with pathological post-mortem cardiac changes including cardiac mass \( (p < 0.0001) \), biventricular wall thickness \( (p < 0.0001 \) left ventricle, \( p = 0.001 \) right ventricle), presence of any coronary disease \( (p = 0.0035) \) and severe coronary disease \( (p < 0.0001) \). CAC score > zero had positive predictive value of 89.7\% for severe coronary stenosis, and specificity of 94.5\%. Sensitivity of CAC score > zero for severe stenosis was only 70\%. Of cases with CAC score of zero, 25.9\% had severe coronary disease \( (p < 0.0001) \).

**Conclusion:** PM CAC scoring is highly feasible. Elevated CAC score in 18–50 years old with sudden death has high positive predictive value and specificity for coronary cause of death. However, CAC score of zero does not exclude a cardiac cause. PM CAC score assessment may be considered as a further tool to determine cause of death when there is objection to post-mortem.

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**006**

**Truncating Variants in the Desmoplakin Gene Cause a Distinct Arrhythmogenic Cardiomyopathy**

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**Background:** Variants in DSP cause arrhythmogenic cardiomyopathy (ACM) and phenotype data derived largely from arrhythmogenic right ventricular cardiomyopathy (ARVC) patient cohorts. We hypothesised that patients with DSP truncating variants (DSP\(_{tv}\)) express a wider phenotype spectrum. We report penetrance, phenotype spectrum and genetic architecture of DSP\(_{tv}\).

**Methods and Results:** Unrelated patients with a DSP\(_{tv}\) and any cardiac phenotype were sought from national centres \( (n = 98) \). Primary diagnosis for \( n = 32 \) included dilated cardiomyopathy \( (n = 19) \), ARVC \( (n = 10) \), ACM \( (n = 1) \), unexplained VF \( (n = 1) \) and Carvajal syndrome \( (n = 1) \). Fifteen \( (47\%) \) experienced sudden cardiac death (SCD) events, including \( 7 \) (22\%) with SCD as the presenting symptom. All had left ventricular (LV) involvement. A family history was reported in 22 (69\%). In the total cohort \( (n = 98) \), there were 68 unique DSP\(_{tv}\) (29 frameshift, 25 nonsense, 12 splice, two insertion/deletions) classified using ACMG criteria (5 pathogenic, 62 likely pathogenic, 1 uncertain significance). We investigated localisation of DSP\(_{tv}\) to key functional gene regions (G1, CR and G2) in cases compared to controls. Case variants were more common in G1 and CR (84–72\% vs 16–28\%) while control variants were more frequent in G2 (58\% vs 42\%) \((p < 0.0001)\). Event-free survival from SCD events was worse when DSP\(_{tv}\) occurred in G1/CR, compared to G2 \((\log-rank p = 0.016)\).

**Conclusion:** In the largest series of DSP\(_{tv}\) carriers, we show a wide phenotype spectrum. It should be considered a distinct gene-specific cardiomyopathy characterised by LV dysfunction with high risk of ventricular arrhythmias. DSP\(_{tv}\) are highly penetrant and gene location is associated with worse clinical outcomes.

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**007**

**Allied Health Technology Prize Finalists (007–010)**

**007**

**A Technical Perspective of Improving Detail and Consistency in the Performance and Reporting of Echocardiography: An Eight-Year Trend from a National Database**

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**Introduction:** Despite the available technology and potential for detailed information to be obtained for non-invasive cardiac assessment, the American College of Cardiology (ACC) note that there has been limited agreement on quality standards for imaging. This can lead to a lack of consistency, detail and quality of echocardiographic reports. The ACC proposed data standardisation, structured reporting with key data elements and imaging registries to address such deficits. These changes were examined in the largest national echocardiographic registry in Australia.

**Methods:** Between 2010 and 2014, there was implementation of direct online entry of echocardiographic studies into an electronic database, identification and auditing of six key data elements (LVEF, AV peak velocity, E/e’, LA area, RVSP and rhythm) along with pathways to improve quality and maximise the completeness of data acquisition and reporting.
There was a significant improvement in the completeness of the six key data elements from 2011–2018 (72.0 ± 26.8% vs 88.2 ± 13.5%; p = 0.02). Inter-practice variability for LVEF fell from 2011 to 2018 (p < 0.02).

Conclusion: Identification of key data elements as well as systematic capture and auditing significantly improved the consistency, detail and quality of echocardiographic reports. A rapid adoption of local quality improvements could be made utilising a national database.

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009

Evaluating Risk Factors for Sonographer Injuries – Lessons for Safer Scanning

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Aim: Performing echocardiography is associated with an elevated risk of injury amongst sonographers. There are guidelines for reducing sonographer injuries. We set out to investigate which of the guidelines had the greatest impact in preventing sonographer injuries.

Method: Following ethics approval, a confidential electronic survey was widely distributed through professional networks. Sonographers were asked to document work injuries, and how well they complied with published guidelines for sonographer safety. The results were then separated into two groups: “pain” and “pain-free”.

Results: 1494 sonographers completed the survey, with 84.3% reporting pain from scanning. The greatest impact on injury prevention was having regular breaks from scanning (36.7%), and assessing patient preference (37.5%). Other factors included having a supervisor assess scanning technique (36.7%), having a flexible room for ambidextrous scanning (22.2%), and daily stretching (22.2%).

Table 1. Reduction in sonographer injury risk for each recommendation.

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Conclusion: We were able to demonstrate the individual benefit of different recommendations on sonographer’s risk of injury. These results can be used to further tailor ergonomic recommendations to improve the risk profile of sonographers.

http://dx.doi.org/10.1016/j.hlc.2019.06.009

009

Exercise Cardiac MRI Unmasks Cardiac Dysfunction in Childhood and Adolescent Cancer Survivors with Reduced Cardiopulmonary Fitness

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Background: Young cancer survivors are at increased risk of reduced fitness and heart failure. Assessment of exercise cardiac reserve may reveal sub-clinical abnormalities that cannot be detected by resting measures of cardiac function.

Methods: 11 patients (age 9 to 24) with recent anthracycline treatment completed a maximal cardiopulmonary exercise test (VO2peak), and assessment of left-ventricular ejection fraction (LVEF), stroke volume (SV) index, heart rate (HR) and cardiac index (CI) measured at rest and peak exercise using exercise cardiac MRI (exCMR). Participants were classified by normal VO2 peak (n = 6, 99 ± 15% predicted) or impaired VO2 peak (n = 5, 60 ± 20% predicted) defined as VO2peak ≥85% age-predicted values.

Results: There was no difference in resting LVEF between the normal or impaired fitness groups (53.5 ± 4.4% vs 52.4 ± 8.0%, P = 0.76). In contrast, exCMR revealed participants with impaired fitness had an elevated increase in CI during exercise (Fig. 1A) due to a reduced SV and blunted augmentation from rest to peak exercise (Fig. 1B), with no difference in HR response (Fig. 1C).

Table 1. Reduction in sonographer injury risk for each recommendation.

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Conclusion: Reduced exercise capacity is associated with impairments in cardiac reserve in young cancer survivors. These measures may aid in the early identification of survivors at increased risk of heart failure.

http://dx.doi.org/10.1016/j.hlc.2019.06.010