Echocardiography and Imaging in Pediatric and Congenital Heart Disease

Utility of Echocardiographic Gradient for Evaluating and Predicting Need for Intervention in Children With Valvar Aortic Stenosis

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Background: In pts with valvar aortic stenosis (VAS), the relative accuracy of peak versus mean echocardiographic gradients (EG) for predicting peak-to-peak catheterization gradient (CG) is controversial, as is the relative accuracy of EG samples obtained from apical (AW) versus suprasternal or high parasternal window (HW).

Methods: We reviewed EG and CG data from 79 children (aged 9.5±5.9 yrs) with isolated VAS, recording the peak (AWpeak, HWpeak) and mean (AWmean, HWmean) EG and compared these with the CG. We recorded the need for intervention at catheterization.

Results: All EG estimates correlated in a linear fashion with the CG and with each other (R²=0.34-0.86; p<0.01). However, the correlations were suboptimal (0.58 – 0.63), demonstrating unpredictable over- or underestimation of the CG. AWpeak provided the closest overall estimate. HWpeak and HWmean were generally higher than the corresponding AWpeak and AWmean at high CG but showed little difference at low CG. AWpeak and HWpeak overestimated the CG in 60% (10 mmHg mean) and 86% (19 mmHg mean), respectively, whereas AWmean and HWmean underestimated the CG in 94% (23 mmHg mean) and 83% (16 mmHg mean), respectively, with errors as high as 70 mmHg. No independent variable (e.g., age, CG, window, peak versus mean) was associated with the magnitude of error. Of the 79 pts, 58 (67%) underwent balloon aortic valvuloplasty. No single EG method was adequately predictive of intervention with the area under the ROC curve = 0.79±0.06. No pt satisfying either of the following equations underwent intervention (AWpeak<107.1-0.5* HWpeak, AWmean<63.1-0.65* HWmean), and all pts satisfying either of the following equations did (AWpeak > 114.5-0.56* HWpeak, AWmean > 64.6-0.55* HWmean). In approximately 50% of pts intervention could not be predicted.

Conclusion: In children with isolated VAS, apical and suprasternal notch samples are not equivalent with HWpeak overestimating CG more than AWpeak. Both peak and mean EG frequently and unpredictably over- or underestimate CG. Although pts at high and low CG who did and did not have intervention could be predicted, we were not able to determine a reliable predictive equation for the 50% of pts with CG in the middle portion of the spectrum.

Myocardial Performance Index in Pediatric Patients With Primary Pulmonary Hypertension: Correlation With Pulmonary Artery Pressure and Response to Vasodilators

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Background: Management of primary pulmonary hypertension (PPH) relies on invasive measures of PA pressure (PAP) and resistance (PVR). Echo myocardial performance index (MPI) ([isovolumic contraction + relaxation time]/ejection time), a measure of global ventricular function, predicts outcome in adult PPH. This study determined the utility of MPI in assessing RV dynamics and PA reactivity in pediatric PPH and monitoring response to vasodilators.

Methods: We reviewed MPI and CG data from 78 children (aged 10.9±4.2 yrs) with isolated PPH. MPI was calculated at baseline (pre-intervention with the area under the ROC curve = 0.79±0.06). No pt satisfying either of the following equations underwent intervention (AWpeak<107.1-0.5* HWpeak, AWmean<63.1-0.65* HWmean), and all pts satisfying either of the following equations did (AWpeak > 114.5-0.56* HWpeak, AWmean > 64.6-0.55* HWmean). In approximately 50% of pts intervention could not be predicted.

Conclusion: In children with isolated VAS, apical and suprasternal notch samples are not equivalent with HWpeak overestimating CG more than AWpeak. Both peak and mean EG frequently and unpredictably over- or underestimate CG. Although pts at high and low CG who did and did not have intervention could be predicted, we were not able to determine a reliable predictive equation for the 50% of pts with CG in the middle portion of the spectrum.

Echocardiographic Screening for Clinically Silent, but Potentially Life-Threatening Coronary Anomalies

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Background: Some congenital anomalies of coronary artery origin may first present catastrophically with sudden cardiac death. In 1992 we introduced in our laboratory a standard comprehensive examination protocol mandating high-resolution magnification and low-velocity optimized color flow mapping of the coronary artery origins and proximal course. We sought to determine the extent and variety of potentially silent coronary anomalies detected by this protocol. Methods and Results: An echocardiographic database review was conducted of all pediatric patients examined from 1992 to 2003. There were 407 pts diagnosed with coronary artery abnormalities. The 358 pts with acquired coronary disease, clinically evident coronary anomalies or constrical malformations commonly associated with coronary anomalies were excluded. The remaining 49 pts had potentially silent coronary anomalies; 25 of these anomalies occurred in structurally normal hearts. Associated simple congenital heart defects in the remainder included atrial septal defect-5, ventricular septal defect-12, aortic coarctation-5, aortoventricular canal-1, aortic stenosis-1, pulmonary stenosis-1 and mitral prolapse-2. The most common anomalies were tiny coronary artery fistulae-14, right coronary artery (RCA) origin from the left sinus of Valsalva (SOV)-11, and high-takeoff of the RCA above the SOV-9, an anomaly associated with potential myocardial injury if undiagnosed prior to undergoing cardiological bypass with cardioplegia. The others were: single RCA-6, circumflex off the RCA-4, left coronary artery (LCA) from the right SOV-2, high takeoff of the LCA-1, high takeoff of both coronaryis-1, and single LCA-1. All wrong sinus coronary origins took an interarterial course. Three single RCA cases were diagnosed prospectively with an intraskeletal course of a left coronary artery. Conclusion: An echocardiographic screening protocol which mandates routine comprehensive examination of the coronary artery ori-