

Ventricular Assist Device Support in Advanced Heart Failure Patient with Congenitally Corrected Transposition of the Great Arteries and Dextrocardia- Case Report and Review

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ABSTRACT:

Congenitally corrected transposition of the great arteries (ccTGA) is congenital cardiac malformation with an incidence of 0.5% of all patients with congenital heart defects. Dextrocardia is reported among these patients up to 20%. We report a 52-year old female patient with ccTGA and dextrocardia with situs solitus. Due to congestive heart failure refractory to medications and high pulmonary vascular resistance multidisciplinary team decided for long term unloading with ventricular assist device as a bridge to heart transplant candidacy. In September 2017 intrapericardial ventricular assist device (centrifugal continuous flow, fully magnetically-levitated technology) was implanted in failing systemic ventricle. Accurate imaging (trans-thoracic echocardiography with contrast, transoesophagic echocardiography, cardiac MSCT) in multidisciplinary heart team is pivotal in successful implantation of small size intrapericardial VAD in patient with complex anatomy as ccTGA and dextrocardia. Because the existing literature consists mainly of case reports, management should include the stepwise introduction of the treatment modalities and close monitoring of the clinical response as well as heart multidisciplinary team.

KEYWORDS: congenitally corrected transposition of the great arteries; dextrocardia; ventricular assist device; heart failure

SAŽETAK:

PODRŠKA ZA VENTRIKULARNE POMOĆNE UREĐAJE KOD PACIJENTA S NAPREDNOM SRČANOM INSUFICIJENCIJOM S KONGENITALNO KORIGIRANOM TRANSPOZICIJOM VELIKIH ARTERIJA I DEKSTROKARDIJOM - PRIKAZ I PREGLED SLUČAJA

Kongenitalno korigirana transpozicija velikih arterija (ccTGA) kongenitalna je srčana malformacija s incidencijom od 0,5% svih bolesnika s urođenim srčanim manama. Dekstrocardija je zabilježena među tim bolesnicima i do 20%. U ovom radu prikazujemo 52-godišnju pacijenticu s ccTGA i dekstrocardijom sa situs solitusom. Zbog kongestivnog zatajenja srca otpornog na lijekove i visokog plućnog vaskularnog otpora multidisciplinarni tim odlučio se za dugoročno rasterećenje s ventrikularnim pomoćnim uređajem kao prijelaznim rješenjem do indikacije za transplantaciju srca. U rujnu 2017. ugrađen je intraperikardijalni ventrikularni pomoćni uređaj. Precizno slikanje (transtorakalna ehokardiografija s kontrastom, transezofagična ehokardiografija, MSCT srca) u multidisciplinarnom srčanom timu ključni su za uspješnu implantaciju intraperikardijalnog VAD-a malih dimenzija kod pacijenta sa složenom anatomijom poput one u ccTGA i dekstrocardija. Budući da se postojeća literatura sastoji uglavnom od izvještaja o slučajevima, odluke u liječenju trebale bi uključivati postupno uvođenje modaliteta liječenja i pomno praćenje kliničkog odgovora, kao i multidisciplinarnog tima za rad srca.

KLJUČNE RIJEČI: kongenitalno korigirana transpozicija velikih arterija; dekstrocardija; ventrikularni pomoćni uređaj; zastoj srca

INTRODUCTION

Congenitally corrected transposition of the great arteries (ccTGA) is congenital cardiac malformation with an incidence of 0.5% of all patients with congenital heart defects. Morphologically, ccTGA comprises a combination of atrioventricular and ventricular-arterial discordance (“double discordance”)¹. CcTGA is accompanied by associated cardiac lesions in up to 80% of cases, ventricular septal defects (VSDs) being most common of all in 60% to 80% of cases and pulmonary stenosis in ≈ 50%^{1,2}. Prevalence of ccTGA is 0.5%, with dextrocardia reported among 20% of them^{3,4}. Patient with isolated ccTGA are often asymptomatic for decades, but as right ventricle (RV) and tricuspid valve (TV) support systemic circulation, higher afterload can over time lead to gradual dysfunction and failure of systemic ventricle and systemic atrioventricular valve. Patients with congenitally corrected transposition of great arteries (ccTGA) present with heart failure commonly in the fourth or fifth decade of life^{3,4}. Because therapeutic options are limited and probability of increasing number of individuals with TGA with longer survival, management of these patients needs an effective strategy^{5,6}.

CASE REPORT

We report a 52-year old female patient with ccTGA and dextrocardia (Figure 1) with situs solitus. In 2008, she was first admitted to our institution because of heart failure, and diagnosed with ccTGA combined with dextrocardia.

For approximately ten years she was treated with heart failure medications, angiotensin-receptor blockers, beta blockers, spironolactone which subsequently changed to pelerine. Optimal therapy improved her condition to well compensated ccTGA over longer period of time. In the January 2017, despite optimal

medications, she developed congestive heart failure and required repeated hospitalisations with symptoms of congestion and low cardiac output with elevated NT-pro-BNP levees up to 10752 pg/mL. Her cardiac functional status decreased to NYHA (New York Heart Association) class IV. Laboratory test results indicated liver and renal dysfunction. She was treated with inotropic support and careful volume management to maintain euvoletic status. Inotropic therapy was administered in addition to intravenous prostaglandine infusion. However, her condition deteriorated to INTERMACS (Interagency Registry for Mechanically Assisted Circulatory Support) Profile 2. Echocardiography confirmed the presence of ccTGA and dextrocardia with situs solitus, with ejection fraction of dilatator and trabeculated systemic ventricle less than 20%. Preoperative echocardiography (Figure 2) systemic ventricular end-diastolic dimension and endsystolic dimension were 6.0/5.5 cm, and a non-systemic ventricular ejection fraction of 40%. Patient also had severe atrioventricular systemic valve insufficiency, morphologically tricuspid valve, and mild atrioventricular non systemic valve insufficiency, morphologically mitral valve. She had mild pulmonic valve insufficiency with mild spumoni obstruction due to protrusion of the membrane of basal part of interventricular septum. No aortic valve insufficiency and no ventricular septal defect was found. She was at that time evaluated for heart transplantation and placed on the cardiac transplant waiting list.

Despite medication her condition deteorated, so invasive haemodynamics measurements were repeated revealing RVP 71/6/19mmHg, PCWP 29mmHg, CI 1,39 L/min/m2, MPAP 45mmHg, PVR 6,24WU. Pulmonary hypertension and pulmonary vascular resistance may have induced right heart failure in the transplanted heart, so multidisciplinary heart transplant team



Figure 1. RTG showing ccTGA and dextrocardia with situs solitus A. before ventricular assist device implantation and B. after ventricular assist device implantation

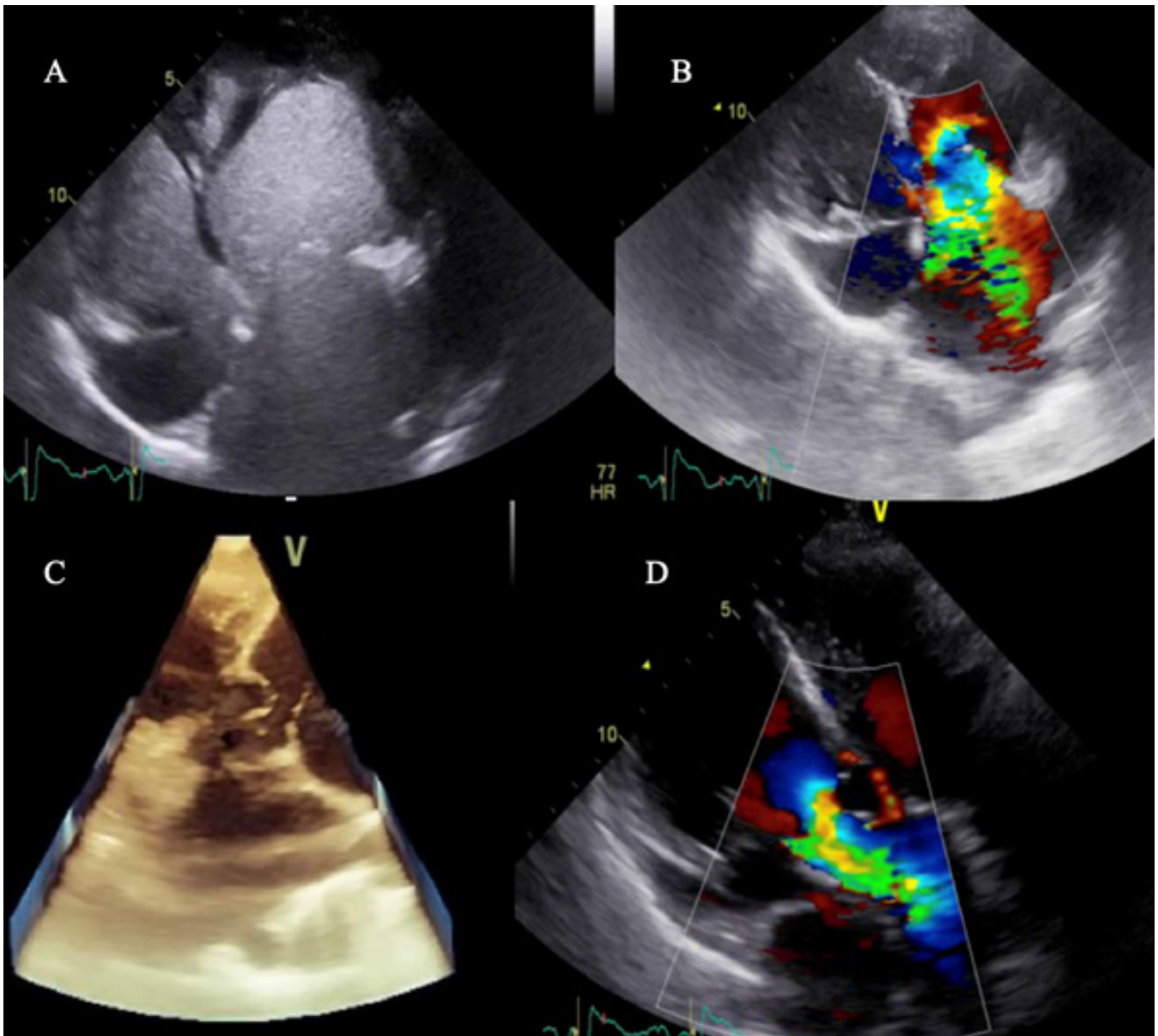


Figure 2. Transsthoracic echocardiography A. ccTGA and dextrocardia with situs solitus, with ejection fraction of dilated and trabeculated systemic ventricle less than 20%. B. severe atrioventricular systemic valve insufficiency (morphologically tricuspid valve) C.D. mild pulmonic valve insufficiency with mild subpulmonic obstruction due to protrusion of the membrane of basal part of interventricular septum.

decided for long term unloading with ventricular assist device as a bridge to heart transplant candidacy. In September 2017 intra-pericardial ventricular assist device (centrifugal continuous flow, fully magnetically-levitated technology) (Figure 4) was implanted at midline position. We used preoperative optimization of subpulmonic ventricle as in all patients with biventricular failure, comprised of aggressive dehydration with diuretics in combination with inotropic therapy levosimendan prior to VAD implantation. We aimed to unload and optimize the subpulmonic ventricle to achieve mechanical circulatory support for systemic ventricle alone by targeting central venous pressure (CVP) of less than 10 mm Hg. Postprocedural hospital course was complicated by mild subpulmonic (nonsystemic) ventricle failure managed by inotropic support and pump adjustment. Operative technique cardiopulmonary bypass was used. The ventricular cavity was closely inspected before cardiopulmonary bypass was on because it is morphologically a right ventricle with many more trabeculations and possible sites for thrombus formation. The device was inserted to the inferior, diaphragmatic surface of the morphologic RV free wall. As an nonobstructed inflow optimal

positioning of the inflow cannula is paramount, we achieved that by using intraoperative transesophageal echocardiography (TOE) to find the optimal device implantation site. We used pre and perioperative transoesophageal echocardiography (TOE) for guidance and positioning the inflow cannula (Figure 3) in the morphological right ventricle (systemic ventricle), positioning and extracting moderatory band which could induce inflow cannula obstruction, complicated by dextrocardia present in our patient. TOE guided placement of the inflow cannula was done when the heart was still full before going on cardiopulmonary bypass and positioned as dextrocardia, as failing systemic morphologic RV was dilated and a globular shape. Transesophageal echocardiography was also used to confirm cannula position after coming off bypass. Careful resection of adequate amount of muscle trabeculation or muscle bands to prevent obstruction to the inflow cannula was done, and papillary muscles were preserved. Outflow cannula was placed beneath the sternum and attached at in the lateral right side of ascending aorta. In early postoperative period, clinical course was complicated with mild failing of subpulmonic ventricle, managed with medication therapy of

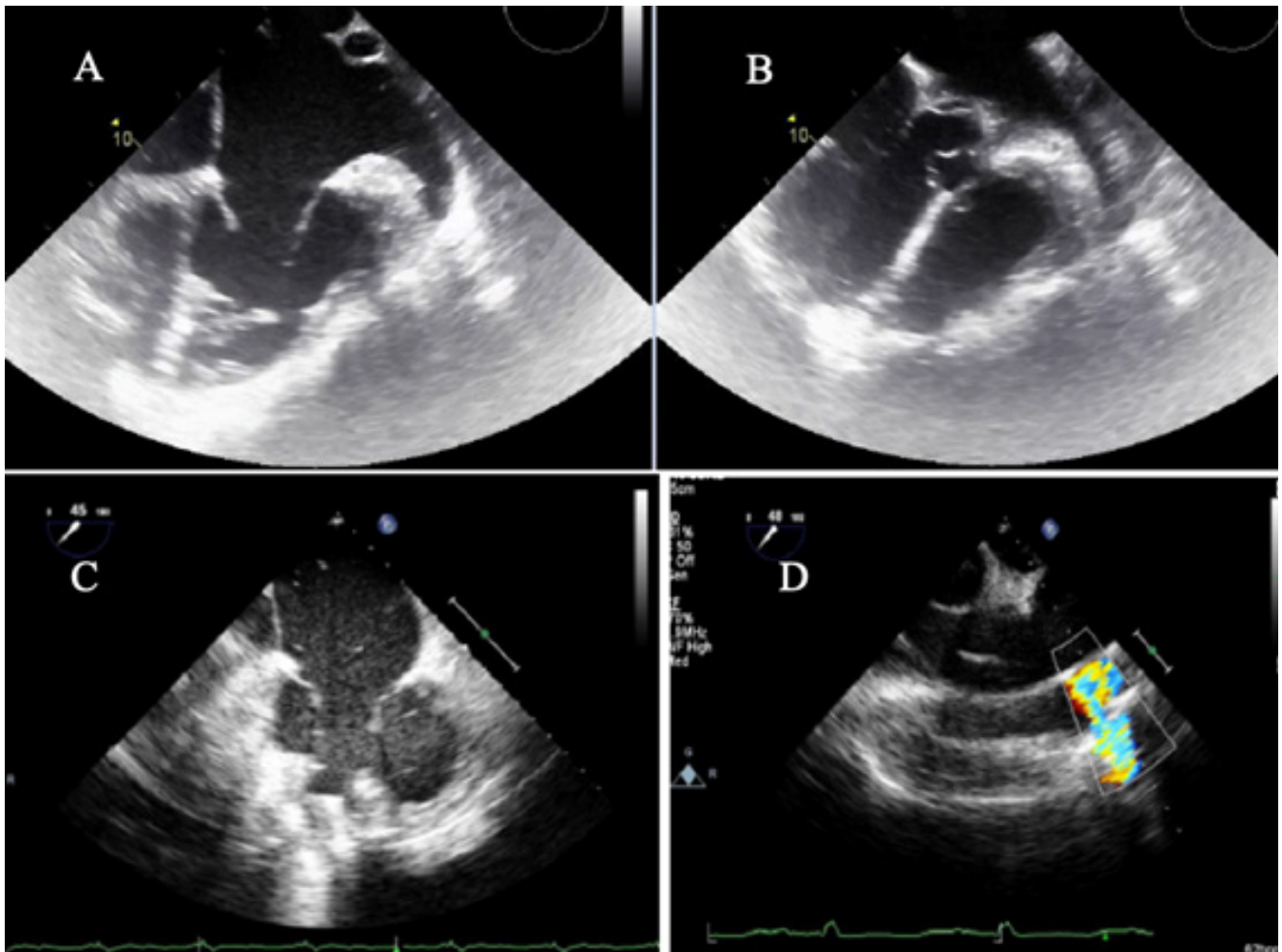


Figure 3. Preoperative A.B. transoesophageal echocardiography –ccTGA and dextrocardia with situs solitus, and C.D. postoperative transoesophageal echocardiography showing the position of the inflow cannula.

dobutamin and diuretic iv. Patient was discharged four weeks after implantation of ventricular assist device, taking pharmacological treatment of varfarin, acetylsalicylic acid, angiotensin-receptor blocker, beta blocker, eplerenon and is doing well, currently in NYHA class II. Eight months right heart catheterization was done revealing now PVR of 3,1 WU as result of good unloading of systemic ventricle by ventricular assist device.

DISCUSSION

Congenitally corrected transposition of the great arteries (ccTGA) is congenital cardiac malformation with an incidence of 0.5% of all patients with congenital heart defects often combined with other cardiac malformation, in few cases with dextrocardia as in our patient.

Patients with transposition of great arteries (TGA) undergone a routine arterial switch operation (ASO) within the first week of life in In Europe in 25–30% of patients⁷. This very early approach is controversial and upper age limit for a primary ASO in TGA cannot be determined. In case of late failure of the systemic ventricle after the Senning or Mustard operation is controversial, and treatment options are medical management or cardiac devices possibly followed by heart transplant, or staged anatomical repair. Medical therapy for congestive heart failure include serial echocardiographic follow-up combined with exercise testing when needed for detection of early ventricular or valvular function changes⁸. Mild systemic ventricular dysfunction with mild-to moderate valvular regurgitation can be treated conser-

vatively with afterload reduction using angiotensin-converting enzyme inhibitors, b-blockers and diuretics^{8,9}. Progressive aortic insufficiency and arrhythmias add to morbidity and mortality of these patients¹⁰.

Also use of left VAD for RV failure following the Mustard operation has been described¹¹. But approximately one-quarter of these patients will need a heart transplant^{8,12}.

Patients presenting with advanced right ventricular or biventricular failure, severe valvular dysfunction, arrhythmias resistant to therapy or heart block should be enrolled in the heart transplant program^{8,12}. Heart transplant is a well-established treatment strategy and is likely to be a superior option to anatomical conversion challenging^{12,13}.

Patients with congenital heart disease, as in our case ccTGA present with unique challenges for mechanical assist device implantation because of their individual specific anatomical features or previous operations. Systemic and pulmonary vascular resistances (PVR) add to the complex interplay of the preceding factors as in our case where we decided to implant VAD in systemic ventricle for unloading the systemic ventricle and thus to decrease PVR over time. Few reports describe the possibility of using long-term VAD support as an alternative treatment option for ccTGA patients with end-stage heart failure either as a bridge to transplant or destination therapy treatment strategy¹³⁻¹⁷.

In our case ventricular assist device implantation was feasible using different specific to our patient cannulation site positioned and verified by echocardiography and pump performance before

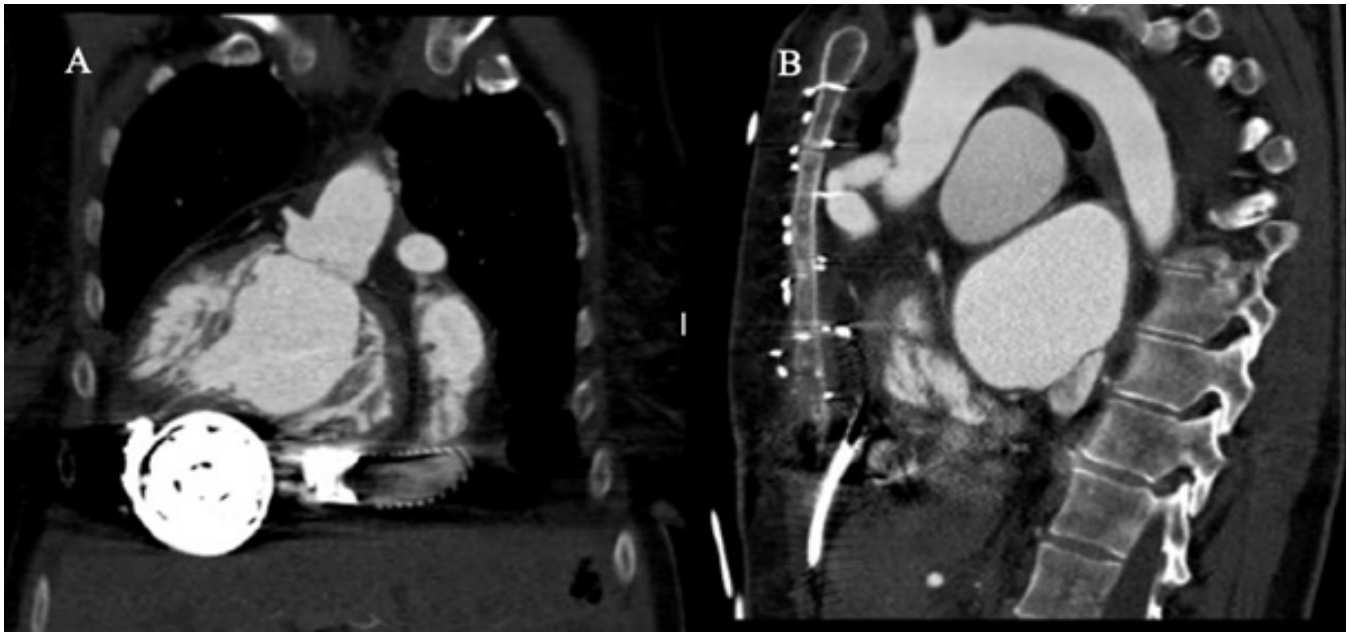


Figure 4. Multi slice Computer Tomography (MSCT) showing A. position of ventricular assist device in systemic ventricle and dextrocardia and B. mild kinking of outflow graft beneath sternum with no hemodynamic significance

leaving the operating room.

CONCLUSION

Implantation of ventricular assist device in systemic ventricle in ccTGA has been described in small groups of patients and in our knowledge in a couple patients with ccTGA combined with dextrocardia¹³⁻¹⁷. Accurate imaging (transthoracic echocardiography with contrast, transoesophageal echocardiography, cardiac MSCT) in multidisciplinary heart team is pivotal in successful implantation of small size intrapericardial VAD in patient with

complex anatomy as ccTGA and dextrocardia. Because the existing literature consists mainly of case reports, management should include the stepwise introduction of the treatment modalities and close monitoring of the clinical response as well as heart multidisciplinary team.

AUTHOR CONTRIBUTIONS:

All authors listed have made a substantial, direct and intellectual contribution to the work, and approved it for publication.

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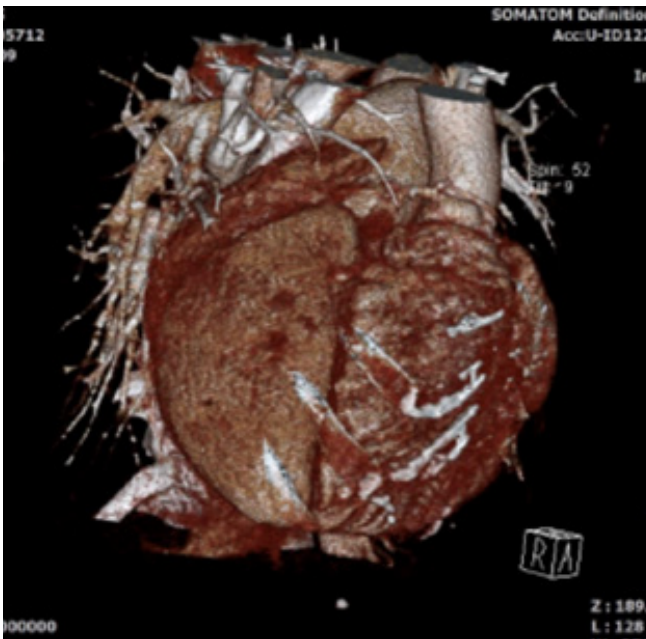


Figure 5. 3-D reconstruction showing systemic, morphologically right ventricle.

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