Although ERT has become the standard of care for patients with Pompe disease, careful monitoring of patients is also essential for optimal care. Guidelines for the care of patients with Pompe disease have been published by a number of countries and the goal of the presentation is to highlight some of the consensus recommendations. The goal of guidelines is to provide a general framework of evidence or consensus-based standards of care for the team caring for patients with Pompe disease. It is important to remember that the guidelines should be flexibly contextualised for the available clinic resources and must cater to the needs of the individual patient. Enrolment in a patient registry (i.e., http://www.pomperegistry.com/) may help in clinically relevant research; offering external support through various support groups (i.e., http://www.worldpompe.org; http://www.unitedpompe.com, etc.) is of interest to some, but not all, patients. Depression and anxiety may be part of the disease process and support via counselling and psychiatry may be needed. A team approach is recommended and members may include: neurology, medical genetics, respirology, cardiology, physiatry, psychiatry, speech and language pathology, occupational therapy, physiotherapy, nutrition and kinesiology. The frequency of assessment is a function of the severity of the disease, complications, comorbidities and patient factors (distance to clinic, finances, interest). Baseline testing would include; bloodwork (below + vitamin D), spirometry, ECG, echocardiography, neurological exam, with DEXA scanning (bone and body composition) and MRI being considered in certain centres. The general q6 month assessments would usually consist of blood tests (CK, glucose, HgbA1c, CK, CBC), neurological exam, and spirometry (sitting and supine). Other assessments should be considered as a function of availability and disease progression including; respirology studies (sleep study, blood gases, MIPs, MEPs, peak cough flow rate), swallowing studies, and quality of life measurements (i.e., SF-36). Patients with Pompe disease should have a diet record assessment and selected blood measurements done (vitamin D and B12, (? folate)) to identify deficiencies and optimise diet (higher protein content). Patients should meet with a physiotherapist and/or kinesiologist to evaluate and plan for exercise interventions including gradually progressive endurance and resistance exercise as well as respiratory muscle training.