We present the case of a 33 years-old patient, G2P1, referred to our center for a dichorionic-diamniotic twin pregnancy. She is now for a Crohn disease treated with Imuran. The first trimester ultrasound at 12 5/7 weeks showed a nuchal translucency for BB1 of 1,4mm associated with a microretrognathia. BB2 was normal with a nuchal translucency of 1,2mm.

Four weeks later, an early morphology ultrasound was made demonstrating a severe retrognathia for BB1 and an estimated fetal weight below the third percentile. Moreover the axis of the heart was deviated and they noted anomalies of both hands. BB2 showed no anomaly.

Selective termination of pregnancy was decided by the couple and performed at 17 weeks of pregnancy without complication.

An amniocentesis was performed and confirmed a Nager syndrome with an heterozygote mutation c.1A >G in the SF3B4 gene. The patient had a vaginal delivery at 40 weeks with no complication.

The mode of transmission is autosomal dominant and the prevalence is unknown. Few cases of Nager syndrome have been reported. We presented a rare and early diagnosis of Nager syndrome and his management in dichorionic twins.