

Recommended techniques for initial/ongoing assessment of cardiac anatomy and function in patients with MPS ¹

ECG

- Surface 12-lead: rhythm, conduction blocks, bundle branch blocks
- Holter 24-hours: rhythm, heart rate (values and variability by time and frequency domain), supraventricular and ventricular arrhythmia

ECHO

- | | | |
|--------------------|---|-----------------------------------|
| • 2D transthoracic | } | Posterior wall thickness |
| • M mode | | Interventricular septal thickness |
| • Doppler | | Left ventricular dimension |
| • Color doppler | | Left ventricular mass/hypertrophy |
| • Tissue doppler | | Left ventricular function |
| | | Valve thickness and function |

Adapted from Braunlin, *J Inherit Metab Dis*, 2011.

Abbreviations: 2D, 2-dimensional; BP, blood pressure; ECG, electrocardiography; ECHO, echocardiography.

Reference: 1. Braunlin EA, Harmatz PR, Scarpa M, et al. Cardiac disease in patients with mucopolysaccharidosis: presentation, diagnosis and management. *J Inherit Metab Dis*. 2011;34(6):1183-1197. doi:10.1007/s10545-011-9359-8.