Announcement

Familial Ovarian Cancer: The Establishment of a European Registry

Background

To date, epidemiological studies have provided only few data useful for the identification of women at risk of developing ovarian cancer. Furthermore, there are no cheap, reliable screening measures, and this malignancy is almost invariably diagnosed at an advanced stage.
During the past decade, evidence consistent with a relevant role for genetics in the etiology of ovarian cancer has rapidly accrued. Three carcinoma-prone conditions have been recognized in which malignant epithelial ovarian neoplasms show significant familial concentration: 1) Site-specific ovarian carcinoma, where familial risk is restricted to ovarian cancer; 2) Breast/ovarian cancer syndrome, where carcinoma of the breast is associated with ovarian carcinoma; 3) Cancer family syndrome (Lynch syndrome II), consisting of hereditary non-polyposis colorectal cancer with proximal colonic cancer predominance, associated with endometrial and/or ovarian carcinoma.
Families with two or more first-degree relatives affected by ovarian cancer have been conventionally considered suitable for epidemiological and genetic analyses. Studies on such families at high risk for ovarian carcinoma showed that the hereditary pattern seems consistent with the segregation of an autosomal dominant mutation with variable penetrance. However, data so far reported have not conclusively clarified the mode of genetic trasmission.
Lynch estimated that the hereditary subgroup represents 5%-10% of all ovarian cancers. Although the real entity of the genetically controlled ovarian cancer may be overestimated, it is very clear that familial ovarian cancer should no longer be looked on as a rare occurrence.
As recent molecular genetic studies have shown that non-hereditary and hereditary forms of the same tumour may share the same genetic pathway of development -such as in retinoblastoma and colon cancer- molecular genetic studies in ovarian cancer families would be relevant to ovarian cancer in general. To this purpose, it has to be stressed that the number of such families should be adequate, and a common effort in their identification is desirable.
Genetic counseling in individuals from such kindreds is obviously of great importance from the standpoint of surveillance, management and adequate approach to psychological problems (sisters and daughters of probands in families with a history of ovarian cancer have approximately a 50% chance of developing the disease compared to 1.4% chance in women without this family history).

Familial Ovarian Cancer European Registry

A U.S. Familial Ovarian Cancer Registry was established in 1981 for the accession of families which contain two or more first-degree relatives with ovarian cancer. This registry, directed by Dr. M.S. Piver (Roswell Park Memorial Institute, Buffalo NY), has been recently renamed The Gilda Radner Familial Ovarian Cancer Registry in honor of the late comedienne, Gilda Radner, with the honorary chairman, Mr. Gene Wilder.
However, data on familial ovarian cancer are still insufficient -most of them being retrospectively collected-, and a further effort is desirable for a larger, prospective recruitment of ovarian cancer families. Moreover, cases from Europe have been only occasionally reported, and, if sufficient suitable families can be collected, a bank of biological samples from family members could be extremely important for marker and molecular genetic studies.
In this respect, a European Registry on Familial Ovarian Cancer has been established in 1989 under the auspices of the E.O.R.T.C. Gynaecological Cancer Cooperative Group and in close collaboration with the U.S. Registry, with the following purposes:
1. Identification and registration of ovarian cancer-prone families (families with at least two first-degree relatives affected by ovarian cancer) in Europe; 2. Knowledge of detailed epidemiological characteristics of familial ovarian cancer; 3. Establishment of a bank of blood and tumour samples from family members of the Registry (for marker and molecular genetic studies); 4. Study of genetic transmission pattern and identification & registration of high-risk subjects; 5. Counseling & surveillance of high-risk subjects; 6. Exchange of data with the U.S. Registry.
Data are elicited regarding: social background, personal and ob/gyn history, personal history of cancer, familial history of cancer, and pedigree, and assumed as strictly confidential. Serum and blood samples from probands and their first-degree relatives are requested for biochemical marker and molecular genetic studies. Genetic counseling is provided together with a follow-up program for high-risk subjects.
To date, about 50 European Institutions have been accepted to participate in the Registry from the following countries: Italy, France, W.Germany, Belgium. Holland, Austria, Denmark, Sweden, Iceland, USSR, Hungary, Yugoslavia, Turkey, Greece. Cases from U.K. are collected by Dr. B.Ponder (Human Genetic Research Group, Cancer Research Campaign) who is conducting a study on a national basis.

For registration forms and any further information, please address requests to: Stefano Greggi, M.D., Ph.D. (Study Coordinator) or prof. Salvatore Mancuso, M.D. (Chairman), Familial Ovarian Cancer European Registry, Dept. of Gynaecology & Obstet., Universita' Cattolica S.Cuore, largo A.Gemelli 8, 00168 Rome, Italy (Tel. No.: 6-33054496; Fax No.: 6-3051343).