

32 year follow up of patients following atrial redirection surgery for transposition of the great arteries

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Abstract

Objective: A significant body of patients who have undergone Mustard or Senning procedure require lifelong follow up. In this retrospective review, we examined the cohort of such patients currently attending our center.

Design: Patients who had undergone either Mustard or Senning procedure were identified. We retrospectively reviewed medical records, recorded demographic information and data regarding the clinical state, NHYA class, cardiopulmonary exercise testing, NT-proBNP measurement, and recent cardiac MRI findings.

Results: Forty-six patients were identified, the mean age was 32.2 years (± 6.1 years), 67.4% were male. Thirty-two patients (69.6%) had undergone a Senning procedure. The median length of the follow-up was 32 years. Thirty-two patients (69.6%) were NHYA class 1. The mean VO_{2max} achieved was 24.2 ± 5.8 mL/min/kg. The mean NT-proBNP was 266.4 pg/mL (± 259.9 pg/mL). The mean right ventricular end-diastolic volume (RVEDV) was $212.4 \text{ mL} \pm 73.1 \text{ mL}$ (indexed $114.2 \text{ mL/m}^2 \pm 34.4 \text{ mL/m}^2$). The mean right ventricular ejection fraction (RVEF) was $53.7\% \pm 7.9\%$. The mean left ventricular end-diastolic volume (LVEDV) was $161.5 \text{ mL} \pm 73.7 \text{ mL}$ (indexed $87.8 \text{ mL/m}^2 \pm 41.1 \text{ mL/m}^2$). The mean left ventricular ejection fraction (LVEF) was $59.8\% \pm 5.7\%$. There was a significant correlation between right ventricular (RV) size on MRI and NT-proBNP level.

Conclusions: We present a relatively well cohort of patients with overall favorable long-term outcome. The majority of patients are NHYA class 1 and the systemic right ventricular function appears to be well preserved as assessed by MRI. The exercise tolerance is reduced, with the majority of patients achieving around 60% of the estimated VO_{2max} . Regular specialist follow-up and assessment with advanced imaging at regular intervals remain important for this group.

KEYWORDS

adult congenital heart disease, Mustard procedure, outcome, Senning procedure, transposition

1 | INTRODUCTION

Transposition of the great arteries (TGA) represents the second most common cyanotic congenital heart defect with a prevalence of 0.2 per 1000 live births worldwide.¹ Until the late 1950s, the outlook for patients with TGA was poor; with 55%, 85%, and 90% mortality at 1 month, 6 months, and 1 year.^{1,2} The atrial switch, first performed in 1958 by Ake Senning and adapted by Mustard in 1963, revolutionized their survival and for over 30 years the atrial redirection surgery was the principal palliation for patients with this diagnosis.¹

Although now superseded by the arterial switch operation, there remains a significant body of patients who have undergone Mustard or Senning procedure that require a follow-up.¹ Although the population will diminish over the next 40 years or so they represent a clinical challenge. During adult life, such patients are at increased risk of complications; including exercise intolerance, ventricular dysfunction, arrhythmia, tricuspid valve insufficiency, problems relating to the intra-atrial baffles, and sudden cardiac death. Failure of the systemic right ventricle is a major determinant of morbidity and mortality.³ In this retrospective review, we examined the clinical progress of our current cohort of patients who have undergone atrial redirection surgery for TGA currently attending the Belfast Trust for the follow-up and related this to cardiopulmonary exercise testing, serum NT-pro BNP measurement, and cardiac MRI data.

2 | METHODS

The adult congenital heart disease (ACHD) population in Northern Ireland comprises around 4000 patients almost all of who visited The Royal Victoria Hospital, Belfast Trust, which is the tertiary center for adult congenital heart disease in the province. We identified patients currently attending the adult congenital heart disease service who had undergone either Mustard or Senning procedure for TGA from the patient management system, HeartSuite (2002–2009 Systeria Ltd, Glasgow, UK). We excluded those who had a Rastelli correction or a palliative Mustard procedure. Medical records were retrospectively reviewed, and demographic information and data regarding clinical state were recorded. NHYA class, cardiopulmonary exercise testing, NT-pro BNP measurement, and most recent echocardiography and cardiac MRI findings were recorded.

Data obtained were analyzed using the IBM SPSS statistics version 21.0 for Mac (IBM Corp 2012). Continuous data were expressed as mean and standard deviation. The frequency was expressed as percentage. An unblinded correlation analysis was performed to examine associations between right ventricular parameters on MRI, VO_{2max} , and NT-pro BNP.

3 | RESULTS

3.1 | Background data

Forty-six patients were identified, the mean age was 32.2 years (\pm 6.1 years), 67.4% were male. The mean age at surgery was 11.2 months

(\pm 6.8 months), with thirty-two patients (69.6%) who had undergone a Senning procedure. The background and demographic data is presented in Table 1. Almost all patients had simple transposition; only 1 patient had TGA with ventricular septal defect. The median length of the follow-up was 32 years. The majority of patients (69.6%) had remained in sinus rhythm, though 21.7% of patients had a history of either atrial or ventricular arrhythmia and nine patients (19.5%) had an implantable pacemaker or implantable cardiac defibrillator in situ. Fourteen patients (30.4%) received some form of angiotensin-converting-enzyme inhibitor and three patients (6.5%) received some form of angiotensin-receptor blocker. Digoxin and beta blocker were initiated as required for management of arrhythmia and heart failure. Seventeen patients (36.9%) were on no cardiovascular medication at all.

3.2 | Exercise data

CPET indices are given in Table 1. Thirty-two patients (69.6%) were NHYA class 1. Eleven patients (23.9%) were NHYA class 2 and (6.5%) were NHYA class 3. The mean VO_{2max} achieved was 24.2 ± 5.8 mL/min/kg. This was $61.3\% \pm 12.4\%$ of the estimated VO_{2max} for the group.

TABLE 1 Background and demographic characteristics

Characteristic	Mean \pm SD
Age (years)	32.2 \pm 6.1
Age at surgery (months)	11.2 \pm 6.8
Height (cm) (n = 39)	169.4 \pm 10.0
Weight (kg) (n = 42)	75.9 \pm 18.4
BMI (n = 39)	26.1 \pm 5.2
NT-proBNP (pg/mL) (n = 36)	266.4 \pm 259.9
VO_{2max} (n = 27)	24.2 \pm 5.9
% VO_{2max} (n = 27)	61.3 \pm 12.4
Respiratory exchange ratio	1.2 \pm 0.1
Anaerobic threshold	14.2 \pm 3.9
RVEDV (n = 32)	212.4 \pm 73.1
RVEDVi	114.2 \pm 34.4
RVESV	101.2 \pm 53.2
RVESVi	55.2 \pm 26.8
RVEF (n = 32)	53.7 \pm 7.9
LVEDV (n = 32)	161.5 \pm 73.7
LVEDVi	87.8 \pm 73.7
LVESV	67.9 \pm 30.3
LVESVi	35.7 \pm 16.6
LVEF (n = 32)	59.8 \pm 5.7

Abbreviations: BMI, body mass index; LVEF, left ventricular ejection fraction; LVEDV, left ventricular end-diastolic volume; LVESV, left ventricular end systolic volume; LVEDVi, left ventricular end-diastolic volume indexed; LVESVi, left ventricular end systolic volume indexed; RVEDV, right ventricular end-diastolic volume; RVEDVi, right ventricular end-diastolic volume indexed; RVESV, right ventricular end systolic volume; RVESVi, right ventricular end systolic volume indexed; and RVEF, right ventricular ejection fraction.

3.3 | NT-pro BNP

NT-proBNP ranged from 34 pg/mL to 1340 pg/mL. The mean NT-proBNP was 266.4 pg/mL (\pm 259.9 pg/mL). Sixteen patients had an NT-proBNP value of more than 200 pg/mL.

3.4 | Echocardiography

Right ventricular systolic function and tricuspid regurgitation were graded qualitatively. Results are presented in Table 2.

3.5 | CMR

Thirty-two patients (69.6%) had undergone at least one recent cardiac MRI. Nine patients could not be scanned due to an implantable device in situ and two further patients could not undergo CMR due to other comorbidities (cerebral palsy and psychiatric illness, respectively). Three patients had refused to attend their offered appointments for MRI.

The mean right ventricular end-diastolic volume was 212.4 mL \pm 73.1 mL (indexed 114.2 mL/m² \pm 34.4 mL/m²). The mean right ventricular ejection fraction was 53.7% \pm 7.9%. The mean left ventricular end-diastolic volume was 161.5 mL \pm 73.7 mL (indexed 87.8 mL/m² \pm 41.1 mL/m²). The mean left ventricular ejection fraction was 59.8% \pm 5.7%. Complete MRI parameters are presented in Table 1.

There was no evidence of baffle obstruction on MRI. There was no delayed enhancement to suggest myocardial fibrosis. Four patients (12.5%) were found to have significant baffle leaks. Of note, all 4 patients were NHYA class 1 with preserved right ventricular function. Their data are shown separately in Table 4 (Supplemental data).

3.6 | Correlation data

The echocardiographic assessment of right ventricular systolic function demonstrated a positive correlation with NHYA class, the correlation coefficient was 0.32 (*P* value .04).

There was a significant correlation between the right ventricular size on MRI and NT-proBNP level (Table 3). There was no correlation between MRI parameters and the indices obtained on cardiopulmonary exercise testing. There was no relationship between VO_{2max} and NT-proBNP level (Table 3). Scatter plots of positive correlations are shown in Figure 1.

4 | DISCUSSION

For the past two decades the arterial switch operation has replaced the atrial redirection surgery for TGA meaning that the population post Mustard or Senning procedure will diminish.⁴ Until then, however, there is still a need to understand what determines morbidity and mortality in this patient group. This may provide a valuable window of opportunity to commence therapeutic strategies before the onset of systemic right ventricular decline and ultimately irreversible right ventricular dysfunction. This group also provides valuable information regarding how the right ventricle copes in the systemic position, which is applicable to patients with a diagnosis of congenitally corrected transposition. The atrial redirection surgery has become a palliative procedure for patients not suitable for the arterial switch but the Senning procedure itself has emerged as an essential part of the anatomic repair for congenitally corrected transposition and similar long-term complications may be anticipated.⁵

We present a relatively well cohort of adult patients who have undergone the atrial redirection surgery for TGA. In this cohort, the majority of patients are NHYA class 1 and while they do exhibit dilated right ventricles as expected, the right ventricular function appears to be well preserved as assessed by MRI. However, this exercise tolerance, as assessed by objective measures, is reduced with the majority of patients achieving around 60% of estimated VO_{2max}.

Authors have previously demonstrated that subjective measures of the functional class such as NHYA class tend to underestimate the degree of impairment experienced by adult congenital heart disease patients.⁶ The reduced exercise capacity is well described in patients who have undergone Senning or Mustard procedure.⁷⁻¹⁰ Reduced aerobic capacity, shorter exercise times, and attainment of lower peak heart rate when compared to controls have all been reported.¹⁰ The poor exercise capacity is an important prognostic factor as it correlates closely with hospitalization and death.¹¹ Attainment of a peak VO_{2max} of \leq 52.3% is predictive of a substantially higher 4-year risk of death or cardiac-related hospitalization.¹¹ In this the cohort mean %VO_{2max} achieved was 61.3% and seven patients had a value \leq 52.3%. Of this subgroup, three patients had a device in situ and therefore did not have a cardiac MRI. Two of the remaining patients did have right ventricular volume above the cohort mean (right ventricular end-diastolic volume 225.4 mL and 248 mL, right ventricular end-diastolic volume indexed 113.1 mL/m² and 134 mL/m², and right ventricular ejection fraction 56% and 49%, respectively). An

TABLE 2 Qualitative grading of echo parameters

	Right ventricular systolic function (n = 45) patients (%)		Tricuspid regurgitation (n = 42) patients (%)
Mildly reduced	7 (15.5%)	None	5 (11.9%)
Mildly to moderately reduced	8 (17.8%)	Trivial	11 (26.2%)
Moderately reduced	26 (57.8%)	Mild	24 (57.1%)
Severely reduced	1 (2.2%)	Mild to moderate	1 (2.4%)
		Moderate to severe	1 (2.4%)

TABLE 3 Correlation coefficient and P value for multiple parameters

	NT-proBNP		VO _{2max}		% VO _{2max}		Respiratory exchange ratio		Anaerobic threshold	
	Correlation coefficient	P value	Correlation coefficient	P value	Correlation coefficient	P value	Correlation coefficient	P value	Correlation coefficient	P value
RVEDV	0.56	0.03	0.00	0.99	-0.04	0.87	0.03	0.89	0.13	0.56
RVEDVi	0.61	0.01	0.18	0.94	-0.11	0.65	0.15	0.53	0.17	0.47
RVESV	0.65	0.00	-0.02	0.94	-0.11	0.64	0.12	0.60	0.17	0.44
RVESVi	0.66	0.00	-0.02	0.93	-0.22	0.34	0.16	0.48	0.15	0.50
RVEF	-0.49	0.01	-0.04	0.85	0.17	0.45	-0.30	0.19	-0.33	0.13
VO _{2max}		0.20								
% VO _{2max}		0.40								
Respiratory ex-change ratio		0.66								
Anaerobic threshold		0.90								

Abbreviations: RVEF, right ventricular ejection fraction; RVEDV, right ventricular end-diastolic volume; RVEDVi, right ventricular end-diastolic volume indexed; RVESV, right ventricular end-systolic volume indexed; RVESVi, right ventricular end-systolic volume indexed. Bold values indicates statistical significance.

impaired ability to recover from the exercise has also been demonstrated and is a further independent predictor of survival.¹²

Reasons for reduced exercise tolerance are multifactorial and certainly raise the question the adequacy of the right ventricle in coping with the stress and the effort demanded in the systemic circulation, and also the efficacy of the tricuspid valve when performing as a systemic atrioventricular valve.^{4,5} In this cohort, only one patient was graded as having a moderate to severe degree of tricuspid regurgitation on echo. VO_{2max} did not correlate with right ventricular dimension or ejection fraction as measured on MRI, perhaps reflecting in a part of general physical deconditioning, but the RV systolic function as graded on echo unsurprisingly appeared to correlate positively with NYHA class, the correlation coefficient was 0.32 (P value .04).

Other factors implicated in reduced exercise tolerance have been impaired chronotropic response (loss of sinus rhythm, pacing, and drugs), inadequate ventricular filling, and lack of increase in stroke volume due to smaller, noncompliant atria, and loss of the atrial kick. This may be compounded by the presence of stenosis, either functional or real in the atrial baffles.¹² The reduced exercise tolerance may also be contributed to the lack of physical activity during an overprotected childhood which is carried throughout the adult life.¹⁰

Buheitel et al demonstrate that the exercise tolerance of patients following the Senning procedure is qualitatively no different to patients who have a Fontan circulation and similar to adult patients with chronic heart failure.⁸

Several authors have reported areas of late gadolinium enhancement of the right ventricle on MRI representing abnormal myocardium and fibrosis, which may contribute to right ventricular dilatation and dysfunction.^{13,14} Significant myocardial perfusion defects have also been noted on the radionuclide scanning.¹⁵ Such areas of focal fibrosis have been found to correlate with right ventricular systolic dysfunction and arrhythmia.¹⁶ No such myocardial structural alterations were noted in the outpatient group.

NT-proBNP is widely accepted as a biomarker of ventricular function and heart failure.¹⁷ An NT-proBNP value of more than 200 pg/mL has been suggested as a cutoff value to predict clinical deterioration.^{3,18} In this series, 16 patients had an NT-proBNP value of more than 200 pg/mL and the mean NT-proBNP was 266.4 pg/mL. In this series, the NT-proBNP correlated strongly with right ventricular dilatation on MRI. This is supported by other works. Koch et al 2008 demonstrate a positive correlation between NT-proBNP and basal right ventricular diameter measured on echo.¹⁸ NT-proBNP continues to have a role as an important clinical parameter for estimating the appearance of cardiovascular complications and potential risk stratification for events before the patient's follow-up to be tailored accordingly.³ Our study supports this and we routinely measure and track serum NT-proBNP levels at our yearly routine clinic appointments.

Assessment of right ventricular function and its implications continues to be challenging in this group. There is a wealth of literature demonstrating the progressive systemic ventricular dysfunction occurring following the atrial redirection surgery.¹⁹⁻²¹ The function of the

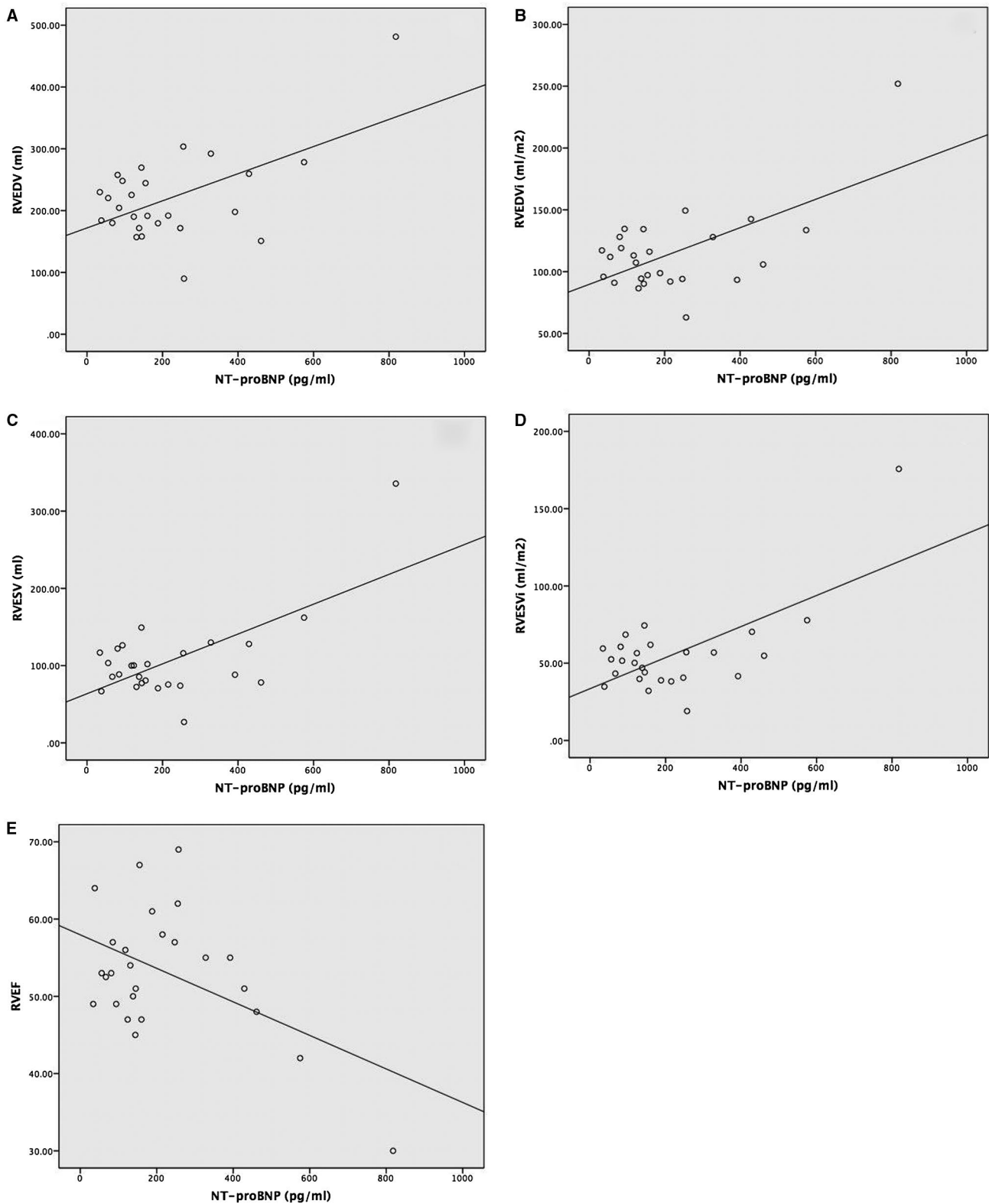


FIGURE 1 Scatter plots demonstrating significant correlations. A, Positive correlation between NT-proBNP and right ventricular end-diastolic volume (RVEDV) correlation coefficient 0.56 (P value .03). B, Positive correlation between NT-proBNP and right ventricular end-diastolic volume indexed (RVEDVi) correlation coefficient 0.61 (P value .01). C, Positive correlation between NT-proBNP and right ventricular end systolic volume (RVESV) correlation coefficient 0.65 (P -value < .01). D, Positive correlation between NT-proBNP and right ventricular end systolic volume indexed (RVESVi) correlation coefficient 0.66 (P value < .01). E, Negative correlation between NT-proBNP and right ventricular ejection fraction (RVEF) correlation coefficient -0.49 (P value .01)

TABLE 4 MRI and CPET data of the four patients with significant baffle leaks

	LVEDV (indexed)	LVESV (indexed)	LVSV (indexed)	LVEF	RVEDV (indexed)	RVESV (indexed)	RVSV (indexed)	RVEF	QP:QS MRI	VO _{2max}	% predicted VO _{2max}	Resp ex- change ration	VO _{2max} - anaero- bic threshold	SaO ₂ peak exercise
Case 1	130.1 (82.7)	55.2 (35.1)	74.9 (47.6)	58	121.4 (77.1)	32.9 (45.7)	75.7 (48.1)	63	1.5:1	20	44	1.17	10.3	94
Case 2	276.7 (151.7)	112.3 (61.6)	164.3 (90.1)	59	171.7 (94.1)	74 (40.6)	97.7 (56.6)	57	2.7:1	27	64	1.21	16.3	90
Case 3	308.5 (197)	125.4 (76)	193.1 (111)	59	191.5 (116.1)	102 (61.9)	89.5 (54.2)	47	2.5:1	33	73	1.28	18.1	96
Case 4	431 (239.4)	172.4 (95.8)	72.4 (39.8)	60	157.1 (86.5)	72.4 (39.8)	84.8 (46.6)	54	3:1	20	44	1.17	21.9	85

Abbreviations: LVEF, left ventricular ejection fraction; LVEDV, left ventricular end diastolic volume; LVESV, left ventricular end systolic volume; QP:QS, pulmonary to systemic flow ratio; RVEDV, right ventricular end diastolic volume; RVEF, right ventricular ejection fraction; RVSV, right ventricular end systolic volume.

systemic right ventricle can be quantified by a cardiac MRI. Not all units have easy access to MRI and many centers depend on echocardiography which continues to be challenging in this patient group due to body habitus, previous surgery, and concomitant respiratory disease. While echo parameters are easily obtainable in the clinical environment, Budts et al demonstrated that the echocardiography alone is not adequate to assess the right ventricular function accurately.⁹ In addition, the traditional transthoracic echo is not always sufficient to precisely interrogate the intra-atrial baffles. The cardiac MRI has emerged as the gold standard investigation for assessing right ventricular dimensions and function. In this cohort, the cardiac MRI demonstrates the right ventricular dilatation with reasonably well preserved ventricular function, the mean right ventricular ejection fraction was $53.7 \pm 7.9\%$. Identifying potential attrition from the late right ventricular failure before it occurs is one of the continuing challenges when looking after this group and reinforces the usefulness of NT-proBNP as a marker. There is evidence that the right ventricular dysfunction may be more frequent in patients with TGA¹⁹⁻²¹; however, the number of patients who had undergone a ventricular septal defect closure in this cohort was not sufficient to permit comparison.

Appraisal of the intra-atrial baffles is a vital component of the follow-up. In this group, no patients had evidence of baffle obstruction. Four patients were found to have significant baffle leaks on MRI, with pulmonary to systemic flow ratios ranging from 1.5:1 to 3:1 by the MRI flow analysis and cardiac catheterization (Table 4). Figure 2 demonstrates a significant baffle leak as observed on MRI. Of note, all four patients were NYHA class 1 and demonstrated the preserved right ventricular systolic function. Following the review of these cases individually with several large centers within the UK and Ireland, our decision has been to monitor the clinical progress of these patients and perform serial MRI scans. To date, there has been no significant deterioration in neither the left ventricular dilatation, nor the systemic right ventricular function at the 3-year MRI follow-up, nor any suggestion of raised pulmonary pressures. In our experience, there is currently no consensus on the optimal management of baffle leaks following atrial redirection surgery.

The incidence of baffle leaks is reported as ranging from 3.3% to 35%.^{19,20} The presence of a baffle leak with significant systemic venous to pulmonary venous shunt may lead to desaturation, exercise intolerance, atrial arrhythmia, and thrombo-embolic events.¹ Conversely, a persistent pulmonary venous to systemic venous shunt may theoretically lead to pulmonary hypertension although this was not evident in any of our cases at catheterization, nor at the 3-year MRI and echocardiographic follow-up. The risk of paradoxical emboli in this population appears to be related to the presence of atrial arrhythmias and those with endocardial pacemakers. The authors believe in these circumstances that the patient should be anticoagulated or the baffle leak should be closed.

A baffle leak generally behaves akin to an atrial septal defect, permitting the blood flow from pulmonary venous to systemic venous baffle; thus, placing a volume load on the posterior subpulmonary left ventricle. In time, there is left ventricular dilatation but this is often well tolerated. We postulate that a baffle leak may actually be beneficial by splinting the ventricular septum and thereby supporting the systemic right ventricular function with improved coaptation of the systemic atrioventricular

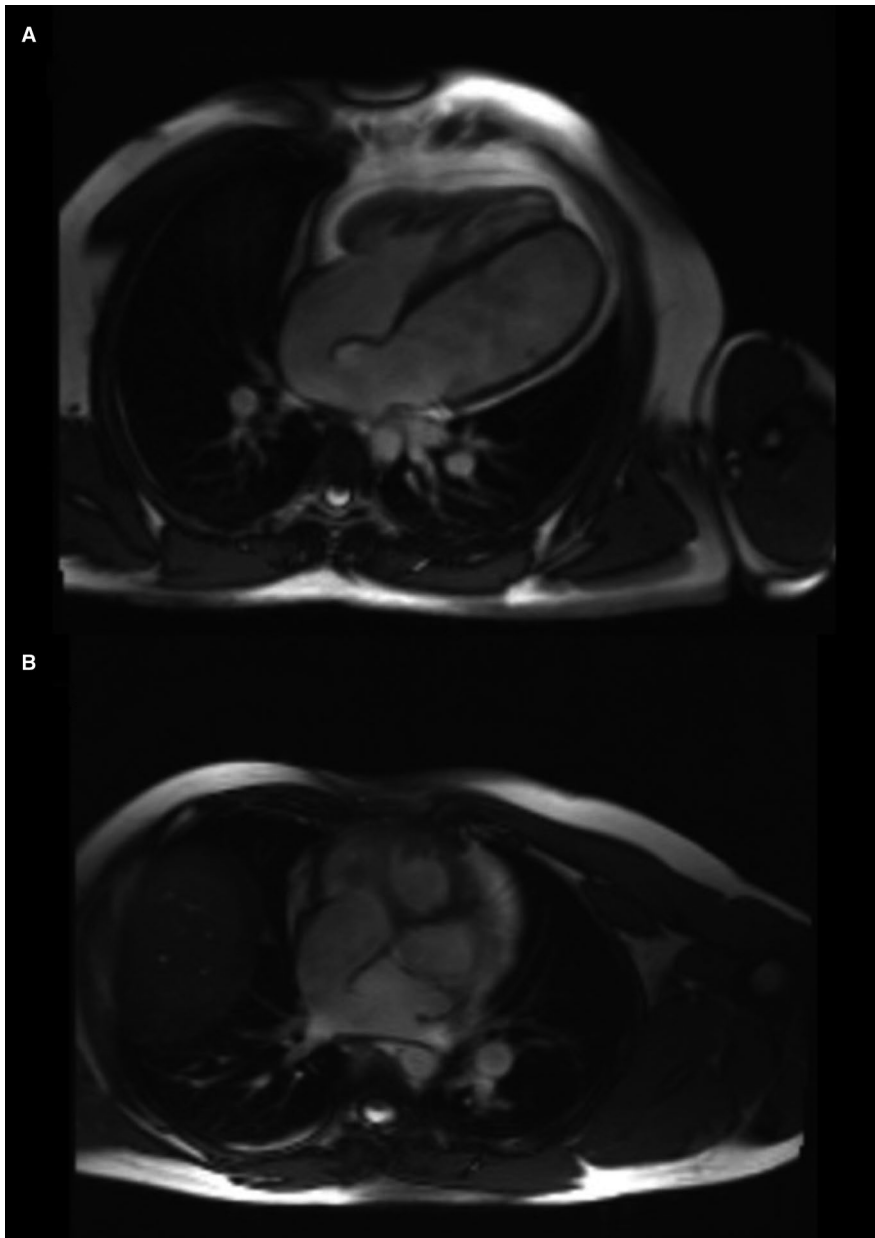


FIGURE 2 Appearances of baffle leak as demonstrated on the cardiac MRI. A, four chamber view with significant sized baffle leak and moderate to severe left ventricular dilatation. B, Para-axial view demonstrating the significant leak and unobstructed pulmonary venous baffle

valve; not dissimilar to the retraining of the left ventricle with a pulmonary artery band.⁵ Despite recognized complications of having a shunt at the atrial level in the setting of a systemic right ventricle, for a subset of patients, we suggest it may prove beneficial. However, meticulous investigation and follow-up are required to appropriately diagnose and counsel this patient group. The authors would assert that in the setting of a baffle leak, if the patient remains asymptomatic and free from complications, then yearly careful follow up and imaging is appropriate.

5 | STUDY LIMITATIONS

This study includes all patient's post atrial redirection surgery attending the adult congenital heart disease service in Northern Ireland; however, there are some limitations. The study is retrospective and

observational in nature; because of this not all of the patients had received all of the investigations. This would not have been possible anyway as patients would have been excluded from certain tests due to comorbidities or the presence of an implantable device. Nor do we have any vocational information or data on day-to-day living due to the retrospective nature of the study. Although this patient cohort is relatively small, they do represent a significant proportion of the adult congenital heart disease population in a single tertiary center within Ireland and remain at risk of future complications.

6 | CONCLUSIONS

This study demonstrates that, the patient's post atrial redirection surgery has reduced the exercise tolerance on objective testing as expected; but

overall a high proportion is NYHA class I (almost 70%). A higher NT-pro BNP correlates with increased right ventricular size and reduced function. The right ventricular impairment on echo did have a positive correlation with NYHA class, although in this cohort did not achieve a significant P value. Overall our cohort has reasonable day-to-day functioning as evidenced by their lack of symptoms and cardiopulmonary exercise tests. NT-pro BNP is an important marker for subtle deterioration and should prompt consideration for therapeutic intervention. The regular specialist follow-up and assessment with advanced imaging at regular intervals remains critical for the optimal long term outcomes in this group of patients.

CONFLICTS OF INTEREST

The authors report no relationships that could be construed as a conflict of interest.

AUTHOR CONTRIBUTIONS

Study concept, data collection and analysis, and manuscript preparation: M. Louise Morrison.

Study concept, data review, and manuscript review: Christopher J. Lockhart & Brian Grant.

Manuscript review: Andrew J. Sands, Brian A. McCrossan, Frank A. Casey, Brian G. Craig, Colum G. Owens & Mark S. Spence.

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