Ovarian cancer arising in endometriotic cyst has recently received much attention, since infertile women with endometriosis seem to be increasing in number and ovarian cancers in endometriosis sometimes appear during laparoscopic surgery for fertility preservation. However, it remains unknown about its incidence, natural course, molecular genetics and adequate clinical management.

It is well known that major histological type of ovarian cancer in endometriosis is either clear cell or endometrioid adenocarcinoma, and the expected incidence is reported to be 0.7% according to the prospective study by Kobayashi et al. Our multicenter study on ovarian cancer patients whose transvaginal ultrasonography (TVS) findings were available 12 months or less prior to the diagnosis showed that such clear cell or endometrioid carcinoma develops during the follow-up of ovarian endometriotic cyst, in the interval of approximately 6 months, along with increase in size and appearance of solid portion by TVS, and that most of them were diagnosed at Stage I. Therefore, close follow-up of the patients with endometriotic cyst using TVS at every 6 months would be mandatory for its early detection.

Our molecular analysis of ovarian clear cell carcinomas has disclosed the characteristic gene expression pattern (OCCC signature), containing up-regulation of de-methylated, stress-related genes, such as HNF-1beta, VAN, SOD2 and STAT3. Further analysis revealed that OCCC signature represents the microenvironment of endometriotic cyst containing much amount of free iron, being stressful to cell survival. Bioinformatics also demonstrated that multikinase inhibitor such as Sorafeniv may be effective for clear cell carcinoma that is resistant to standard taxane-platinum chemotherapy. Therefore, we started clinical trial using Sorafeniv for patients with recurrent ovarian clear cell carcinoma.