Conjoined twins are a rare form of monozygotic twins: they occur in approximately one in 50,000 gestations and one in 250,000 live births. In most cases, twinning fusion defects are severe enough to preclude survival; anyway, the prognosis is strictly related to the type of fusion and encouraging results of separation after surgical correction are reported. Thus, identifying an effective first trimester screening technique in order to provide an early diagnosis of these malformations is mandatory. With the introduction of high resolution transvaginal ultrasound, and in particular first trimester 3D-imaging, an accurate diagnosis of conjoined twins becomes viable early in pregnancy. We report the case of a 30 years old 0-para woman who was referred to our department because of a first trimester ultrasound screening performed during the ninth week of amenorrhoea. The visualization of a sole embryo with a bifid appearance under transvaginal 2D-ultrasound made the physician suspect a twinning fusion anomaly. Thus, a 3D-sonography was scheduled at tenth weeks of gestation and pointed out a monochorionic, monoamniotic abdominal conjoined twin pregnancy (thoraco-omphalopagus). After adequate counselling, the patient decided to interrupt pregnancy without undergoing any further diagnostic procedure. The ultrasonographic diagnosis was confirmed by the histo-pathological findings. In this case, the identification of first-trimester sonographic indicators and the further evaluation of foetal anatomy under 3D-sonography allowed appropriate diagnosis of conjoined twin pregnancy. As reported in literature, 3D-ultrasound screening during the first trimester of pregnancy may be an helpful tool to diagnose twinning fusion defects, thereby improving clinical management and counselling options.