Elevated QT interval variability (QTV) has been associated with increased cardiac mortality, but the underlying mechanisms are incompletely understood. Sympathetic activity is thought to be a main contributor to QTV. The aim of this study was to investigate the relation between cardiac sympathetic integrity and QTV in 15 patients with type 2 diabetes mellitus and varying degrees of cardiac autonomic neuropathy. Cardiac sympathetic innervation was assessed by 123I-mIBG scintigraphy based on heart-to-mediastinum ratio of 123I-mIBG uptake 4 hours after infusion. To assess QTV high resolution ECGs (1000 Hz) were recorded in the supine position and during standing. Beat-to-beat QT intervals were calculated over a period of 5 minutes, using a template-stretching algorithm. QTV was quantified as the logarithm of beat-to-beat QT interval variance. The group mean and standard deviation of HMR values were 1.84 ± 0.16. During standing QTV tended to be higher than in the supine position (1.07 ± 0.48 vs. 0.83 ± 0.37, p = 0.07). QTV during standing was significantly inversely correlated with HMR (r = -0.79, p < 0.001), but there was no significant correlation between QTV in the supine position and HMR. In conclusion, the amount of QTV measured during a period of sympathetic activation, as induced by orthostatic challenge, is reflective of cardiac sympathetic integrity in patients with diabetes mellitus. Progressive cardiac sympathetic dysfunction is associated with increased QTV. This might partly explain the association between QTV and cardiac mortality.