

Assessment of left ventricular functions with tissue Doppler, strain, and strain rate echocardiography in patients with familial Mediterranean fever

Echocardiography can give insight in both global as well as regional myocardial function. Fractional shortening and ejection fraction are well-known parameters for a considerable period of time for assessing left ventricular (LV) systolic function. However, these parameters only measure global systolic function. Using Tissue Doppler Imaging (TDI) allows assessment of both global as well as regional myocardial performance of the right (RV) as well as the left ventricle. Previous studies demonstrated its usefulness in the evaluation of myocardial function in patients after correction of their congenital heart defect (1-4). However, TDI has a number of disadvantages. No distinction can be made between active and passive motion using TDI. Furthermore, as TDI uses the Doppler principle it can only be used along the direction of the Doppler beam. Finally, conflicting results considering the load dependency of TDI exist.

Speckle tracking strain analysis makes use of the speckles, natural acoustic markers, present in the myocardium. Speckle tracking strain analysis is angle-independent and can assess both global as well as regional myocardial function. It can be used multidirectionally; radial, circumferential, and longitudinal. In addition, it can be used to assess rotational motion, including twist and torsion. Speckle tracking strain analysis has been used in several studies to assess myocardial dysfunction at an early phase. Disorders that have been focused upon include twin-to-twin transfusion syndrome (5) and anthracycline cardiomyopathy (6).

Familial Mediterranean Fever (FMF) is an autosomal recessively inherited disease characterized by recurrent self-limited attacks of fever accompanied by aseptic inflammation of serosal spaces, joints, and skin (7). Although acute attacks of this disease are self-limited, some patients develop amyloidosis, leading to end-stage renal disease. Other long-term complications are peritoneal adhesions, causing intestinal obstructions and female infertility, and amyloidosis of the gastrointestinal tract, liver, and spleen. Involvement of the myocardial tissue has been studied for some period. Although overt cardiomyopathy has not been described, more subtle changes in both diastolic and systolic function may develop. Development of the newer echocardiographic techniques as described above made it possible to study these subtle changes. Both the use of TDI (8, 9) and speckle tracking strain analysis (10) unmasked myocardial dysfunction in patients affected by FMF.

The study of Ceylan et al. (11) presents the results of different methods of echocardiography in pediatric FMF patients. The main findings of their study include a higher myocardial performance index (MPI), lower values for different TDI parameters, and a lower global longitudinal strain (GLS) as compared with those of normal controls. These results largely confirm the results of previous studies in both adults (9, 10) and pediatric patients (8). However, the finding of lower GLS, even at a young age is interesting. It underscores the cardiac involvement in FMF patients at an early stage. The next step in this research topic would be a longitudinal multicenter study to evaluate if a lower GLS has any prognostic significance. Such a study can also elucidate if the cardiac involvement in FMF patients usually is a stable decrease in overall myocardial function or if there are certain factors that can induce a continuous deterioration in myocardial function, ultimately leading to overt clinical heart failure. If the latter is the case, preventive strategies have to be developed.

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