The year 2017 in the European Heart Journal – Cardiovascular Imaging.

Part II.

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Abstract

European Heart Journal – Cardiovascular Imaging was launched in 2012 as a multimodality cardiovascular imaging journal. It has gained an impressive impact factor of 8.366 during its 5 first years and is now established as one of the top 10 cardiovascular journals and has become the most important cardiovascular imaging journal in Europe.

The most important studies from 2017 will be highlighted in 2 reports. Part I of the review will focus on studies about myocardial function and risk prediction, myocardial ischaemia, and emerging techniques in cardiovascular imaging, while Part II will focus on valvular heart disease, heart failure, cardiomyopathies, and congenital heart disease.
European Heart Journal – Cardiovascular Imaging has successfully consolidated as a multimodality journal during its first 5 years. The journal is now on top 10 of all cardiovascular journals according to the recent 2017 impact factor ratings. It has now an important role as a significant resource for cardiologists, specialists in all imaging modalities, and other physicians working in the field of cardiovascular imaging. The tradition of highlighting the most important studies that were published in the last year is continued.\textsuperscript{1,2} In two articles we will summarize the most important papers from the journal in 2017. Part II will focus on cardiomyopathies, congenital heart diseases, valvular heart diseases and heart failure (HF).

**Recommendations and expert consensus documents from the European Association of Cardiovascular Imaging**

One important assignment of European Heart Journal – Cardiovascular Imaging is to publish position papers, recommendations and expert consensus papers from the European Association of Cardiovascular Imaging (EACVI).\textsuperscript{3} The journal published 7 recommendations and expert consensus papers in the field of multimodality imaging\textsuperscript{4–10}, 2 from echocardiography in 2017,\textsuperscript{11,12} and 1 about nuclear imaging and computed tomography (CT).\textsuperscript{13} These papers are commented in more detail elsewhere in the two documents.

**Appropriateness criteria**

The EACVI also has acknowledged the need to develop documents on appropriateness criteria for cardiovascular imaging use in clinical practice.\textsuperscript{14} Two important reports on literature and current practice review were published in 2017. One on the use of transthoracic echocardiography in adults and one on cardiovascular imaging in heart valve diseases.\textsuperscript{15,16}
Cardiomyopathies

The knowledge about cardiomyopathies is steadily increasing and the interest on how cardiac imaging can assist in risk prediction of adverse outcome and in diagnostics is growing. The articles presented last year in our journal added much to this important knowledge. Arrhythmogenic cardiomyopathy is a progressive disease with high risk of life-threatening ventricular arrhythmias. An EACVI expert consensus document by Haugaa and co-workers was published in early 2017 with clinical recommendations for how to use multi-modality imaging in the different aspects of arrhythmogenic cardiomyopathy disease, including diagnosis, family screening, follow-up, risk assessment, and differential diagnosis. Figures 1 and 2. EACVI also support a change in the name of this disease from arrhythmogenic right ventricular cardiomyopathy to arrhythmogenic cardiomyopathy as there is overwhelming evidence that both ventricles are involved in most patients.

Differentiation between early-phase arrhythmogenic cardiomyopathy and right ventricular outflow tract (RVOT)-ventricular tachycardia (VT) can be challenging, and correct diagnosis is important for correct treatment. Saberniak et al. investigated 44 patients with RVOT VT and 121 early phase arrhythmogenic cardiomyopathy patients and found that patients with early-phase arrhythmogenic cardiomyopathy had structural abnormalities with lower RVEF, increased RV diameter, and pronounced RV mechanical dispersion compared with RVOT-VT patients.

The most prevalent cardiomyopathy is hypertrophic cardiomyopathy. Kauer and co-workers assessed diastolic LV strain and untwist in 41 consecutive genotyped family members of patients with hypertrophic cardiomyopathy by speckle-tracking echocardiography. Results showed that LV untwist and diastolic strain were delayed, and that untwist rate and diastolic strain rate were decreased.
The prevalence of aortic dilation in patients with hypertrophic cardiomyopathy was identified by Yousefzai et al. They reported a novel observation with 9% prevalence of dilated aorta in patients with hypertrophic cardiomyopathy.\textsuperscript{19}

Whether LV global longitudinal strain (GLS) measured early during treatment with anthracyclines can predict subsequent alterations in left ventricular (LV) ejection fraction was investigated by Charbonnel and coworkers.\textsuperscript{20} They performed a prospective study on 86 patients after 150 mg/m\textsuperscript{2} of anthracycline therapy. GLS greater than –17.5% was an independent predictor of future anthracycline-induced cardiotoxicity. This study was commented in an editorial by Dr Mohty and co-workers.\textsuperscript{21}

Santoro and coworkers compared standard echo, 2D, and 3D speckle tracking echocardiography for detection of subclinical anthracycline cardiotoxicity in breast cancer patients.\textsuperscript{22} They demonstrated potential superiority but also suboptimal feasibility of 3D EF and 3D strain in diagnosing subclinical anthracycline cardiotoxicity in these patients. 2D GLS was superior to standard echo and showed a good feasibility. E/e’ ratio also offered advantages in revealing cardiotoxicity.

Wing-yi Li et al investigated if myocardial stiffness as assessed by diastolic wall strain was altered in adult survivors of childhood leukemia with preserved LV ejection fraction (EF), and explored its association with myocardial fibrosis and diastolic deformation.\textsuperscript{23} They found that despite the preservation of LV EF, increased stiffness of the LV myocardium was evident and was associated with myocardial fibrosis and impaired ventricular diastolic function.

Salinaro and coworkers determined whether echocardiographic longitudinal systolic strain parameters could identify short-term improvement following chemotherapy for light-chain cardiac amyloidosis\textsuperscript{24}. The findings suggested that longitudinal systolic strain was a sensitive measure of pre-treatment cardiac functional impairment in these patients, could predict survival
over and above that of cardiac biomarkers, and could detect early cardiac functional improvement following chemotherapy.

Nochioka and coworkers assessed left atrial (LA) function by speckle-tracking echocardiography in 124 patients with cardiac amyloidosis. LA function was severely impaired and highly correlated with LV deformation. Differences in LA function between amyloid subtypes suggested that amyloid aetiology played a role in the pathophysiology of cardiac dysfunction in cardiac amyloidosis.

Lo Iudice and coworkers assessed the contributors of LV strain components, using 3D speckle tracking echocardiography in endurance athletes. Endurance athletes had an increased myocardial function at rest when compared with normal controls, this being elicited mainly by subendocardial and mid-wall fibres. Sinus bradycardia, LV mass, and afterload were independent determinants of supernormal myocardial deformation at rest.

Rundqvist and coworkers explored the effects of long-term endurance exercise on atrial and ventricular size and function in adolescents and examined whether these changes were related to maximal oxygen uptake (VO2max). They found an increase in atrial as well as in ventricular dimensions which were associated with better maximal oxygen uptake. The physically active group also demonstrated functional remodeling with a higher TAPSE and systolic RV wall velocity.

The EACVI consensus document on restrictive cardiomyopathies by Habib and coworkers provided comprehensive information for the appropriateness of all non-invasive imaging techniques for the diagnosis, prognostic evaluation, and management of patients with restrictive cardiomyopathies.
A joint procedural position statement on imaging in cardiac sarcoidosis was published from the European Association of Nuclear Medicine, the EACVI, and the American Society of Nuclear Cardiology. This joint position paper illustrated the role and the correct use of echocardiography, radionuclide imaging with 18F-fluorodeoxyglucose positron emission tomography, radionuclide myocardial perfusion imaging and cardiovascular magnetic resonance imaging for the evaluation and management of sarcoidosis patients.

**Heart failure**

The Euro-Filling report by Lancellotti and coworkers compared the diagnostic accuracy of the 2009 and 2016 echocardiographic grading algorithms for predicting invasively measured LV filling pressure (LVFP). Figure 3. The study demonstrated that the new 2016 recommendations for assessing LV filling pressure non-invasively are fairly reliable and clinically useful, and superior to the 2009 recommendations in estimating invasive LVEDP.

Chan and co-workers assessed the predictive value of combined $E/e'$sr ratio and GLS for prognosis in systolic heart failure. Study end points were defined as all-cause mortality or heart transplantation. The $E/e'$sr ratio was stronger than $E/e'$ ratio in predicting prognosis of patients with systolic HF. Combined assessments of GLS and $E/e'$sr by speckle-tracking longitudinal strain facilitated risk stratification of these patients.

The LA function index was evaluated as a predictor of long-term survival in outpatients with heart failure with reduced ejection fraction (HFrEF) by Sargento and coworkers. The endpoint was all-cause death. LA function index in HFrEF stable outpatients was a predictor of long-term survival and provided increased prognostic value over a wide range of confounder risk factors.
Donal and co-workers studied the value of estimated pulmonary pressure and LA size for diagnosing and determining a prognosis for patients with heart failure with preserved ejection fraction (HFpEF) in a prospective multi-centric cohort. The combination of enlarged left atrium and elevated estimated pulmonary pressure had a strong prognostic impact in patients suffering from HFpEF.

Another study explored if markers of diastolic dysfunction are associated with atrial fibrillation development among patients with heart failure with preserved ejection fraction (HFpEF). O’Neal and coworkers found that diastolic parameters of LA function possibly were more important markers of atrial fibrillation risk than LA dilation in HFpEF. The paper was commented in an editorial by Longobardo et al. stating that left atrium function plays a pivotal role in this context.

Kosmala and co-workers investigated the association of changes of loading parameters with changes in LV global longitudinal and circumferential strains. LV longitudinal and circumferential strains in a population without apparent heart disease were relatively insusceptible to changes in LV afterload within physiological range, which, therefore, seemed unlikely to be a significant confounder in repeated global longitudinal or circumferential strains observations.

Stankovic and coworkers investigated the association of the extent of LV remodeling, mechanical dyssynchrony, and survival in patients undergoing cardiac resynchronization therapy (CRT). Volumetric response assessed at 1-year after CRT was strongly associated with long-term mortality. However, an optimal cut-off could not be established. The association of the correction of mechanical dyssynchrony with survival was stronger than that of any volumetric cut-off. This study was commented in an editorial by Haugaa and Edvardsen, highlighting
mechanical dyssynchrony as a resurrected flashing and rocking parameter to predict prognosis after CRT.\textsuperscript{36}

Menet et al. investigated the relationship between septal deformation patterns studied by longitudinal speckle tracking and clinical outcome following CRT.\textsuperscript{37} The identification of septal deformation patterns provided important prognostic information in CRT candidates in addition to ordinary clinical, and echocardiographic predictors of outcome in heart failure patients.

**Valvular heart diseases**

Transcatheter aortic valve replacement (TAVR) and mitraclips have been the subject of several papers in 2017. These percutaneous approaches are imposing new challenges for imaging techniques. Also imaging these new implantable devices is mandatory to insure the efficacy and the safety of these new treatments. Cases reports have been published like one on the treatment of a tricuspid regurgitation in presence of a pacing lead.\textsuperscript{38}

Winter et al did an impressive work: out of 240 abstracts, 155 studies reporting echocardiographic parameter for twelve different valves prosthesis in a total of 27,159 patients were included in a meta-analysis. The means and standard deviations of peak velocity, peak gradient, mean gradient and effective orifice were extracted and pooled from the included studies. The pooled means and standard deviations for all available TAVR prosthesis were classified according to implanted valve size and time since implantation. The present study describes normal values for all available TAVR prosthesis.\textsuperscript{39} Another observational study explored follow-up parameters of 162 (79 Lotus and 83 SAPIEN3) consecutive patients. Authors are reporting that the frequency of mild (13.9% vs. 31.3%) and at least moderate (1.3% vs. 3.6%) paravalvular aortic regurgitation was less frequently found after Lotus than after SAPIEN3 implantation.
Lotus valve required less oversizing and was associated with less paravalvular regurgitation than SAPIEN3. One paper is reporting pre-discharge transthoracic echocardiography in 223 consecutive patients with severe aortic stenosis who underwent TAVR with a Medtronic CoreValve. Echocardiographic stent inflow frame eccentricity was defined as major–minor diameter in a short-axis view >2 mm. Eccentricity correlated with the Agatston score. This eccentric shape results in more para-valvular leakage. In another study, at 5-year follow-up, considering that echocardiographic follow-up was available in 96 patients, 30% of patients who underwent TAVR with a balloon-expandable valve showed initial structural valve deterioration (leaflet thickening >3mm, presence of calcification and abnormal leaflet motion). However, structural valve deterioration was not associated with severe stenosis in most of the patients and had no significant impact on and clinical outcome.

Discussing the 2017 manuscript dedicated to mitral valve regurgitation (MR), van Wijngaarden et al assess mitral annulus dynamics in primary and secondary mitral regurgitation (MR) with 3-dimensional transesophageal echocardiography (3D TEE) and its impact on MR quantification. The mitral annulus is enlarged and stiff in secondary mitral regurgitation patients, whereas in fibroelastic deficiency and Barlow disease, it is characterized by excessive dynamicity during systole. Enhanced annular dynamics leads to significant increase in grade of MR. Also, 76 patients with non-ischaemic cardiomyopathy and moderate to severe chronic secondary MR treated successfully with transcatheter or surgical mitral valve repair were evaluated. Transthoracic echocardiography was performed at baseline, discharge and 6 months post-repair. After correction of the MR, LVEF, and LV GLS corrected for LV end-diastolic volume remained unchanged over time. The stroke volume increased thanks to LV reverse remodeling.

Dumont et la looked at the determinants of significant tricuspid regurgitation (TR) 1 year after the correction of the aortic valve stenosis in patients treated for an aortic valve stenosis.
Significant TR was present pre-operatively in 11% of patients. Post-operative progression was observed in 27% of patients. Only tricuspid annulus size >40 mm was an independent echocardiographic predictor of moderate to severe TR at the 1-year follow-up. 45

**Cardiac magnetic resonance**

Cardiac magnetic resonance (CMR) is a technique which excels in the ability to characterize myocardial tissue with high spatial resolution.

The important role of CMR for tissue characterization was particularly emphasized in a remarkable study by Csepe et al. 46 They performed high resolution (1 mm³) late gadolinium enhanced images of the left atrium to study the ultrastructure of the human sinoatrial node in healthy volunteers *in vivo* allowing to identify fibrotic areas in this sino-atrial node. This identification of fibrosis in the sinoatrial node by CMR was validated by comparing optical mapping, ex vivo contrast-enhanced CMR and histological validation in ex-vivo perfused explanted human hearts. Thus this original study for the first time demonstrated the feasibility of in vivo imaging or, 3D human SAN fibrotic structure.

Another important paper was published by Hilbert et al on the use of CMR in patients with devices and pacemakers. 47 Indeed use of CMR in patients with devices has been limited, not only because of device compatibility, but also because artifacts induced from the generator- or leads which can impair image quality of cardiac images. In this setting the study by Hilbert et al evaluated image quality of all conventionally employed diagnostic CMR imaging modules depending on device type. Then they proposed strategies of CMR imaging sequences depending on the device type allowing to acquire diagnostic images in the majority of patients even with
non-MR-conditional devices. Thus this was an important study allowing to increase the use of CMR to diagnose cardiac disorders in patients with devices and pacemakers.

Another exciting feature of CMR is myocardial characterization of diffuse myocardial fibrosis by T1 mapping. For this purpose different sequences have been proposed. The important study by Child et al compared diagnostic performance of T1 and ECV measurements by 3 different sequences: MOLLI, shMOLLI and Sasha for discriminating between health and disease, and their associations with histologically derived collagen vascular fraction from endocardial biopsies. The study demonstrated that the differences sequences differ in their bioequivalence for detection of abnormal myocardium, which is characterized by diffuse interstitial myocardial fibrosis. Native T1 with MOLLI sequences was found to provide the strongest discriminatory accuracy in characterization of human myocardium.

There was also much interest in new T1, T2 mapping techniques, and evaluation of extracellular distribution volume to Gadolinium which allow respectively non-invasive detection of inflammation, edema and fibrosis in the myocardium in different diseases.

Biesbroeck et al evaluated the additional diagnostic value of CMR to ESC position statement criteria in myocarditis. In a retrospective evaluation of 303 patients hospitalized in two academic centers for suspected acute myocarditis, CMR provided a definite diagnosis in 158 patients (52%), including myocarditis in 104 (34%), myocardial infarction in 44 (15%), and other pathology in 10 patients (3%). As opposed to ESC position statement criteria alone, CMR reclassified diagnosis to alternative cardiac disease that could explain the clinical syndrome in 18% of cases, and confirmed myocarditis in 44% of patients in 122 patients in which ESC PSC criteria were insufficient for diagnosis. Thus this study provided strong support for the routine use of CMR in the work-up of suspected acute myocarditis.
Also in myocarditis, Bohnen et al used tissue T1 and T2 times to monitor myocardial inflammation during the healing process of this disease. They demonstrated that both native myocardial T1 and T2 provide an excellent performance for assessing the stage of myocarditis, by demonstrating increases in acute phase of myocarditis, and normalization during the healing stage. Both parameters had higher diagnostic accuracy than other global or regional CMR parameters, and in particular classic Lake Louise criteria for acute myocarditis.

In patients with different stages of systolic LV dysfunction in dilated cardiomyopathy, Spieker et al used T2 mapping to T2 mapping for the non-invasive assessment of myocardial inflammation in comparison with endomyocardial biopsy. They found that Global T2 time was significantly increased in patients with dilated cardiomyopathy relative to controls. This increase of T2 time correlated with the presence of inflammatory cells in endomyocardial biopsy. They concluded that T2 mapping may facilitate the identification of patients with suspected inflammation which may benefit from endomyocardial biopsy for therapeutic decision-making.

Heck et al. used CMR to measure ECV fraction, total ECV, and total cellular volume in 69 women undergoing anthracycline based adjuvant chemotherapy for breast cancer. Then in a 22 factorial, placebo controlled, double-blinded trial the evaluated the effect of candesartan and metoprolol on these parameters. They demonstrated that higher doses of anthracycline chemotherapy were associated with greater increase of ECV fraction and total ECV, indices of oedema, and diffuse myocardial fibrosis. Treatment with candesartan, which alleviated a reduction in LV systolic function, was associated with a greater decline in total cellular volume than without candesartan treatment while metoprolol did not affect myocardial composition.

Similarly Storz et al compared native T1 times, ECV, fibrosis and cell volume in the KORA S4 study, a large sample of the general population in Germany. They compared diabetic and prediabetic subjects with non-diabetic patients and while ECV was decreased, cell volume
was significantly increased in patients with diabetes and prediabetes, demonstrating higher LV remodeling as early detectable changes in this disease process.

Kozor et al. evaluated mass and T1 times of papillary muscle in different phenotypes of LV hypertrophy. The study observed that disproportionate hypertrophy of LVPMs in LVH positive hearts occurred in Fabry disease and hypertrophic cardiomyopathy, but not in other causes of hypertrophy such as aortic stenosis, amyloidosis or hypertension. Further they observed that patients with Fabry disease did not always have reduced T1 times as in septum, suggesting that other causes than sphingolipid storage may account for papillary muscle hypertrophy.

Several other studies evaluated other CMR derived parameters on prognosis in several diseases. Foley et al evaluated qualitative deformation by strain and torsion computed from tissue tagging to differentiate patients with ischemic from non-ischemic etiology of heart failure in the VINDICATE study. The found that torsion and twist were significantly different in non-ischemic vs ischemic cardiomyopathy despite similar volumes, circumferential and longitudinal deformation and EF, thereby allowing to differentiate the etiology of such heart failure.

Cao et al evaluated central transit time from first pass perfusion images normalized to cardiac cycle length intervals in heart failure with preserved and reduced ejection fraction and demonstrated significant prolongation relative to controls. Increases of central transit time correlated with ejection fraction in HFrEF and increased pulmonary capillary pressure in HFpEF. Similarly, Ricci et al evaluated pulmonary blood volume as product of pulmonary transit beats form first-pass contrast-enhanced perfusion CMR and anterograde indexed right ventricular (RV) stroke volume index and demonstrated the prognostic value of this parameter in heart failure outpatients in the Prove HF study.

Pedrotti evaluated the prognostic impact of late gadolinium enhancement in the risk stratification of heart transplant patients. They found that the total amount of late gadolinium
enhancement identifying myocardial fibrosis was the only other independent predictor other than cardiac allograft vasculopathy which allowed to predict major cardiac adverse events and mortality in these patients.

Several other studies evaluated the possibilities of CMR perfusion imaging. In a large multicenter study including 5 European centers and 416 patients, Hamada et al evaluated the diagnostic performance of a new whole-heart dynamic 3D CMR first-pass perfusion imaging technique relative to quantitative coronary angiography and fractional flow reserve according to gender. They observed that the sensitivity and specificity of the approach were similarly good both in female 89% (95% CI: 77–96) and 82% (95% CI: 70–90) and male patients 83% (95% CI: 77–86) and 79% (95% CI: 71–86) relative to QCA and FFR 95% (95% CI:82–99) and 84% (95% CI: 73–92) for females and males 83% (95% CI: 76–89) and 82% (95% CI: 74–88) respectively. They concluded that whole-heart dynamic 3D CMR stress perfusion imaging has a high diagnostic accuracy for the detection of significant CAD irrespective of gender and is therefore a suitable non-invasive testing tool to detect myocardial ischaemia in both genders.

Also, Hosking et al evaluated the usefulness of lack of splenic switch off for detecting false-negative adenosine stress perfusion cardiac magnetic resonance (CMR). Lack of splenic switch off was present in 11% and predicted higher rates of false negative stress tests, indicating that this parameter has potential to improve on haemodynamic criteria as a marker of adenosine understress in CMR perfusion scans.

**Congenital Heart Disease**

Morgan et al examined the mechanism for branch pulmonary artery stenosis after the arterial switch operation for transposition of the great arteries. Neo-pulmonary to neo-aortic
geometry as well as post-operative compression of the left pulmonary artery stenosis by an enlarged aorta impacted left pulmonary artery size and perfusion of the left lung.

Huttin et al aimed to assess and quantify mitral valve chordae, leaflets, and LV myocardial interactions using speckle tracking echocardiography. Speckle tracking echocardiography was a useful tool in the assessment of interplays between mitral valve leaflets and myocardium and helped to demonstrate changes in temporal pattern of myocardial deformation.

The diagnostic accuracy of echocardiographic measures of great vessels in patients before bidirectional cavopulmonary connection (BCPC) was compared with CMR by Krupickova et al. They showed that echocardiography cannot substitute CMR for reliable identification of great vessel stenoses in complex patients prior to the BCPC, particularly those with Blalock–Taussig shunts.

Bhat et al sought to identify predictors of change in RV function and exercise capacity in adolescents following repair for tetralogy of Fallot. Their results suggested that early monitoring with magnetic resonance imaging might identify those at highest risk for progressive disease.

Hauck et al aimed to define TAPSE across aetiologies of paediatric pulmonary hypertension and assess the correlation between TAPSE and measures of disease severity. Reduced TAPSE after repair of congenital heart disease did not correlate with functional status and may reflect post-operative changes rather than poor function primarily due to pulmonary hypertension.

Pasquale et al determined the prevalence of baffle leaks in adults after atrial switch operations for transposition of the great arteries, as these may predispose to paradoxical embolic events, particularly in patients with transvenous pacemaker or defibrillator leads. They
proposed routine screening with agitated saline contrast, particularly prior to implantation of transvenous pacemaker or defibrillator leads.

Stephensen et al determined the effect of stress on left-to-right shunting in patients with atrial septal defect and investigated if the degree of shunting, cardiac output, and RV volumes were related to exercise capacity. Pulmonary to systemic flow ratio and shunt volume per heartbeat decreased during stress in atrial septal defect patients. A high systemic cardiac output during stress was a strong predictor of exercise capacity.

Moceri et al compared global ventricular function assessed by speckle-tracking in adult patients with Eisenmenger syndrome, other pulmonary artery hypertension aetiologies, and healthy controls and assessed the relationship between ventricular function and survival. RV remodelling differed between adults with Eisenmenger syndrome and other pulmonary artery hypertension aetiologies. Eisenmenger syndrome and increased RV free wall transverse strain were associated with better survival.

Hanneman et al. explored CMR quantified extracellular volume in patients with tetralogy of Fallot and its association with major adverse cardiovascular outcomes such as death, out-of-hospital cardiac arrest, heart failure requiring admission for escalation of therapy, or haemodynamically significant ventricular tachycardia. They found that right but not LV extracellular volume was significantly increased with increased risk of MACE, suggesting that this parameter could be an interesting biomarker for risk stratification and targeted therapeutic intervention in this population.

Finally, Ntsinjana demonstrated the utility of adenosine stress perfusion CMR as a screening test in pediatric patients (aged 14 years) with acquired or congenital coronary artery disease, such as anomalous origin of the left coronary artery arising from the pulmonary artery (ALCAPA) syndrome, Kawasaki syndrome, and transposition of great vessels corrected by
arterial switch operation. They found that adenosine stress had 100% sensitivity 98% specificity and 93% PPV and 100% PPV when compared with invasive coronary angiography. This suggests that stress adenosine perfusion CRM is adequate as an initial, non-invasive screening test for the identification of significant coronary artery lesions, with anatomical imaging used to confirm the extent of the culprit lesion in pediatric patients with congenital heart disease.
Figure legends:

Figure 1 Cardiac Magnetic resonans (CMR) right ventricular outflow tract (RVOT) in and out view, cine image in end diastole (A) and end systole (B). Micro-aneurysms of the RVOT and RV diaphragmatic wall are present (white arrows). From Haugaa et al Comprehensive multi-modality imaging approach in arrhythmogenic cardiomyopathy-an expert consensus document of the European Association of Cardiovascular Imaging. 2017;18:237-253

Figure 2 Proximal right ventricular (RV) outflow diameters. (Parasternal long axis (PLAX) and parasternal short axis (PSAX) and RV basal diameter (RVD) (diastole). Courtesy: Dr J. Saberniak. From Haugaa et al Comprehensive multi-modality imaging approach in arrhythmogenic cardiomyopathy-an expert consensus document of the European Association of Cardiovascular Imaging. 2017;18:237-253

Figure 3. Relationship between left ventricular (LV) filling grade and invasive left ventricular end diastolic pressure (LVEDP). Undefined corresponds to ‘indeterminate or cannot determine’ category. From Lancellotti P et al. Echo-Doppler estimation of left ventricular filling pressure: results of the multicentre EACVI Euro-Filling study. Eur Heart J Cardiovasc Imaging 2017;18:961–968.


