Monochorionic (MC) twins represent a significant proportion of perinatal morbidity and mortality. Overall, 1 of 3 MC twin pregnancies will develop complications in relation to the MC placenta and the presence of interfetal placental anastomoses. From a clinical standpoint, these complications can be grouped into four main types of clinical problems: chronic transfusion, acute transfusion, growth discordance, and discordant malformation. Differential diagnosis of MC twin complications is still challenging due to frequent overlap of their clinical signs and the complex relationships between them. Clinical experience demonstrates that most instances of wrong management derive from failure to comply with a basic set of rules for diagnosis and management. In this review, we propose a simplified approach to the understanding of MC twin pregnancy, its potential complications, and the key concepts allowing adequate differential diagnosis and targeted management.