Autism spectrum disorder (ASD) is a neurodevelopmental disorder characterized by deficits in social communication and the presence of restricted interests and repetitive behaviors. There have been recent concerns about increased prevalence, and this article seeks to elaborate on factors that may influence prevalence rates, including recent changes to the diagnostic criteria. The authors review evidence that ASD is a neurobiological disorder influenced by both genetic and environmental factors affecting the developing brain, and enumerate factors that correlate with ASD risk. Finally, the article describes how clinical evaluation begins with developmental screening, followed by referral for a definitive diagnosis, and provides guidance on screening for comorbid conditions