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Prevalence and prognostic implication of restenosis or dilatation at the aortic coarctation repair site assessed by cardiovascular MRI in adult patients late after coarctation repair☆

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ABSTRACT

Background: Cardiovascular magnetic resonance (CMR) is ideal for assessing patients with repaired aortic coarctation (CoA). Little is known on the relation between long-term complications of CoA repair as assessed by CMR and clinical outcome. We examined the prevalence of restenosis and dilatation at the repair site and the long-term outcome in patients with repaired CoA.

Methods and results: CMR imaging and clinical data for adult CoA patients (247 patients aged 33.0 ± 12.8 years, 60% male), were analyzed. The diameter of the aorta at the repair site was measured on CMR and its ratio to the aortic diameter at the diaphragm (repair site-diaphragm ratio, RDR) was calculated. Restenosis (RDR ≤ 70%) was present in 31% of patients (and significant in 9% [RDR < 50%]), and dilatation (RDR > 150%) in 13.0%. A discrete aneurysm at the repair site was observed in 9%. Restenosis was more likely after resection and end–end anastomosis, whereas dilatation after patch repair. Systemic hypertension was present in 69% of patients. Of the hypertensive patients, blood pressure (133 ± 20/73 ± 10 mm Hg) was well controlled in 93% with antihypertensive therapy. Mortality rate over a median length of 5.9 years was low (0.69% per year, 95% CI: 0.33–1.26), but significantly higher than age-matched healthy controls (standardised mortality ratio 2.86, CI 1.43–5.72, $p < 0.001$).

Conclusion: Restenosis or dilatation at the CoA repair site as assessed by CMR is not uncommon. Medium term survival remains good, however, albeit lower than in the general population. Life-long follow-up and optimal blood pressure control are likely to secure a good longer term outlook in these patients.

1. Introduction

Coarctation of the aorta (CoA) accounts for up to 10% of all congenital heart defects. It is associated with premature morbidity and mortality if left untreated even after repair. Restenosis and aneurysmal dilatation at the level of the repair site have been recognized as complications after repair [1–4], their precise prevalence and potential adverse impact on outcome are largely unknown.

Cardiovascular magnetic resonance (CMR) has been widely used in recent years for assessing aortic coarctation, whether native or after repair and isolated or with coexisting lesions [5–8]. The lack of radiation exposure makes it particularly suitable for periodic follow-up in this relatively young patient population.

In this study, we examined the prevalence of restenosis and dilatation at the repair site, as assessed by CMR, in a large single centre contemporary adult population. We also investigated the potential relation between these 2 complications and clinical outcomes.

2. Methods

Adult patients with previous repair of CoA from our CMR database were identified and their earliest available CMR (the first set of CMR data in this present study was in 2004) was examined by 1 blinded (to patient characteristics and outcome) experienced investigator for restenosis and/or dilatation at the repair site. Patients with CoA are routinely referred for CMR as part of our clinical protocol, unless there are contraindications for it. As our study was a retrospective review of existing CMR data and medical records, individual patient consent was not required but our Ethics Committee approved it.

All CMR studies were performed on a 1.5 T scanner (Siemens Sonata or Siemens Avanto, Siemens Medical Solutions, Erlangen, Germany; maximum gradient field strength, 40 mT/m; slew rate, 200 T m^{-1} s^{-1} on each axis independently) using an 8-element phased-array receiver coil. In all studies, images of the aorta were acquired using ECG-

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gated single shot fast spin echo (multi-slice HASTE, matrix 2.3 × 1.3 × 6.0 mm), steady state free precession gradient echo cine (matrix 1.7 × 1.7 × 7.0 mm), and phase contrast flow mapping (matrix 2.5 × 1.3 mm) sequences. Further interrogation of the aorta, when needed, was achieved using turbo-spin echo T2 (matrix 2.2 × 1.3 × 6.0 mm) images (used in those with stent implantation who had metallic artefact on cine-imaging) and contrast enhanced magnetic resonance angiography (matrix 1.1 × 0.9 × 1.3 mm, used if there was significant stenosis or dilatation at the repair site). Stenosis or dilatation at the site of repair was determined by comparing the aortic diameter at the repair site relative to the aortic diameter at the level of the diaphragm, the "Repair site-diaphragm Ratio", or "RDR" and expressed as a percentage (Fig. 1). Any stenosis was defined as RDR ≤ 70% or less and significant (moderate-severe) stenosis as RDR ≤ 50% (Fig. 1, panel A). Dilatation was defined as RDR > 150% [9] (Fig. 1, panel B). Discrete dilatation or aneurysm formation was defined as a "bulge" at the site of repair. The dimensions of discrete dilatations were measured in 2 orthogonal orientations and the largest diameter was used in our analyses (Fig. 1, panel C). A routine set of LV and RV short-axis cine images 7 mm slice thickness, was acquired at 10 mm intervals from base to apex using a breath-hold retrospective vector cardiography-gated balanced steady state free precession (SSFP) gradient echo sequence. Volumetric analysis including left ventricular ejection fraction and mass (indexed to body surface area) was performed offline using CMRtools (Cardiovascular Imaging Solutions, UK).

Hypertension was defined as a resting cuff blood pressure of >140/90 mm Hg on at least 3 occasions or 24-hour ambulatory blood pressure >135/85 mm Hg. Anti-hypertensive therapy was increased (addition of another drug or increase in the dose of the current drug) if the patient was found to be persistently hypertensive despite current therapy. Date of death was extracted from our institution's database which is linked to the Office of National Statistics. Attempts were made to identify the mode of death, which was obtained for patients who died in-hospital and from available medical records.

3. Statistical analysis

Statistical analysis was performed using R version 2.15.1 (R Foundation for Statistical Computing, www.R-project.org). Continuous variables are expressed as mean ± SD, or median [range], while categorical variables as numbers (percentages). Comparison between groups was performed using Wilcoxon rank sum test or chi-square test as appropriate. The association between stenosis or dilatation and clinical characteristics was assessed using univariate logistic regression analysis. Standardised mortality ratio was estimated using data available for the UK population. Univariate Cox proportional hazards survival analysis was performed to identify the relation between death, hospitalisation or the composite endpoint of death or hospitalisation and clinical parameters (restenosis, dilatation, systemic hypertension, left ventricular ejection fraction and mass indexed to body area). Only hospitalisation events beyond the first year from CMR were considered in the analyses, to avoid hospitalisations, which occurred as a direct result of CMR. For all analyses, a p-value < 0.05 was considered indicative of statistical significance.

4. Results

4.1. Demographics

CMR and clinical data of 247 patients (mean age 33.0 ± 12.8, 59.5% male) were analysed. Demographic and clinical characteristics of these patients are presented in Table 1.

4.2. First repair

In the vast majority of patients, primary repair of coarctation was surgical (81.4%, Table 1). The most common type of initial surgical repair was resection and end-end anastomosis (E–E, n = 77, 31.1%), followed by subclavian flap repair (SCF, n = 53, 21.5%, Fig. 2). Of the 30 patients who underwent a percutaneous procedure as primary repair, 26 (87.6%) received primary stenting (Fig. 2C). Patients with a primary repair before 1980 were more likely to have stenosis or dilatation, (chi-square p = 0.04). Patients born after 1980 were significantly more likely to have undergone SCF (n = 35, 14.2%) or E-E (n = 32, 13.4%). Very young children who underwent first repair at the age of 2 years or less (n = 78, 31.5%) were more likely to have received SCF (n = 38) followed by E–E (n = 25), whereas older children (3–16 years, n = 71, 28.7%) were more likely to have received E–E (n = 34). Patients who were older at first repair (>16 years, n = 72, 29.1%) commonly received primary percutaneous coarctation stenting (n = 26) and, when surgically repaired, interposition graft repair (n = 12). The age/type of initial repair was not available in 26 (10%) patients with subsequent repairs, most having undergone surgery in the 1950–70s. The type of repair may be identified without previous records if it was a SCF, ascending-descending aortic bypass or interposition graft repair, but E–E and patch repairs may be difficult to recognise.

4.3. Re-intervention and most recent repair

One quarter of our repaired patients had required subsequent surgical or percutaneous re-interventions (Table 1, Fig. 3). Patients with a primary repair performed prior to the median year of repair (1985) were not at a higher risk of reoperation. Patients who had needed subsequent interventions had been significantly younger at first repair compared to those with a single operation (p < 0.0001, Fig. 4). At the time of CMR, the most recent type of repair overall was surgical (E–E in the majority of cases, Table 2).

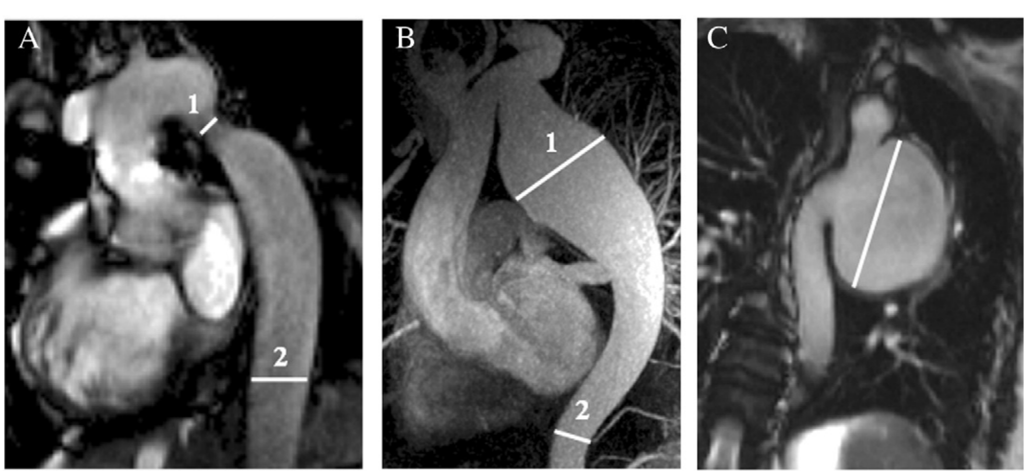

Fig. 1. Repair site-diaphragm ratio (RDR). The RDR is defined as the diameter at the repair site [1] relative to the aortic diameter at the level of the diaphragm [2]. RDR measurement in patients with stenosis (panel A) and dilatation (panel B) at the repair site. Measurement of the maximal diameter of discrete "bulging" at the repaired site (panel C).