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Central Registration of Multiple Endocrine Neoplasia Type 2 Families in the Netherlands

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If means for both early detection and treatment are available, periodic surveillance of families known to have a genetic predisposition to disease improves prognosis and life expectancy, especially if screening is started at an early age. However, periodic examination of large groups requires special administrative and outpatient facilities, and the responsibility for the continuity of screening programs cannot be assumed by an individual physician or general internist. To solve these problems, a central registration center can be established to monitor the expression of the disease in families at risk, although central registration raises ethical and legal problems. The privacy of patients and their relatives is involved, and costs are high. Nevertheless, advantages clearly outweigh the disadvantages.

In 1981, a pilot study was initiated to determine the possibility for decentralized screening and central registration of large groups of patients with the MEN-2A syndrome and their relatives. This syndrome is especially suited for such a study since reliable screening tests exist and, if the disease is found in an early stage, curative surgery is possible.

Patients and Methods

The 20 to 30 known kindreds with the MEN-2A syndrome in the Netherlands are composed of approximately 400 patients and 2,500 relatives to be examined annually (1-3). Over 300 patients are already known, with 100 more expected to come from the present screening efforts of relatives within these kindreds. An additional 100 patients are expected to be found in new kindreds. At present only about 33% of the at-risk individuals are being examined each year.

In November 1983 a foundation was established for the early detection of inheritable tumors. Its primary objectives are to encourage early detection and to establish a registration system. A periodic screening program is being developed for individuals who belong to families with inheritable tumors such as MEN-2 syndromes and familial polyposis coli. In addition, the foundation will promote scientific research connected with inheritable tumors.

A registration center has been established to collect data on patients and families as provided by their physicians. The manager of the center is responsible to the foundation council, composed of physicians who have placed their patient data at the disposal of the center. Each year the center will ask the physician to arrange periodic examinations for patients and their relatives. Data will be sent to the center so that a record of the annual screening will exist. In this way, decentralization of screening programs has been ensured so that each individual can choose his or her own physician who will perform tests locally. If physicians leave their practices, the center will arrange for other physicians to take over the screening and supply information about "new" patients. To protect the rights of the patients and their family members, the center is not permitted to give out information to third parties without their consent. Medical data might be used for scientific research only after consent of the physicians concerned.

Discussion

At present, approximately 300 patients and 300 family members are being screened for the MEN-2 syndrome; an approximately equal number are being screened among polyposis coli families. Ideally, a group of 2,500 individuals of the MEN-2 families and 6,000 individuals of the polyposis families should be examined periodically; however, it is expected that the size of the number to be regularly examined will grow.

Within a few years new diagnostic tests (eg, demonstration of marker genes or coupling to restriction enzyme fragment length polymorphisms) will make it possible to assess predisposition for the disease before

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expression occurs. High-risk individuals will be identified, and the number of family members to be examined periodically will decrease. For the MEN-2 syndrome in the Netherlands, only 500 to 700 high-risk individuals are likely to remain; for polyposis coli, the number is probably 1,500.

At present the activities of the foundation are limited to MEN-2 syndromes and polyposis coli. Later, other autosomal dominant inherited diseases will be admitted by the foundation council and monitored by the central registration center.

Conclusions

After a trial period of three years, the following conclusions about the registration system are possible:

- Large groups of patients and their relatives can be handled properly.
- The rights of patients and their relatives can be protected.
- Physicians and families are cooperative.
- Other syndromes can be included if sensitive tests and curative treatment are available.

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