Non-compaction (NC) cardiomyopathy is characterized by prominent ventricular trabeculae with intertrabecular recesses with a thin compacted myocardial layer. It is clinically diverse, ranging from no symptoms to heart failure, arrhythmia, and thromboembolism. 30-50% of cases are familial with autosomal dominant and x-linked inheritance and several sarcomeric, cytoskeletal and NOTCH signaling pathway gene involvement. It is commonly associated with congenital heart defect

**Case report:**

We describe the prenatal diagnosis of NC cardiomyopathy. She is the second child of healthy, nonconsanguineous Ashkenazi Jewish parents. Her three-year-old sister has a mild form of NC cardiomyopathy with left ventricular (LV) dilatation, apical thickening and a mild global hypokinesis. Our patient's fetal ultrasound revealed biventricular hypertrophy with mildly reduced contractility, heavily trabeculated RV apex and prominent LV trabeculations. Color Doppler showed that the deep intertrabecular recesses are filled with ventricular blood, compatible with NC cardiomyopathy. A large 6mm membranous septal defect (VSD) was seen. Amniocentesis for chromosomal microarray was normal. She was born full-term weighing 2950gr, not dysmorphic. Echo confirmed the prenatal diagnosis.

**Conclusion:** The prenatal diagnosis of the unique non-compaction cardiomyopathy with the diverse clinical outcome enables an appropriated genetic and cardiologic counselling. Additional congenital heart malformation are common and should be looked for