Interactive CardioVascular and Thoracic Surgery 2022, 35(3), ivac135
https://doi.org/10.1093/icvts/ivac135 Advance Access publication 28 May 2022

ORIGINAL ARTICLE

CONGENITAL

Cite this article as: Patukale A, Shikata F, Marathe SS, Patel P, Marathe SP, Colen T *et al.* A single-centre, retrospective study of mid-term outcomes of aortic arch repair using a standardized resection and patch augmentation technique. Interact CardioVasc Thorac Surg 2022; doi:10.1093/icvts/ivac135.

# A single-centre, retrospective study of mid-term outcomes of aortic arch repair using a standardized resection and patch augmentation technique

Aditya Patukale [a,b], Fumiaki Shikata[c], Shilpa S. Marathe[a,b], Pervez Patel[a], Supreet P. Marathe [a,b,d], Timothy Colen [a,b], Prem Venugopal [a,b,d], QPCR Group[†] and NelsonAlphonso [a,b,d,*]

[a] Queensland Paediatric Cardiac Service (QPCS), Queensland Children's Hospital, Brisbane, QLD, Australia
[b] School of Clinical Medicine, Children's Health Queensland Clinical Unit, University of Queensland, Brisbane, QLD, Australia
[c] Kitasato University, Kanagawa, Japan
[d] Children's Health Research Centre, University of Queensland, Brisbane, QLD, Australia

* Corresponding author. Department of Cardiac Surgery, Clinical Directorate Level 7F, Queensland Children's Hospital, 501 Stanley Street, South Brisbane, QLD 4101, Australia. Tel: +61-7-3068-5775; e-mail: n.alphonso@uq.edu.au (N. Alphonso).

Received 11 March 2022; received in revised form 19 April 2022; accepted 27 May 2022

**Key question**

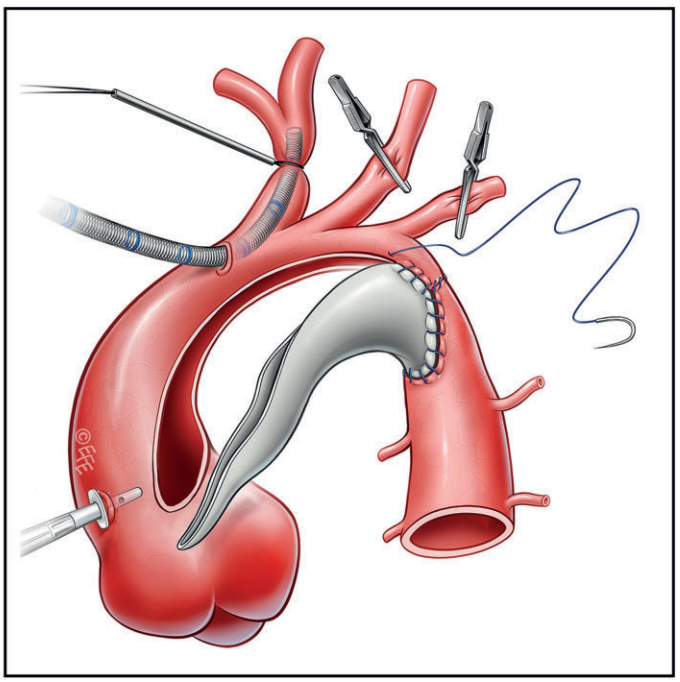
What are the outcomes after repair of aortic arch using a standardised patch augmentation technique?

**Key finding(s)**

Freedom from reintervention at 5 years was 93%. The technique is applicable irrespective of arch anatomy.

**Take-home message**

Repair of hypoplastic/interrupted aortic arch by excision of ductal tissue and standardised patch augmentation provides good mid-term durability.



## Abstract

**OBJECTIVES:** The aim of this study was to evaluate the mid-term outcomes after the repair of aortic arch using a standard patch augmentation technique.

[†]The collaborators in the QPCR Group are listed in the Acknowledgements section.

Presented at the 35th Annual Meeting of the European Association for Cardio-Thoracic Surgery, Barcelona, Spain, 13–16 October 2021.

**METHODS:** The study included all patients who underwent repair of a hypoplastic/interrupted aortic arch (IAA) in a single institute from June 2012 to December 2019 by a standardized patch augmentation (irrespective of concomitant intra-cardiac lesions). End points evaluated were reintervention for arch obstruction and persistent/new-onset hypertension.

**RESULTS:** The study included 149 patients [hypoplastic aortic arch, *n* = 92 (62%), IAA, *n* = 9 (6%), Norwood procedure, *n* = 48 (32%)]. The patch material used for augmentation of the aortic arch included pulmonary homograft (*n* = 120, 81%), homograft pericardium (*n* = 18, 12%), CardioCel® (*n* = 9, 6%) and glutaraldehyde-treated autologous pericardium (*n* = 2, 1%). The median age and weight at surgery were 7 days [interquartile range (IQR) 5–17 days] and 3.5 kg (IQR 3–3.9 kg), respectively. The median follow-up was 3.27 years (IQR 1.28, 5.08), range (0.02, 8.76). Freedom from reintervention at 1, 3 and 5 years was 95% [95% confidence interval (CI) = 89%, 98%], 93% (95% CI = 86%, 96%) and 93% (95% CI = 86%, 96%) respectively. One patient (0.6%) had persistent hypertension 8 years after correction for interrupted arch with truncus arteriosus.

**CONCLUSIONS:** Repair of hypoplastic/IAA by transection and excision of all ductal tissue and standardized patch augmentation provide good mid-term durability. The freedom from reintervention at 5 years is >90%. The incidence of persistent systemic hypertension following arch reconstruction is low. The technique is reproducible and applicable irrespective of underlying arch anatomy.

**Keywords:** Surgery for congenital heart disease • Hypoplastic aortic arch repair • Norwood procedure • Interrupted arch repair

**ABBREVIATIONS**

| | |
|---|---|
| ACP | Antegrade cerebral perfusion |
| CI | Confidence interval |
| CPB | Cardiopulmonary bypass |
| CT | Computed tomography |
| DHCA | Deep hypothermic circulatory arrest |
| DKS | Damus–Kaye–Stansel |
| HLHS | Hypoplastic left heart syndrome |
| IAA | Interrupted aortic arch |
| IQR | Interquartile range |

## INTRODUCTION

Hypoplasia of the aortic arch ranges from narrowing of the distal arch and isthmus near the insertion of the ductus arteriosus to hypoplasia of the entire aorta. The most extreme form includes complete interruption of the aortic arch (IAA). While coarctation of the aorta and hypoplasia of the distal arch are usually repaired via a thoracotomy approach avoiding the detrimental effects of cardiopulmonary bypass (CPB), hypoplasia involving the proximal aortic arch is usually repaired through a median sternotomy on CPB using hypothermia and circulatory arrest with or without antegrade cerebral perfusion (ACP). Repair of IAA may not always be possible by direct native aortic anastomosis and patch augmentation may be required for a tension free anastomosis. The goals of surgical repair of hypoplastic aortic arch are to completely relieve obstruction, while preserving growth potential of the repaired aorta and minimizing the risk of re-obstruction.

Recurrent arch obstruction is one of the most important complications after arch surgery and can lead to hypertension, systemic ventricle hypertrophy, raised pulmonary artery pressures and eventually, cardiac failure. The incidence of reported recurrent arch obstruction after univentricular and biventricular repair is 2–40% [1–4]. There is no established standard technique for repair of a hypoplastic arch.

In this study, we present the mid-term outcomes of our standardized patch augmentation technique for repair of hypoplastic/interrupted aortic arch (IAA) (central figure).

## METHODS

### Ethics approval

The study was approved by the hospital Human Research Ethics Committee (HREC number LNR/19/QCHQ/53807 dated 16 May 2019). The need for written consent was waived in view of retrospective nature of the study.

### Study design

We retrospectively identified all patients who underwent repair of a hypoplastic/IAA using a standardized patch augmentation technique in a single institute from June 2012 to December 2019. The study included patients undergoing biventricular repair as well as single ventricle palliation.

### Operative technique

The operative technique is illustrated in Fig. 1a–e. All procedures were performed via median sternotomy using CPB and deep hypothermia at 20°C with ACP. Deep hypothermic circulatory arrest was utilized for a short period during Norwood procedures as per the surgeon's preference. Arterial cannulation was performed either through the distal ascending aorta or a side graft sutured to the innominate artery, depending on the diameter of the ascending aorta. Venous cannulation was determined by the need for concomitant procedures. For isolated aortic arch repair, a single venous cannula was inserted into the right atrial appendage. The pulmonary and aortic ends of the ductus arteriosus were ligated after commencing CPB. The ductus was divided and extensive dissection of the arch, its branches and the descending thoracic aorta was performed while cooling. The descending thoracic aorta was dissected for at least 2 cm distal to the ductal insertion (Fig. 1a). ACP was performed at 20°C at a flow of 30 ml/kg/min. The arch was transected above and below ductal insertion, and all visible ductal tissue excised (Fig. 1a). The subclavian artery was sacrificed if there was ductal tissue encroaching into its origin. The descending thoracic aorta was splayed open medially for 8–10 mm and the corners trimmed. The undersurface of the arch was incised and the aortotomy carried along the lesser curvature of the arch down the medial side of the ascending aorta, stopping just above the sinotubular junction (Fig. 1b).

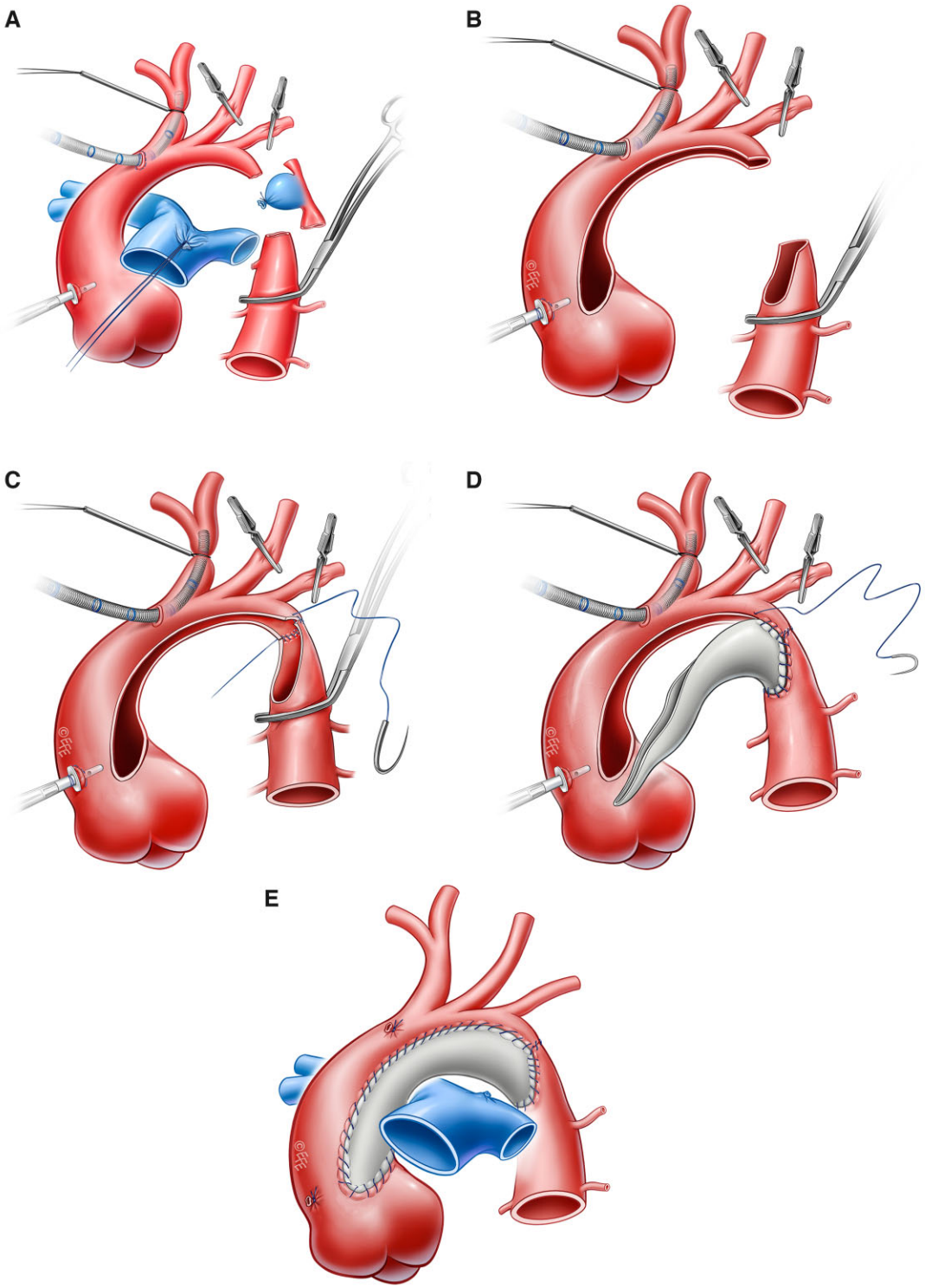



**Figure 1:** (**a**) Resection of all ductal tissue. (**b**) Incisions on the medial surface of the descending thoracic aorta and the under surface of the arch extending to the sino-tubular junction. (**c**) Native tissue anastomosis between the descending thoracic aorta and distal aortic arch posteriorly. (**d**) Augmentation of the descending thoracic aorta, under surface of the aortic arch and ascending aorta using a patch. (**e**) Completed repair.

The descending thoracic aorta was sutured to the distal arch posteriorly creating a posterior anastomosis of native tissue (Fig. 1c). A patch of tissue substitute, preferably pulmonary homograft, was trimmed to a finger shape approximately 10–12 mm wide with a rounded leading edge for the distal descending aorta. Other materials were used only when pulmonary homograft was not available. The patch was used to augment the descending thoracic aorta, under the surface of arch and the medial aspect of the ascending aorta. The arch reconstruction was started at the descending thoracic aorta progressing proximally towards the transverse arch and ascending aorta. The medial edge of the patch was tailored at the transition from the aortic arch to the

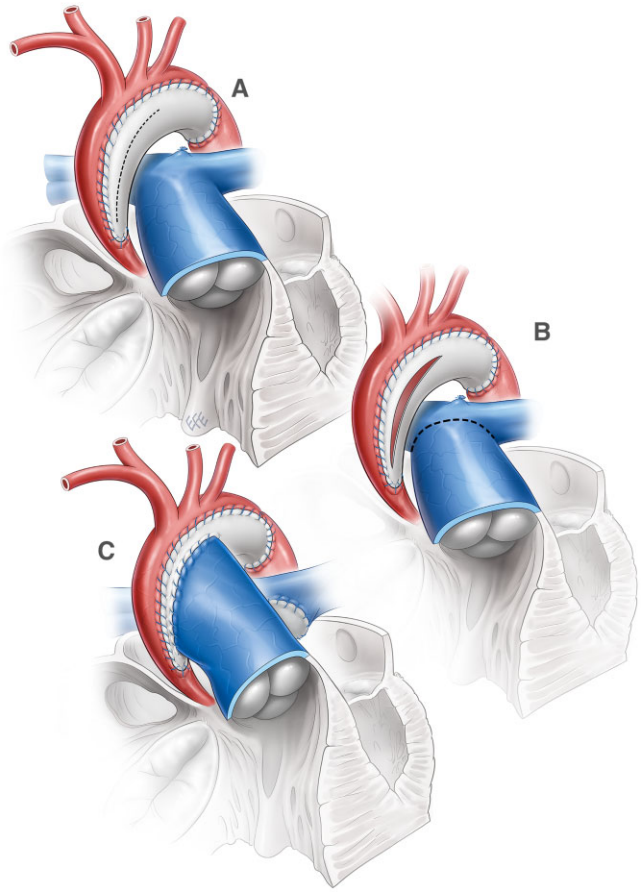



**Figure 2:** Creation of the Damus–Kaye–Stansel anastomosis. (**a**) Incision in the patch on the under surface of the reconstructed arch. (**b**) Division of pulmonary artery at the bifurcation. (**c**) End-to-side anastomosis of the main pulmonary artery to the incision in the patch.

ascending aorta to prevent kinking, twisting or redundancy (Fig. 1d).

For the Norwood procedure, the patch on the undersurface of the arch was incised and the distal end of the main pulmonary artery sutured to the opening in an end-to-side fashion to create the Damus–Kaye–Stansel (DKS) anastomosis (Fig. 2) [5]. The source of pulmonary blood flow was determined as per the cardiac anatomy.

Repair of IAA follows the same 3 principles of arch repair. All ductal tissue was resected. A native tissue anastomosis was created posteriorly between the medial edge of the descending thoracic aorta and the posterior edge of a counter incision along the medial surface of the distal ascending aorta. The descending aorta was splayed open laterally. One or 2 short incisions were made into the anterior margin of the ascending aortotomy to enlarge the opening and a similar finger shaped patch was used the augment the descending thoracic aorta and the anterior part of the anastomosis.

## Endpoints and definitions

Pre-operative evaluation of the aortic arch was performed using echocardiography, and more recently contrast enhanced computed tomography (CT). Hypoplastic aortic arch was defined as:

**Table 1:** Patient characteristics

| | |
|---|---|
| Total patients | 149 |
| Male, *n* (%) | 89 (60) |
| Diagnosis, *n* (%) | |
| Hypoplastic aortic arch | 92 (62) |
| Single ventricle morphology | 48 (32) |
| HLHS (aortic stenosis/mitral stenosis) | 26 |
| HLHS (aortic atresia/mitral atresia) | 6 |
| Unbalanced CAVSD | 6 |
| Double inlet left ventricle | 5 |
| HLHS (aortic stenosis/mitral atresia) | 2 |
| Tricuspid atresia + hypoplastic aortic arch | 2 |
| HLHS (aortic atresia/mitral stenosis) | 1 |
| Interrupted aortic arch | 9 (6) |
| Concomitant procedures | 123 |
| Damus–Kaye–Stansel anastomosis with BT shunt or RV–PA conduit | 48 |
| VSD closure | 34 |
| PA band | 12 |
| Division of subclavian artery | 7 |
| Arterial switch + VSD closure | 6 |
| Aorto-pulmonary window repair | 4 |
| Truncus arteriosus repair | 3 |
| Repair of total anomalous pulmonary venous drainage | 2 |
| Repair of partial anomalous pulmonary venous drainage | 2 |
| Repair of supra-valvular aortic stenosis | 2 |
| Reduction aortoplasty | 1 |
| Cortriatriatum repair | 1 |
| Arterial switch operation + PA band | 1 |
| Median age (days) (IQR) | 7 (5-17) |
| Median weight (kg) | 3.5 (3-3.9) |
| Median cardiopulmonary bypass time (min) | 153 (125-185) |
| Median myocardial ischaemia time (min) | 77 (53-98) |
| Median antegrade cerebral perfusion duration (min) | 42 (37-49) |
| Patch material, *n* (%) | |
| Pulmonary homograft | 120 (81) |
| Homograft pericardium | 18 (12) |
| Bovine pericardium (CardioCel®) | 9 (6) |
| Autologous pericardium | 2 (1) |
| Morbidity, *n* (%) | |
| Vocal cord dysfunction | 40 (27) |
| Chylothorax | 16 (11) |
| Acute renal failure requiring temporary peritoneal dialysis | 10 (7) |
| Stroke | 2 (1) |
| Re-exploration for bleeding | 1 (<1) |
| Mediastinitis | 1 (<1) |

BT: Blalock–Taussig; CAVSD: complete atrioventricular septal defect; HLHS: hypoplastic left heart syndrome; IQR: interquartile range; PA: pulmonary artery; RV: right ventricular; VSD: ventricular septal defect.

1. Diameter of the transverse aortic arch less than the patient's body weight at repair plus 1 mm [6] and/or
2. *z*-score of aortic arch diameter ≤-2.0.

The primary end point was reintervention (surgical or transcatheter) for recurrent obstruction of the aorta. Patients were also evaluated for persistent systemic hypertension after repair. Recurrent aortic arch obstruction was defined as a peak gradient >25 mmHg across the aortic arch by echocardiography along with obvious structural narrowing. Systemic hypertension was defined as systemic blood pressure >95th centile for the age and weight of the patient. Early death was defined as that occurring within 30 days of the index procedure or before hospital discharge.

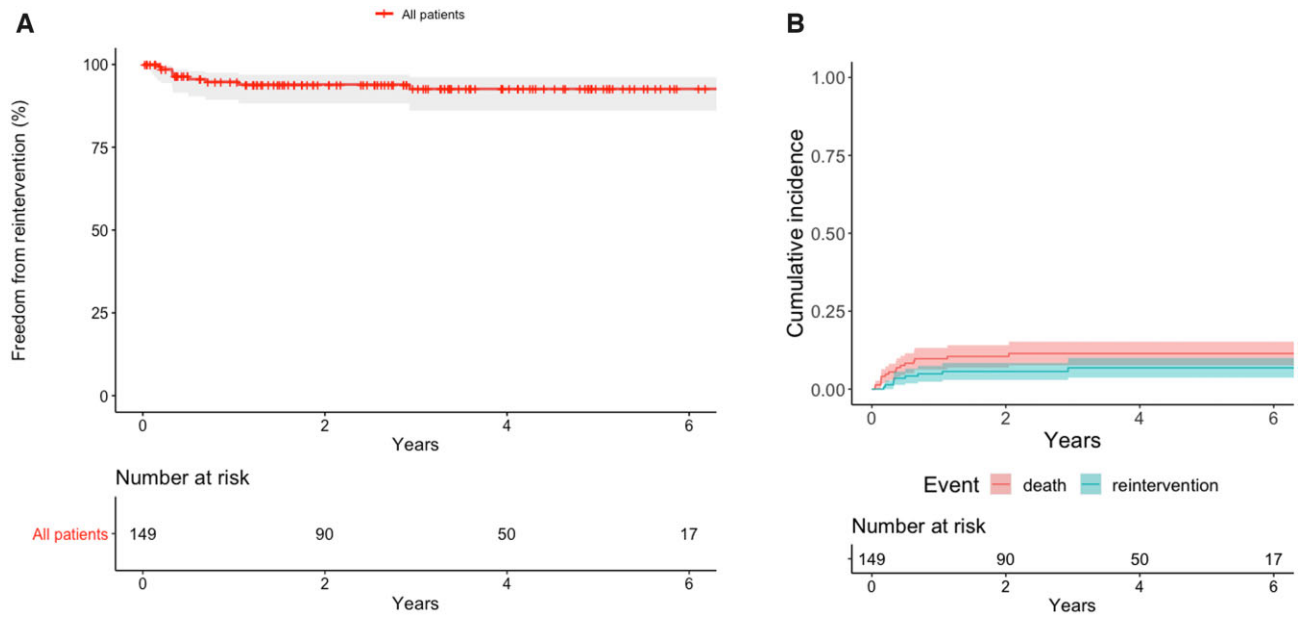



**Figure 3:** (**a**) Kaplan–Meier analysis for freedom from re-intervention. Freedom from reintervention at 1, 3 and 5 years was 95% (95% confidence interval = 89%, 98%), 93% (95% confidence interval = 86%, 96%) and 93% (95% confidence interval = 86%, 96%), respectively. (**b**) Kaplan–Meier analysis for a cumulative incidence function of the risks of reintervention and death. (Note: No patient with a reintervention died.)

## Follow-up

Perioperative and follow-up data including echocardiography for all patients was extracted from hospital medical records. Cardiac catheterization and/or CT were performed when recurrence of aortic obstruction was suspected. In addition, 2 cardiologists (Shilpa S. Marathe and Pervez Patel) reviewed the latest follow-up echocardiogram on every patient and measured the dimensions of all parts of the aorta and the gradient across the reconstructed arch. Aortic dimension z scores were calculated using criteria described by Lopez *et al.* [7]. All single ventricle pathway patients who underwent a BCPS had a CT angiogram or cardiac catheter study preoperatively. All patients who underwent a Fontan procedure had a cardiac catheter study preoperatively.

## Statistical analysis

Patient characteristics were summarized as mean and standard deviation for approximately normally distributed continuous variables, median and interquartile range (IQR) for non-normally distributed continuous variables, and percentages for categorical variables. Next, median and IQR was compared for several variables (patient age, weight, height at surgery, gender, and arch morphology, CPB time and myocardial ischaemia time) among the 3 morphology groups, with *P*-values calculated from a Kruskal–Wallis test. Univariable Cox regression was then used to test the associations between the patient characteristics and surgical factors with time to reintervention. Kaplan–Meier curves were also plotted to show the overall time to reintervention among the full sample, and to compare time to reintervention by morphology type and patch type (separately), with p-values for these comparisons tested with a log-rank test. We also performed a Kaplan–Meier analysis for a cumulative incidence function of the competing risks—reintervention and death. However, for all remaining plots and analyses, we censored patients at the time of their death.

# RESULTS

Between June 2012 and December 2019, 149 patients underwent repair of the aortic arch using the technique under evaluation. Baseline characteristics of all patients are shown in Table 1.

The median follow-up (clinical + echocardiographic) was 3.27 years (IQR 1.28, 5.08), range (0.02, 8.76). One patient (single ventricle palliation) was lost to follow-up after hospital discharge ($n = 140$). Follow-up was complete in 139 patients (139/140 early survivors; 99.3%).

## Deaths

There were 16 deaths (11%). The median interval from surgery to death was 3 months (IQR 2–6). In all 16 patients, the aortic arch was unobstructed on the latest echocardiogram prior to death. There were 9 (6%) early deaths and 7 (5%) late deaths. The causes of mortality are summarized in Supplementary Material, Tables S3 and S4. No death was related to aortic arch obstruction.

## Morbidity

Early morbidity is shown in Table 1. No patient had evidence of left main bronchus or left pulmonary artery compression.

## Reintervention

Nine patients (6%) underwent reintervention for recurrent aortic arch obstruction (Supplementary Material, Table S1). Five patients underwent patch augmentation of the distal arch [pulmonary homograft, $n = 3$, bovine pericardium (CardioCel®), $n = 2$]. Four patients underwent balloon dilatation after which 1 patient required further surgical augmentation and 1 patient underwent 2 further balloon dilatations. The median interval from index procedure to first reintervention was 4 months (IQR 4–9) with a

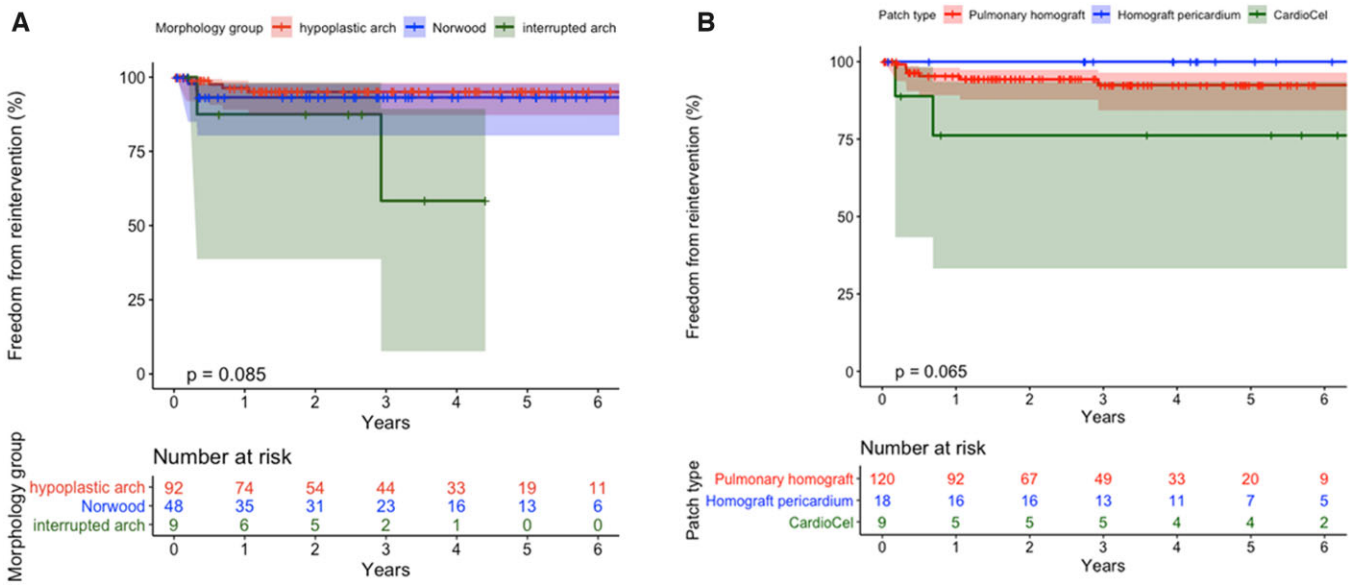



**Figure 4:** Kaplan–Meier analysis for freedom from reintervention stratified by. (**a**) Underlying morphology. (**b**) Patch material used for arch augmentation. (Note: The 2 patients with autologous pericardium were excluded.)

range of 2–35 months. Of the 9 patients who required reintervention, 4 patients had undergone a repair of the hypoplastic aortic arch, 3 had arch repair as a part of single ventricle palliation and 2 underwent IAA. All 3 patients with single ventricle palliation underwent aortic arch reintervention concurrently with stage II repair.

Follow-up details are given in Supplementary Material, Table S1. Freedom from reintervention at 1, 3 and 5 years was 95% [95% confidence interval (CI) = 89%, 98%], 93% (95% CI = 86%, 96%) and 93% (95% CI = 86%, 96%), respectively (Fig. 3a and b). Univariate analysis identified IAA to be associated with a higher incidence of reintervention ($P = 0.047$) (Supplementary Material, Table S2). Kaplan–Meier analysis of freedom from reintervention did not reveal significant differences based on the underlying arch morphology ($P = 0.085$) or type of patch material used for arch augmentation ($P = 0.065$) (Fig. 4).

## Aneurysm formation

There was no aneurysm of the pulmonary homograft patch noted intraoperatively during a subsequent operation or on any follow-up echocardiogram, CT angiogram or cardiac catheter study during the follow-up period.

## Norwood procedures

The outcomes after the Norwood procedure are summarized in Supplementary Material, Fig. S1. There was no gradient across the DKS anastomosis in any patient on echocardiography, CT angiography or cardiac catheter study preoperatively or during follow-up. No patient required revision of the DKS anastomosis (the longest follow-up of our Norwood cohort was 8.76 years).

## Hypertension

One patient (0.7%) had persistent hypertension 8 years after correction for IAA with truncus arteriosus.

## Echocardiographic follow-up

A total of 131 (88%) patients were evaluated for echocardiographic dimensions of the aorta during follow-up. The 9 (6%) patients who died before hospital discharge and 9 (6%) patients who underwent reintervention were excluded. Of these, 26 patients (18%) did not have a follow-up echocardiogram with measurements of the aortic arch after hospital discharge. Thus, follow-up echocardiograms suitable for evaluation of aortic dimensions and gradients were available in 105 patients (80%). Details of latest aortic dimensions are presented in Supplementary Material, Table S1.

The peak gradient at last follow-up ($n = 100$) was a median of 10 mmHg (IQR 8–13, range 3–37 mmHg). Eight patients had a peak gradient of >20 mmHg including 4 patients who had a peak gradient of >25 mmHg. However, all these patients had adequate transverse arch and isthmus dimensions ($z$ score >0.53) and no obvious anatomical obstruction.

# DISCUSSION

Our study demonstrates that a standardized technique of excision of ductal tissue and patch augmentation provides good mid-term durability with >90% freedom from reintervention at 5 years. Importantly, the technique is applicable irrespective of the underlying arch anatomy.

Surgery for coarctation of the aorta with aortic arch hypoplasia is invariably performed in the neonatal period. While extended end-to-end anastomosis in neonates with coarctation and distal arch hypoplasia has demonstrated 'catch-up' growth of the aortic arch [8] more extensive surgery via a sternotomy is required to address proximal arch hypoplasia [9]. Multiple techniques have evolved to repair the arch, with diverse opinions on the management of the coarctation segment and the posterior shelf at the isthmus as well as the use of patch augmentation versus 'autologous' or native tissue anastomoses.

Some studies have shown lower probability of restenosis after coarctectomy in univentricular and biventricular repairs [3, 10]

compared to those in which the posterior peri-ductal aorta is preserved [11, 12]. Histological evaluation of resected specimens of the isthmus in hypoplastic left heart syndrome (HLHS) have shown that ductal tissue can extend along the entire circumference of the aortic lumen as well as for variable lengths proximal and distal to the ductal orifice [13]. Techniques which include aggressive excision of periductal tissue have shown a similarly lower incidence of recurrence of arch obstruction [2, 3]. In contrast, others have reported that the type of aortic arch reconstruction did not affect the need for arch intervention [14]. The preoperative echocardiographic demonstration of a definite segment of coarctation or the presence of a posterior shelf is also difficult in cases with a large ductal insertion and turbulence which is often generated at this site [10]. We perform aggressive excision of the entire circumferential and visible longitudinal extent of ductal tissue (Class I recommendation: EACTS/APEC HLHS Task Force Guidelines) [15].

We sacrificed the left subclavian artery in 7 patients in whom the ductal tissue was encroaching into the subclavian artery origin to ensure the adequacy of excision of ductal tissue. Furthermore, the ductal tissue in the origin of the subclavian artery is fragile and can result in compromise of the integrity of the posterior anastomosis of the distal arch and the descending thoracic aorta. We did not note any acute ischaemic changes in our patients. Despite our extensive resection strategy, 1 patient who had the left subclavian artery sacrificed during a Norwood procedure had recurrent distal arch obstruction and underwent patch augmentation of the arch concomitantly with a bidirectional cavopulmonary shunt procedure. Data on patients in whom the left subclavian artery is sacrificed have shown that while anthropometric and blood pressure disparity is noted in the upper limbs after the subclavian artery is sacrificed, no acute or chronic functional, vascular or neurologic impairment is noted on long-term follow-up [16, 17].

In addition, the descending aorta is incised medially so that the augmentation of the aortic arch extends distally into the descending aorta (Class I recommendation as per EACTS/APEC HLHS Task Force Guidelines) [15]. Also, the distal end of the patch is rounded to minimize turbulence at the transition from the arch to the descending aorta.

The resultant geometry of the reconstructed arch has also shown to have important long- term consequences with a 'Romanesque' arch having the most favourable outcome [18]. Patch augmentation of the arch using a variety of materials [10, 12, 19, 20] preserves the geometry of the arch and prevents narrowing of the retro-aortic space and compression of the left pulmonary artery and left main bronchus. Recurrent arch obstruction after patch augmentation has been reported in 13–28% of patients [2, 3, 12, 19] and is thought to be partly due to the lack of growth potential of the patch material. Techniques potentially avoiding patch augmentation such as ascending aortic advancement [21] and the interdigitating technique [2, 3] for distal arch reconstruction combined with excision of the entire ductal segment have, in comparison, shown a lower recurrence of 0–3% after the Norwood procedure. These techniques may, however, occasionally require patch augmentation of the proximal aorta and ascending aorta. We prefer using pulmonary homograft because the natural curvature of the pulmonary bifurcation lends itself to be trimmed to resemble the lesser curvature of the arch. It is easy to handle, there is minimal bleeding through the suture holes and the thickness is similar to the native neonatal aorta. However, limited availability is the major drawback and was the main reason for the use of other substitutes in our patients. However, in our study, univariate analysis did not demonstrate patch material to be a predictor of restenosis.

While the haemodynamic benefit of augmenting the lesser curvature of the aorta has been previously debated, we have found that it resulted in a low recurrence of aortic arch obstruction, but also did not compromise the retro-aortic space and cause left pulmonary artery or left bronchial compression [12, 22]. The resulting 'Romanesque' geometry of the arch may also be the reason for better haemodynamics and a low incidence of hypertension in our series.

The incidence of recurrent arch obstruction has been reported to range from 2% to 40% [1–4, 23–25]. Recurrent aortic arch obstruction has been detected in the first few months after surgery in both univentricular and biventricular repairs, irrespective of the technique used for aortic arch repair [12, 14, 19, 26]. These observations indicate that recurrent aortic arch obstruction is likely to be influenced by the operative technique rather than the lack of growth potential of the native aorta [10]. We observed recurrent arch obstruction in 6% of our patients which is lower than the incidence of 13–14% seen when the posterior wall of the aorta opposite the ductal insertion is preserved [10, 12]. Most of our recurrent aortic obstructions were observed in the region of the distal arch, which may be a result of inadequate ductal resection.

In patients undergoing the Norwood procedure, we believe that by augmenting the entire ascending aorta till the sinotubular junction, the risk of coronary malperfusion is reduced even in cases with a diminutive ascending aorta [15]. Anastomosing the main pulmonary artery (DKS) distally to the patch on the lesser curve of the arch instead of the aortic root minimizes distortion of the aortic root which may impair coronary blood supply [15]. Our technique of constructing the DKS anastomosis is straightforward, reproducible and can be used irrespective of the size of the ascending aorta or the anatomy of the arch. There was no gradient across the DKS anastomosis in any patient on echocardiography, CT angiography or cardiac catheter study during follow-up. No patient has required a revision of the DKS anastomosis (the longest follow-up of our Norwood cohort was 8.76 years). Longer follow-up will be needed to ascertain the long-term risk of stenosis of the DKS anastomosis using this technique.

A CHSS (Congenital Heart Surgeons Society) study on IAA repair reported a lower mortality (18% vs 30%) and higher survival without reintervention (59% vs 47%) after repair using direct anastomosis with patch augmentation, compared to repair using only direct anastomosis [27]. Aortic arch advancement technique for IAA avoiding a patch augmentation demonstrated a freedom from reintervention of 100% at 5 years [28]. The incidence of reintervention following IAA repair was 22%, which compares favourably to previous reports. One failure was caused by intimal proliferation related to the CardioCel® patch used for augmentation of the anastomosis, and the other was related to incomplete resection of ductal tissue at the lower end of the patch.

The augmented native aorta has been shown to grow at a rate similar to the normal population [29]. The native aortic tissue has shown to be primarily responsible for this growth, with little contribution from the patch material [29]. Our technique incorporates the native aorta as a part of the circumference of the entire length of the aorta from the sinotubular junction to the descending thoracic aorta to provide the potential for growth. Follow-up echocardiograms of over two-thirds of our patient cohort have

shown adequate dimensions of all segments of the aortic arch (median *z* scores over 0).

Another long-term complication repair of aortic coarctation or hypoplastic aortic arch is systemic hypertension, which is seen in 10–20% of patients even after repair of coarctation of aorta in infancy [30]. Hypertension may be secondary to residual or recurrent arch obstruction or unfavourable geometry of the arch. The incidence of postoperative hypertension in our study was 0.6% though this must be taken in the context of a relatively short follow-up.

### Limitations

Our study bears all the drawbacks of a retrospective study. Follow-up evaluation was based on clinical examination and transthoracic echocardiography. Cardiac imaging studies and catheterization were performed only when there was a suspicion of re-obstruction and at the time of the different stages of the univentricular palliation. Blood pressure measurements were performed using cuff pressures during outpatient visits which are less accurate in children as compared to adults. Standard echocardiographic visualization and measurements of the reconstructed aorta, especially the distal arch, and consequently gradients across the arch were not available in all patients. Pulmonary homografts were used in 81% of patients, which impacts the validity of statistical comparison of the performance of the different patch materials. Finally, long-term follow-up information for many patients in our series is still not available.

## CONCLUSIONS

Repair of hypoplastic/IAA by transection above and below ductal insertion, excision of ductal tissue and standardized patch augmentation provide good mid-term durability. The freedom from reintervention at 5 years is >90%. The incidence of persistent systemic hypertension following arch reconstruction is low. The technique is reproducible and applicable irrespective of underlying arch anatomy.

## SUPPLEMENTARY MATERIAL

Supplementary material is available at *ICVTS* online.

## Funding

None.

**Conflict of interest:** The authors have no conflicts of interest to declare.

## ACKNOWLEDGEMENTS

The authors wish to thank our medical artist, Dr. Levent Efe CMI, for the illustrations in Figs. 1 and 2. The collaborators in Queensland Paediatric Cardiac Research (QPCR) Group are as follows: Jessica Suna, Kim Betts, Tom R Karl, Janelle Johnson and Kathryn Versluis.

## Data Availability Statement

The data underlying this article will be shared on reasonable request to the corresponding author.

## Author contributions

**Aditya Patukale:** Data curation; Formal analysis; Investigation; Methodology; Project administration; Writing—original draft; Writing—review & editing. **Fumiaki Shikata:** Data curation; Methodology. **Shilpa S. Marathe:** Data curation; Investigation; Validation. **Pervez Patel:** Data curation; Validation. **Supreet P. Marathe:** Data curation; Formal analysis; Project administration; Writing—original draft; Writing—review & editing. **Timothy Colen:** Resources; Supervision; Validation; Writing—review & editing. **Prem Venugopal:** Conceptualization; Formal analysis; Methodology; Project administration; Resources; Supervision; Validation; Writing—review & editing. **QPCR Group:** Data curation; Formal analysis; Investigation; Project administration; Software; Writing—review & editing. **Nelson Alphonso:** Conceptualization; Formal analysis; Investigation; Methodology; Project administration; Resources; Supervision; Validation; Writing—original draft; Writing—review & editing.

## Reviewer information

Interactive CardioVascular and Thoracic Surgery thanks Carl L. Backer, Katarzyna Januszewska, Hitendu Hasmukhlal Dave and the other, anonymous reviewer(s) for their contribution to the peer review process of this article.

## REFERENCES

[1] Zeltser I, Menteer J, Gaynor JW, Spray TL, Clark BJ, Kreutzer J *et al.* Impact of re-coarctation following the Norwood operation on survival in the balloon angioplasty era. J Am Coll Cardiol 2005;45:1844–8.

[2] Lamers LJ, Frommelt PC, Mussatto KA, Jaquiss RDB, Mitchell ME, Tweddell JS. Coarctectomy combined with an interdigitating arch reconstruction results in a lower incidence of recurrent arch obstruction after the Norwood procedure than coarctectomy alone. J Thorac Cardiovasc Surg 2012;143:1098–102.

[3] Burkhart HM, Ashburn DA, Konstantinov IE, De Oliveira NC, Benson L, Williams WG *et al.* Interdigitating arch reconstruction eliminates recurrent coarctation after the Norwood procedure. J Thorac Cardiovasc Surg 2005;130:61–5.

[4] Ishino K, Stümper O, De Giovanni JJ, Silove ED, Wright JG, Sethia B *et al.* The modified Norwood procedure for hypoplastic left heart syndrome: early to intermediate results of 120 patients with particular reference to aortic arch repair. J Thorac Cardiovasc Surg 1999;117:920–30.

[5] Sakurai T, Rogers V, Stickley J, Khan N, Jones TJ, Barron DJ *et al.* Single-center experience of arch reconstruction in the setting of Norwood operation. Ann Thorac Surg 2012;94:1534–9.

[6] Karl TR, Sano S, Brawn W, Mee RB. Repair of hypoplastic or interrupted aortic arch via sternotomy. J Thorac Cardiovasc Surg 1992;104:688–95.

[7] Lopez L, Colan S, Stylianou M, Granger S, Trachtenberg F, Frommelt P *et al.* Relationship of echocardiographic Z scores adjusted for body surface area to age, sex, race, and ethnicity: the Pediatric Heart Network Normal Echocardiogram Database. Circ Cardiovasc Imaging 2017;10:e006979.

[8] Karamlou T, Bernasconi A, Jaeggi E, Alhabshan F, Williams WG, Van Arsdell GS *et al.* Factors associated with arch reintervention and growth of the aortic arch after coarctation repair in neonates weighing less than 2.5 kg. J Thorac Cardiovasc Surg 2009;137:1163–7.

[9] Tulzer A, Mair R, Kreuzer M, Tulzer G. Outcome of aortic arch reconstruction in infants with coarctation: importance of operative approach. J Thorac Cardiovasc Surg 2016;152:1506–13.e1.

[10] Bautista-Hernandez V, Marx GR, Gauvreau K, Pigula FA, Bacha EA, Mayer JE Jr, *et al.* Coarctectomy reduces neoaortic arch obstruction in hypoplastic left heart syndrome. J Thorac Cardiovasc Surg 2007;133:1540–6.

[11] Cleuziou J, Kasnar-Samprec J, Hörer J, Eicken A, Lange R, Schreiber C. Recoarctation after the Norwood I procedure for hypoplastic left heart

syndrome: incidence, risk factors, and treatment options. Ann Thorac Surg 2013;95:935–40.
[12] Ghani MOA, Raees MA, Harris GR, Shannon CN, Nicholson GT, Bichell DP. Reintervention after infant aortic arch repair using a tailored autologous pericardial patch. Ann Thorac Surg 2021;111:973–9.
[13] Machii M, Becker AE. Nature of coarctation in hypoplastic left heart syndrome. Ann Thorac Surg 1995;59:1491–4.
[14] Hill KD, Rhodes JF, Aiyagari R, Baker GH, Bergersen L, Chai PJ *et al.* Intervention for recoarctation in the single ventricle reconstruction trial: incidence, risk, and outcomes. Circulation 2013;128:954–61.
[15] Alphonso N, Angelini A, Barron DJ, Bellsham-Revell H, Blom NA, Brown K *et al.*; Chaiman HLHS Guidelines Task Force. Guidelines for the management of neonates and infants with hypoplastic left heart syndrome: the European Association for Cardio-Thoracic Surgery (EACTS) and the Association for European Paediatric and Congenital Cardiology (AEPC) Hypoplastic Left Heart Syndrome Guidelines Task Force. Eur J Cardiothorac Surg 2020;58:416–99.
[16] Barreiro CJ, Ellison TA, Williams JA, Durr ML, Cameron DE, Vricella LA. Subclavian flap aortoplasty: still a safe, reproducible, and effective treatment for infant coarctation. Eur J Cardiothorac Surg 2007;31:649–53.
[17] Pandey R, Jackson M, Ajab S, Gladman G, Pozzi M. Subclavian flap repair: review of 399 patients at median follow-up of fourteen years. Ann Thorac Surg 2006;81:1420–8.
[18] Ou P, Celermajer DS, Mousseaux E, Giron A, Aggoun Y, Szezepanski I *et al.* Vascular remodeling after "successful" repair of coarctation: impact of aortic arch geometry. J Am Coll Cardiol 2007;49:883–90.
[19] Morell VO, Wearden PA. Experience with bovine pericardium for the reconstruction of the aortic arch in patients undergoing a Norwood procedure. Ann Thorac Surg 2007;84:1312–5.
[20] Roussin R, Belli E, Lacour-Gayet F, Godart F, Rey C, Bruniaux J *et al.* Aortic arch reconstruction with pulmonary autograft patch aortoplasty. J Thorac Cardiovasc Surg 2002;123:443–8; discussion 9-50.
[21] Mery CM, Guzmán-Pruneda FA, Carberry KE, Watrin CH, McChesney GR, Chan JG *et al.* Aortic arch advancement for aortic coarctation and hypoplastic aortic arch in neonates and infants. Ann Thorac Surg 2014; 98:625–33, discussion 33.
[22] Hasegawa T, Oshima Y, Maruo A, Matsuhisa H, Tanaka A, Noda R *et al.* Aortic arch geometry after the Norwood procedure: the value of arch angle augmentation. J Thorac Cardiovasc Surg 2015;150:358–66.
[23] Tworetzky W, McElhinney DB, Burch GH, Teitel DF, Moore P. Balloon arterioplasty of recurrent coarctation after the modified Norwood procedure in infants. Cathet Cardiovasc Intervent 2000;50:54–8.
[24] Zellers TM. Balloon angioplasty for recurrent coarctation of the aorta in patients following staged palliation for hypoplastic left heart syndrome. Am J Cardiol 1999;84:231–3.a9.
[25] Wong JSY, Lee MGY, Brink J, Konstantinov IE, Brizard CP, d'Udekem Y. Are more extensive procedures warranted at the time of aortic arch reoperation? Eur J Cardiothorac Surg 2017;52:1132–8.
[26] Whiteside W, Hancock HS, Pasquali SK, Yu S, Armstrong AK, Menchaca A *et al.* Recurrent coarctation after neonatal univentricular and biventricular Norwood-type arch reconstruction. Ann Thorac Surg 2016; 102:2087–94.
[27] McCrindle BW, Tchervenkov CI, Konstantinov IE, Williams WG, Neirotti RA, Jacobs ML *et al.*; Congenital Heart Surgeons Society. Risk factors associated with mortality and interventions in 472 neonates with interrupted aortic arch: a Congenital Heart Surgeons Society study. J Thorac Cardiovasc Surg 2005;129:343–50.
[28] Morales DL, Scully PT, Braud BE, Booth JH, Graves DE, Heinle JS *et al.* Interrupted aortic arch repair: aortic arch advancement without a patch minimizes arch reinterventions. Ann Thorac Surg 2006;82:1577–83, discussion 83–4.
[29] Mahle WT, Rychik J, Weinberg PM, Cohen MS. Growth characteristics of the aortic arch after the Norwood operation. J Am Coll Cardiol 1998;32: 1951–4.
[30] Gillett C, Wong A, Wilson DG, Wolf AR, Martin RP, Kenny D. Underrecognition of elevated blood pressure readings in children after early repair of coarctation of the aorta. Pediatr Cardiol 2011;32:202–5.