Objectives: To evaluate the frequency of diagnosis of Congenital Zika Syndrome (CZS) during pregnancy and the prevalence of confirmed cases through RT-PCR for ZIKV in amniotic fluid.

Methods: A study with pregnant women with exanthema submitted to ultrasonography (prospective arm) and children with SZC (retrospective arm). Pregnant women with sonographic findings suggestive of SZC performed using Samsung WS80 Elite were submitted to amniocentesis for ZIKV research. Neurosonography (prospective arm) and Computed Tomography were performed in the neonate (retrospective arm). Chi-square test was used to compare the presence of symptoms between the prospective and the retrospective arm and the Mann-Whitney test to compare the number of ultrasonographies when intrauterine diagnosis was performed or not.

Results: 102 pairs of mothers and fetuses/children with CZS were included (28.7% prospective and 71.3% retrospective). Intrauterine diagnosis was performed in 52.9% of the total sample and in 100% of the prospective arm.

The median sonographic examination was 3 vs 4 (had no intrauterine diagnosis vs the had intrauterine diagnosis, p<0.001). Amniocentesis was performed in 19 pregnant women, with RT-PCR positive in 11 (57.9%). Among the ultrasound findings, ventriculomegaly was the main finding (43.1%), followed by microcephaly (42.2%), subcortical calcifications and/or nuclei of the base (30.4%), posterior fossa alterations (22.5%), arthrogryposis (7.8%) and corpus callosum dysgenesis (5.9%).

Conclusions: Despite having a high frequency, microcephaly was not present in all CZS cases, and the diagnosis of the syndrome should take into account intracranial findings. The percentage of cases without intrauterine diagnosis was high.