**Objective**
To describe dizygotic twins concordant for defects in neural tube closure.

**Methods**
Case report.

**Results**
We present the case of a 29-year-old pregnant woman referred to our center due to dichorionic diamniotic twin pregnancy (concordant with sex), hypothyroidism and fetal spina bifida associated to Arnold-Chiari malformation type II. She denied the use of folic acid supplementation during pregnancy. The previous pregnancy was uneventful. The patient's husband was a 32-year-old and nonconsanguineous man with unremarkable medical history. The patient's sister had myelomeningocele. Ultrasound examination performed at 22 weeks' gestation showed, in both fetuses, lemon sign, flattened cerebellum (banana sign), and lumbosacral myelomeningocele. Ventriculomegaly was observed only in the second twin. Fetal echocardiography of the twins was normal, and fetal MRI confirmed these findings. The babies were born by cesarean section at 38 weeks of pregnancy. Clinical evaluation of them confirmed the prenatal diagnosis. The additional examination through cerebral ultrasonography showed the presence, in both newborns, of dilated ventricular system. The correction of myelomeningocele was carried out in the first day after birth in both neonates.

**Conclusion**
Structural defects, including concordant malformations, are more frequently observed in monozygotic than in dizygotic twins. The association observed in our patients is considered rare. However, we can not exclude the possibility that the positive family history of defects in neural tube closure, considering a model of multifactorial disease, and the lack of maternal supplementation with folic acid may have contributed to a greater chance of involvement of both fetuses. Events related to the twinning itself, as well as other intrinsic not known genetic factors, can not be discarded.