EPIDEMIOLOGY OF PATIENTS WITH WILMS TUMOUR REGISTERED IN SUCCESSIVE UK-TRIALS THROUGH 38 YEARS

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Background
Since 1979, 2,510 patients with Wilms tumour (WT) have been registered in five consecutive UK clinical trials (UKW1-3, SIOP-2001, and IMPORT). We reviewed all trial data with focus on congenital abnormalities (CA) and laterality.

Methods
We categorised patients with WT into five mutually exclusive groups; WT and aniridia with/without urogenital malformations (WA), urogenital/renal malformations including Denys-Drash syndrome (UM), hemi-hypertrophy including Beckwith-Wiedemann syndrome (HH), other congenital abnormalities (other-CA), and without CA (non-CA). We compared distribution of sex, age at diagnosis, stage, histology and 5-year overall survival (OS) of each group to those of non-CA. Bilateral vs unilateral tumours were compared for similar outcomes.

Results
Numbers and proportions in each group were: WA (n=24, 1%); UM (n=79, 3%); HH (n=65, 3%); other-CA (n=116, 5%); non-CA (n=2,226, 88%). Bilateral (n=181, 7%); unilateral(n=2,309, 92%).
Patients in WA, UM and other-CA groups had younger median age of diagnosis and higher proportion of bilateral disease compared to non-CA (20m, 21m and 30m vs 39m, and 33%, 19%, 17% vs 6%, respectively). HH patients showed no significant difference compared to non-CA group (41m and 11%). UM had male predominance (M/F ratio 1.72 vs 0.88 in non-CA).

There were no significant differences in 5-year OS for each group compared to non-CA [83% (WA), 88% (UM), 91% (HH), 87% (other-CA), vs 88% (non-CA)].
Compared to unilateral disease, the bilateral group had female excess (M/F ratio 0.62 vs 0.93), younger age at diagnosis (24m vs 39m) and poorer 5-year OS (82% vs 89%, p=0.003) that had improved to 90% for cases diagnosed since 2002.

Conclusions

About one in nine patients with WT had congenital abnormalities. These patients are diagnosed at earlier age, have more frequent bilateral tumours but 5-year survival rate seems very similar. Compared to unilateral, bilateral WT were younger and had poorer survival.