

## X Tumori delle cellule germinali e altri tumori gonadici

### Germ-cell trophoblastic and other gonadal neoplasms

I tumori maligni delle gonadi e delle cellule germinali rappresentano complessivamente il 3% dei casi di neoplasia nell'età pediatrica registrati nella banca dati AIRTUM incidenti nel 1998-2002, con un **tasso di incidenza** (TI) standardizzato per età pari a 4,9 casi per milione di bambini per anno; (IC 95% 3,6-6,1) (TI standardizzato sulla popolazione europea 5,1; IC 95% 2,8-7,4).

La classe raggruppa diverse entità nosologiche, tra cui la più frequente è costituita dalle neoplasie gonadiche derivanti da cellule germinali, che rappresentano il 43%.

Una diagnosi istologica è disponibile per 83,3% dei casi, senza nessun DCO. Il **rischio cumulativo** fino a 14 anni di età è 0,07 per mille (IC 95% 0,06-0,09). In entrambi i sessi il tasso di incidenza ha presentato un andamento per età irregolare, con l'incidenza inferiore per la classe di età 5-9 anni.

La **frequenza** dei tumori maligni delle gonadi e delle cellule germinali è simile tra i due sessi e non presenta variazioni significative tra le diverse aree geografiche.

L'**andamento per periodo** dell'incidenza non mostra variazioni statisticamente significative, ma suggerisce un possibile andamento diverso nei due sessi: aumento dell'incidenza per i maschi (APC 5,8%; IC 95% da -0,07 a 13,8%) e tassi stabili tra le femmine (APC 0,8%; IC 95% da -0,5 a 7,0%).

La **sopravvivenza** ha mostrato nel corso dello studio solo modeste variazioni. Non si osservano variazioni statisticamente significative tra i diversi periodi di diagnosi. Per i casi diagnosticati nel 1998-2002, la sopravvivenza cumulativa osservata a 5 anni è 81,9% (IC 95% 71,6-92,2%). La sopravvivenza è risultata inferiore per i casi diagnosticati sotto i 5 anni di età.

*Germ-cell malignant tumours and other malignant tumours of the gonads accounted for 3% of paediatric (age 0-14 years) malignancies incident in 1998-2002 and recorded in the AIRTUM database. Age-standardised incidence rate (IR) was 4.9 cases per million year (95% CI 3.6-6.1). The IR standardised by age on the European population was 5.1 (95% CI 2.8-7.4). The category groups different nosological entities: the most frequent were gonadal neoplasms, that accounted for 43% of cases. Diagnosis was histologically confirmed for 83.3% of cases. No cases were included only on the basis of the cause of death.*

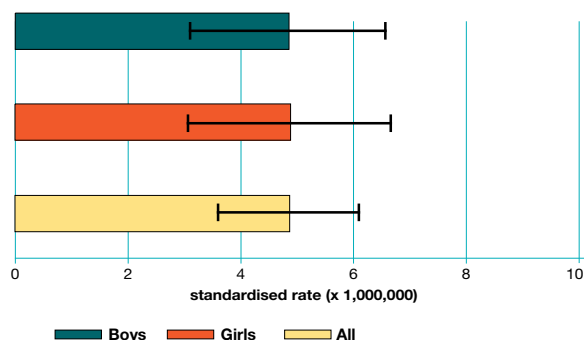
*Cumulative incidence up to age 14 was 0.07 per thousand (95% CI 0.06-0.09). In both genders the incidence curve by age was U-shaped, with the*

*lowest incidence in the 5-9 years age class. Incidence was similar in the two genders and did not present relevant differences by area of residence.*

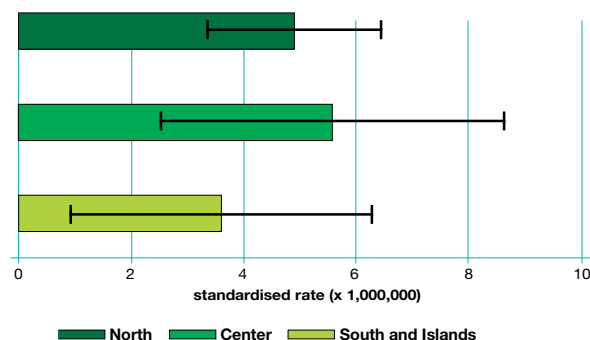
*Overall, no incidence trends over period of diagnosis were observed, but when data were analyzed by gender, different results were observed. An increasing trend, close to statistical significance was observed among boys (APC 5.8%; 95% CI from -0.07 to 13.8%) while constant rates were measured among girls (APC 0.8%; 95% CI from -0.5 to 7.0%).*

*Survival presented only modest variations during the study period, which were not statistically significant. Cumulative survival at 5 years after diagnosis for the cases diagnosed in 1998-2002 was 81.9% (95% CI 71.6-92.2%). The lowest survival was observed for the cases diagnosed below age 5.*

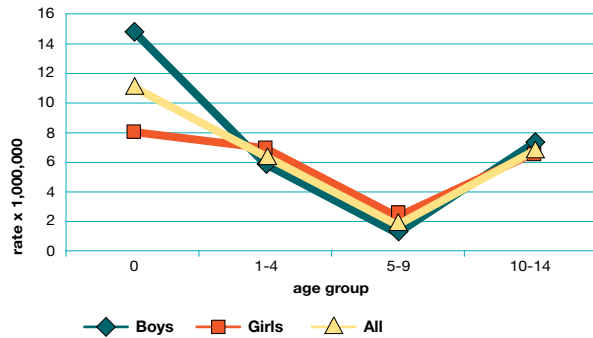
Incidence rate by gender



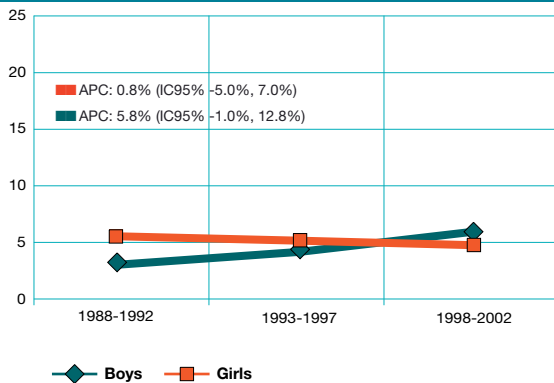
Incidence rate by area



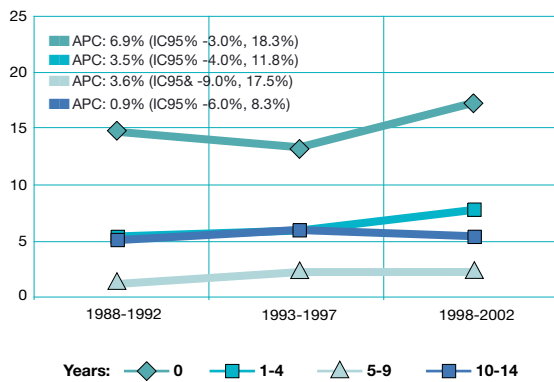
## Incidence rate by age and gender



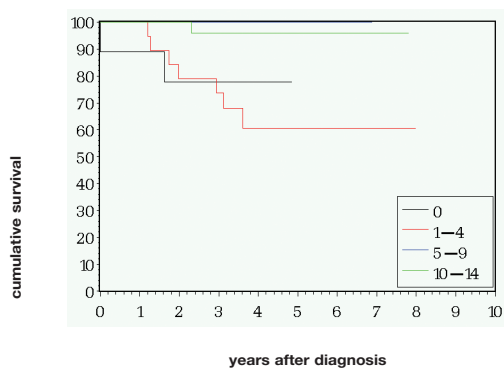
## Incidence trend by gender



## Incidence trend by age



## Cumulative survival by age



Distribuzione percentuale e numero di casi per i diversi tipi di tumori maligni delle gonadi e delle cellule germinali nella banca dati AIRTUM, 1998-2002, secondo le categorie della International Classification for Childhood Cancer (ICCC).

Frequency distribution (number of cases and percentage) of germ-cell, trophoblastic and other gonadal neoplasms in the AIRTUM database, 1998-2002, by type according to the ICCC classification and age.

| ICCC   | 0 | 1-4 | 5-9 | 10-14 | n  | %   |
|--|---|-----|-----|-------|----|-----|
| Xa Intracranial and intraspinal germ-cell tumours      | 3 | 0   | 3   | 6     | 12 | 20  |
| Xb Other and unspecified non-gonadal germ-cell tumors  | 4 | 8   | 0   | 1     | 13 | 22  |
| Xc Gonadal germ-cell tumors                            | 2 | 10  | 1   | 13    | 26 | 43  |
| Xd Gonadal carcinomas                                  | 0 | 0   | 0   | 0     | 0  | 0   |
| Xe Other and unspecified malignant gonadal             | 0 | 1   | 2   | 6     | 9  | 15  |
| X Germ-cell, trophoblastic and other gonadal neoplasms | 9 | 19  | 6   | 26    | 60 | 100 |

Sopravvivenza cumulativa a 1, 3 e 5 anni dalla diagnosi per i casi di tumori maligni delle gonadi e delle cellule germinali diagnosticati nel periodo 1998-2002, per classe di età. Fonte: banca dati AIRTUM.

Cumulative survival at 1, 3, and 5 years after diagnosis for children affected by germ-cell, trophoblastic and other gonadal neoplasms incident in 1998-2002. Data from the AIRTUM database.

| Age group | n. cases | Cumulative survival |                     |                     |
|-----------|----------|---------------------|---------------------|---------------------|
|           |          | 1 y (95%CI)         | 3 y (95%CI)         | 5 y (95%CI)         |
| 0         | 9        | 88.9 (68.4-100.0)   | 77.8 (50.6-100.0)   | <sup>a</sup>        |
| 1-4       | 19       | 100.0 (100.0-100.0) | 73.7 (53.9-93.5)    | 60.5 (37.0-83.9)    |
| 5-9       | 6        | 100.0 (100.0-100.0) | 100.0 (100.0-100.0) | 100.0 (100.0-100.0) |
| 10-14     | 26       | 100.0 (100.0-100.0) | 96.0 (88.3-100.0)   | 96.0 (88.3-100.0)   |

Log-rank test: p= 0.0212 <sup>a</sup> Censored data

Sopravvivenza cumulativa a 3, 5, 10 e 15 anni dalla diagnosi per i casi di tumori maligni delle gonadi e delle cellule germinali diagnosticati nel periodo 1988-2002, per periodo di diagnosi. Fonte: banca dati AIRTUM.

Cumulative survival at 3, 5, 10 and 15 years after diagnosis for children affected by germ-cell, trophoblastic and other gonadal neoplasms incident in 1988-2002, by period of diagnosis. Data from the AIRTUM database.

| Period | n. cases | Cumulative survival |                  |                  |                  |
|--------|----------|---------------------|------------------|------------------|------------------|
|        |          | 3 y (95%CI)         | 5 y (95%CI)      | 10 y (95%CI)     | 15 y (95%CI)     |
| 1988   | 36       | 88.9 (78.6-99.2)    | 88.9 (78.6-99.2) | 88.9 (78.6-99.2) | 88.9 (78.6-99.2) |
| 1992   | 56       | 87.5 (78.8-96.2)    | 87.5 (78.8-96.2) | 87.5 (78.8-96.2) | -                |
| 1993   | 56       | 87.5 (78.8-96.2)    | 87.5 (78.8-96.2) | 87.5 (78.8-96.2) | -                |
| 1997   | 60       | 86.3 (77.5-95.2)    | 81.9 (71.6-92.2) | -                | -                |
| 1998   | 60       | 86.3 (77.5-95.2)    | 81.9 (71.6-92.2) | -                | -                |
| 2002   | 60       | 86.3 (77.5-95.2)    | 81.9 (71.6-92.2) | -                | -                |

Log-rank test: p= 0.7138

## Cumulative survival by period

