Wilms Tumor 1

Alternative Names
WT1
Nephroblastoma

WHO International Classification of Diseases
Neoplasms

OMIM Number
194070

Mode of Inheritance
Autosomal dominant, somatic mutation

Gene Map Locus
Xq26, 13q12.3, 11p13

Description
Wilms tumor is one of the most common solid tumors of childhood, occurring in 1 in 10,000 children and accounting for 8% of childhood cancers. It is believed to result from malignant transformation of abnormally persistent renal stem cells which retain embryonic differentiation potential. The risk of Wilms tumor is increased in association with several recognizable congenital malformation syndromes, although these cases account for less than 5% of all clinical patients with Wilms tumor.

Synchronous bilateral Wilms tumor occurs in about 5% of all patients with Wilms tumors. Patients with synchronous bilateral Wilms tumor are known to present at younger age than those with unilateral Wims tumor. Whereas nephrectomy has remained the mainstay of treatment for patients with unilateral Wilms tumor, this is not true for bilateral cases. Patients with bilateral Wilms tumor pose a complex therapeutic problem, and the emphasis is currently on maximal preservation of functioning renal parenchyma. The role of preoperative chemotherapy is now well-established in the management of patients with synchronous bilateral Wilms tumor.

Molecular Genetics
Germline mutations cause less than 5% of Wilms tumors; most WTs are sporadic. However, numerous instances of multiple sibs with Wilms tumor have been described. Several genes or chromosomal areas have been associated with the development of WT: WT1 at 11p13, WT2 at 11p15.5, WT3 at 16q, WT5 at 7p15-p11.2, and WT4 at 17q12-q21.

Epidemiology in the Arab World

Egypt
Zaghloul et al. (1994) reported 112 children with pathological diagnosis of Wilms' tumor that were treated during the period 1979-1989. They were postoperatively staged as follows: stage I, 25 patients; stage II, 27 patients; stage III, 48 patients; and stage IV, 9 patients. Three patients were preoperatively classified as stage V. Except for Stage V patients, the treatment regimens consisted of nephrectomy followed by radiotherapy and/or chemotherapy according to stage of disease and pathology. Stage I, II, and III patients with favorable histology enjoyed 94 +/- 6%, 86 +/- 8%, and 71 +/- 8% 10-year actuarial survival, respectively. Stage IV patients and those in stages I, II, and III with unfavorable histology had a 10-year actuarial survival of 36 +/- 8%. Zaghloul et al. (1994) noted that massive tumor rupture at surgery increased the incidence of local relapse but not of distant metastasis and did not affect the overall survival rates.

United Arab Emirates
Abou-Chaaban et al. (1997) studied the pattern of pediatric renal diseases among children in the Dubai Emirate during the period from 1991 to 1996. In this period, a total of 712 pediatric patients, including 230 nationals of the United Arab Emirates, were seen with various renal problems. In their study, Abou-Chaaban et al.
(1997) observed one case of an Emirati national with nephroblastoma. In 1999, Nawaz et al. presented a prospective review of case notes on children treated for Wilms tumor at Tawam Hospital between January 1982 and December 1996. Of 34 cases diagnosed with Wilms tumor, 17 had tumor on the left side, 10 on the right, and seven (20.6%), including five males, were bilateral. This is significantly higher than the expected frequency of bilateral Wilms tumor. The age of the patients varied between 1.3 to 5 years. None of the patients had any signs suspicious of familial Wilms tumor. Preoperative chemotherapy proved to be effective in debulking and down staging the tumors. In one of the patients of Nawaz et al. (1999) there was total disappearance of the Wilms tumor on one side.

Al Talabani et al. (1998) studied the pattern of major congenital malformations in 24,233 consecutive live and stillbirth at Corniche hospital, which is the only maternity hospital in Abu Dhabi, between January 1992 and January 1995. A total of 401 babies (16.6/1,000), including 289 Arabs, were seen with major malformation. Sporadic conditions accounted for 26% of the cases. In their study, Al Talabani et al. (1998) observed two patients with renal tumors (teratoma and Wilms) in families from the United Arab Emirates. Recurrence was not reported in the families. Al Talabani et al. (1998) concluded that their study was very close to representing the true incidence of congenital abnormalities in the whole United Arab Emirates, as they investigated over 98% of deliveries in Abu Dhabi, the capital of United Arab Emirates.

References

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