch obstruction.14

Current clinical and echocardiographic criteria for the diagnosis of recurrent or residual arch obstruction have limitations. The oldest and most widespread definition for recoarctation is a 20-25 mmHg peak systolic upper to lower limb blood pressure gradient.20 Assessment of the repaired arch using this technique has two weaknesses. There is a relatively long distance between the two measuring sites, and this is of particular relevance as the arterial pressure wave changes throughout progression along the arterial bed.21 This leads to consistently higher blood pressure readings in the lower limb than the upper making this approach potentially inaccurate.21 In addition accurate blood pressure measurements are usually difficult in small children, in this
study despite efforts to carefully obtain BP measurements, a large proportion are likely to be inaccurate due to poor subject cooperation.

Echocardiography undoubtedly improves sensitivity in the diagnosis of residual or recurrent arch obstruction but for an accurate estimation of the degree of obstruction across the repair site an assessment of flow velocities just proximal to the stenosis is essential. In practice given the geometry of the transverse arch this measurement is very difficult to obtain. Nearly all reported series rely on gradients derived from Doppler interrogation across the site of repair alone which will inevitably lead to an over estimation of the true gradient. In addition work by De Mey et al using simulated models of the aortic arch after coarctation repair showed that increased Doppler velocities occurred as a manifestation of the change in compliance of the repaired vessel wall in the absence of stenosis. Therefore the large area of scarring related to extensive dissection in extended arch repairs may result in higher systolic descending aortic velocities in the absence of stenosis when compared other forms of arch repair potentially due changes in compliance over a long segment.

It is clear, therefore that individual criteria all have limitations. In this study all clinical and echocardiographic factors, not least clear 2D visualisation of the aortic arch were taken into account when assessing the repaired arch and we are convinced that no patient with important recurrent or residual arch obstruction was missed. It is interesting to note that in all patients requiring reintervention to the aortic arch the time to half peak diastolic velocity was >100 m/s whilst this measurement was less than 100 m/s for the rest of the group (mean 70 m/s). This was true also for the small number of our patients (10) who had systolic descending aortic velocities marginally in excess of 3 m/s but with no other evidence of recurrent arch obstruction (including cardiac catheter (1) or magnetic resonance imaging (3) showing a completely unobstructed aorta). Carvalho et al found diastolic parameters improved sensitivity and negative predictive value of echocardiography compared with single measurements of systolic velocity in patients with unrepaired coarctation. It is possible that this parameter may improve accuracy in the diagnosis of recurrent or residual arch obstruction after extended arch repair.

**Conclusion**

Survival after extended arch repair is excellent. At long follow up times recurrent or residual arch obstruction are rare with prematurity the only significant risk factor.

**Competing interests:** None
**Figure legend**

Figure 1: Extended end to end repair (see text for detailed description of surgical technique)

Figure 1a: Basic anatomy including lines of incision
- AAo  Ascending aorta
- DA   Ductus arteriosus
- DAo  Descending aorta

Figure 1b: Surgical procedure

Figure 1c: Final repair including lines of suture

Figure 2: Extended end to side repair (see text for detailed description of surgical technique)

Figure 2a: Basic anatomy including lines of incision
- AAo  Ascending aorta
- DA   Ductus arteriosus
- DAo  Descending aorta

Figure 2b: Surgical procedure

Figure 2c: Final repair including lines of suture

Figure 3: Kaplan-Meier survival curves

Figure 4: Freedom from reintervention (Kaplan-Meier) in term and premature infants
References

Figure 1

AAo
DA
DAo

a  b  c