Aicardi–Goutieres syndrome (AGS) is a rare genetic disease arising during the first year of life and affect mainly the CNS. Fetal AGS mimics in utero infection, and presents with microcephaly and cerebral calcifications.

**Case report:**
We report a case of prenatal myocardial calcifications as the first manifestation of AGS. The parents are consanguineous with two healthy children and one affected, with severe intellectual disability, epilepsy and diffuse brain calcifications on CT scan. He died at 4 years of age without a definitive diagnosis.

At 21st week of the current pregnancy, fetal myocardial calcification with premature atrial beats and pericardial effusion were observed. No calcifications were detected in the brain, liver, spleen, and placenta. Maternal serology for intrauterine infection and tests for ANA, anti-SSA and anti-SSB were negative. Ultrasound at 27 week revealed calcifications in myocardium, caudothalamic groove and liver. At 32 weeks, reduced cardiac function, multiple bilateral thalamic, brainstem, and striatal calcifications and a unilateral parenchymal temporal lobe cyst were detected. The couple chose to terminate the pregnancy. Genetic analysis of the amniotic fluid revealed biallelic mutations in TREX1 gene compatible with AGS. The same mutation was detected in the DNA of the previous affected sibling.

**Conclusion:** AGS is a genetic disease associated with a high risk of recurrence. It mimics congenital infection and should be considered in cases with negative TORCH workup. Myocardial calcifications and arrhythmia are unusual in AGS but may be the earliest manifestations of AGS due to TREX1 mutation.